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Contents

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Growing Bigger and Better

Dear colleagues and friends!!


Dr. (Prof.) Subhash C. Dadeya

As we bid goodbye to the chilly winters and welcome the scorching summers with open arms, we now bring the next issue of the DOS Times in our sub speciality series. Like a welcome summer rain, this new series will cleanse and cherish our minds.

This issue highlights a plethora of articles from ophthalmic plastic surgery. This subspeciality involves eyelid diseases and tumours, orbital diseases as well as aesthetics. So we have tried to incorporate articles from all these streams, reviewing the most important reviews for residents and fellows. Moreover our knowledge about these disorders is incomplete without an accurate and adequate understanding of the radiology of the eye.

Our special edition in oculoplastics aims to help residents identify all signs of an orbital disease as well as provide a review of literature and appropriate references. The edition is designed to serve as a residents primary source of information on the detailed topics.

I am indebited to my team and all individuals who assisted with the preparation of this manuscript.

Wishing you a joyful reading.

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Ptosis: Management Pearls

Dr. Ashok K. Grover

Dr. Sushil Kumar

Dr. Gangadhar Sundar

Dr. Anuj Mehta

Blepharoptosis surgery is a common oculoplastic procedure done primarily to clear the visual axis, reduce amblyopia in young patients, and improve superior visual fields. The secondary goal is to improve cosmesis by producing symmetric lid crease and contour in the upper lids. The choice of surgical procedure varies depending on the amount of ptosis and levator function. However the choice of procedure can also vary from surgeon to surgeon. This is specially true for sling surgery where different materials and methods of passing the slings are available.

We therefore asked a panel of eminent oculoplasty surgeons from around the globe about their opinion and views on various aspects of management approach and treatment options through this questionnaire.

(AKG) Dr. Ashok K. Grover: is a prominent name in the realm of ophthalmology. He is an alumnus of Dr. Rajendra Prasad Eye Centre, New Delhi. He pursued his degree of MD from AIIMS. He started his professional life with the Maulana Azad Medical College, New Delhi as an Associate professor in the year 1984. In 1992, Dr. Grover joined Sir Ganga Ram Hospital as a senior consultant and is currently chairman of Dep't of Ophthalmology. He was awarded the prestigious Padamshri by the President of India in year 2009. He is Past President of AIOS, OPAI and APSOPRS. He is also chairman of Vision Eye Centres.

(SK) Dr. Sushil Kumar: *(Director, Professor and Head of Oculoplasty Services)* Guru Nanak Eye Centre, New Delhi.

(GS): Dr. Gangadhar Sundar: after obtaining his degrees from Madras Medical College, India and advanced subspecialty training from the Henry Ford Hospital, Detroit, specializes in the diseases and surgery of the Eyelids, Lacrimal system, Orbit & Oculofacial diseases and Ophthalmic Oncology. A strong believer of multidisciplinary, multimodality approaches to complex ocular and orbitofacial disorders, he was instrumental in starting various multidisciplinary services including the NUH Orbitofacial trauma service (with Craniomaxillofacial Surgeons), NUH Retinoblastoma service (with pediatric oncologists, neuro-interventional radiologists and pathologists), NUH Thyroid Eye Disease service (with Endocrinologists & immunologists) and the NUH Lacrimal service (with Rhinologists). He has trained Fellows from Singapore, Malaysia, Myanmar, Philippines, India, Thailand & the United Kingdom.

(AM) Dr. Anuj Mehta: is an alumnus of the Maulana Azad Medical College and Guru Nanak Eye Centre, from where he completed his graduation and postgraduation both. He had the privilege of being awarded the prestigious WHO fellowship for Oculoplasty from the L.V.Prasad Eye Institute, Hyderabad. He is presently working at the V.M.M.C & Safdarjung Hospital, New Delhi as Professor and Consultant and is the incharge of the Oculoplasty and Ocular Oncology clinic.

Q1: Of all the cases of ptosis encountered at your centre, how many of these are congenital or acquired? What is the proportion of severe, moderate or mild ptosis?

AKG: The spectrum of ptosis cases operated has undergone a great change over the last 2 decades. From about 90% of cases being congenital earlier to about 70% congenital and 30% acquired now. About 60% of all congenital cases are moderate, 30% severe and 10% mild. About 80% of acquired cases are severe.

SK: Most of the cases of blepharoptosis presenting in our centre are of congenital variety and they constitute 90-95% of all ptosis cases. Acquired variety of ptosis remain comparatively less. Severity wise division of congenital ptosis cases shows mild ptosis (<2mm) accounting for roughly 10-15%; moderate ptosis (>2 mm <4 mm) constitute the maximum number (approx. 65%) and the severe ptosis (>4 mm) accounting for (15-25%).

GS: Acquired 85% Congenital 15%, Severe 15%, Moderate 65%, Mild 20%.

AM: We see about 50-60 patients of ptosis in a year. Approximately 80% of these patients have congenital ptosis, 70-75% of patients attending our clinic have severe ptosis, 20% have mild ptosis and 5-10% have moderate ptosis.

Q2: What are the key essential points in a ptosis examination?
How do you assess severity of congenital ptosis especially in kids less than 3 years of age where cooperation and comprehension can pose a major problem?

AKG: I would like to stress the two key inputs in decision making – amount of ptosis and levator action by Berke's method. Of course ocular motility, Bell's phenomenon are other essential ingredients. In children, younger than 3 years the important assessment is whether the pupil is covered or not and whether the head posture necessary to use the eye is unacceptably high. Simple observation in the clinic usually suffices.

SK: Detailed workup of a ptosis case is must for the best outcome of any ptosis procedure that is chosen. Amount of ptosis, levator muscle action and the Bell's phenomenon status must be assessed along-with other parameters which include visual acuity/ amblyopia, ocular motility imbalance/strabismus especially the vertical squint, tear film status, jaw-winking phenomenon, any diurnal variation in the amount of ptosis etc. For this detailed history, neurologic workup and some pharmacologic tests if required needs to be done.

If the patient is a child where cooperation cannot be sought a careful observation of the child is made for any chain elevation, head tilt, movement of the upperlid on sucking and also the lid covering of the pupil region. For this an ambient environment for the child is must. Surgeon should be patient enough for the observations. If possible a note should be made of behaviour of the child on occlusion of one eye and also behaviour of the everted upperlid in up and down gaze by showing a toy.

GS: Unilateral or bilateral?

MRD1, Levator function (mild, moderate or severe dysfunction), Simple vs Atypical/Complex congenital ptosis, normal Bell's vs reverse Bell's phenomenon, Visual disability vs Cosmesis, Parents/patient expectations –realistic and unrealistic, acceptance of possible undercorrection/ overcrrection/revision 15-20%.

Children: Chin elevation/AHP, Eyelid crease (absent vs moderate), mental make up/ of child/parents.

AM: The key points in examination of a ptosis patient are chin elevation, frontalis action. whether pupillary area is covered/uncovered, any associated amblyopia/ refractive errors, extraocular movements and any abnormal innervation. In children of less than 3 years of age, the primary aim of examination is to assess any risk of amblyopia and severity of ptosis. We look for any chin elevation, level of brow to assess frontalis use, whether pupillary area is covered or uncovered, presence or absence of lid crease, any associated extraocular movement abnormality or Marcus Gunn phenomenon.

Q3: What are your preferred choices as the primary modalities of management in cases of mild, moderate and severe ptosis?

AKG: I prefer Fasanella Servat Surgery for mild cases. Levator resection for the moderate cases and Frontalis Sling for the severe cases, with bilateral Fascia Lata Sling Surgery being the gold standard.

SK: Choice of procedure for the correction of ptosis is guided by the levator muscle action, grade of the ptosis and status of Bell's Phenomanon. For severe bilateral ptosis cases suspension procedure/sling surgery is preferred. Fasial Lata is the preferred suspension material. If this can not be harvested silicone rods or ePTFE (Gore Tax) may be tried. For moderate ptosis cases with fair to good levator function, LPS resection is considered. For mild ptosis with good or excellent levator function, Fasanella-Servat operation is one option, otherwise a small resection of LPS (12mm) may be the other choice.

GS: Adult: Mild – Conj mullerectomy with/without blepharoplasty/crease formation
Moderate: Levator advancement
Severe: levator resection
Congenital: Mild – Observation, Levator advancement/resection
Moderate – Levator resection, Supramaximal levator resection
Severe – Supramaximal levator resection/Frontalis suspension

AM: In severe cases with poor LPS action, we do frontalis sling with silicon rod. In children less than 3years of

age, frontalis sling with 4-0 Ethibond suture.
In moderate cases, LPS resection.
In mild cases with positive phenylephrine test, Fasanella Servat surgery and in patients with negative phenylephrine test, LPS resection/ advancement.

Q4: What would you consider as the appropriate age to operate a child with mild, moderate or severe congenital ptosis?

AKG: The preferred age for surgery is 3 years onwards, except in cases where induction of amblyopia is a consideration. In those cases surgery may be done at the age of a few months, as soon as the general anaesthesia is considered safe. Mild cases often present a little late for surgery. Severe cases where autologous Fascia Lata is the material of choice, we would wait till 4 years of age for adequate development of the fascia.

SK: A child with severe ptosis presenting with chin elevation and pupil covered with the upperlid should be taken up for temporary sling surgery as early as possible. It is for the prevention of amblyopia. Here surgeon must examine the sleeping child to evaluate the Bell's Phenomenon or ask the parents for the rolling up of the eyes during sleep.
Children with mild to moderate ptosis should undergo refraction test and they may be followed up on 6 monthly basis for any change in the seventy of ptosis as well as levetor muscle function.
Moderate ptosis cases may be operated around the age of 5 years when the child is cooperative for the assessment.
Mild ptosis cases may be followed up to the age of adolescence.

GS: Mild – as late as possible, pre primary school 5-6 yrs.
Moderate – 'age of awareness' by the child, generally girls earlier than the boys.
Severe: whenever indicated.

AM: A child with severe ptosis and at risk of developing amblyopia as early as possible and in moderate or mild ptosis, around school going age (4-5years).

Q5: What is the procedure of choice in mild congenital ptosis with good LPS action? Why?

AKG: For cases of mild congenital ptosis with levator action exceeding 10 mm and a good lid crease, my first choice is the modified Fasanella Servat surgery (Betharia, Grover & Kalra, BJO 1982). The reason I choose this procedure is its high predictability, excellent contour and absence of any dry eye symptoms or lid instability in over 300 cases performed.

SK: I practice both Fasanella-servat operation and also small LPS resection. If you want to avoid the double upper eyelid creasing after FS open. LPS resection is the right option.

GS: Adult mild ptosis: Conj mullerectomy with bleph/ crease formation.
Simpler, faster, predictable, less morbidity, good outcomes, patient acceptance, etc.

AM: In mild ptosis with good LPS action, positive phenylephrine test and tarsal height of more than 8mm: Fasanella Servat surgery.
If tarsal height is less than 8mm: Mullerectomy
In case of negative phenylephrine test, LPS advancement.

Q6: Which approach do you prefer for LPS resection? Conjunctival or skin? Why?

AKG: Skin approach has been my preference. The reasons are easy orientation, applicability to all cases including traumatic ptosis, resurgery patients and cases requiring Whitnall's sling. The lid fold creation is excellent and the tarsal show can be matched quite well.

SK: I prefer skin approach for most of the cases of LPS resection as you can match the skin crease with the fellow eye and the amount of resection you wish to do can be gauged.
Conjunctival approach is used for the revision cases only.

GS: Transcutaneous lid crease approach.
Familiarity, training, exposure, ability to perform a bleph/crease formation at the same time.

AM: Skin approach easier, with full view of relevant anatomy and good dynamic crease.

Q7: In which group of patients would you like to do frontalis sling surgery?

AKG: I would do a Frontalis Sling surgery in severe congenital simple ptosis with levator action of 4 mm or less, jaw winking ptosis (after levator excision) and blepharophimosis syndrome. I also choose it in acquired neurogenic and myogenic ptosis.

SK: Frontalis sling surgery is done for bilateral severe ptosis cases with poor LPS action and good Bell's Phenomenon. It is also done for Marcus Gunn ptosis cases where Jaw Winking is severe and the scleral show is prominent.

GS: Pediatric/Congenital: Severe ptosis, with absent <2mm Levator Function, surgery <2yr (Silicone rod), Surgery > 3 yrs Autologus Fascia Lata (AFL),
Adult: Myopathic ptosis.

AM: In patients with severe ptosis and poor LPS and those who are adept to using their frontalis muscle.

Q8: Do you assess the frontalis action prior to surgery ? In cases of severe ptosis with poor LPS where frontalis action is also poor or the patient is not adept to using frontalis muscle, which procedure do you prefer?

AKG: I have not really found Frontalis action to be a useful predictor in decision making and do not use it regularly.

SK: Any severe ptosis case will show forehead Winking

(implying that frontalis is being used) and indistinct upper eyelid crease. Person not using frontalis shall have chin-elevation to keep the pupillary area clear of any obstruction. So in any situation sling surgery is desirable.

GS: May combine levator resection with frontalis suspension or periosteal fixation of Frontalis suspension material.

AM: Yes we assess frontalis action prior to sling surgery. In cases where patient is not adept to using frontalis, the ideal surgery would be Whitnall Sling but we have very limited experience with this procedure.

Q9: In cases of unilateral severe ptosis with poor LPS, do you perform unilateral or bilateral sling?

AKG: My first choice in unilateral severe ptosis is contralateral levator disabling (excision) with bilateral Fascia Lata Sling Surgery to provide the best possible symmetry. In the cases that elect unilateral surgery, silicone sling is the procedure chosen.

SK: Theoretically one may consider unilateral sling surgery, but I prefers combined procedure of LPS resection alongwith tarsectomy if unilateral surgery is consented. Otherwise to avoid any cosmetic blemish due to the condition or unilateral procedure performed (especially the sling surgery), choose bilateral suspension procedure.

GS: Seldom, only if parents agreeable.

AM: Unilateral sling with silicon rod.

Q10: Which material do you prefer for sling? And why?

AKG: Fascia Lata is the material of first choice in all bilateral congenital cases. The reason for the choice is almost universal take, very low rates of recurrence, granuloma formation or infection. The appearance of the lids is very close to the natural. Silicone is the material of choice for unilateral surgery, for cases with restricted ocular motility and relatively poor Bell's phenomenon (greater elasticity, adjustability and reversibility).

SK: Fascial Leta is the standard for the sling surgery. Other option is the silicon rod which must be passed in the submuscular ePTFE may also be tried plane and its knots must be deeply buried.

GS: Temporizing/Myopathic ptosis: Silicone rod – simple, adjustable, repeatable, LA/shorter GA Permanent/non myopathic ptosis: AFL – inert, predictable, long term outcomes

AM: Silicon rod because it is stretchable, elastic, easily adjustable, avoids extra surgery for harvesting fascia lata, less time consuming.

Q11: Which technique of passing the sling do you commonly use? How do you manage the cases where the droop is predominantly lateral or medial droop?

AKG: I prefer to use the double triangle technique with 4 eyelids incisions (Modified Crawford's) for Fascia Lata and a Fox's pentagon technique for the silicone sling. One does not really need to modify the technique of passing the sling for variations in the contour of ptosis.

SK: Crawford's double triangle or Fox's pentagon method for sling surgery may be used. Double triangle techniques helps in the correction of unequal ptosis on either end of the palpebral tissue.

GS: Pentagonal sling.
Intraoperative adjustment with a deeper pass at the brow/rim at the corresponding side.

AM: We use the "Mehta's Modification of Modified Crawford technique" (Mehta A, Abrol S, Garg P. Mehta's modification of Crawford's technique for frontalis sling surgery with silicone rod. Delhi J Ophthalmol 2015;26:115-17) which gives better control of curvature, a very good crease, avoids forehead scar and can take care of predominant medial or lateral droop.

Q12: How do you manage cases of Marcus Gunn Phenomenon? Do you perform unilateral or bilateral slings for unilateral Marcus Gunn Phenomenon? Do you only disinsert the LPS or do resection also?

AKG: I prefer bilateral levator disabling (excision of a large piece of levator aponeurosis, including a thorough cutting of horns) with bilateral fascia lata sling as the first choice for cases of jaw winking.

SK: If the Jaw-winking is minimal, one may attempt the LPS resection. If the Jaw-winking is very prominent, always choose the bilateral sling surgery alongwith excision of LPS muscle on both the sides.

GS: Mild - Moderate: routine levator resection.
Severe: levator extirpation with frontalis suspension- unilateral or bilateral.

AM: We prefer LPS disinsertion with resection with frontalis sling using silicon rod. We have done only unilateral slings in such cases.

Q13: What is your preferred mode of treatment in cases of CPEO, CFOEM, MED with severe ptosis?

AKG: In cases of CPEO, CFOEM and MED associated with severe ptosis, a conservative functional silicone sling is my first choice as it has a greater elasticity and results in a better eye closure. Also silicone sling correction may be adjusted or reversed in the rare case where exposure keratopathy cannot be control by conservative therapy.

SK: If Bell's Phenomenon is not good, choose critech-glasses alongwith topical lubricants. Option of small resection of LPS may be there theoretically but postoperative corneal explosive appreciation must be weighed against correction to be achieved.

GS: When possible levator resection with lower eyelid tightening/lateral canthoplasty.
Frontalis suspension with Silicone rod along with

aggressive blepharoplasty and lid tightening and crease formation.

AM: After taking care of the extraocular movement part, we prefer to do the frontalis sling with silicon rod just uncovering the pupillary area. We have 15 patients in our follow-up with good results.

Q14: At what level do you leave the upper lid at the end of the sling surgery? Is it different for patients who are operated under LA or GA?

AKG: With Fascia Lata I would tighten it to level where I achieve maximum height without lifting it off the globe. With Silicone Sling one has to control the tightening to get a small overcorrection, while maintaining a good contour. No I do not differentiate between cases under local and general anaesthesia except compensating for rotation of the globe under general anaesthesia.

SK: Keep the upperlid margin at the limbus. Most of the time lid comes down a little because of orbicularis being used by the patient and who has habitual national of small palpebral fissure.

As regards G.A. it depends upon the level of anaesthesia, the agent being used and the technique by anaesthesia being going. But the practice is to keep at the limbus.

GS: Overcorrect – 2-3 mm.
Generally no.

AM: We have not been able to develop a normogram.
In simple unilateral cases done under local anaesthesia, we ask the patient to sit and then finalise the height leaving the lid about 1-1.5mm higher than the normal eye.
In cases done under general anaesthesia, the lid is left at a level 2-3mm above the level of the normal eye.

Q15: In how many percent of patient undergoing sling surgery, do you experience post-op over-correction or under-correction and how many of these require adjustment?

AKG: Under or overcorrections are fairly rare with fascia lata sling. With Silicone one does get overcorrections occasionally (? 10%), which do tend to decrease with time. Most undercorrections in silicone sling usually appear later. Some of the unilateral cases , that are undercorrected, really are habitual undercorrections due to inadequate use of Frontalis.

SK: Over and under corrections are part and parcel of the ptosis procedures. But after sling surgery under correction is most of the time, due to slippage of the material and is obvious within 1 or 2 weeks of the surgery. Regarding over creation it is really to be assessed after the lid inflammation has decreased. One may wait for 2-3 weeks and after that only decide for the intervention. And this is very-very occasionally needed.

GS: Overcorrection – Seldom
Undercorrection – 10% approx.
Adjustment – 10% approx.

AM: The incidence of over correction is very low and not more than 2-3%. However, under correction is not uncommon. Approximatelty15% of patients are undercorrected and require readjustment.

Q16: In what percentage of cases have you noticed recurrence of ptosis due to slippage of breakage of sling and what steps do you take during surgery to avoid it?

AKG: The recurrence of ptosis due to slippage occurs in about 10% of cases with silicone sling. It is best avoided by ensuring adequate tissue between the eyelid incisions and the lid crease incision and avoiding excessive tightening. Breakage of sling is much rarer.

SK: Reported incidence is 7-44%.
If there is a slippage or breakage of the sling, it is obviously noticed within 2 weeks of the surgery. Slippage occurs if anchoring of final knot on the forehead to the underlying frontalis muscle and also on tarsus is not proper. Proper burying of the sling knot is must to avoid exposure and infection. Autologous fascia-lata used with proper width of the strip made shall not show this and the fibrovascular tissue adhesion of fascia-lata also helps in the prevention of slippage.

GS: Only rarely, avoiding tight sutures to secure it eg prolene, nylon , avoid skin crease incision whenever possible, avoiding unnecessary dissection along sling tracks

AM: Rarely. The sling material should not be held with toothed or sharp instruments to prevent breakage. The sleeve should be tightened by passing a prolene suture around it to avoid slippage.

Q17: In how many percentage of cases, do you come across granuloma formation following sling surgery and how do you manage them?

AKG: You virtually never get a granuloma formation with fascia lata, provided you bury your knots deep. It is quite uncommon with silicone while with ePTFE or mersilene it was commoner. A persistent granuloma usually requires removal of the sling.

SK: It may be around 7-10% in cases with synthetic material Granuloma formation/abscess formation cases are seen if aseptic precautions not followed. Patients fascial hygiene is also an important factor. It the sling material is not buried properly, the skin infection may be the contributing factor.
If granuloma has formed one may attempt local lavage with Betadine alongwith systemic antibiotics. If frank abscess is there, it should be drained and the sling material removed. Follow up these patient and re-surgery should be considered after 3 months only.

GS: Seldom. Usually either due to an inflammatory suture material eg silk or if the silicone rod is either too superficial or the cut ends are not sufficiently buried in a deep pocket at the midforehead.

AM: During the initial phase of transition from fascia lata to silicon sling (2008-2009), the granuloma formation rate was about 10-15% but now it has come down to less than 5%. The single most important step to prevent granuloma formation is proper burial of the silicon sling and the prolene suture in the tunnel.

In a small study conducted in our dep't (Mehta A, Naik M, Abrol S, Garg P, Joshi M: Granuloma after sling surgery: an attempt to answer the? Why? and? What to do next? International Ophthalmology 2016, DOI: 10.1007/s10792-016-0342-0) in all cases of granulomas, the common finding was exposure of either sling material or prolene suture. These patients do not respond to oral antibiotics or steroids and definitive treatment is explantation of the sling. The repeat sling surgery can be done after 3 months.

EYELID RECONSTRUCTION

Dr. A.K. Grover MD, MNAMS, FRCS (Glasglow), FIMSA, FICO, **Dr. Shaloo Bageja** DNB, MNAMS,
Dr. Amrita Sawhney DNB, MNAMS, FAICO Oculoplasty

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Abstract: *Eyelid reconstruction is required for wide variety of indications that may vary from congenital to acquired defects. Eyelids are complex structures hence their reconstruction poses a great challenge. They play an important role in in drainage of tears, maintaining the tear film and protecting the globe.*

Congenital eyelid colobomas are either isolated or associated with Fraser syndrome, Goldenhar syndrome or cleft disorder (Figure 1).

Causes of acquired eyelid defects are:

a) Surgical resection of tumor (Figure 2): 5–10% of all skin cancers occur in the eyelid[1]
b) Traumatic tissue loss
c) Burns
d) Irradiation
e) Following previous complicated surgery like Entropion correction

Malignant neoplasms represent the leading cause of eyelid reconstruction followed by cicatricial retraction, post-traumatic tissue loss and congenital colobomas[2].

EVALUATION OF EYELID DEFECTS

- Eyelid involved - upper/lower
- Depth - Superficial/deep/full thickness
- Lid margin defects/non lid margin defects
- Size of defect
- Shape of defect - horizontal, vertical, irregular
- Location – medial, lateral or central
- Age of the patient and elasticity of lids
- Condition of the contralateral and opposing eyelids
- Medial or lateral canthus involvement
- Lacrimal apparatus involvement
- Condition of conjunctiva

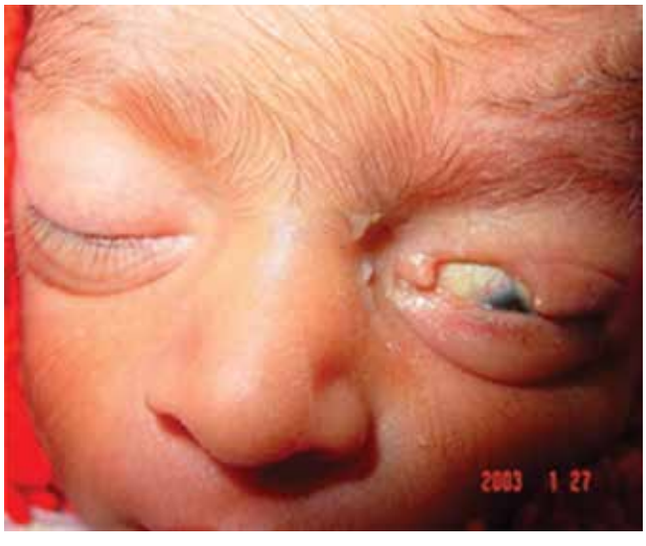


Figure 1: *Showing upper eyelid coloboma*

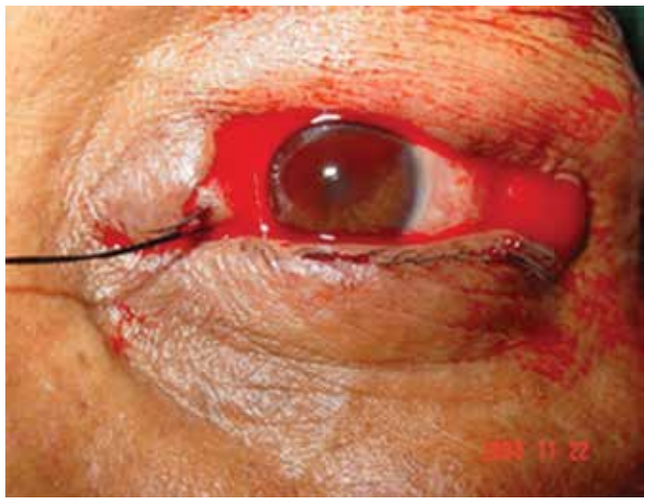


Figure 2: *Showing upper eyelid defect after surgical resection of tumor*

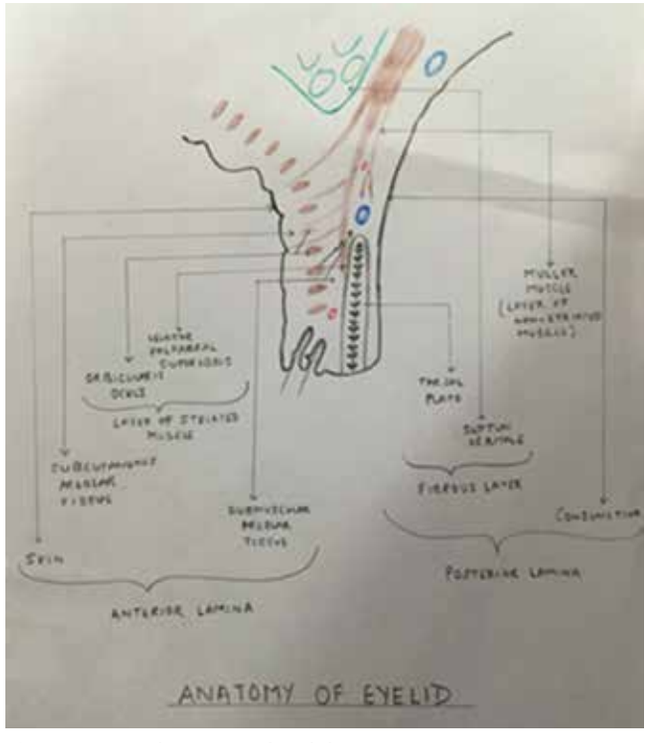


Figure 3: *Normal anatomy of eyelid*

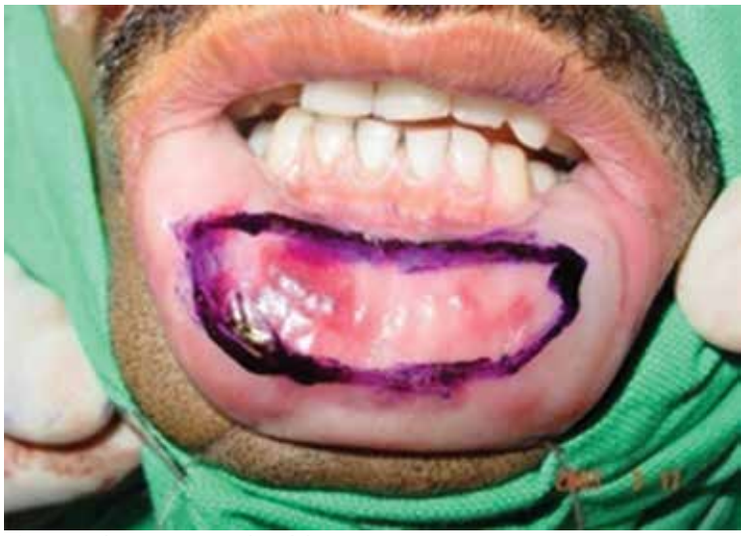


Figure 4: *Showing mucosal graft harvesting*

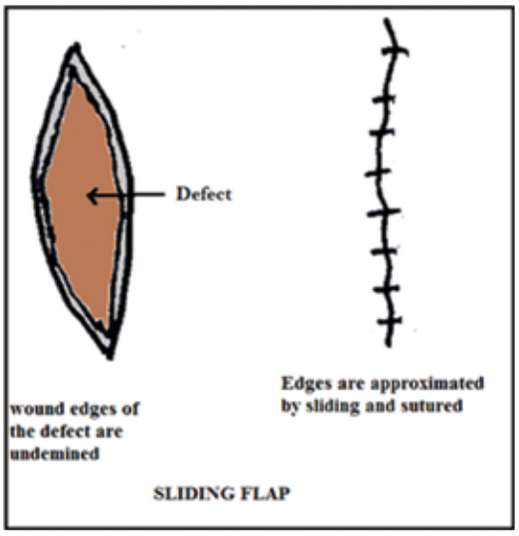


Figure 5: *Showing sliding flap technique*

Showing advancement flap technique

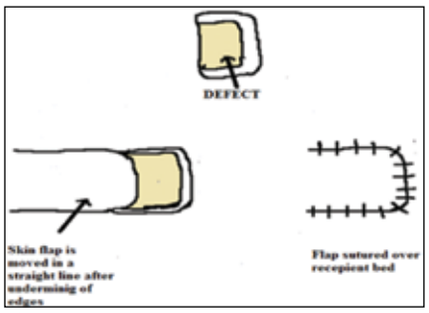


Figure 6(a)

Figure 6(b)

Figure 6(c)

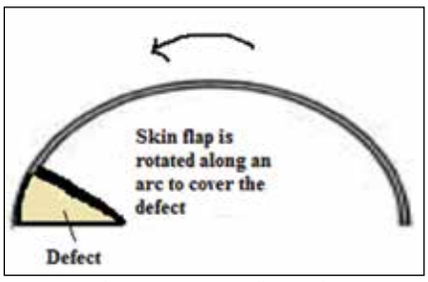


Figure 7: *Showing rotation flap technique*

GOALS OF EYELID RECONSTRUCTION

- To restore physiologic functioning of eyelids
  - Vision, lid closure, mobility, tear drainage
- To restore anatomic integrity
- To provide best possible cosmesis with minimal scars.

PRINCIPLES OF EYELID REPAIR

Lid reconstruction should provide skin – muscle lamina anteriorly with cartilagenous framework and smooth mucous membrane lining posteriorly.

Note: Figure 3 shows normal anatomy of eyelid which is divided into anterior lamina- skin, orbicularis muscle and posterior lamina – tarsus, conjunctiva.

- Aim should be to provide stable mucocutaneous lid margin with intact cilia and good apposition to the globe.
- Medial and lateral canthi should be reconstructed wherever required.
- One lamina should have adequate blood supply to support the other lamina. Hence, two flaps or a flap and a graft can be sutured together but never two grafts.
- Avoidance of undue wound tension
- Atraumatic tissue handling
- Elimination of dead space
- Ensure complete hemostasis

S.No.	Name of the graft	Thickness	Instrument used to harvest it
1.	Wolfe-Krause graft	0.80–1.00 mm	Scalpel
2.	Padgett	0.60–0.80 mm	Dermatome
3.	Blair-Brown	0.40–0.60 mm	Dermatome
4.	Ollier-Thiersh graft	0.20–0.35 mm	Dermatome

Showing transposition flap technique

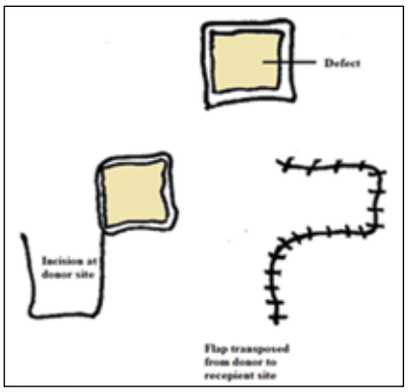


Figure 8(a)

Figure 8(b)

- Avoid overlapping of wound edges and ensure everted wound margins.
- Appropriate selection of sutures &

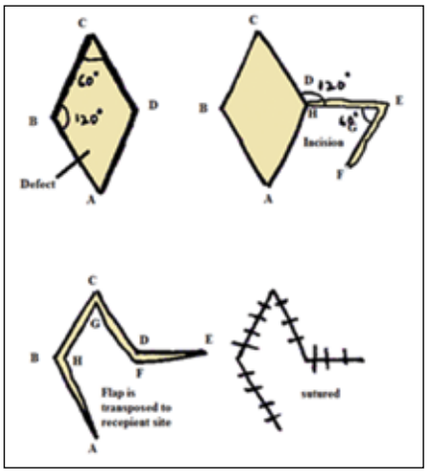


Figure 9: *Showing Rhomboid flap technique*

e) Nasolabial fold
f) Contralateral upper lid skin

2) Free grafts used to repair posterior lamella of eyelid replace either tarsus, conjunctiva or both.
   a) Free Tarsoconjunctival graft
   b) Sclera graft
   c) Oral mucosa (cheek or lip) (Figure 4)
   d) Fascia lata
   e) Auricular cartilage
   f) Palatal mucoperichondrium
   g) Nasal mucosa with septal cartilage
   h) Free conjunctival autograft from contralateral eye

minimal contraction.

Skin flaps can be classified into the following types:

a) Sliding flap
b) Advancement flap
c) Rotation flap
d) Transposition flap
e) Pedicle flap

a) Sliding flap (Figure 5)
b) Advancement flap - It is moved from donor to recipient site in a straight line without any lateral or rotational movement (Figure 6a,b,c)[6].
c) Rotation flap - Movement of rotation flap is in the direction of an arc around a fixed point (Figure 7).

Figure 10(a)

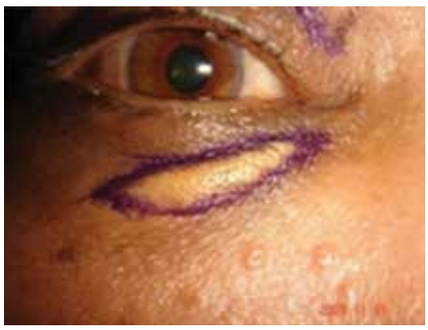


Figure 10(b)

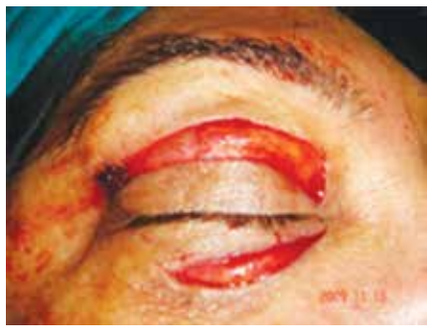


Figure 10(c)

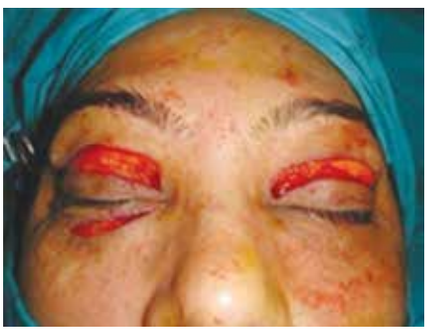


Figure 11(a)

Figure 11(b)

needles

- The tissue deficiency in the forehead and cheek must be assessed preoperatively to avoid secondary contracture.

FREE GRAFTS IN LID RECONSTRUCTION

Free skin grafts are devoid of blood vessels and are used in cases of extensive eyelid defects which cannot be repaired by direct suturing or with use of flaps alone[3].

1) Full thickness skin grafts used for anterior lamella can be obtained from following donor sites:
   a) Postauricular
   b) Supraclavicular
   c) Medial side of upper arm
   d) Groin

Skin grafts can be classified according to their thickness as follows[4,5]:

SKIN FLAPS

Skin flap consists of skin, subcutaneous tissue and subdermal plexus of vessels. It is completely raised from the underlying tissue but still it is connected by one side to the surrounding skin and fat. It is because of the vessels in this pedicle that the flap can preserve its own blood supply.

Term flap is derived from dutch word "flappe" meaning ' anything that is hung and loose, fastened only by one side'.

Skin flaps provide better cosmetic results as compared to skin grafts as they maintain their original colour and texture, provide their own blood supply, maintain surface contour and undergo

d) Transposition flap – This flap is rotated around a pivot point (Figure 8a,b).

Rhomboid flap – It is a type of transposition flap which is specially designed for rhombic shaped defects. Defect must form 60 and 120 degree angles (Figure 9)[7].

NON-MARGINAL LID DEFECTS

Principles of repairing non-marginal lid defects are:

A) Convert defect into elliptical shape (Figure 10 a,b,c).
B) There should be no tension or vertical pulling effect of suturing on the lid margins. Vertical tension can be avoided by adequate undermining of wound edges.
C) Upper lid - all incisions should be parallel to eyelid margin (Figure 11 a,b).
D) Lower lid - convert defects in a direction perpendicular to eyelid margin (prevent ectropion or sclera show) (Figure 12 a,b).
E) Full thickness defects should be closed in 2-3 layers.
F) If defect is large and direct closure is not possible then skin flaps or skin grafts are used to repair the defect (Figure 13,14,15).

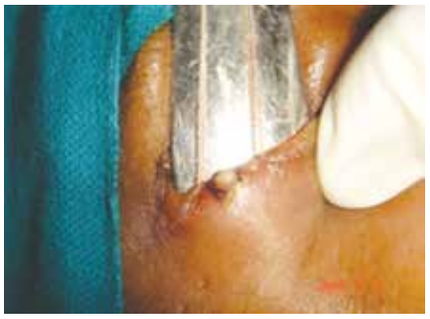


Figure 12(a)

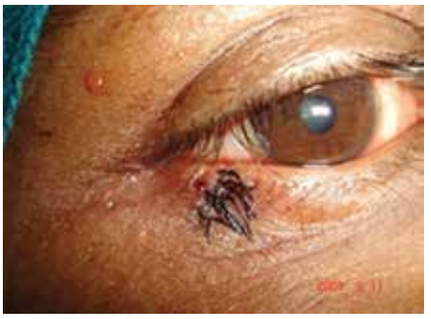


Figure 12(b)

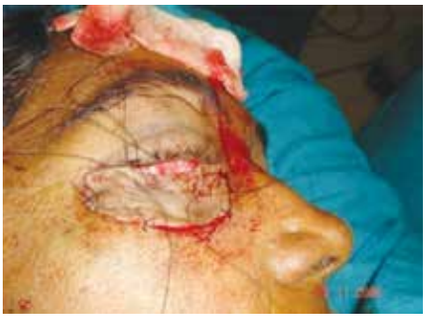


Figure 13

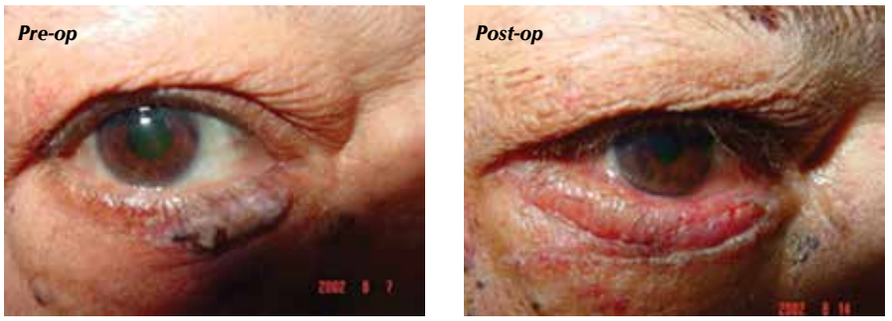


Figure 14: *Non marginal eyelid defect - Repair with advancement flap*

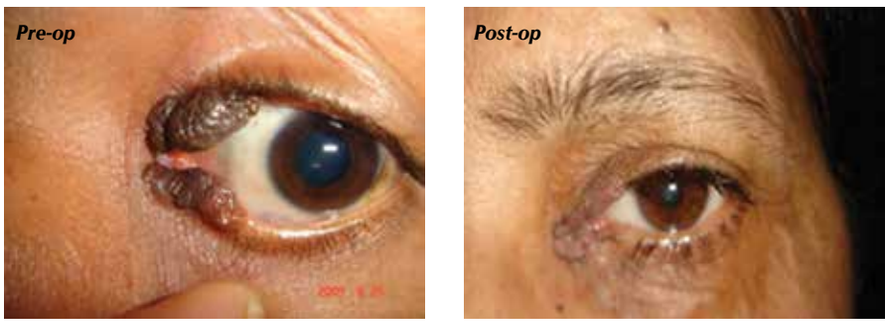


Figure 15: *Medial canthus Nevus excision with skin grafting*

FULL THICKNESS LID MARGIN DEFECTS

Upto one fourth of lid defect

Direct layered closure is done in lid defects measuring upto 25% in younger individuals. This technique can also be employed to repair 30-40% lid defects in older individuals or in patients with lid laxity.

Direct layered closure (Figure 16 a-g)

- Eyelid defect is converted into a pentagon with vertical sides covering height of tarsal plate and the two arms converging at the fornix. This is done so that tarsal plate is sutured together and tension is evenly distributed across the wound (Figure 16a,b).
- Vertical mattress sutures are passed at the grey line, posterior lid margin and anterior lid margin.
  The sutures are passed 3mm from the cut edge and 3mm depth , coming back 1mm from the cut edge and 1 mm depth to complete the vertical mattress sutures (Figure 16 c,d).
- The tarsal plate and skin are sutured in separate layers (Figure 16e).

Skin sutures are removed after 5 days and lid margin sutures are removed after 10 days.

Advantages

a) It is the simplest and single stage procedure to repair lid defect.
b) It provides lid margin with intact cilia.
c) Repaired lid is stable as there is direct suturing of tarsal plate.

Upto one half of lid defect (50%)

A) Lid lengthening procedures like lateral cantholysis and lateral canthotomy along with direct suturing.

Lateral cantholysis: method

- Mark a skin incision overlying the area between two limbs of lateral canthal tendon.
- Make skin incision and separate skin and conjunctiva from lateral canthal tendon limbs.
- The lid being repaired is made taut and corresponding lateral canthal tendon limb is cut leaving the other limb intact .
- Close the skin incision after direct lid repair.

B) Tenzel's lateral semicircular rotation flap (Figure 17a-i)

It is a periorbital rotation flap which can be used to repair both upper and lower lids.

- Semicircular skin muscle incision, with concavity towards the eyelid defect, is given starting from lateral canthus extending horizontally 18-20mm and vertically 22mm forming a high arch (Figure 17b).

Note: Incision should not extend beyond lateral part of eyebrows as the branches of seventh nerve lie lateral to this area.

The eyelid defect is converted into a pentagon (Figure 17c).

- Raise the flap and perform lateral canthotomy along with lateral cantholysis of the corresponding limb of lateral canthal tendon (Figure 17d).
- Pull the lid medially by mobilization of conjunctiva to approximate the edges of the defect and carry out direct repair of the pentagon shaped defect (Figure 17e).
- Lateral canthus is formed and the repaired eyelid is suspended to the intact limb of lateral canthal tendon especially in cases of repaired lower eyelid in order to prevent ectropion (Figure 17f).
- Orbicularis muscle and skin on donor site must be closed in separate layers (Figure 17g).
- Temporal portion of the reconstructed lid is formed by the flap which lacks conjunctival covering. Hence, conjunctiva obtained from pentagon excision of the lid defect is sutured to the bare

Showing direct layered closure

Figure 16(a)

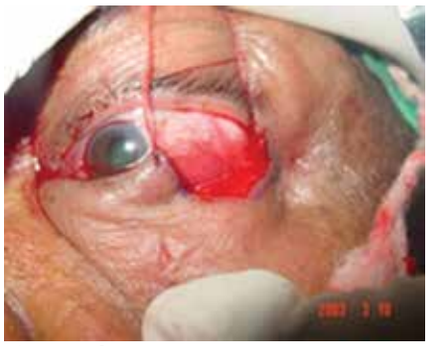


Figure 16(b)

Figure 16(c)

Figure 16(d)

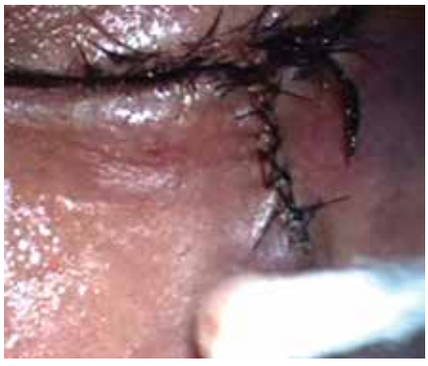


Figure 16(e)

Figure 16(f)

Figure 16(g)

flap area.

Advantages

1) It is a one stage procedure and especially useful for infants with congenital lid coloboma as it does not cause stimulus deprivation amblyopia.
2) Good cosmetic results are obtained.
3) Surgery is confined to periorbital area only.
4) No graft required to strengthen the repaired part of eyelid.

Disadvantages

1) Lateral part of eyelid formed by semicircular flap lacks cilia.
2) Lateral portion of eyelid lacks the rigid support hence notching of lid can be seen in this part.

C) Cutler Beard technique

It is also called bridged flap technique or bucket handle technique.

It is a two stage procedure and an important lid sharing technique in which large upper lid full thickness defect is repaired by utilizing full thickness advancement flap from lower lid.

METHOD: STAGE 1 (Figure 18 a-k) Showing Cutler Beard technique stage 1

- Upper eyelid defect is measured and marked using marking pen (Figure 18b).
- The upper lid defect is converted into rectangular shape (Figure 18c).
- Lower lid traction sutures are passed and incision line is marked 4-5 mm below lid margin with horizontal extent corresponding to the upper lid defect and vertical incisions marked towards the fornix at each end of horizontal incision (Figure 18d).

Note: Lower lid incision is made 4-5 mm below lid margin to preserve marginal artery and thus prevent necrosis of lower lid bridge.

- Full thickness lower lid incision is given along the marked lines with lid spatula underneath to protect the globe (Figure 18e).
- Skin, muscle and tarsoconjunctival layers of flap are separated (Figure 18f).
- Flap is pulled towards the upper lid defect under the bridge and sutured to the defect in 3 layers – tarsoconjunctiva, muscle and skin separately (Figure 18 g,h).
- Pad and bandage is done for 24 hours.
- Suture removal is done after 7 days.

METHOD – STAGE 2 (Figure 19 a- f) Showing Cutler Beard technique stage 2

Second stage surgery is carried out 6-8weeks after first stage surgery mainly to divide the flap.

- Bridge is retracted and incision is marked according to the desired level of lid height (Figure 19a).
- The bridge flap is divided layer by layer thereby leaving more conjunctiva than skin attached to the upper lid (Figure 19b).
- Excess conjunctiva is sutured to the skin anteriorly in order to form round and smooth lid margin and also to prevent entropion.
- The borders of the host flap area in the lower eyelid are freshened and sutured in separate layers (Figure 19 c,d).

Disadvantages of Cutler Beard technique are:

1) Upper lid entropion due to instability of the lid.

Showing Tenzel's lateral semicircular rotation flap

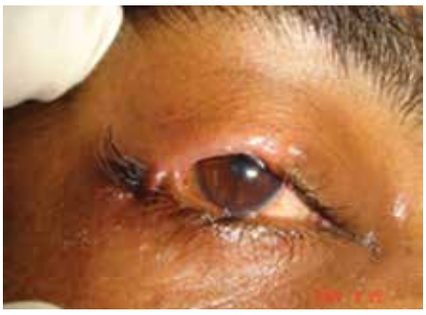


Figure 17(a)

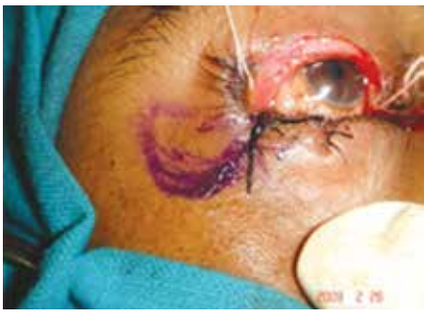


Figure 17(b)

Figure 17(c)

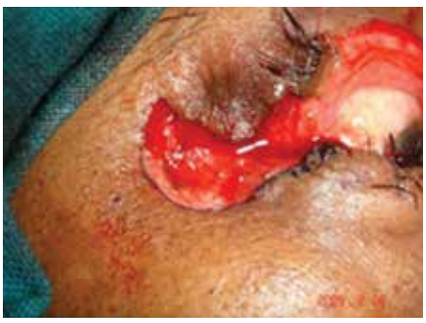


Figure 17(d)

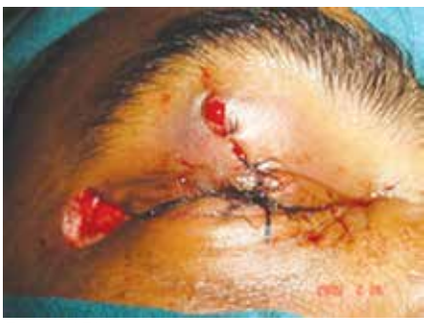


Figure 17(e)

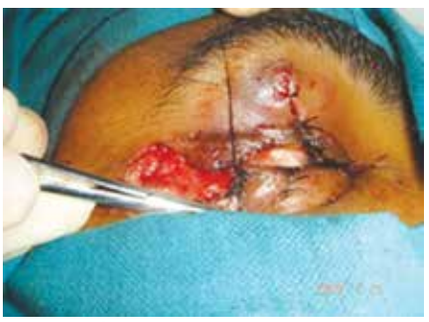


Figure 17(f)

Figure 17(g)

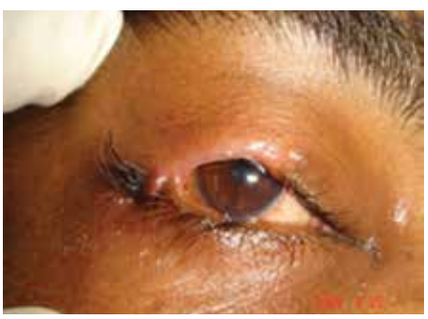


Figure 17(h)

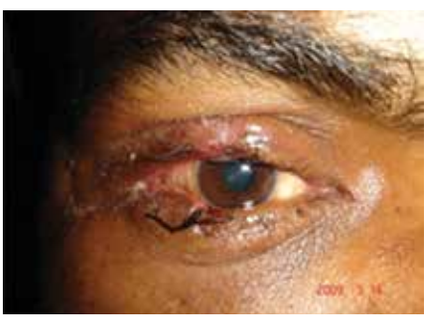


Figure 17(i)

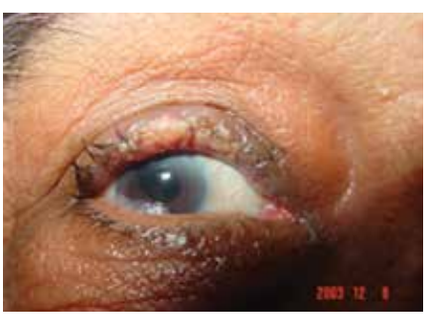


Figure 18(a)

Figure 18(b)

Figure 18(c)

Figure 18(d)

Figure 18(e)

Figure 18(f)

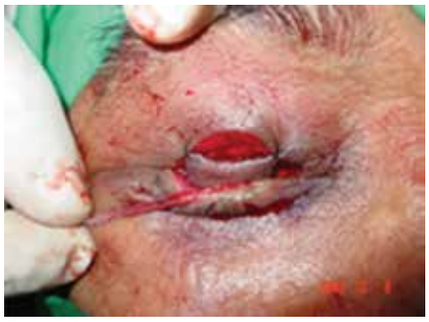


Figure 18(g)

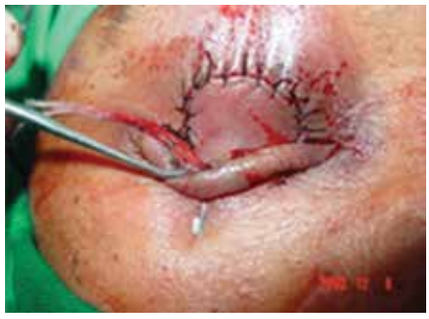


Figure 18(h)

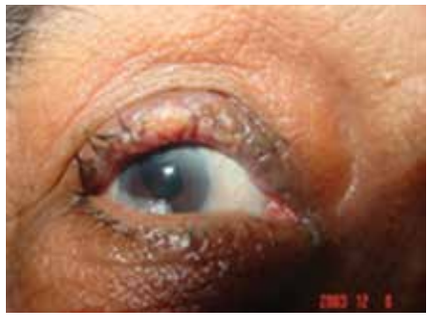


Figure 18(i) - *Preop*

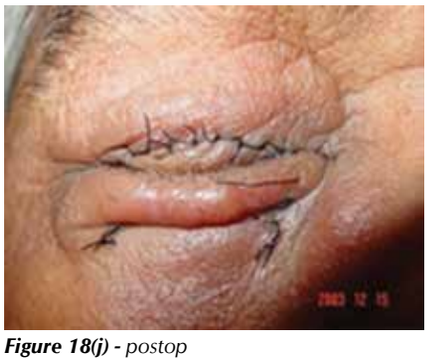


Figure 18(j) - *postop*

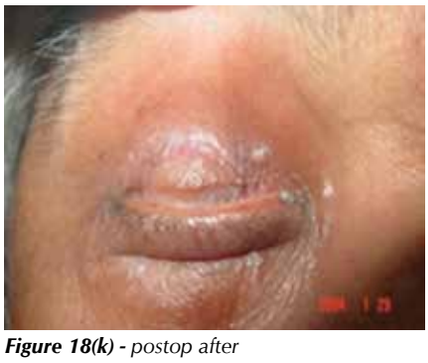


Figure 18(k) - *postop after*

Figure 19(a)

Figure 19(b)

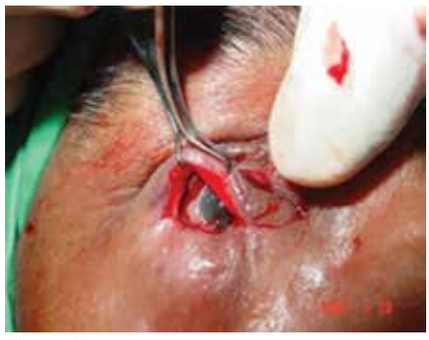


Figure 19(c)

Figure 19(d)

Figure 19(e)

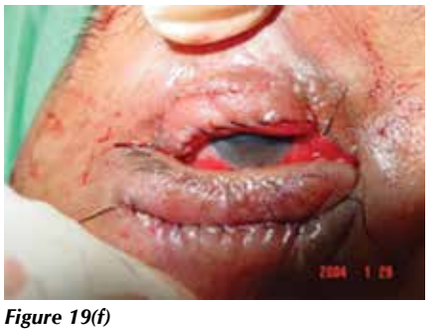


Figure 19(f)

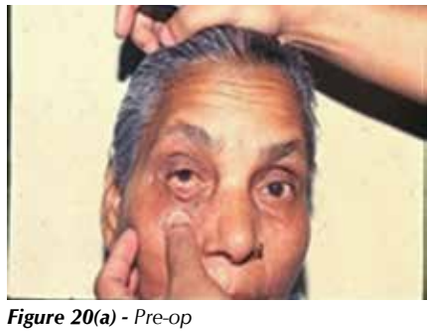


Figure 20(a) - *Pre-op*

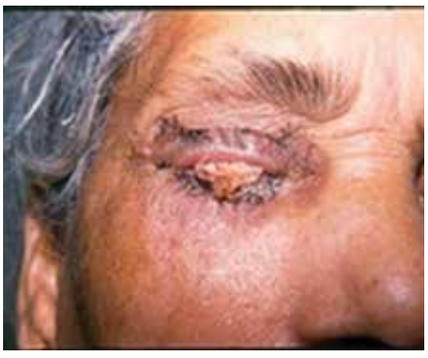


Figure 20(b): *Postop (1st stage)*

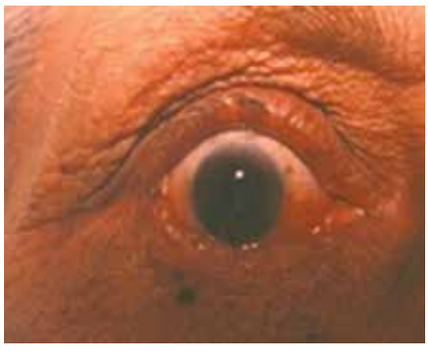


Figure 20(c): *Final postop*

Figure 21(a): *Pre-op*

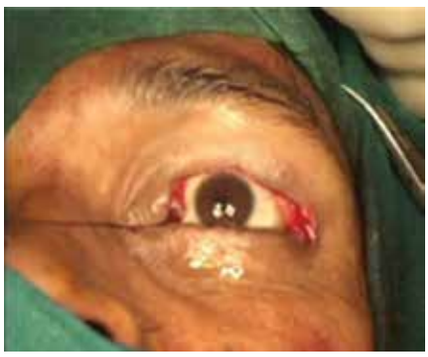


Figure 21(b)

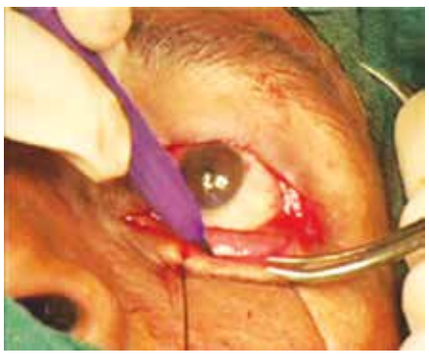


Figure 21(c)

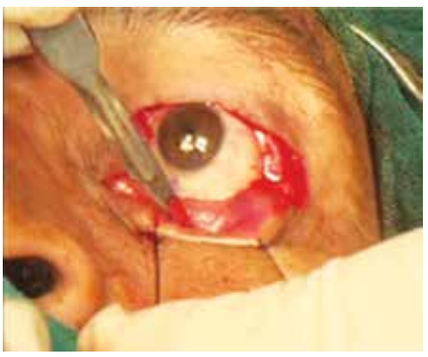
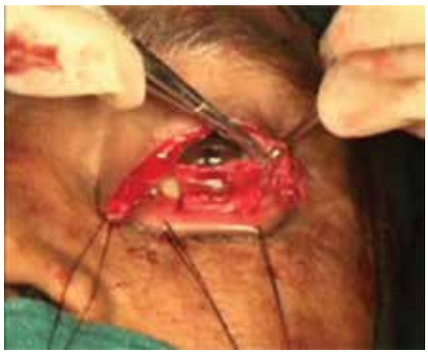


Figure 21(d)



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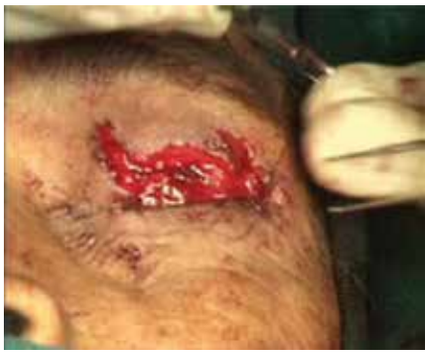


Figure 21(f)

Figure 21(g)

Figure 21(h)

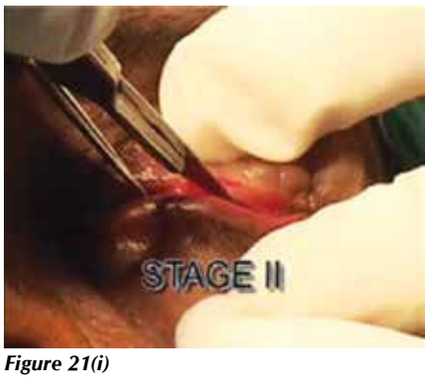


Figure 21(i)

Figure 21(j)

2) Lack of cilia in the reconstructed area of lid.
3) Lid edema and notch formation.
4) Upper lid shortening leading to lagophthalmos.
5) Ischaemic necrosis of bridge flap.

D) Reverse Cutler Beard technique (Figure 20 a,b,c)

When upper lid flap is utilized to repair lower lid defect then it is called Reverse Cutler Beard technique.

E) Hughes Tarsoconjunctival flap technique with skin mobilization or skin graft (Figure 21 a-k).

Stage 1

- Horizontal extent of lid defect is measured.(Figure 21b)
- Evert the apposing intact lid and make horizontal incision, corresponding to the lid defect, through tarsal plate away from lid margin. Vertical incision should extend into conjunctiva (Figure 21c,d).
- Blunt dissection is carried out to free the tarsoconjunvtival flap and advance the flap and suture it into the defect (Figure 21e).
- Anterior surface of this flap is covered with mobilized skin (Figure 21f,g,h).

Stage 2

- After 3 weeks conjunctival pedicle is divided and the new lid margin is formed (Figure 21 I,j).
- The defect in the intact lid is left open to heal by granulation.

F) Marginal pedicle rotational flap (Mustarde) (Figure 22 a- d)

Upper lid defect can be repaired by rotating lower lid flap into the upper lid defect.

- Upper lid defect is measured and accordingly lower lid incision is marked (Figure 22 c).
- Full thickness lower lid incision is given leaving a small pedicle laterally.
- Lower lid flap is rotated and sutured to the upper lid defect (Figure 22 d).

More Than Half To Near Total Defect

a) Cutler beard procedure/reverse cutler beard
b) Tarso conjunctival flaps (Hughes') with skin grafts
c) Free tarso conjunctival graft + myocutaneous advancement flap
d) Cheek rotation flap (Mustarde's) with posterior lamella graft (Figure 23a,b).

Lower lid defect can be repaired using cheek rotation flap with posterior lamella graft preferably nasal mucocartilage graft.

- Lower lid defect is measured.
- Skin incision is marked which extends as a curved line from lateral canthus upwards temporal to lateral eyebrow hairs continuing over the temple and finally curve downwards in front of the ear. Lower limb of lateral canthal tendon is cut and mobilize the flap by dissecting

Figure 21(k): *Postop after stage 2*

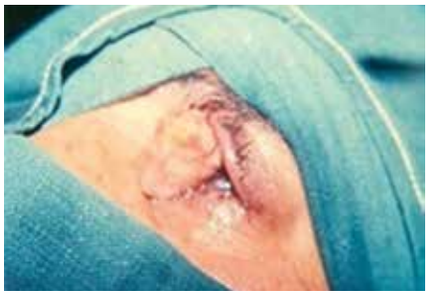


Figure 22(a): *Pre-op*

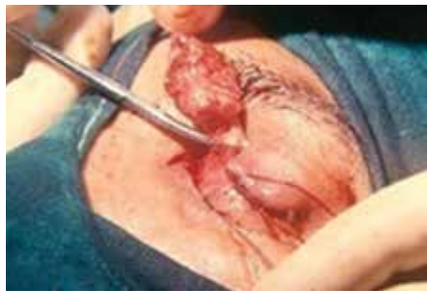


Figure 22(b)

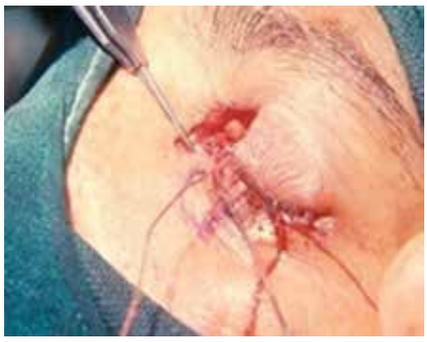


Figure 22(c)

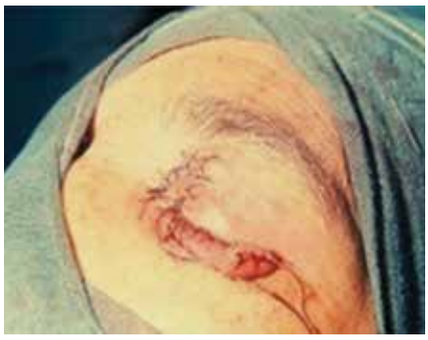


Figure 22(d)

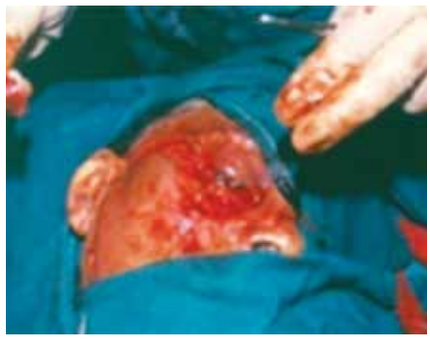


Figure 23(a)

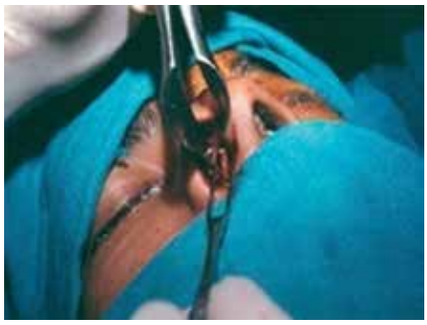


Figure 23(b)

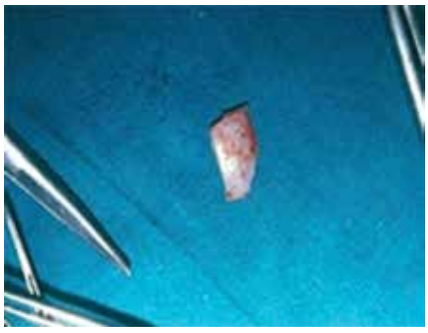


Figure 23(c)

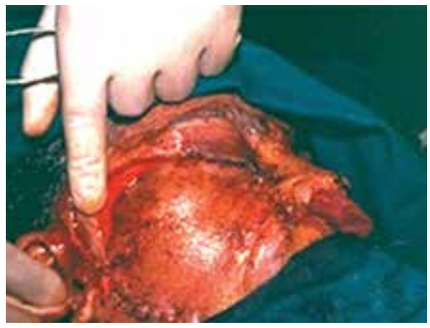


Figure 23(d)

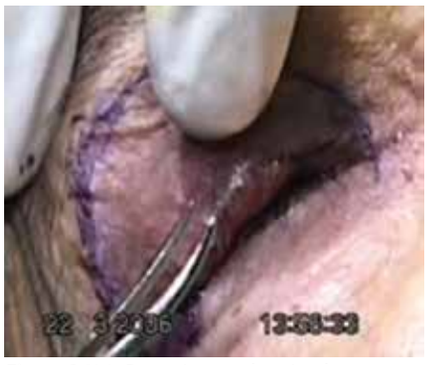


Figure 24(a): *Preop image showing large upper eyelid tumor*

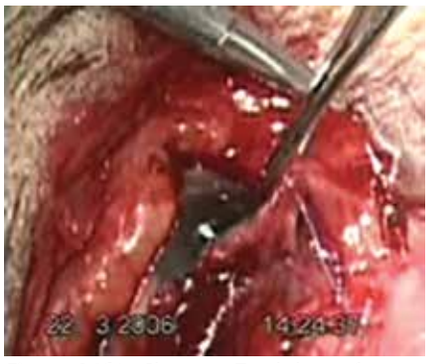


Figure 24(b): *Shows - Total loss of upper lid. Posterior lamella is being formed by tarsoconjunctival flap from lower lid.*

Figure 24(c): *The skin is formed by temporal forehead transposition flap.*

Figure 24(d): *The transposition flap is sutured to the posterior lamella. Donor site is closed using (5-0) non-absorbable sutures.*

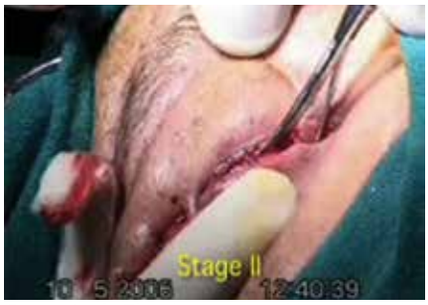
Figure 24(e): *Postop image after stage 1.*

inferior orbital septum (Figure 23 a).

- Nasal septal cartilage and mucosal graft is harvested (Figure 23 b, c).
- Nasal mucocartilage graft is sutured to remnants of conjunctiva to form posterior lamella of the lid being repaired.

Flap is sutured to lateral orbital rim and subcutaneous tissue.

Skin edges are sutured and free mucosa of graft is brought anteriorly over the edge of flap and sutured to flap anteriorly (Figure 23 d).



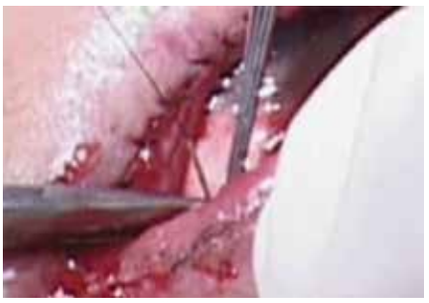


Figure 24(f): *The skin is formed by temporal forehead transposition flap.*

Figure 24(g): *Pedicle flap is divided and skin closure is completed.*

Figure 24(h): *Postop image after stage 2.*

Total Loss of Upper Lid

a) Mustarde's pedicle rotation flap + cheek rotation flap along with posterior lamella graft
b) Temporal forehead/glabellar flaps combined with posterior lamellar grafts (tarso conjunctival or muco cartilagenous)

GLABELLAR FLAP

AV to Y flap is rotated from glabellar region to repair medial part of upper lid and medial canthus area.

Superficial Temporal artery based temporal forehead pedicled flap combined with posterior lamellar graft.

CONCLUSION

Eyelid defects should be properly assessed before chosing any particular technique for lid reconstruction. There are a variety of techniques each having its advantages and disadvantages hence no fixed rules can be laid down regarding any case. The procedure of choice should be one which gives best cosmetic and functional results with minimum chances of complications.

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EYELID TUMORS: A REVIEW

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Abstract: *Tumors of the eyelid are frequently benign in nature. The commonest benign tumors are cystic in nature while the commonest malignant tumors are epithelial in origin. An eyelid tumor generally presents at an early stage as a visible eyelid mass, and wide excision with primary lid reconstruction is curative. Those presenting with extensive lesions require careful planning for excisional surgery followed by lid reconstruction with grafts or flaps. Additionally, radiotherapy maybe necessary in bony involvement. Extensive orbital extension necessitates exenteration.*

The eyelid is composed of two lamellae-the anterior lamella comprising the skin and the orbicularis, and the posterior lamella comprising the tarsus and the palpebral conjunctiva. The eyelid margin is a composite structure on its own, containing hair follicles and the glands associated with it, openings of meibomian glands, and the transition from the tarsal conjunctiva to the skin of the lid. All these structures present within the lid can give rise to neoplastic lesions of different varieties. Commonest are the ones arising from the epithelial layer of the lids. Herein, we discuss the common benign and malignant lesions of the eyelid (Table 1).

CYSTIC TUMORS OF THE EYELID

Various cystic lesions arising in the skin of the eyelids simulate neoplasms. These include eccrine hidrocystoma, apocrine hidrocystoma, sebaceous cyst and epidermoid cyst. Eccrine hidrocystoma is a retention cyst of the eccrine sweat glands. It appears as a clear cyst located near the eyelid margin (Figure 1A). Apocrine hidrocystoma is a retention cyst of the apocrine glands most commonly occurring near the eyelid margin. It arises from the glands of Moll and has a bluish color. In contrast to eccrine hidrocystoma which can be multiple, apocrine hidrocystoma is usually solitary. Sebaceous cyst is a retention cyst of the sebaceous glands, either meibomian glands or Zeis glands. It appears as a yellow, opaque lesion near the eyelid margin or in the periocular skin (Figure 1B). Epidermoid cyst is a retention cyst caused by obstruction of the orifices of the pilosebaceous units, which clinically resembles a sebaceous cyst. However, the cyst largely contains desquamated keratin. Management of these cysts can be just observation or surgical excision. Carbon dioxide laser-assisted or radiofrequency-assisted vaporization may also be performed.

EYELID SQUAMOUS PAPILLOMA

Squamous papilloma is histopathologically characterized by benign hyperplasia of squamous epithelium. It is one of the most common eyelid lesions and unlike the conjunctival papilloma, no strong association with human papilloma virus has been found. Seen in elderly individuals, eyelid papillomas can be sessile or pedunculated, solitary or multiple with smooth or convoluted surface (Figure 2A). Management of these lesions can be just observation or shave excision. Carbon dioxide laser-assisted or radiofrequency-assisted vaporization may also be performed.

EYELID SEBORRHEIC KERATOSIS

Seborrheic keratosis generally occurs in the periocular region of older individuals. The lesion appears as a minimally elevated tan to brown plaque which is frequently solitary (Figure 2B). However, the sudden appearance of multiple lesions may indicate the presence of an internal malignancy, more specifically gastrointestinal adenocarcinomas. This is called the "Sign of Leser-Trelat". Treatment is observation or

Figure 1: *Benign tumors of the epidermis (A) A transparent, cystic eccrine hidrocystoma located near the eyelid margin (B) A typical sebaceous cyst*

Figure 2: *Benign tumors of the epidermis (A) A single pedunculated papilloma with a convoluted surface (B) A tan colored plaque representing an actinic keratosis*

Figure 3: *Pre-malignant periocular skin lesions (A) Xeroderma pigmentosum (B) A geographic pattern of pigmentation representing a sebaceous nevus*

Table 1: Benign and Malignant Tumors of the Eyelid			
Tumors of the eyelid	**Benign**	**Pre-Malignant**	**Malignant**
1. Cystic tumors	Eyelid eccrine hidrocystoma Eyelid apocrine hidrocystoma Eyelid sebaceous cyst Eyelid epidermal inclusion cyst		
2. Tumors of the epidermis	Eyelid squamous papilloma Eyelid seborrheic keratitis Eyelid inverted follicular keratitis Eyelid keratoacanthoma	Eyelid actinic keratosis Xeroderma Pigmentosum Sebaceous Nevus	Eyelid squamous cell carcinoma Eyelid basal cell carcinoma
3. Sebaceous gland tumors	Eyelid sebaceous adenoma		Eyelid sebaceous carcinoma
4. Sweat gland tumors	Eyelid syringoma Eyelid eccrine hidradenoma		Eyelid sweat gland adenocarcinoma
5. Hair follicle tumors	Eyelid trichoepithelioma Eyelid trichofolliculoma Eyelid trichoadenoma Eyelid tricholemmoma		Eyelid trichilemmal carcinoma
6. Melanocytic tumors	Eyelid melanocytic nevus Oculodermal melanocytosis	Eyelid lentigo maligna	Eyelid malignant melanoma
7. Neural tumors	Eyelid neurofibroma Eyelid schwannoma		Eyelid Merkel cell carcinoma
8. Vascular tumors	Eyelid congenital capillary hemangioma Eyelid acquired capillary hemangioma Eyelid varix Eyelid lymphangioma		Eyelid Kaposi's sarcoma Eyelid angiosarcoma Eyelid glomus tumor
9. Histiocytic and fibrous tumors	Eyelid xanthelasma and xanthoma Eyelid xanthogranuloma Eyelid angiofibroma		
10. Lymphoid tumors	Eyelid eccrine hidradenoma		Eyelid lymphoma
11. Tumors of the lacrimal drainage system	Eyelid trichoepithelioma		Lacrimal gland carcinoma Lacrimal sac melanoma

shave excision. Carbon dioxide laser-assisted or radiofrequency-assisted vaporization may also be performed.

XERODERMA PIGMENTOSUM

Xeroderma pigmentosum is an autosomal-recessive disorder in which there is a defect in DNA-repairing enzymes. Such patients are extremely sensitive to ultraviolet radiation from the sun that can predispose them to a variety of cancers, including tumors of the eyelid and conjunctiva. The skin of the affected individuals shows variegated pigmentation, scaling and telangiectasia (Figure 3A). Multiple tumors develop by the end of 1st decade including squamous cell carcinoma, basal cell carcinoma, malignant melanoma, and sarcomas. Management mainly includes protection from sunlight with use of topical sunscreen applications, protective clothing, ultraviolet blocking spectacles, and surgical excision of small premalignant and malignant skin lesions.

SEBACEOUS NEVUS

Sebaceous nevus can be an isolated lesion in the eyelid area, or a part of a systemic syndrome, organoid nevus syndrome. Organoid nevus syndrome primarily has neurologic manifestations with arachnoid cysts and cerebral atrophy leading to seizures and mental retardation. The ocular findings include large pigmented patch (tan to brown) in the scalp, eyelids, face and retroauricular area and conjunctival choristoma (Figure 3B). This cutaneous lesion can frequently give rise to BCC, which is reported in approximately 20% of the patients. Regarding management, small lesions can be excised. Lesions which are too extensive for excision can be simply observed. Any tumor arising within the lesion should be completely excised.

EYELID SQUAMOUS CELL CARCINOMA

Squamous cell carcinoma (SCC) accounts for 5% to 10% of periocular cutaneous tumors, and is the second most common cancer of the eyelids. These tumors can arise either de novo or from precursor lesions, including actinic keratosis, Bowen's disease, xeroderma pigmentosa or albinism. Patients with xeroderma pigmentosa tend to be younger, have multiple and recurrent cutaneous lesions in the eyelids and other parts of the body. The lower eyelid is the most common periocular site to be involved. SCC in the eyelid tends to appear sessile or elevated, erythematous with indurated borders and with a scaly surface (Figures 4A-B).

Figure 4: *Squamous cell carcinoma (SCC) of the eyelid (A) presenting as a sessile necrotic lesion (B) and as a nodular lesion with conjunctival extension*

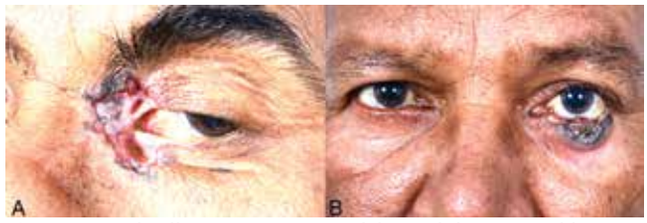

Figure 5: *Basal cell carcinoma (BCC) of the eyelid (A) Typical location of an eyelid BCC causing ectropion of the lower lid (B) An early nodular pigmented BCC of the upper lid*

The lesions frequently ulcerate, become friable, and tend to bleed on touch. Sometimes the central part of a necrotic lesion may develop a secondary infection. SCC of eyelid shows neurotropism and regional lymph node metastasis. Histopathologically, the tumor consists of nests of squamous epithelial cells arising from the epidermis and extending into the dermis. These cells with eosinophilic cytoplasm can also contain keratin pearls.

SCC of the eyelid is known to be aggressive with prognosis correlating with local recurrence and metastatic rate. Treatment options include excision biopsy in conjunction with histological monitoring of tumor margins, radiation, cryotherapy, intralesional chemotherapy and intralesional interferon. Radiation may be especially useful in cases with perineural invasion as an adjunctive therapy. Exenteration is reserved for cases where there is an evidence of extensive orbital invasion.

EYELID BASAL CELL CARCINOMA

Basal cell carcinoma (BCC) is the most common malignant tumor of the periocular skin. It most commonly arises from the lower lid (65%) followed by medial canthus (15%) and upper eyelid (15%). Exposure to UV radiation is the most important risk factor. BCC is associated with genetic syndromes including Gorlin-Goltz syndrome and xeroderma pigmentosa. It characteristically presents as a well-circumscribed, pearly, waxy or translucent tumor with telangiectasia visible near the border of the lesion. Most common clinical types of BCC are nodular and nodulo-ulcerative (Figures 5A-B). Other varieties include pigmented, cystic, superficial and morpheaform BCC. Histopathologically, the tumor cells consist of nests of well-differentiated basal cells arranged in a palisading pattern. Although BCC of the eyelid is known to have a gradual clinical course with a low incidence of metastasis and mortality, advanced BCC can invade the orbit, nasal cavity and sinuses.

Complete surgical excision under frozen section control offers a long-term cure in early lesions. Cryotherapy has also been tried in smaller lesions with comparable outcome. Immunomodulator agents, as a cure for small periocular BCC, have gained popularity in the recent past. Topical application of 5% Imiquimod offers an excellent alternative treatment for small to medium sized periocular BCC. Although some authors have advocated the use of radiotherapy in small lesions, it is now generally offered in patients as a palliative treatment in recurrent lesions or in advanced BCC with orbital, intranasal or intracranial extension. Vismodegib is a new FDA approved treatment option for metastatic and locally advanced BCCs that are not amenable to surgery or radiation. Vismodegib, given orally at a dose of 150 mg once a day, inhibits the Hedgehog signalling pathway, which is abnormally up regulated in more than 90% of BCC.

EYELID SEBACEOUS CARCINOMA

Sebaceous gland carcinoma of the ocular adnexa can arise from the meibomian glands, Zeis glands of the cilia, the glands of the eyebrows, the caruncle, and also the glands of the fine hair follicles on the skin of the eyelid. Periocular region accounts for 75% of all sebaceous gland carcinoma, although extra-ocular sites are also well-known; this is due to the fact that there is an abundance of sebaceous glands in this area. In the west, sebaceous gland carcinoma has been reported to account for approximately 2–7% of all eyelid malignancies. In contrast, several studies from India, China and Japan have reported that sebaceous gland carcinoma accounts for up to 30% of malignant eyelid neoplasms, and is the second most common malignant eyelid neoplasm in these regions after basal cell carcinoma.

Eyelid sebaceous gland carcinoma presents either as a solitary nodule or diffuse lid thickening, in which case it may often be misinterpreted as chronic blepharitis (Figures 6A-D). The latter presentation of sebaceous gland carcinoma often leads to a delay in accurate diagnosis and treatment, often leading to an unfavorable prognosis. The tumor typically causes distortion of the posterior lid margin, blockage of meibomian gland orifices, loss of cilia, with surrounding telangiectasia and sometimes ulceration in advanced cases. Upto 35% of the patients can have a pagetoid spread on histopathology, and it may involve both eyelids and conjunctival epithelium.

Etiological factors leading to sebaceous gland carcinoma are not well-established. Sebaceous gland carcinoma occurs in patients with Muir– Torre syndrome, a rare autosomal dominant disorder characterized by neoplasms of the sebaceous glands and visceral malignancies that is caused by mutation of DNA mismatch repair genes, leading to the propagation of genetic defects within replicating cells and predisposition to tumor formation. Histopathologically, the tumor consists of malignant proliferation of sebaceous cells with vacuolated cytoplasm owing to the presence of lipid.

Standard treatment for sebaceous gland carcinoma of eyelid consists of wide local excision with frozen section or Mohs microsurgery. When a pagetoid conjunctival spread is suspected, several small map biopsies are done to determine the extent of the lesion, followed by a definitive surgical planning. Orbital Exenteration is considered when there is spread into the anterior orbit. Radical neck dissection is necessary in patients with locoregional metastases as in other malignant tumors of the eyelid. Alternative treatments include cryotherapy, topical or systemic chemotherapy and radiotherapy. Cryotherapy is a useful adjunctive treatment for epibulbar and pagetoid extension of sebaceous gland carcinoma. Topical Mitomycin C has been tried for eyelid sebaceous gland carcinoma with or without pagetoid spread with variable success. Systemic chemotherapy as a neoadjuvant treatment in advanced periocular sebaceous gland carcinoma

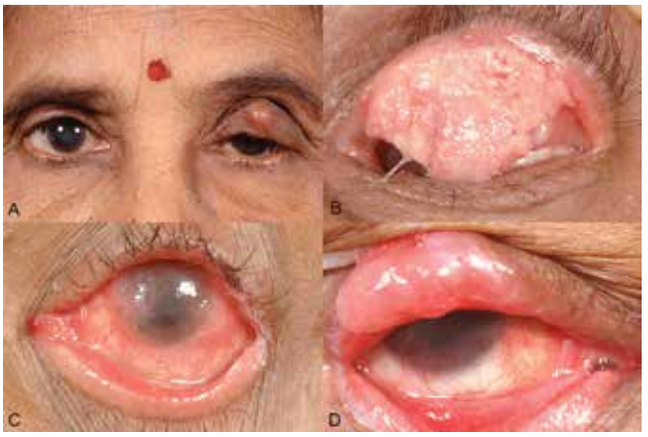

Figure 6: *Sebaceous carcinoma of the eyelid (A) Chalazion-like presentation of an eyelid sebaceous gland carcinoma (SGC) (B) Same patient on lid eversion shows an extensive tumor (C) Diffuse conjunctival involvement by SGC presenting as chronic conjunctivitis (D) Typical presentation of SGC with diffuse thickening of the lid margin, erythema, and loss of cilia*

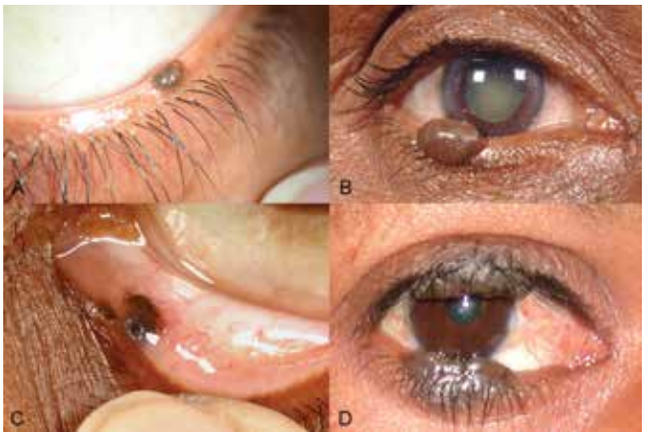

Figure 7: *Melanocytic tumors of the eyelid: different presentations of a nevus (A) A small, flat marginal nevus (B) A hypertrophied marginal nevus (C) An atypical peripunctal location of an eyelid nevus (D) A "kissing" nevus*

is evolving. Although sebaceous gland carcinoma is considered to be radiosensitive, radiation therapy for this has been described in very few short case series, mostly as an alternative to surgical treatment in patients who may be poor surgical candidates.

EYELID MELANOCYTIC NEVUS

Melanocytic nevus of the eyelid, like any other nevus, comprises of melanocytes derived from the neural crest that migrate to the skin during embryonic development. A nevus can be acquired or congenital. Depending on the location, nevi are divided into junctional, compound, and intradermal types. In general, childhood nevi are junctional and in adulthood, there is a tendency towards migration into the dermis. The clinical features vary with patient age and stage of the disease. The nevus can vary in size, location and pigmentation (Figures 7A-C). Completely amelanotic nevi commonly occur in the eyelids. A variant of congenital nevus is the "kissing" nevus of the upper and lower lid which occurs due to the formation of the nevus before the lid separation during embryological development (Figure 7D). Management is generally observation. Surgical excision maybe performed in those causing cosmetic blemish.

OCULODERMAL MELANOCYTOSIS

Oculodermal melanocytosis is a bluish-black pigmentation of the periocular skin, uveal tract, and sometimes ipsilateral orbital soft tissues, ipsilateral pinna, ipsilateral meninges, and ipsilateral hard palate (Figures 8A-B). The pigmentation is congenital, and the eyelid pigmentation is known as the nevus of Ota. It tends to follow the distribution of the first and second divisions of the trigeminal nerves. Bilaterality is seen in about 10% of cases. In the uveal tract, this condition predisposes to formation of uveal melanoma. Malignant transformation of the eyelid component into cutaneous melanoma is rare. Histopathologically, nevus of Ota is characterized by increase in the number of scattered dendritic melanocytes in the dermis. Management is generally close observation.

EYELID LENTIGO MALIGNA

Lentigo maligna or melanotic freckle of Hutchinson is an acquired pigmentation that usually occurs on sun-exposed areas. It can rarely involve the eyelid as a small localized lesion. It is rare in darkly pigmented individuals. Clinically, the lesion appears as a flat, tan to brown pigmentation with well-demarcated borders. It enlarges slowly over years. Lentigo maligna is the precursor lesion of lentigo maligna melanoma. A melanoma secondary to lentigo maligna is also flat or minimally elevated in the early stages. Management is by wide surgical resection.

EYELID MALIGNANT MELANOMA

Periocular malignant melanoma is a rare condition, accounting for less than 1% of malignant eyelid neoplasms. It can occur in the eyelid as a primary lesion or as an extension of a conjunctival melanoma, or rarely, as a metastasis from a distant primary (Figures 8C-D). Majority of patients present in their sixth and seventh decade. The most common type in the periocular area is Lentigo maligna melanoma, followed by superficial spreading melanoma and nodular melanoma. Lentigo maligna melanoma and superficial spreading melanoma are characterized by radial growth confined to the epidermis in the early stages, followed by invasion of subepidermal structures. In contrast, nodular melanoma exhibits early invasion of the subepidermal tissue.

Patients with malignant melanoma most commonly present with a sudden change in the appearance of a preexisting nevus. Increase in size and pigmentation, elevation, tenderness, and ulceration point towards malignant transformation. Lower lid is the most common site for eyelid melanoma, followed by upper lid, lateral canthus, and medial canthus. Histopathologically, the malignant cells are of three types- spindle cells, epithelioid cells, or nevus-like cells. Wide local excision is the treatment of choice for malignant melanoma. Advanced cases with orbital involvement may require orbital exenteration. The patient should be checked for preauricular and submandibular lymphadenopathy.

EYELID CAPILLARY HEMANGIOMA

Capillary hemangioma can be congenital (infantile capillary hemangiomas or strawberry hemangiomas) or acquired. Acquired capillary hemangiomas are very tiny lesions which are red-blue in color

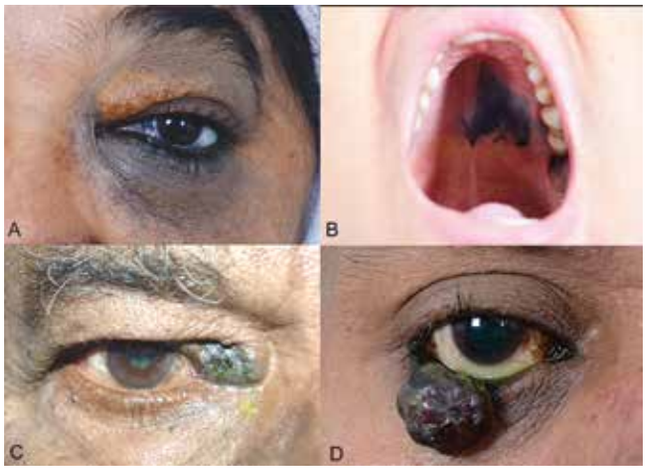

Figure 8: *Melanocytic tumors of the eyelid (A) Oculodermal melanocytosis with slate-grey pigmentation of the periocular skin and episclera (B) Ipsilateral palatal pigmentation in the same patient (C) An eyelid malignant melanoma with caruncular extension (D) A large eyelid malignant melanoma secondary to extension from the conjunctiva*

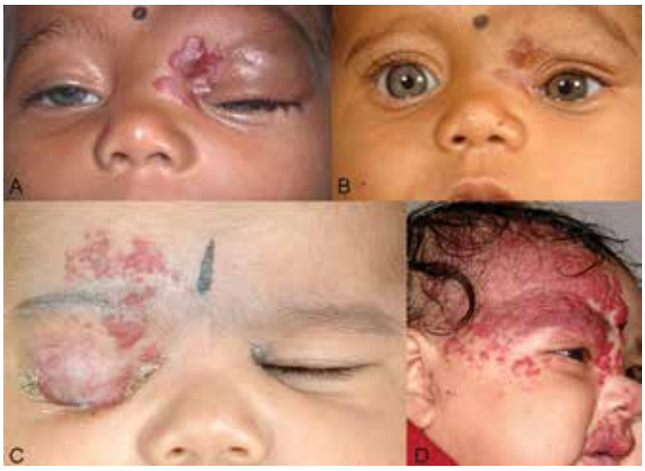

Figure 9: *Capillary hemangioma of infancy (A) Large periocular capillary hemangioma causing ptosis (B) Same patient after treatment with intralesional steroids (C) Complete ptosis secondary to a large capillary hemangioma (D) Extensive hemangioma involving the ipsilateral scalp, periocular and perioral region*

(cherry hemangiomas) seen in elderly individuals. These generally do not require any treatment. Congenital capillary hemangioma develops either at birth or within the first year of life. It can be located superficially (anterior to the orbital septum), deep (posterior to the orbital septum), or both. Regarding its etiopathogenesis, due to the similar immunohistochemical characteristics with the placenta, it is believed that infantile hemangiomas could be of placental origin.

Eyelid capillary hemangiomas are not present at birth, but develop in the first few months of life and continue to enlarge over the first 6-12 months after the first year (proliferative phase), with 90% resolution occurring within 8 years of life (involutional phase). Periocular capillary hemangioma of infancy may be seen in association with Kasabach-Merritt syndrome, which is characterized by large visceral hemangiomas, platelet entrapment and thrombocytopenia. Some of the superficial hemangiomas can lie in the subcutaneous tissues with little or no involvement of the epidermis (Figures 9A-D). The skin overlying the lesion has a bluish hue, and the lesion becomes more apparent with crying or straining.

Lesions greater than 1 cm in diameter are more likely to cause complications, with an incidence of amblyopia upto 60%. The amblyopia can either be from pupil obstruction or from refractive errors induced by the compression of the globe by the tumor. Periocular capillary hemangioma can also cause strabismus secondary to tumor compressing the recti muscles or amblyopia.

Most tumors can be managed by observation, although those causing amblyopia should be treated with refraction and occlusive patching. Oral use or local injection of corticosteroids can hasten regression of the lesion. Oral prednisolone 2 to 4 mg/kg/day for 2-4 weeks is administered under the supervision of a pediatrician. The major risks include adrenal suppression and growth retardation. Intralesional corticosteroids are administered as a combination of triamcinolone 1 mL (40 mg/mL) and dexamethasone 1 mL (4 mg/mL). Interferon α-2a upto 3 million units/m2 of body surface area can be given as daily subcutaneous injections for vision-threatening hemangiomas to cause complete regression of the lesions. Propranolol is a non-selective beta blocker that can be used systemically for eyelid capillary hemangiomas with high efficacy. The recommended dosage of oral propranolol is 2 to 3 mg/kg/day until regression and additionally for a month to prevent recurrence. Surgical treatment is rarely necessary, but can be considered in those with visual symptoms not responding to pharmacologic modalities.

EYELID KAPOSI'S SARCOMA

Kaposi's sarcoma is an endothelial cell malignancy seen more commonly in immunosuppressed individuals. Kaposi's sarcoma of the eyelid is generally seen in association with AIDS. The lesion presents as a red, purple or blue flat subcutaneous nodule (Figures 10A-B). It is frequently well-circumscribed, but can be diffuse and large. Histopathologically, it is composed of proliferating groups of endothelial cells that contain blood-filled spaces. Management is generally for palliative purpose. When the lesion is diffuse, chemotherapy is more effective than radiation. For smaller lesions, low-dose radiotherapy (15–20 Gy) in fractionated doses is curative.

EYELID HISTIOCYTIC TUMORS

Xanthelasma is a common, benign subcutaneous, minimally elevated eyelid lesion. When it is nodular, it is called a xanthoma. Xanthelasma tends to be bilateral and is more common in the elderly. It occurs in 1-3% of individuals and more frequently in women. Half of the patients with xanthelasma are normolipemic while the other half has essential or secondary hyperlipidemia. It appears as a yellow, placoid lesion that affects the medial aspect of the eyelids. It is frequently bilateral and symmetrical. Microscopically, xanthelasma and xanthoma is composed of foamy histiocytes infiltrating the dermis. Surgical excision should be considered for larger lesions causing cosmetic blemish. Topical application of 35% trichloroacetic acid is proven to be effective. Carbon dioxide laser-assisted or radiofrequency-assisted vapourization may also be performed.

Juvenile xanthogranuloma is an idiopathic granulomatous inflammation that affects older children. It appears as an orange nodule which generally regresses on its own. It is typically composed of histiocytes, lymphocytes, mononuclear cells, eosinophils, and Touton giant cells. Management is generally by observation.

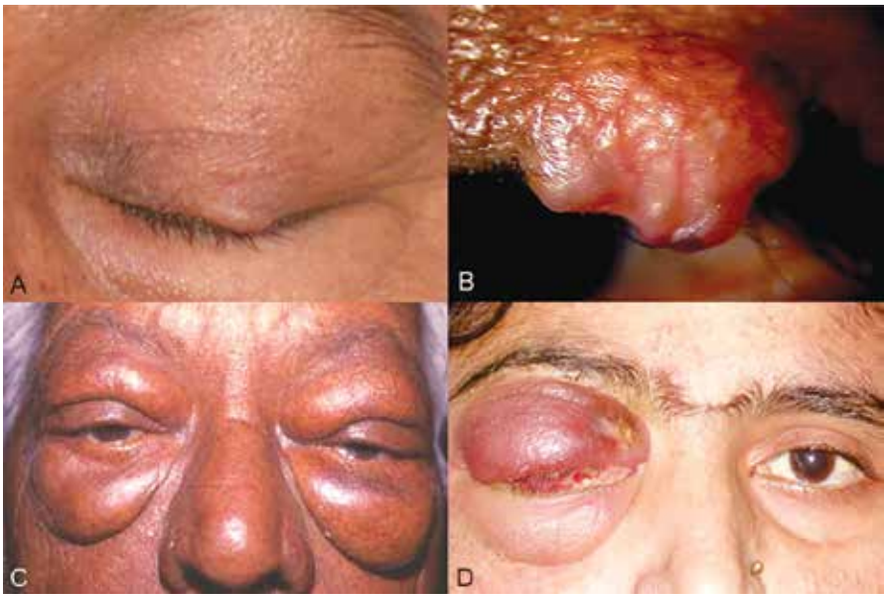

Figure 10: *Other rare tumors of the lid (A) Eyelid Kaposi's sarcoma presenting as a small mass in the upper lid (B) Rapid growth of the tumor in the same patient with a reddish-blue hue to the tumor (C) Typical appearance of a xanthogranulomatous tumor with bilateral lid involvement imparting the skin a yellowish color (D) Eyelid lymphoma with orbital extension*

Systemic or intralesional corticosteroids can be effective in refractory cases. Surgical excision may be performed if the lesion fails to respond to corticosteroids. The adult form of xanthogranuloma can occur as a solitary lesion often in patients with severe asthma (Figure 10C).

EYELID LYMPHOMA

Primary eyelid lymphomas are extremely rare. Like orbital lymphomas, eyelid lymphomas are also of B-cell type. It presents as a painless subcutaneous mass (Figure 10D). Rarely, cutaneous T-cell lymphoma (mycosis fungoides) may be seen, and this generally occurs in immunocompromised individuals. It appears as an eczema or in the form of ulceration, causing cutaneous inflammation with induration. The lesion can be solitary or multiple. Localized lesions can be controlled with radiation therapy, whereas systemic disease is treated with chemotherapy.

In conclusion, the commonest lesions affecting the eyelid are benign in nature. Primary malignant tumors affecting the lid are frequently epithelial in origin, and metastatic eyelid tumors are extremely rare. Eyelid tumors generally present in early stages, and wide excision with primary lid reconstruction is curative. Those presenting with extensive lesions require careful planning for excisional surgery followed by lid reconstruction with grafts or flaps. Additionally, radiotherapy maybe necessary in bony involvement. Orbital extension of these tumors requires exenteration.

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Botulinum Toxin A and Dermal Fillers in Oculofacial Aesthetics: An Overview

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Abstract: *With the ever-increasing demand for beauty, the importance of facial rejuvenation need not be overemphasized. Correction of facial rhytides and volume loss forms the fundamental principle in aesthetic rejuvenation of the face. Botulinum toxin injections are primarily indicated for correction of dynamic rhytids while dermal fillers are meant for volume augmentation as well as for improvement of static rhytids. The aim of this review article is to briefly discuss the important aspects of botulinum toxin and dermal fillers in the field of ocular and facial aesthetics.*

Key words: *Facial rejuvenation, rhytides, botulinum toxin, fillers.*

The ageing face results from the combined effects of loss of tissue elasticity, collagen loss, soft tissue atrophy, the continued use of facial muscles and the effect of gravity. All these lead to volume loss and the development of facial wrinkles[1]. Photo damage and smoking also contributes to the ageing process[2]. The fundamental principles of facial rejuvenation include improvement of contour, control of movement and volume augmentation[3].

BOTULINUM TOXIN

Botulinum toxin, a powerful neurotoxin causes chemo denervation of the muscles. The word "botulinum" is derived from the Latin word "botulus" which means sausage which came into existence following the many cases of sausage poisoning due to the neurotoxin produced by the bacteria Clostridium botulinum. In 1946, Schantz isolated the crystalline form of the Botulinum Toxin A. The first clinical use of Botulinum toxin injection in ophthalmology dates back to 1981 when Dr. Alan Scott injected botulinum toxin into the extraocular muscles for correction of strabismus following which it has been increasingly used for both cosmetic and therapeutic purposes.

STRUCTURE AND MECHANISM OF ACTION

Botulinum toxin A (BoNTA) is one of the eight exotoxins produced by the anaerobic bacteria Clostridium botulinum and is most commonly used toxin in clinical practice. It consists of two heavy and light chains linked with a disulphide bond. Botulinum toxin acts by inhibiting the release of acetylcholine (Ach) at the neuromuscular junction. Normally, at the junction, SNARE (soluble N-ethylmaleimide-sensitive factor attachment protein receptor) complex, facilitates the release of the Ach. The SNARE proteins are responsible for fusion of vesicle of Ach with nerve cell membrane causing its release into the synaptic cleft and resulting in propagation of the action potential. After injection of the toxin into a target tissue, it is divided into 100kD of the heavy chain and 50kD of the light chain. The heavy chain is responsible for binding with the nerve terminal, which leads to internalization of the molecule into the cytoplasm of the motor nerve terminal. Light chain acts by cleaving and inactivating the SNARE proteins leading to the inhibition of release of the Ach from the junction and paralysis of the muscle. The effects of Botulinum toxin are temporary as new axonal sprouting and turnover of neuromuscular junctions occur[5,6]. They typically last for 3-6 months.

PREPARATION AND TECHNIQUE

The two most common commercially marketed preparations of Botulinum toxin injection are "Botox" and "Dysport" both of which are Botulinum toxin type A. Dysport is thought to have increased diffusion as well as shorter duration of action, which is why Botox is preferred in most centres. These also differ in terms of molecular weight, dosing and units per vial[7]. For most procedures, 1 U of Botox is equivalent to 3-4 U of Dysport[8].

Botox is available in sterile freeze-dried powder containing 50, 100 or 200 units. This has to be reconstituted with preservative free normal saline. Depending on the volume of saline injected and the indication for its use, a solution containing 1.25 to 5 U/0.1 ml is made. This can be stored under refrigeration for up to a week, with some advocating its efficacy up to 6 weeks following reconstitution[9].

Injection is typically given with 30 or 32 G needle to reduce the pain and risk of bruising. Topical lidocaine gel or ice packs are usually given prior to injection which helps in decreasing the pain. However, lidocaine injection should not be given as it can potentiate the effect of BoNTA.

INDICATIONS

BoNTA injection can be used both for aesthetic purpose and therapeutically. The various indications for its use in ophthalmic community are listed in (Table 1). It is FDA approved for the treatment of Blepharospasm, hemi facial spasm, strabismus, glabellar lines and periocular rhytides. Mesobotox is a term used to describe multiple intradermal injection of botulinum toxin in diluted doses. This technique improves skin texture as well as facial contour in the injected area. (Figure 1A,1B,1C) shows the use of BoNTA Inj. for crow's feet (hyperkinetic

Table 1: Ophthalmic indications of Botulinum toxin injection

Aesthetic
- Glabellar lines (Frown lines)
- Orbicularis rhytids (Crows feet)
- Bunny lines
- Smoker lines
- Marionette lines
- Masseter hypertrophy
- Brow lift

Therapeutic
- Essential blepharospasm
- Hemifacial spasm
- Cervical dystonias
- Frey's syndrome
- Strabismus
- Thyroid eye disease
- Temporary chemical tarsorraphy
- Spastic lower lid entropion

Table 2: Contraindications of Botulinum toxin injection

Contraindications
- Allergy to human albumin
- Previous allergic reaction
- Pregnancy / Lactation
- Drugs -Aminoquinolones, calcium channel blockers, cyclosporine and D-penicillamine
- Neuromuscular disorders – Myasthenia, Lambert Eaton syndrome
- Infection at injection site
- Urinary tract infection, urinary retention

orbicularis oculi). Figure 2A to 2D shows the four points injection site for masseter hypertrophy. Figure 3A and 3B shows pre and post injection images for horizontal forehead lines. Figure 4A and 4B shows injection point for forehead frown lines.

CONTRAINDICATIONS

Table 2 lists the contraindications of BoNTA injection. It should not be used in children less than 12 years of age.

ADVERSE REACTIONS

Any treatment is not without its risks and complications. The following adverse reactions can be seen after BoNTA injection

- Pain and eyelid edema
- Diplopia if toxin spreads to the extraocular muscles
- Ptosis if levator palpebrae superioris is affected

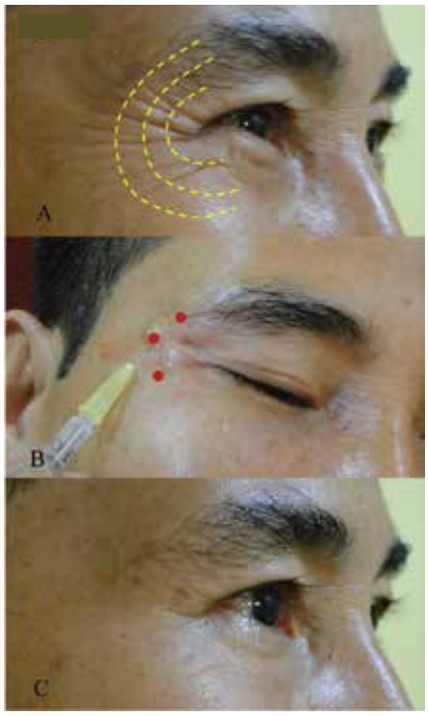

Figure 1A: *Crow's feet (pre-injection BoNTA injection),* ***Figure 1B:*** *Inj. BoNTA sites* ***Figure 1C:*** *Crow's feet (post-injection BoNTA injection).*

- Lagophthalmos causing corneal ulceration
- Ectropion, epiphora
- Mouth droop
- Brow asymmetry, Lid retraction
- Systemic absorption causing - Dysphonia, Dysarthria, Dyspnoea, Dysphagia
- Anaphylactic reactions
- Death

DERMAL FILLERS

Paraffin was the first filler used for the face in 1907[10]. Due to intolerable side-effects its use was abandoned. Subsequently many substances like mineral oil, lanolin, beeswax, vegetable oil, rubber and purified latex were used for cosmetic purpose but were found to have too many undesirable adverse effects[11,12]. Liquid silicone was used off-label for facial augmentation during the 1960s. It was not until 1980s that bovine collagen came to the market under the brand name "Zyderm I" and it became the first FDA approved dermal filler for use in facial rejuvenation[13]. The role of dermal fillers for facial aesthetics has revolutionised with the introduction of Hyaluronic acid (HA) fillers. The first HA filler approved by FDA was Restylane in 2003. Since then, HA fillers have been the cornerstone for volume augmentation of the face.

CLASSIFICATION[14]

Fillers can be classified depending on the duration of effect, material of origin, and reversibility (Table 3). Depending on duration of effect they may be classified as short (less than 3 months), medium (3-12 months), long (12-24 months), or very

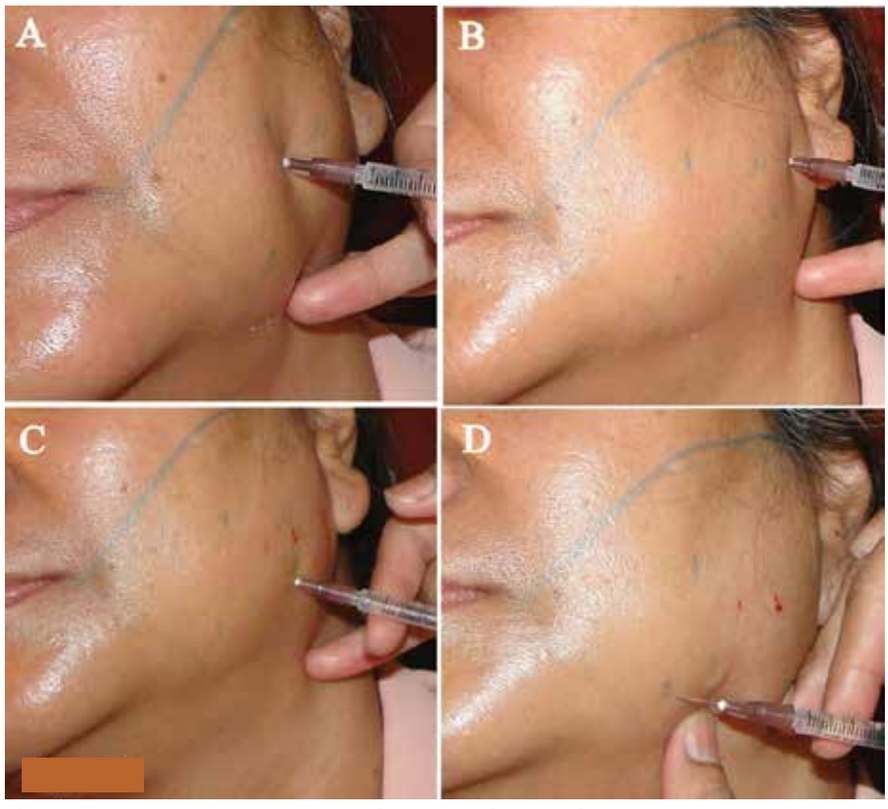

Figure 2A to 2D: *Four point Inj. BoNTA for masseter hypertrophy.*

Table 3: Classification of dermal fillers[14]	
Source	**Example**
Autologous	Fat
Biological	Collagen, Hyaluronic acid
Synthetic	Hydroxyapatite, silicone oil, polymethamethylacrylate microspheres, polyacrylamide hydrogel, hydroxyethyl methylacrylate/ ethyl methacrylate, poly-L-lactic acid
Duration of cosmetic benefit	
Temporary short duration	Saline
Short duration	Bovine collagen
Reversible(medium to long duration)	HA
Nonreversible long duration	Hydroxyapatite, polyacrylamide hydrogel, porcine collagen
Nonreversible very long duration	Silicon oil, PMMA microspheres, hydroxyethylmethacrylate, ethylmethacrylate, poly-L-Lactic acid, Fat
Nonreversible variable duration	Fat
Risk profile	
Low	Saline, HA
Medium	Collagen, hydroxylapatite, PMMA microspheres, poly-L-Lactic acid, fat
High	Hydroxylapatite, polymethylmethacrylate microspheres, poly-L-lactic acid, fat, silicone, polyacrylamide hydrogel, hydroxyethyl methacrylate/ethyl methacrylate
Level Of Physician Skill, Training, Experience, and Judgment	
Low	Saline
Medium	HA, collagen
High	Hydroxylapatite, polymethylmethacrylate microspheres, poly-Llactic acid, fat, silicone, polyacrylamide hydrogel, hydroxyethyl methacrylate/ethyl methacrylate

long acting (more than 24 months). Only Hyaluronic acid fillers will be discussed in brief.

INDICATIONS

Fillers are primarily indicated for volume augmentation and correction of static rhytides. They restore symmetry and are used for mid-face lift. An ideal filler is safe, volumizing, biocompatible, does not migrate, long lasting, easy to inject and cost effective. The following are some common indications for dermal fillers.

1. *Upper face:* Glabellar lines, forehead lines, superior sulcus deformity, temporal fossa hollowing
2. Mid- face: midface lift, tear trough deformity, cheek augmentation, nose augmentation and contouring
3. *Lower face:* Lip augmentation, marionette lines, perioral rhytids, downturned oral commissures, and irregular chin lines, pre-jowl sulcus, chin augmentation

TECHNIQUE

Preoperative consent is a must and like any cosmetic procedure, the patient should be given a mirror and asked to point out the areas where he/she feels needs treatment. Lignocaine topical cream should be applied 30 minutes prior to the filler injection. Ice packs and dental blocks have also been used for anaesthesia[15]. The desired areas are then cleaned with isopropyl alcohol. Different techniques of injection are described. These are serial puncture, linear, crosshatching and fanning techniques. In the serial puncture technique, multiple punctures are made and small boluses

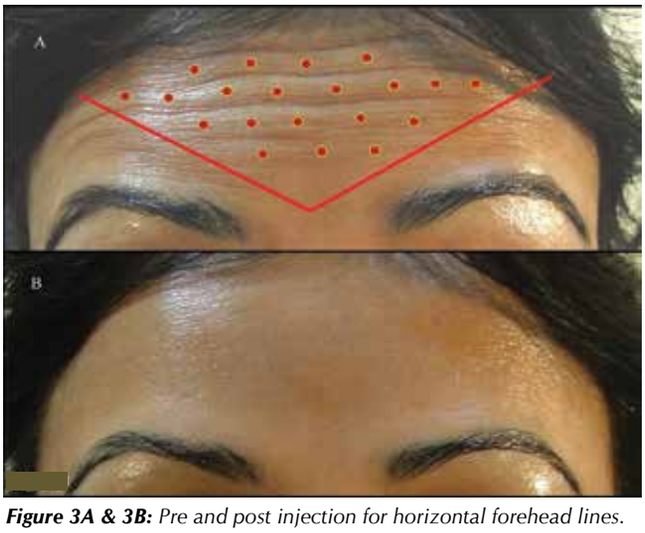

Figure 3A & 3B: *Pre and post injection for horizontal forehead lines.*

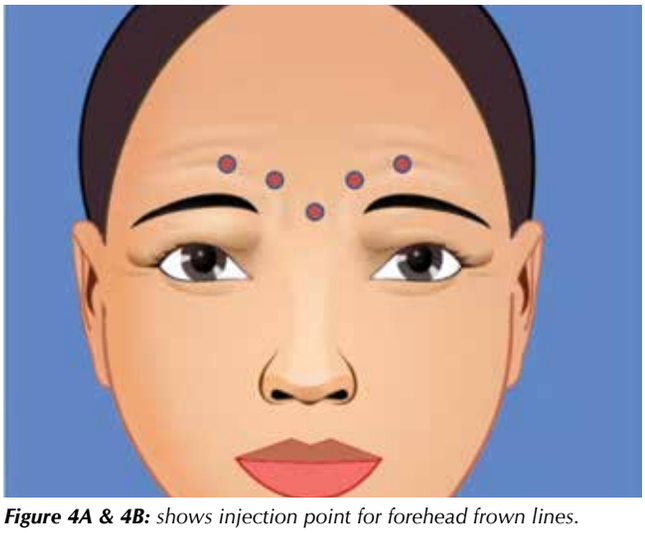

Figure 4A & 4B: *shows injection point for forehead frown lines.*

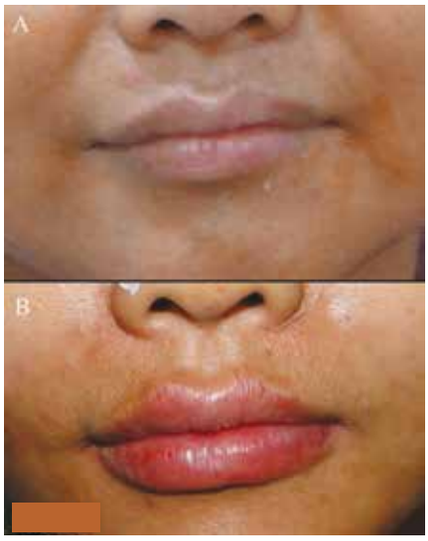

Figure 5A & 5B: *Lip augmentation using HA fillers.*

are injected in close proximity to each other. This is particularly useful for acne scars. The linear technique involves advancing the needle to its full extent into the dermis. Cross hatching again involves two perpendicularly placed injections in a linear fashion. While fanning technique utilises a single injection with multiple linear threads emanating from that point[16].

HYALURONIC ACID FILLERS (HA)

Hyaluronic acid is a naturally occurring compound, which forms a part of the normal extracellular matrix of the dermis and connective tissue[17]. Thus they are highly biocompatible with no immunogenicity. They are hydrophilic and have the ability to imbibe water up to 1000 times its volume. It is rapidly degraded in its natural form and needs to be cross-linked for stabilisation. Various commercial preparations of HA differ on the basis of the following aspects: the source, concentration, particulate size, cross-linking, type of crosslinking agent being used, and whether the HA is monophasic (more cohesive & do not migrate) or biphasic (customized for particular anatomic area), and whether an anaesthetic has been added. The injection technique as well the filler type should be customised to which area is to be treated. For example for treatment of superficial fine wrinkles, less viscous HA should be given while for volume augmentation of the malar area or naso-labial fold, more viscous agents should be preferred. It is always better to undertreat, as HA fillers will expand as they imbibe water overnight. Figure 5A and 5B shows lip

Figure 6A to 6D: *Classic HA fillers injection points for mid face lift.*

Figure 7A & 7B: *Pre and post op images after HA fillers for mid face lift and contouring.*

augmentation using HA fillers. Figure 6A to 6D shows classic HA fillers injection points for mid face lift, and figure 7A and 7B shows pre and post op images after HA fillers for the same.

Some of the commercially available HA fillers are Juvederm XC, Juvederm Ultra XC, Voluma, Vollure.

Advantages of HA fillers are –

- non immunogenicity
- non requirement of skin testing
- reversibility
- long lasting

Adverse effects include pain, bruising, edema, nodule formation, accidental intravascular injection, Tyndall effect (if the injection is given too superficial and the skin is very thin), granuloma, scarring, CRAO (causing blindness)

CONCLUSION

Facial rejuvenation has become an important subject of psychosocial well being in this era of fashion and beauty. BoNTA injections and dermal fillers offer an excellent non surgical method of facial rejuvenation. These procedures should be performed by a skilled facial aesthetics surgeon to meet the patient's expectations. Important knowledge of facial anatomy is a must for the aesthetic surgeon interested in these methods. It is always better to under correct than overcorrect while giving dermal fillers.

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IMAGING OF THE ORBIT: COMPUTED TOMOGRAPHY

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Modern medicine has advanced significantly in part due to the ability to non-invasively identify, diagnose, and follow-up lesions with high precision. Computed tomography (CT) was the first cross-sectional modality introduced by Sir Godfrey Hounsfield and Alan M Cormack in 1971. The maturation of technology has remarkably improved the speed, efficiency and ubiquity of this modality making it the primary imaging choice for the orbit[1]. It is important for the clinician to be competent in the analysis of radiological investigations to achieve a better understanding of the disease pathology resulting in better treatment. Herein, we discuss the underlying basic principles of computed tomography (CT) and its applications.

BASIC PRINCIPLES OF CT

Computed tomography (CT) is an x-ray based modality with 2 basic hardware components – an x-ray tube and a detector array housed within a rotating CT gantry. A basic understanding of CT physics requires understanding of its 3 basic phases: x-ray production, x-ray beam detection and image reconstruction.

X-ray production: The CT x-ray tube is similar to that of conventional radiography with a cathode and an anode for x-ray production, which produces beams of a fixed energy (120kV) and variable amplitude (200-350mAs)[2].

Detector Array: The x-ray beam traverses the anatomy and undergoes differential attenuation depending on the electron density of the individual tissues. These attenuated rays then fall on detectors within the detector array that produce electrical signals in response to photon stimulation.

CT Gantry: The x-ray tube and detector array are housed diametrically opposite to each other on a circular CT gantry which surrounds the patient.

Image Reconstruction Algorithm: When the x-ray beam passes through the anatomy of interest, each individual ray is attenuated by the tissue in its path to a varying extent. With the information of attenuation along the paths of multiple rays, it is possible to mathematically reconstruct the individual structures of the entire area as a cross-sectional image by a computer algorithmic method. The resultant image can be described in terms of its image quality (pixel size, resolution, image noise) and image content (tissue Hounsefield units), and can be manipulated using post-processing methods (3D and multiplanar reconstructions).

Image Quality - Pixel/Voxel sizes & Image Resolution: The reconstructed image of the cross-sectional volume of interest is divided into multiple 'pixels', and each pixel has a finite thickness, termed 'slice thickness' giving rise to a three dimensional 'voxel'. Today's helical CT scanners are able to achieve this isotropic voxel sizes (same size in all 3 axes), as small as 0.3–0.4mm. This allows for high resolution multiplanar reconstructions in any plane, obviating the need for separate acquisitions. However, image noise or graininess increases with smaller voxel sizes, and therefore, the thickness can be retrospectively increased to give an increased slice thickness resulting in better image quality. In the brain, the slice thickness is hence set to 5-10 mm for optimal image noise, while in the orbit with its smaller structures, a slice thickness of 1-3 mm is usually better. For most indications, a 2 mm slice thickness is sufficient. However, in orbital trauma or suspected foreign body, a slice thickness of 1-1.5 mm is ideal for detection of small foreign bodies and additionally, this allows for high quality 3D reconstructions for surgical planning[3].

Image Content - Linear Attenuation Coefficients and Hounsefield Units (HU)/CT Units: An important advantage of CT is its improved tissue characterization as compared to radiography. This is possible because different tissues have different x-ray attenuation which is dependent on the electron density of these tissues. In the Hounsefield scale, the radiodensity of water is set empirically at 0 and is represented in a shade of grey. Air, being least radioattenuating, has a value of -1000HU and appears a dense black. In the positive direction however, the scale may extend to about +1500HU (cortical bone) to +4000HU (for dense metals) appearing a bright white (Figure 1).

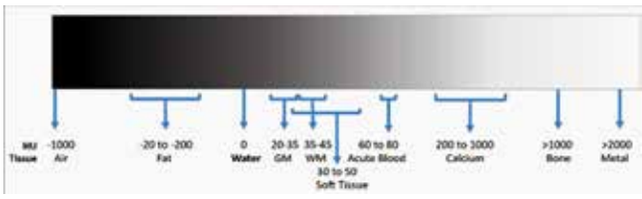

Figure 1: *The Hounsefield scale. Water is represented at 0 as a shade of grey. Hypoattenuating substances are represented as hypodense (dark) while hyperattenuating substances are represented as hyperdense (white).*

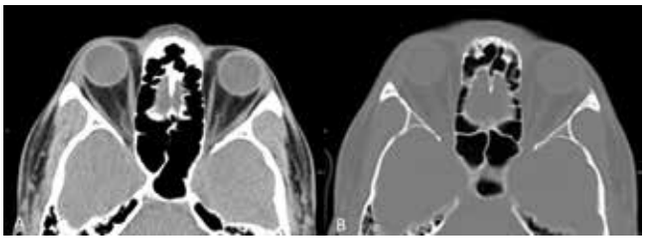

Figure 2: *(A) Soft Tissue & (B) Bone window. Note loss of bone structure in the soft tissue window and loss of soft tissue details in the sharper bone window.*

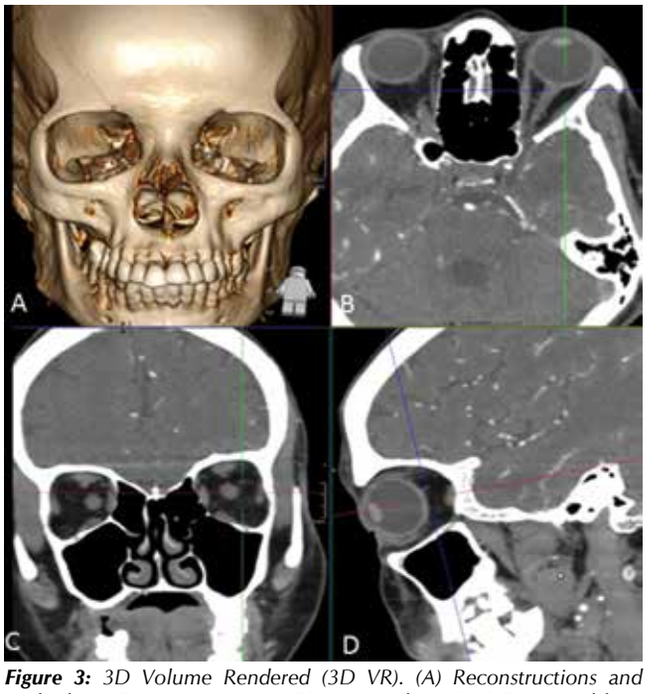

Figure 3: *3D Volume Rendered (3D VR). (A) Reconstructions and Multiplanar Reconstructions (MPR) in any plane (B, C,D) created from thin axial sections of CT can be used to demonstrate pathology more clearly.*

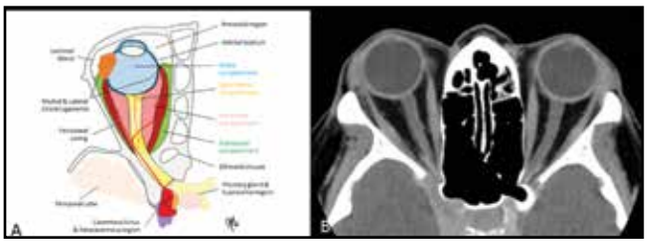

Figure 4: *(A) Diagrammatic representation of orbital anatomy and (B) Plain CT orbit. Normal appearance and densities of the orbital tissues are shown. Fat appears very hypodense, while fluid filled structures such as the vitreous chamber slightly hypodense compared to soft tissue. Bone and air are at the extremes of hyperdensity and hypodensity respectively.*

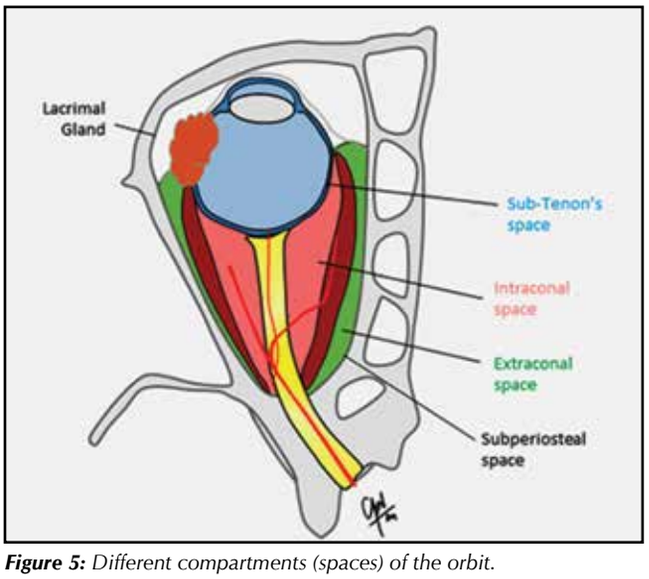

Figure 5: *Different compartments (spaces) of the orbit.*

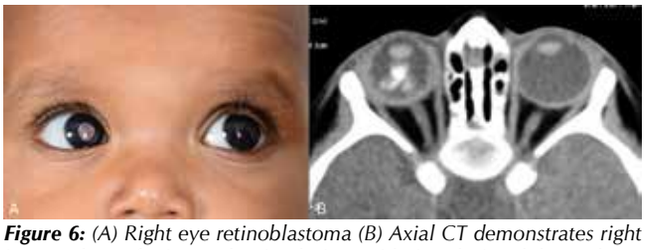

Figure 6: *(A) Right eye retinoblastoma (B) Axial CT demonstrates right intraocular soft tissue density lesion with intralesional hyperdense specks of calcification.*

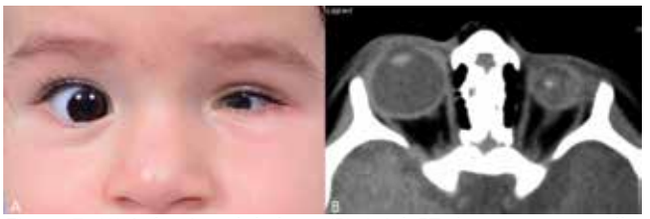

Figure 7: *(A) Left microphthalmos (B) Axial CT demonstrates smaller left eye and orbit in comparison to the contralateral side.*

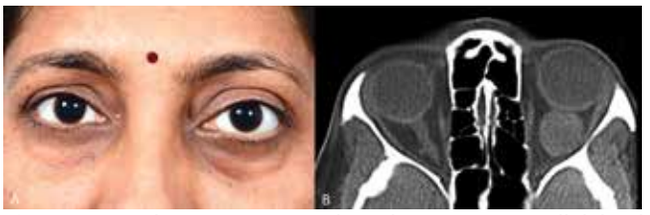

Figure 8: *(A) Left proptosis (B) Axial CT demonstrates a well-defined isodense intraconal lesion which on biopsy was proven to be cavernous hemangioma.*

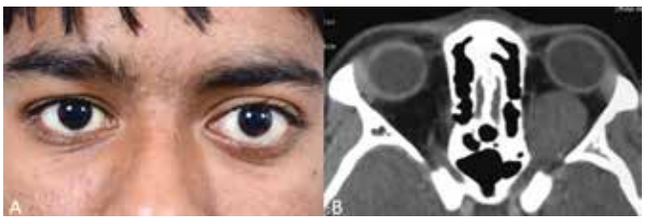

Figure 9: *(A) Left proptosis (B) Axial CT demonstrates a well-defined isodense intraconal lesion with optic foramen widening which on biopsy was proven to be optic nerve glioma.*

Figure 10: *(A) Right proptosis (B) Axial CT demonstrates a well-defined isodense intraconal lesion with optic foramen widening which on biopsy was proven to be optic nerve sheath meningioma.*

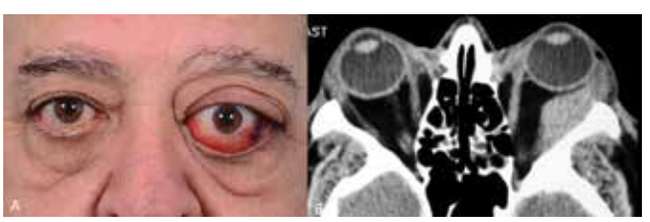

Figure 11: *(A) Left proptosis (B) Axial CT demonstrates lateral rectus thickening with an isodense lesion which on biopsy was proven to be lymphoma.*

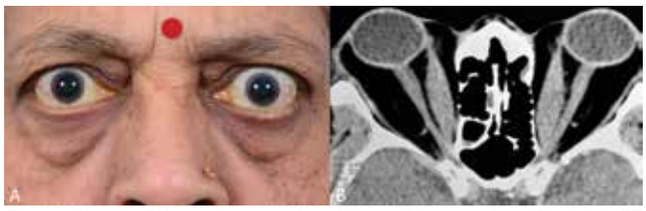

Figure 12: *(A) Bilateral proptosis with eyelid retraction (B) Axial CT demonstrates bilateral medial rectus thickening without involving the tendon, characteristic of Grave's myopathy.*

Figure 13: *(A) Right proptosis (B) Coronal CT demonstrates inferior rectus thickening and the presence of a cyst with a hyperdense central scolex, characteristic of myocysticercosis.*

Figure 14: *(A) Left proptosis (B) Coronal CT demonstrates diffuse thickening of superior and lateral rectus muscles involving the tendons (not shown in the images) which on biopsy was proven to be non-specific orbital inflammatory disease (myositis).*

Figure 15: *(A) Left internal angular orbital mass (B) Axial CT shows a cystic lesion in the left superomedial orbit, suggestive of dermoid.*

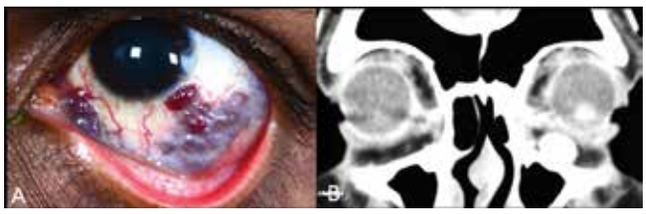

Figure 16: *(A) Left eye conjunctival venous malformation (B) Coronal CT shows the presence on an irregular isodense mass in the inferior left orbit with a hyperdense well-circumscribed lesion along the orbital floor, suggestive of phlebolith within the orbital varix.*

Figure 17: *(A) Right proptosis with hypoglobus (B) Coronal CT shows a large right superomedial isodense orbital lesion displacing the globe downward and outward, which on biopsy was proven to be rhabdomyosarcoma.*

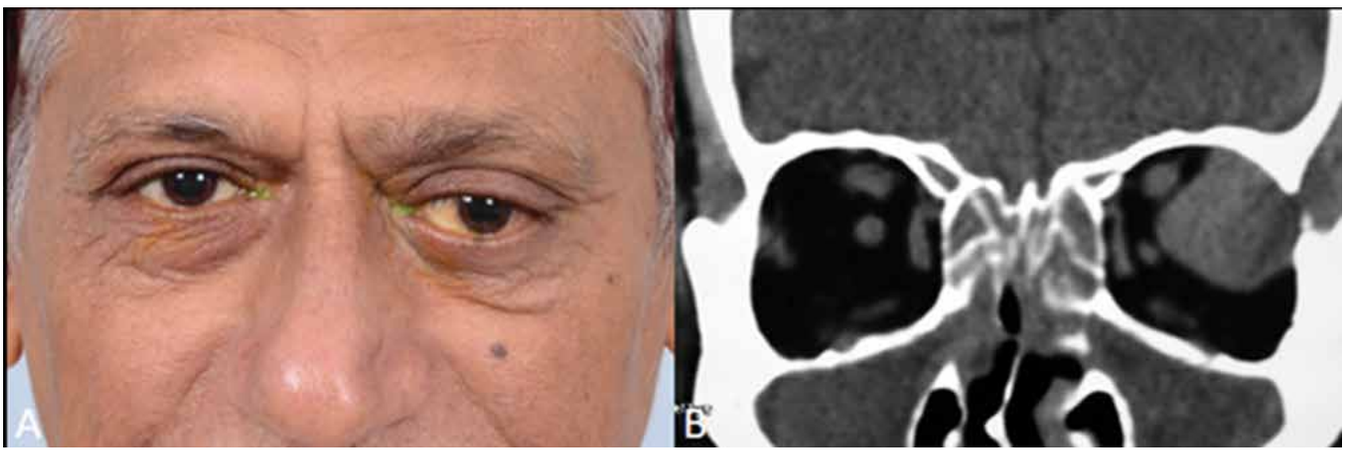

Figure 18: *(A) Left proptosis (B) Coronal CT reveals a left supertemporal isodense orbital mass arising in the lacrimal fossa, with bony indentation and no bone destruction (apparent bony discontinuity is due to overlapping of the soft tissue mass, which was not present on axial scans, not seen in the image) which on biopsy proved to be lymphoma.*

Display Window: Tissues in the body have a large range of attenuations which can be represented only as shades from black to white. Thus, only a range of densities are displayed and the densities at the extremes are displayed in black and white. This range between the minimum and maximum attenuations that are represented as black and white respectively is termed the 'window width' (WW). Hence, for perceiving soft tissues which are all in a similar range of attenuations, a narrow window level of 100-300HU will suffice, labelled as the 'soft tissue window' (Figure 2). On the other hand, for tissues that have a much wider of attenuations for example bone (2000-3000HU) and air (-1000HU), a much wider window width of 3000-4000HU will be required and this is labelled as the 'bone window'. The window width thus adjusts the contrast level of the image. A 'window level' (WL) represents the midpoint of the range of the window width.

Post-processing: Newer modalities like the Multiplanar Reconstructions (MPR) allow display of anatomy in any plane. Maximum Intensity Projection (MIP) is another method of reconstruction in which multiple slices are summated together to highlight structures that have a high density. Three-dimensional volume rendered (3D VR) scan is a method of reconstructing the entire volume of 2D CT acquisitions in a 3D view. MPR & 3D-CT reconstructions (Figure 3) have been proved to be helpful in the evaluation of comminution fracture, displaced

and complex fractures, and pathologies involving multiple planes which are difficult to analyse in a single plane[4].

Contrast Enhanced CT: Contrast medium is not required in the evaluation of foreign bodies, uncomplicated orbital fractures, uncomplicated thyroid ophthalmopathy or other pathologies of the extraocular muscles, uninfected/ ruptured dermoid cysts or benign bony lesions such as osteoid osteoma, fibrous dysplasia or Paget's disease. On the other hand, the contrast enhanced scan is useful to evaluate adjacent vascular structures, evaluate any intracranial extension of an orbital lesion, look for the presence of metastasis or concurrent lesions (trilateral retinoblastoma), and rule out other unsuspected lesions such as meningiomas, aneurysms, or arteriovenous malformations. It also plays a major role in the staging, treatment planning and follow up of malignant tumors like osteogenic or chondrogenic sarcomas or metastatic bone disease[5,6].

Radiological Approach to Orbital Disease:

A radiological approach to the orbit consists of the following steps:

- Knowledge of normal anatomy (Figure 4) including normal measurements
- Detection of an abnormality
- Compartmental localization of the abnormality and probable structure of origin (Figure 5)
- Assessment of extent of lesion including associated maxillofacial, paranasal sinus, or intracranial extension as well as mass effect
- Characterization of the lesion and probable diagnosis and differentials (Table 1)

In conclusion, with its fast scanning speed and isotropic spatial resolution, CT allows diagnosis of emergent conditions more quickly, safely and accurately. In general, CT is useful in evaluating bony structures and evaluating calcifications. CT angiography may be used in the evaluation of vascularity. The radiologist can be a highly invaluable partner in both the diagnosis and treatment of orbital diseases. A succinct summary of the clinical findings and possible clinical diagnosis must be included in the requisition for the imaging study. This input is of significant value to the radiologist in choosing an imaging protocol and providing a reliable diagnosis.

Table 1: Characterization of the lesion and probable diagnosis and differentials

Ocular Lesions	**Chorioretinal lesions:** Retinoblastoma (Figure 6) Melanoma Choroidal osteoma **Globe abnormalities:** Microphthalmos (Figure 7)
Intraconal Lesions	**Tumors:** Cavernous hemangioma (Figure 8) Lymphangioma Hemangiopericytoma Glioma (Figure 9) Meningioma (Figure 10) Optic nerve sheath cyst Arteriovenous malformation (AVM) Lymphoma **Inflammation:** Optic Neuritis Granulomatous disease (sarcoid, TB) Non-specific orbital inflammatory disease (NSOID)
Enlarged Extraocular Muscles	**Tumors:** Lymphoma (Figure 11) Rhabdomyosarcoma Alveolar soft part sarcoma Vascular lesions (cavernous hemangioma, AVM) **Inflammation:** Graves's myositis (Figure 12) Cysticercosis (Figure 13) Granulomatous myositis including sarcoidosis NSOID (Figure 14) Acromegaly
Extraconal Lesions	**Tumors:** Dermoids (Figure 15) Capillary hemangioma Lymphangiomas Varix (Figure 16) Fibrous Histiocytoma Hematic cyst Lipoma Cholesterol granuloma Peripheral nerve sheath (PNS) tumors Orbital encephalocoele Plasmacytomas Rhabdomyosarcomas (Figure 17) Lacrimal gland lesions (lymphoma, epithelial tumors, inflammation) (Figures 18,19,20) **Inflammation:** Wegener's granulomatosis (Figure 21) Amyloidosis NSOID
Subperiosteal Lesions	Subperiosteal cellultis & abscess Hematoma, Hematic cyst Neoplastic infiltration from PNS En plaque meningioma Lymphomas, leukemias, plasmacytoma Lacrimal gland tumors Dermoid & epidermoid Cholesterol granuloma Fibrous histiocytoma Osseous & cartilaginous tumors Metastasis (neuroblastoma)
Bony Orbit abnormalities	Contracted socket (Figure 22) Orbital fracture (Figure 23)

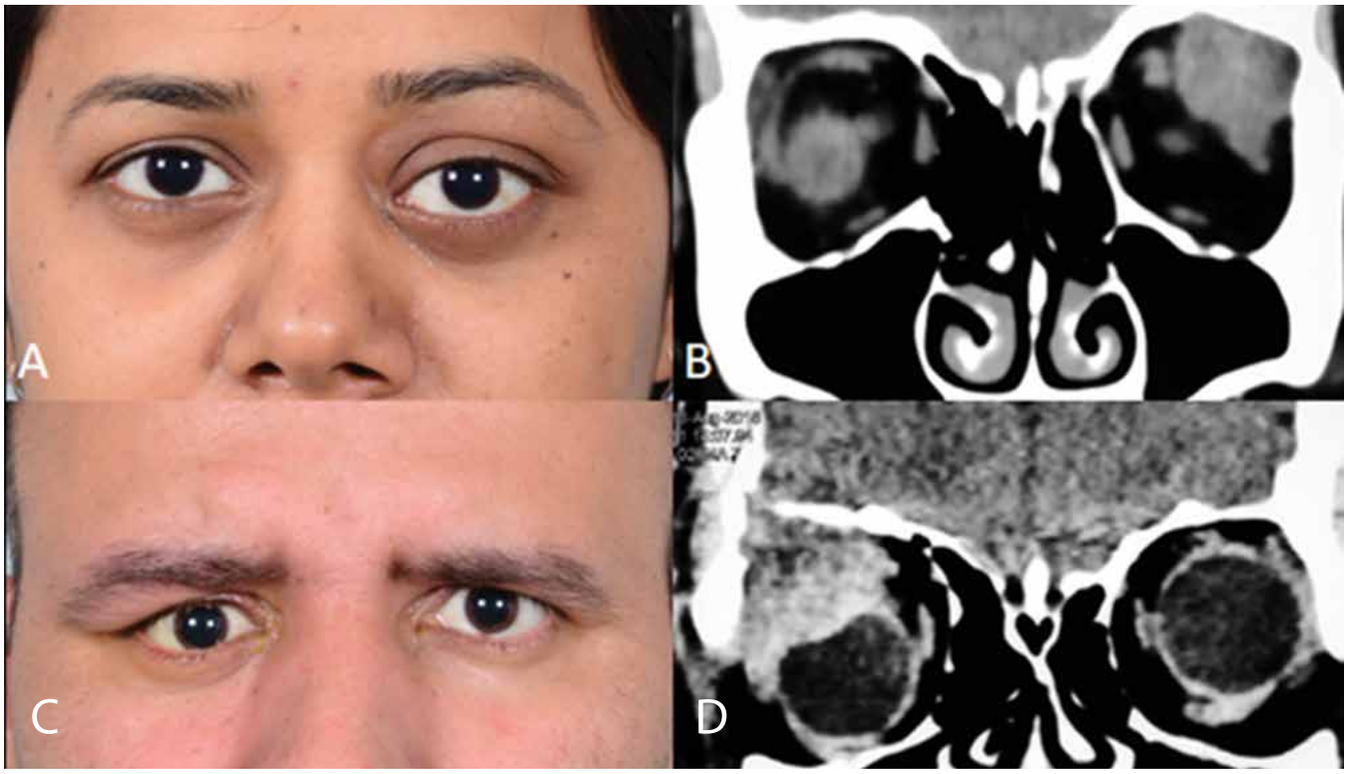

Figure 19: *(A) Left proptosis (B) Coronal CT reveals a left supertemporal homogenous orbital mass in the lacrimal fossa, with bony indentation and no bone destruction which on biopsy proved to be pleomorphic adenoma. (C) Right proptosis (D) Coronal CT shows a non-homogenous ill-defined mass lesion in the right lacrimal fossa with evidence of bone destruction which on biopsy proved to be adenoid cystic carcinoma of the lacrimal gland.*

Figure 20: *(A) Left proptosis with superotemporal mass (B) Coronal CT reveals non-homogenous ill-defined mass lesion in the left lacrimal fossa which on biopsy proved to be to be orbital tuberculosis.*

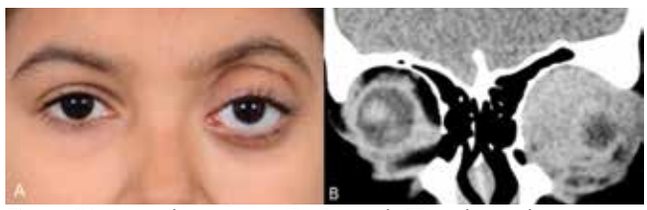

Figure 21: *(A) Left proptosis (B) Coronal CT with non-homogenous diffuse isodense lesion in the left orbit which on biopsy proved to be Wegener's granulomatosis.*

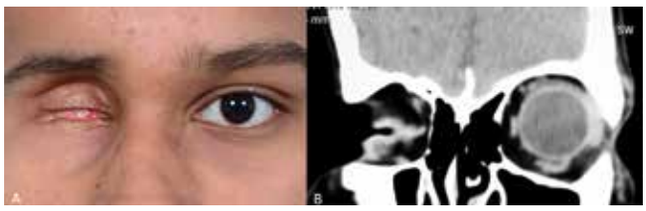

Figure 22: *(A) Right contracted socket after radiotherapy for retinoblastoma (B) Coronal CT shows contracted right socket in comparison to the contralateral side.*

Figure 23: *(A) Right upgaze restriction after blunt trauma (B) Coronal CT shows right orbital floor fracture with incarceration of the inferior rectus muscle in the fracture.*

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Blepharoplasty for Periorbital Rejuvenation: Current Concepts

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Abstract: *Blepharoplasty is a surgical procedure performed to rejuvenate aging eyelids and periorbital region. The goal is to reverse anatomical changes that occur with aging and to restore the youthful contours in a natural fashion while providing an aesthetically pleasing, lasting result. As the eye is the main focus of attention on a face during communication, cosmetic rejuvenation of the periorbital area has received maximum attention by multiple specialities involved in facial enhancement. The improved understanding of the anatamophysiology of facial aging has led to continuous advancement, a surge in demand and a variety of surgical as well as nonsurgical options for rejuvenation. With an in-depth knowledge and thorough understanding of the anatomy, physiology and function of the eyelids and surrounding structures, the oculoplastic surgeons are expanding their horizons and embracing the rewarding field of facial aesthetics.*

Periocular area is the first area of the face to show signs of aging because of the unique anatomy and functional dynamics of the region[1]. According to the American Society of Plastic Surgeons, blepharoplasty is the third most common plastic surgery procedure performed in the United States, with over 216,000 eyelid operations performed in 2013[2]. Although nearly 85 percent of patients undergoing cosmetic eyelid operations are women, it is the third most common aesthetic procedure in men. There are no statistical figures available for India but blepharoplasty procedure has seen a remarkable increase in popularity and frequency in recent years.

The basic cause of aging process is a sum of gravitational descent of tissues , volume loss and deflation along with loss of elasticity and laxity of soft tissues. Lambros[3] observed that gravitational soft tissue descent is not a major aging consequence by documenting that the position of moles, wrinkles and other markers on the upper, midface and malar region remain stable in their relative position over time. Later studies concluded that aging is a multifactorial 3-dimensional process and many causative factors have a role to play.

Currently cosmetic blepharoplasty is not just an operation of the eyelids with simple excision of skin and fat, but includes a plethora of procedures to restore youthful contours of the periorbita and midface so that the eyelids naturally blend into the brow and cheek without any demarcation, curves and grooves. This requires technical expertise and a surgeon focussing on aesthetic precision. A thorough understanding of the structural changes that take place with aging and a mastery of the anatomy of the orbit, midface, forehead and brow is critical. This article provides a comprehensive discussion of eyelid surgical rejuvenation tools and techniques currently practised.

Preoperative evaluation of a blepharoplasty patient should include:

Complete medical history including systemic disorders specially thyroid or bleeding disorders, collagen vascular diseases, prior eyelid or facial surgery and trauma. Hypertension should be well controlled and anticoagulants or platelet inhibiting medications, NSAIDs or supplements should be discontinued before surgery.

A complete ocular examination should be performed to assess visual acuity, extraocular muscle movement and fundus examination and findings should be documented. Schirmer's test and TBUT must be done to rule out dry eye which can get aggravated by blepharoplasty and calls for a more conservative excision of skin and judicious removal of orbicularis muscle. Bell's phenomenon, presence of lagophthalmos and corneal sensations must be assessed to prevent problems like exposure keratitis. Photographs of the patient must be taken to document the preoperative appearance.

UPPER LID BLEPHAROPLASTY

Can be performed for both cosmetic and functional indications.

Functional Indications

- Dermatochalasis that overhangs eyelid margin (pseudoptosis) affecting vision and causing restriction of superior visual field
- Concurrently with ptosis repair (frontalis sling/ LPS advancement)
- To correct associated lash ptosis or entropion,
- Blepharochalasis
- Floppy eyelid syndrome
- Asthenopic symptoms and persistent blepharoconjunctivitis due to excessive redundant skin
- For use of full thickness skin graft elsewhere

PERTINENT ANATOMY AND PHYSIOLOGY OF AGING UPPER EYELID

The upper lid structures going from superficial to deep

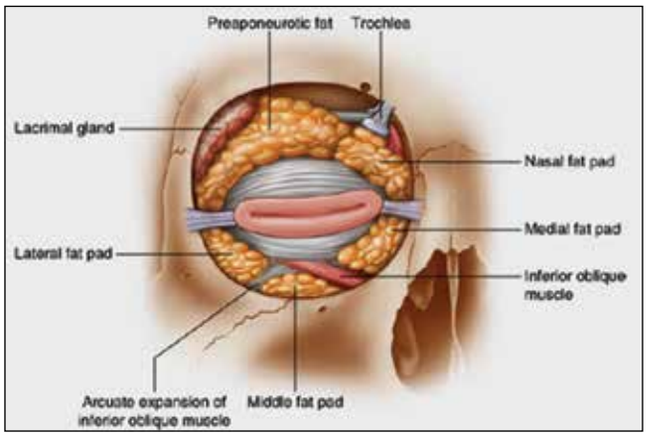

Figure 1: *Upper and lower eyelid fat pads. From KS Tan et al.*

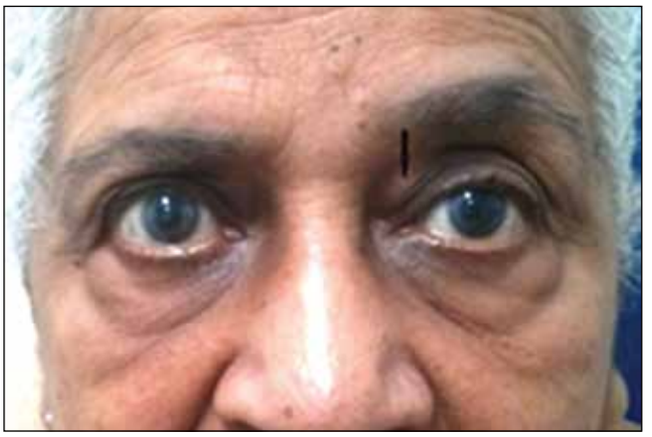

Figure 3: *Prominent nasal fat pad (arrow) is seen. Atrophy of central fat pad with deep superior sulcus and a high lid crease, which is accentuated by ptosis of left upper lid with a compensatory brow elevation. Also, prolapse of all three fat pads is seen in the lower eyelid.*

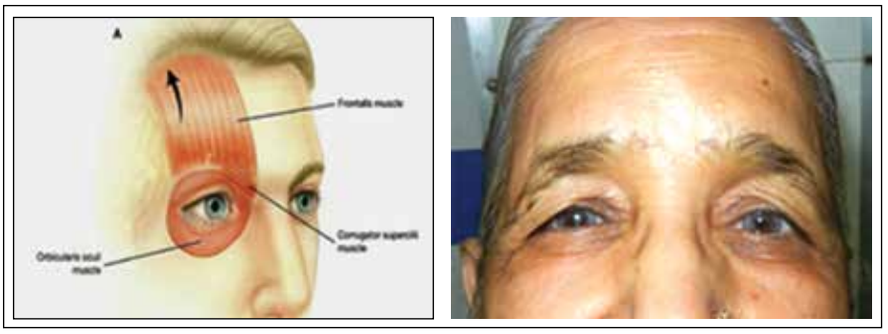

Figure 2: *Frontalis muscle is deficient in the lateral part of the brow. In this patient much of lateral hooding of the upper lid is secondary to brow ptosis. Upper lid blepharoplasty alone will worsen the ptotic brow and must be combined with browplasty.*

include the skin and orbicularis muscle. The next deeper layer is the orbital septum. Just deep to the orbital septum, in the preaponeurotic plane, are the fat compartments. There are two, the nasal and central fat compartments in the upper lid. The lateral compartment is occupied by the lacrimal gland (Figure 1).

Aging is a dynamic process that involves changes in each layer of the facial tissues. Skin loses elastin and collagen resulting in laxity, redundancy, sagging and wrinkles. Volume loss from adipose and muscle mass atrophy causes further tissue deflation. Brows begin to descend due to relaxation of ligamentous attachments along with resorption and subsequent thinning of the superomedial and inferolateral orbital rim. Since the frontalis muscle, which is the only elevator of the brow, is deficient laterally, lateral brow ptosis is more apparent. (Figure 2). This is aided by atrophy of Retro-orbicularis oculi fat (ROOF) pad. The ROOF pad, also called the brow fat pad, is located deep to the orbital orbicularis oculi and frontalis muscles at the brow overlying the superior orbital rim and is responsible for the youthful fullness of the brow.

As we age, shift is noted in orbital fat volume. The nasal fat pad increases and becomes more prominent whereas the central preaponeurotic fat pad involutes and retracts. As a result, the superior tarsal sulcus deepens and the lid crease elevates (Figure 3). This fat loss from all compartments accentuates the typical 'skeletonised' hollowed appearance which is the hallmark of aging. Hence the current emphasis on volume augmentation (by use of Fillers/fat injection and conservative fat and muscle excision) as an essential component of rejuvenation.

PREOPERATIVE ASSESSMENT AND PLANNING FOR UPPER LID BLEPHAROPLASTY

The eyebrow and upper eyelid are so intimately intertwined in their function and esthetics that they are considered 2 parts of a continuum and assessed together[4].

Brow position and symmetry is carefully noted. The brow is manually repositioned at the supraorbital rim in males and slightly above in females to accurately assess and mark for blepharoplaty. The decision is taken whether a brow lift or browpexy needs to be performed adjunctively. If upper blepharoplasty is performed without correction of a lax or ptotic eyebrow, there are residual upper lid folds and narrowing of brow- lash distance postoperatively, which defeats the aesthetic goal.

Upper lid position and symmetry is assessed while carefully stabilising the brow. Margin-reflex distance (MRD) is measured on each side and compared to rule out presence of eyelid ptosis. Even an unnoticeable, small degree of preexisting ptosis gets unmasked after blepharoplasty because compensatory frontalis activity raises the lid crease height on the affected side leading to asymmetry and compromising the aesthetic resul (Figure 4).

A fullness or bulge in the lateral part is due to a prolapsed lacrimal gland which should be repositioned and fixated to the periosteum of the lacrimal gland fossa with sutures.

SURGICAL TECHNIQUE

The first and the most critical step is marking the skin incision. The marking is preferably done with the patient seated upright. If a native eyelid crease is present, it is marked on both the eyes . Crease height is measured on both eyes for symmetry. If the crease is indistinct or needs to be revised, the central height is marked typically at 8-11mm in females and 6-9mm in males. Laterally, beyond the lateral canthus, the mark is tapered superiorly upto the lateral orbital rim to include lateral hooding of skin if present.

Toothless forceps are used to pinch the redundant skin. The inferior tooth of the forceps corresponds to the lid

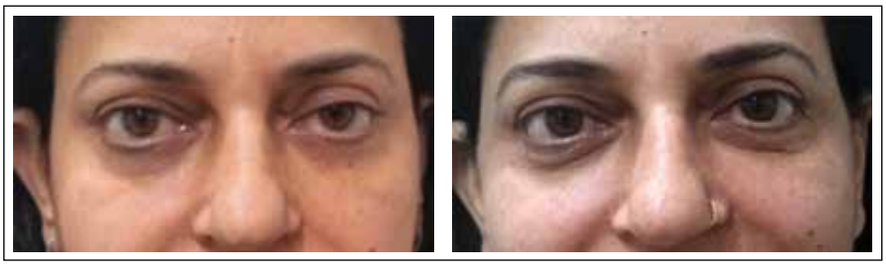

Figure 4: *A mild preexisting ptosis missed at the time of upper blepharoplasty became highlighted postoperatively giving unacceptable result. The patient was happy after Ptosis surgery was performed.*

Figure 5: *Skin marking for upper blepharoplasty is the most crucial step. The superior and inferior marks should blend into the crease on eyelid opening.*

Figure 6: *Skin marking for upper blepharoplasty.*

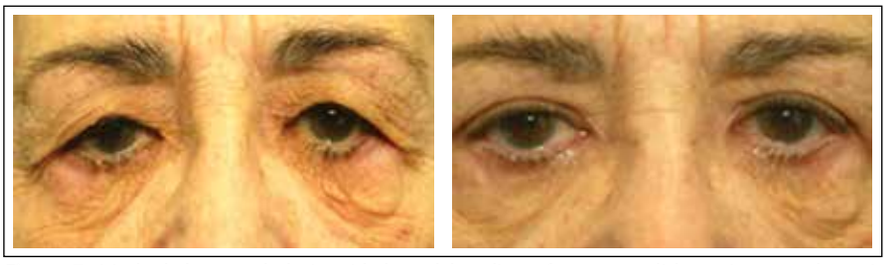

Figure 7: *Marked dermatochalasis with lateral hooding of upper eyelid causing pseudoptosis and obstruction of superior visual field. 6 month postoperative result after upper blepharoplasty performed mainly for functional indication.*

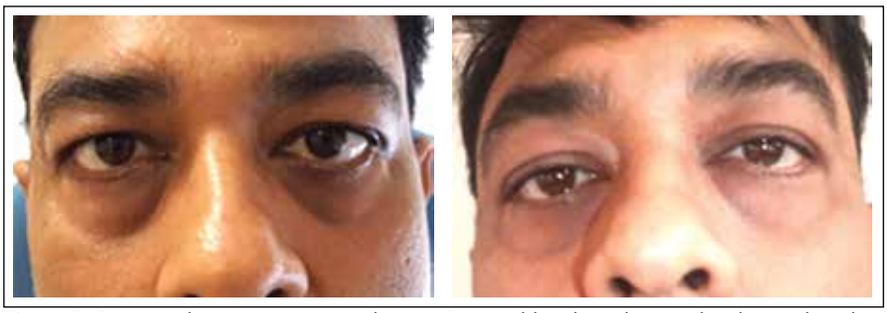

Figure 8: *One month postoperative result in a 40 year old male. Lid crease height was kept low for a better cosmetic result.*

crease marking and the upper tooth delineates the maximum extent of skin to be removed. There should be no lifting of lashes while the skin is pinched. The redundant skin is thus marked (Figure 5).

Many shapes and designs of the skin marking have been described in literature. The basic principle is to excise extra skin while carefully leaving at least 20 mm of skin from inferior brow to the lid margin essentially. The curve of the upper mark should follow the contour of the brow remaining equidistant from it (Figure 6).

Second step consists of making skin incision using a surgical blade, RF cautery, electrocautery or Laser. Only skin is excised along the mark leaving orbicularis muscle intact. The benefit of preserving orbicularis is the volumising effect it has, providing a more youthful enhancement to the upper lid besides reducing chances of inducing lagophthalmos. The orbicularis can be selectively trimmed if appears bulky.

As per the plan formulated at the time of assessment, the nasal fat pad is exposed after opening the septum medially. Newer techniques focus on preservation of fat in the eyelid, especially the central fat pad. The nasal fat pad is either modestly excised or redistributed and anchored in the central compartment in cases of hollowing to fill the deep supratarsal hollow.

Meticulous hemostasis is achieved. Excessive and deep cautery should be avoided to prevent injury to the trochlea which is situated between the medial and the central fat pads. The lacrimal gland may be seen prolapsed out of the orbit in the lateral compartment. Care should be taken to avoid mistaking it with fat. The gland should be reposited in the lacrimal gland fossa by taking a suture bite from the capsule to the periosteum.

Orbicularis muscle closure is performed using vicryl suture followed by skin closure using continuous or interrupted sutures.

Figure 7 and 8 depict postoperative results of functional and cosmetic upper blepharoplasty respectively.

ANATOMY AND AGING OF THE LOWER LID AND MIDFACE

Our understanding of aging has evolved to reflect the concept that the lower eyelid contour does not stop at the inferior orbital rim and is a continuum with the cheek[5]. In youth, the lid-cheek junction is blended together without any

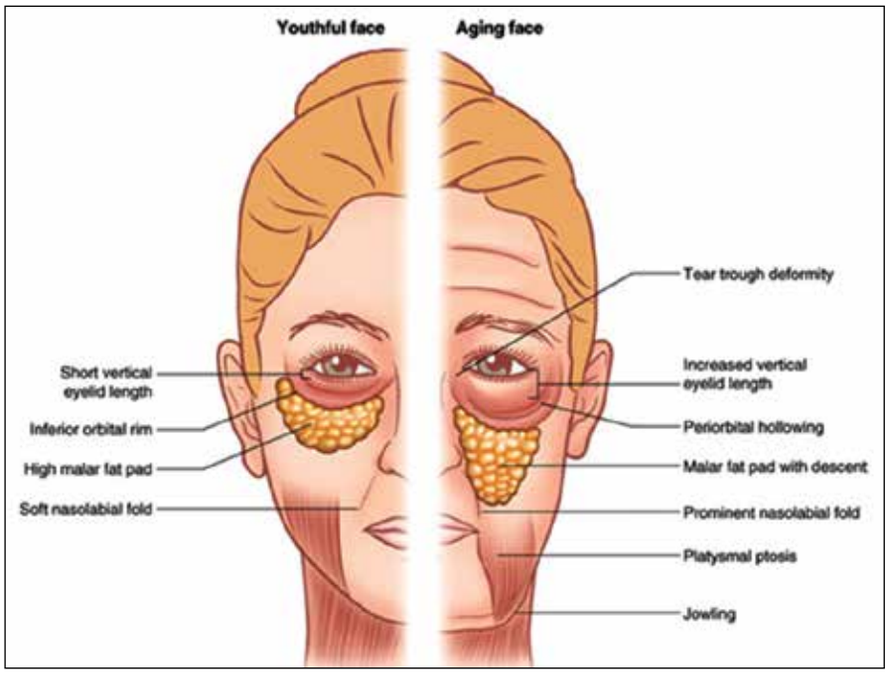

Figure 9: *In youth, the malar fat pad overlies the malar eminence. With age, malar fat pad descends, baring the inferior orbital rim. Vertical length of lower lid appears to increase. Ptosis is also evident in the brow and lower face. Wulc. AE et al.*

The retaining ligaments of the face are important in understanding the concept of facial aging and rejuvenation. These are strong fibrous attachments that originate from the periosteum and travel perpendicularly through fibrous layers to insert onto the dermis. They anchor and support the skin and superficial musculoaponeurotic system (SMAS) to the underlying bone[7]. There are two important ligaments in the periorbital region that are responsible for the curves and grooves that are so characteristic of aging, the orbicularis retaining ligament and the zygomaticofacial ligaments (Figure 10).

The trademark hollow that appears between the lower lid and upper cheek with age is called the orbitomalar sulcus (Figure 11). This hollow represents the attachment of the orbicularis retaining ligament (ORL) laterally and centrally and the insertion of the orbicularis muscle medially. (The medial third of

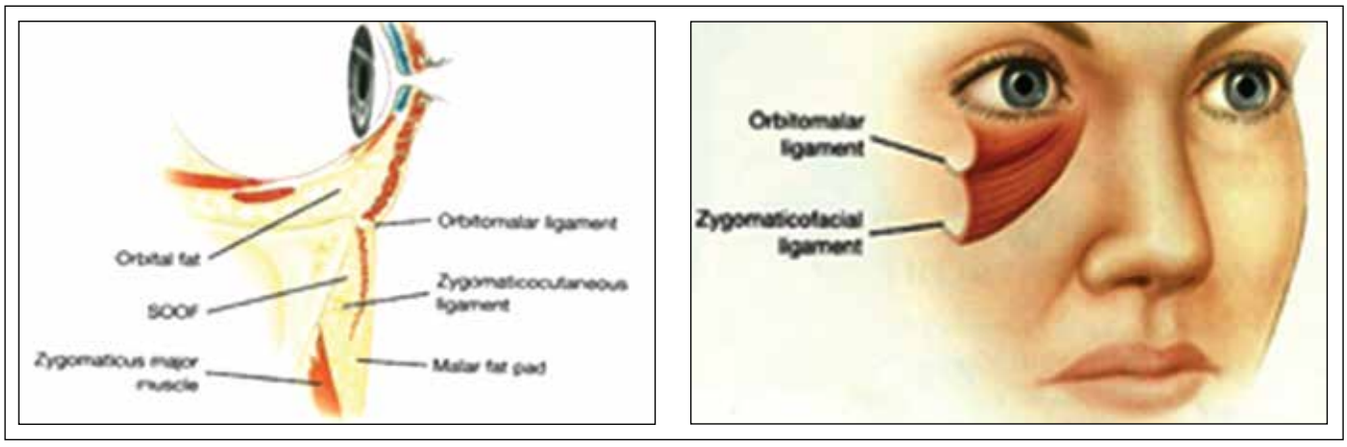

Figure 10: *The normal position of the orbital malar and zygomaticofacial ligaments supporting the soft tissue of the midcheek. When lax, they allow the soft tissues to descend, creating a groove where the ligament is attached to the skin and a bulge in the area around it. Codner M. McCord C[21].*

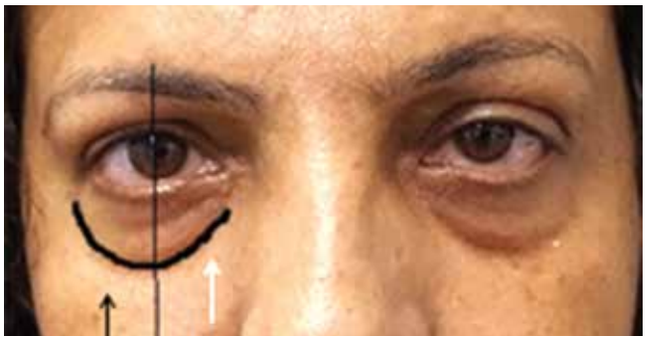

Figure 11: *A 40-year-old woman with prominent orbitomalar sulcus (black line), which is a combination of a tear-trough deformity (white arrow) and a prominent lid-cheek junction (black arrow). It represents the attachments of the orbicularis muscle and ORL respectively.*

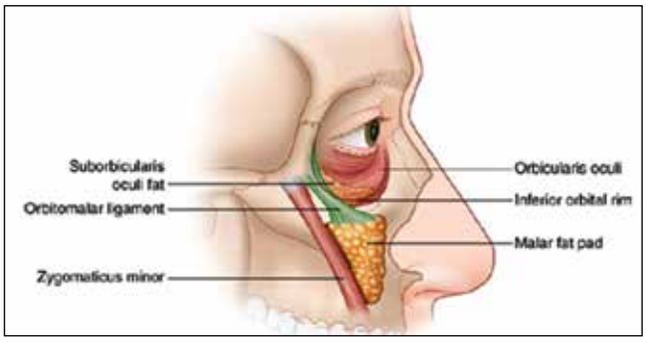

Figure 12: *With aging, laxity of the orbitomalar ligament (green) leads to inferior descent of SOOF and skeletonization of inferior orbital rim. KS Tan et al.*

demarcation and has a uniform smooth convex contour. As we age, changes that occur combine to create a vertically elongated lower lid, ptotic malar/cheek complex, a lax lower eyelid and protrusion of orbital fat[6] (Figure 9). Each of these changes can be attributed to specific changes in the retaining ligaments and soft tissues and need to be understood if we aim to reverse them.

the orbitomalar sulcus is called the Tear Trough). The ORL originates 4-6 mm below the inferior orbital rim , traverses through the Orbicularis Oculi Muscle (OOM) to insert in the dermis at the

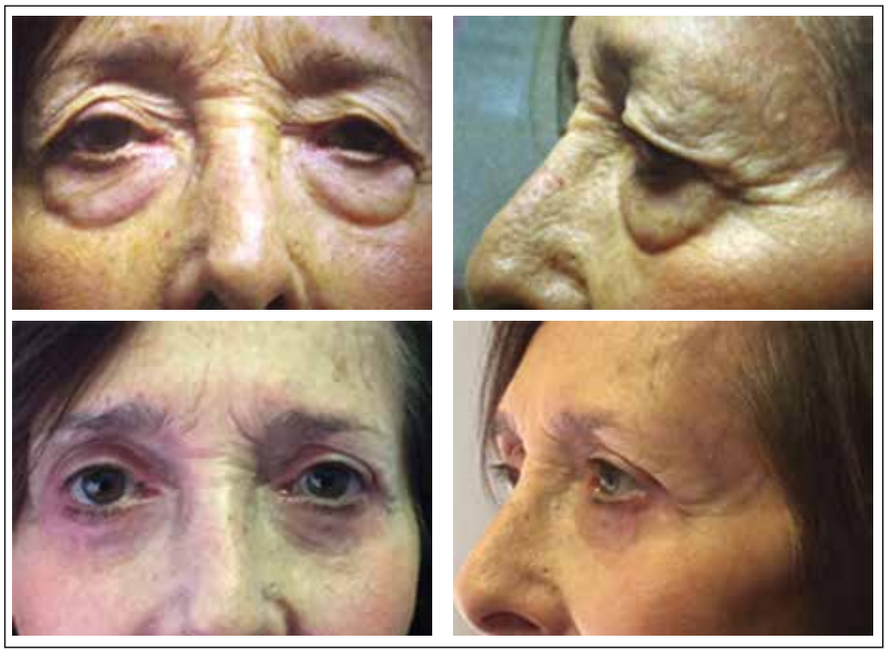

Figure 13: *This 72-year-old woman is an excellent example of a patient with significant periorbital fat pseudoherniation (above); her lower blepharoplasty involved resection of periorbital fat with excision of redundant skin with midface contouring with SOOF lift. The postoperative photographs (below) demonstrate a smooth lid-cheek junction and no residual bulging of fat in the lower eyelids. Her upper blepharoplasty was combined with internal browpexy.*

junction of the lower lid and cheek. As the ligament becomes attenuated and lax with age, tethering of the skin to the zygoma and maxilla just inferior to the orbital rim creates the sulcus which is further accentuated by the orbital fat prolapse above and descent and atrophy of malar fat pad below (Figure 12).

Bone loss in the malar and periorbital region contributes to the overall volume loss[8]. The orbital septum weakens, orbicularis atrophies and the skin becomes lax allowing pseudoherniation of the orbital fat outside the orbital rim. There is also some increase in the actual volume of orbital fat with age[9] which suggests that in many patients the removal of a judicious amount of lower lid fat is warranted.

APPROACHES TO LOWER LID REJUVENATION

Numerous techniques have been described in literature to perform the procedure with no evidence-based consensus regarding the "ideal" approach to lower lid blepharoplasty[10,11,12,13]. Each patient has a different presentation with varying amounts of orbital fat prolapse, skin and orbicularis redundancy, lower lid margin and canthal tendon laxity, cheek descent and volume loss, globe prominence, orbitomalar sulcus formation and midface projection or retrusion. Therefore one single procedure or technique cannot be effective in all the patients. The surgeon should be able to analyse the aging midface and customize the procedure for each patient. This is what makes lower blepharoplasty a challenging procedure to learn.

There are two main approaches to lower lid blepharoplasty.

- Transcutaneous approach
- Transconjunctival approach

The earliest blepharoplasties were performed by the transcutaneous approach using a subciliary incision[14]. The pretarsal orbicularis muscle was violated in this approach which compromised tarsal support leading to rounding of lateral canthus and increased lateral scleral show in many cases.

In 1973, Tessier[15] described the transconjunctival incision for removal of fat which has gained wide acceptance in lower lid rejuvenation Loeb[16,17].

is credited with introducing the concept of repositioning orbital fat along the medial infraorbital rim to address the tear trough deformity. Since then many authors have described a myriad variations in technique to reposition lower lid fat subperiosteally or supraperiosteally through various approaches and anchoring techniques.

Most favoured current technique of lower blepharoplasty favours a transconjunctival incision with a conservative excision of prolapsed fat in the lateral and central compartments and conservation with transposition of fat in the medial compartment. Overresection of fat is avoided as it produces a hollowed look which is less youthful and more skeletonized.

Pinch technique is used to excise excess skin via a subciliary incision removing skin only without disturbing the orbicularis oculi muscle.

This is accompanied by a suspension or lifting procedure for contour correction and effacement of the lid-cheek junction often with elevation of the descended or deflated malar fat pad while simultaneously adequately tightening the lower lid.

The most common suspension procedures performed include orbicularis suspension, Sub Orbicularis Oculi Fat (SOOF) lift and subperiosteal or supraperiosteal midface lift (Figure 13).

A detailed description of each of these procedures is beyond the scope of this article.

SURGICAL TECHNIQUE OF TRANSCONJUNCTIVAL BLEPHAROPLASTY

Placement of transconjunctival incision varies in different reports but most prefer it midway between the lower border of tarsus and fornix. Once conjunctiva has been incised, further dissection is carried out through the retractors until fat is seen. The three fat pads are identified and dissected free. A mild pressure applied posteriorly on the globe by the assistant helps in making identification of the fat pads easier.

Care is taken while dissecting medially to avoid injury to the inferior oblique muscle which traverses between the nasal and the central fat pad.

The temporal herniated orbital fat is isolated and the fat that prolapses outside the orbital rim with gentle pressure is clamped with a hemostat, cut and stump cauterised to ensure complete hemostasis.

If fat redraping is to be performed, a preperiosteal dissection is performed along the arcus marginalis to expose the inferior orbital rim. The medial or both medial and central fat pads depending on the case, are dissected free, pedicles created and redraped in the preperiosteal pocket created earlier.

6-0 vicryl suture passed through the centre of the pedicle in a horizontal mattress fashion is then passed horizontally several millimetres below

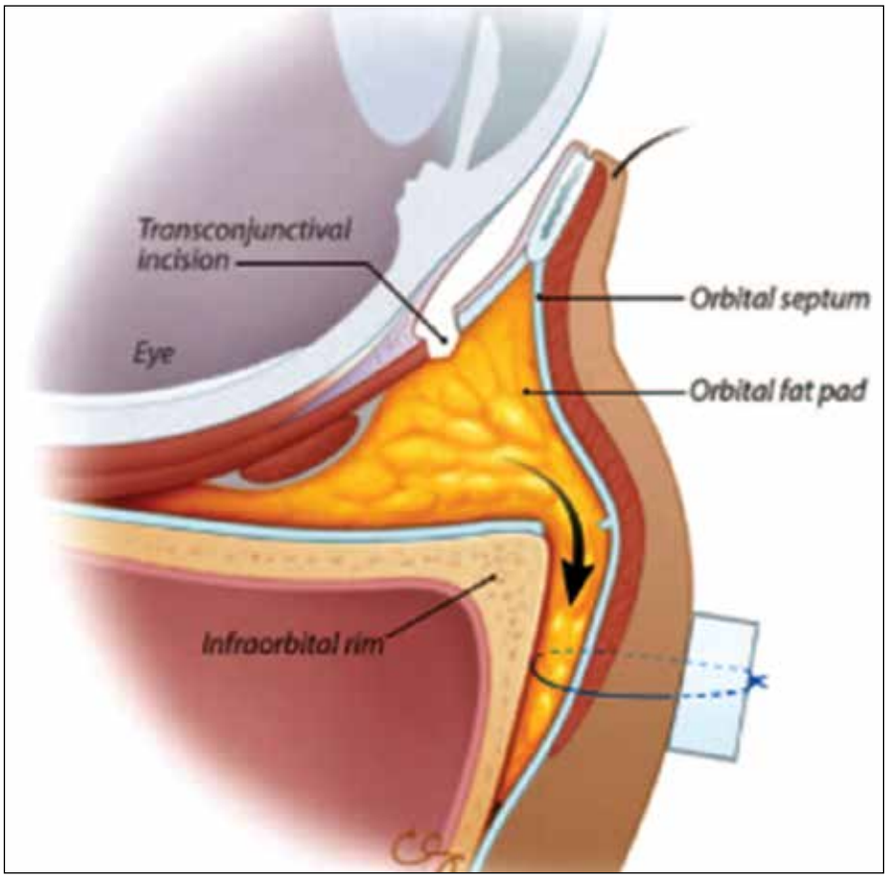

Figure 14: *Illustration of the lower lid and periorbital structures. Sagittal view of a subperiosteally repositioned fat pedicle below the infraorbital rim that is secured in place with a percutaneous bolster. From : Zoumalan et al.*

the arcus marginalis on the inferior orbital rim. To fill a deep tear trough, the suture passed through the nasal fat pedicle is exteriorized to keep the pedicle in place and tied over a bolster to be removed after a week (Figure 14). Ensure that the orbital septum and inferior oblique are not tethered to the orbital rim.

The suspension procedure is performed as planned.

The lower lid is then reassessed for any laxity which must be corrected by lateral canthopexy or canthoplasty depending on the extent of laxity to prevent eyelid malposition.

Excessive redundant skin is excised by the pinch technique using a subciliary incision elevating a skin only flap. This is done in a conservative and tension free manner to avoid shortening the anterior lamella. The skin excision is greatest at the lateral canthus and gradually tapers as it progresses medially.

POSTOPERATIVE CARE

Patients are instructed to use cool packs to the affected area for the first 24 hours to minimize swelling. Severe pain is unusual following a blepharoplasty, and patients should be evaluated immediately to rule out retrobulbar hematoma in cases of severe pain and/or vision changes. Head position is usually maintained at or above the heart level to reduce edema. Antibiotic drops with or without a steroid component are used in cases where a conjunctival incision is made in the first week. Aggressive corneal lubrication achieved with eye drops and ointment. Sutures are removed, usually on postoperative days 5 to 7. Most of the swelling usually subsides in 2 weeks after surgery but occasionally may last longer.

COMPLICATIONS

The most dreaded early complication is orbital hemorrhage, which must be identified and treated immediately, as this can result in permanent vision loss and even blindness[22]. If vision is threatened, immediate treatment should be provided by starting IOP lowering medications and exploration of the wound and/or lateral canthotomy/cantholysis to help reduce orbital pressure.

Intermediate- and long-term complications include dry eyes, lower lid malposition, lagophthalmos, ptosis.

Most of these complications can be avoided with careful preoperative planning and appropriate surgical technique.

Lid malposition is one of the more feared complications of the lower lid blepharoplasty and frequently requires surgical management.

CONCLUSION

The techniques of blepharoplasty are evolving to reflect the concept that eyelid rejuvenation is not achieved by just excision of redundant skin and excess fat. The goal is greater volume preservation and volume augmentation. Brow restoration and volume enhancement is an integral part of upper lid blepharoplasty. Lower lid blepharoplasty can use a transcutaneous or a transconjunctival approach to address herniated fat pads while blending the lid-cheek junction through release of the orbitomalar ligament and volume augmentation with fat (by repositioning and/or grafting) or injectable fillers. Through an algorithmic approach that meets the needs of each individual patient, the approach to blepharoplasty is customized with consistent and predictable results.

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Thyroid Eye Disease-an Update

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Thyroid eye disease (TED), also called Graves' orbitopathy (GO) is a potentially sight-threatening ocular disease, mostly occurring in patients with hyperthyroidism or a history of hyperthyroidism due to Graves' disease (GD). However, it can occur in patients with euthyroid or hypothyroid chronic autoimmune thyroiditis as well and about 5-10% of patients with TED are euthyroid at presentation[1]. Thyroid eye disease is the most common cause of both bilateral and unilateral proptosis in an adult and is the most common orbital pathology in adults.

DEMOGRAPHICS

Prevalence of thyroid eye disease ranges from 20 to 42 % among patients of Graves' disease. However, the reported prevalence in various series shows a wide variability because of the selection bias, lack of standardized ocular assessment criteria, non-uniform definition of Graves' orbitopathy etc. A prevalence of 34.7% has been reported in the in the Asian population and 28% prevalence has been reported in Indian studies among the Graves' disease patients[2,3]. Females are more commonly affected than males. However, for severe disease, this ratio reverses and severe thyroid eye disease is approximately 4 times more common in males than females[4].

Etiology and pathogenesis

Thyroid eye disease is strongly associated with autoimmune thyroid diseases such as Graves' disease (90%) or Hashimoto's thyroiditis. Majority of patients with thyroid eye disease develops eye symptoms within 18 months of onset of the autoimmune thyroid disease. However, in 13% patients the ocular symptoms might present 2 years after the diagnosis and in 3% the diagnosis can precede the onset of GD by more than 12 months[5]. A fraction of these patients may also present with skin involvement in the form of pretibial myxedema and thyroid acropachy suggesting a single underlying systemic process.

Our understanding of the pathogenesis of TED is still incomplete. Pathological changes of the thyroid orbitopathy appear to involve both the extraocular muscles and the orbital fat, with most patients having a combination of extraocular muscle enlargement and orbital fat expansion[6]. Histopathological studies have found extensive deposition of hyaluronan in between the extraocular muscles, mixed inflammatory infiltrate and abundance of cytokines causing interstitial edema and soft tissue expansion and proptosis[7]. Recent studies also support the role of Insulin like growth factor 1(IGF-1) receptor in the pathogenesis of TED[8].

There exists substantial evidence to suggest that the principle cell involved in the pathogenesis of TED is the orbital fibroblast[9]. Activation of orbital fibroblasts leads to proliferation, hyaluronan secretion and soft tissue expansion. The proposed mechanism of fibroblast activation is centred around the fact that the orbit contains two subpopulations of fibroblasts, Thy1/CD90 surface marker positive and Thy1/CD90 negative fibroblasts, which have different structures and functions[10,11]. The process of activation of the orbital fibroblast and consequent mechanism of muscle or fat expansion is summarised in (Figure 1). The relative proportion of the activated Thy1 positive or negative fibroblasts determine whether fibrosis or adipogenesis predominates.

Orbital fibroblasts can be activated in TED in both an antigen dependent and antigen independent manner. The two significant autoantigens involved are the thyrotropin (TSHR) and IGF-1 and orbital fibroblasts have robust expression of receptor to these autoantigens. Autoantibodies against TSHR can be detected in up to 98% of patients with TED and has an

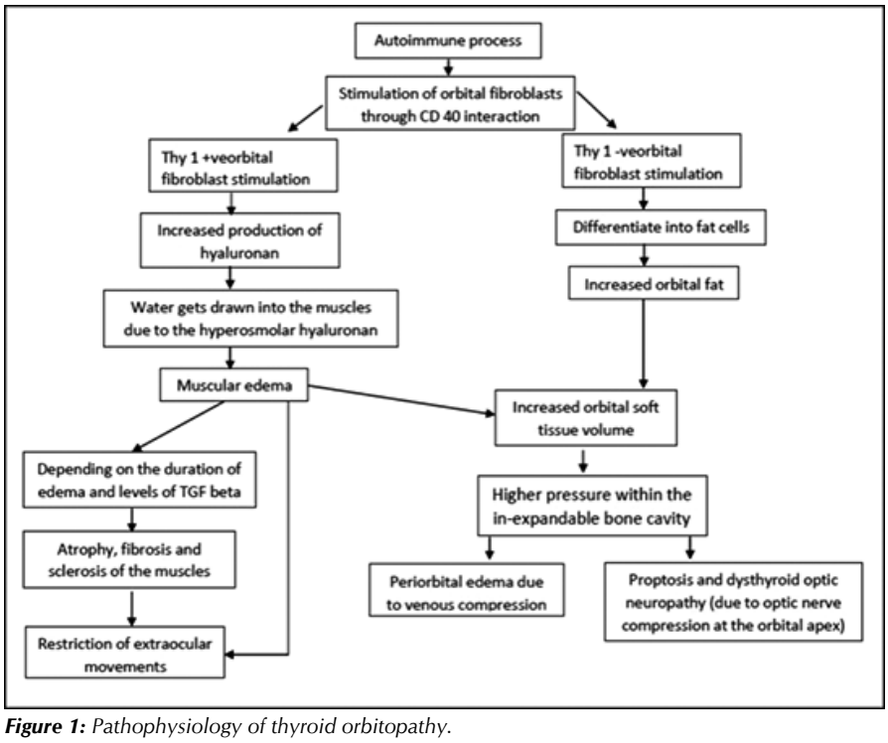

Figure 1: *Pathophysiology of thyroid orbitopathy.*

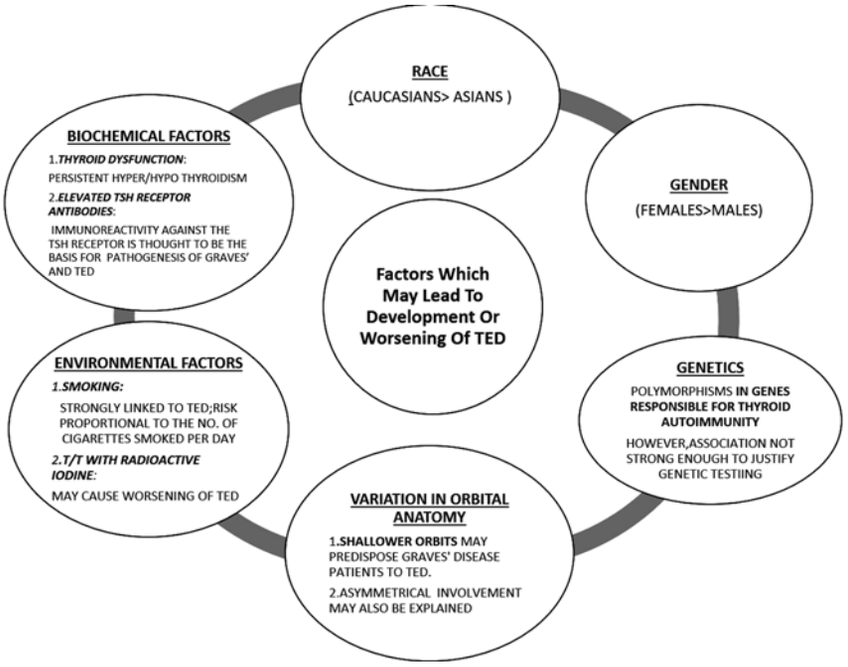

Figure 2: *Risk factors for onset and progression of thyroid eye disease.*

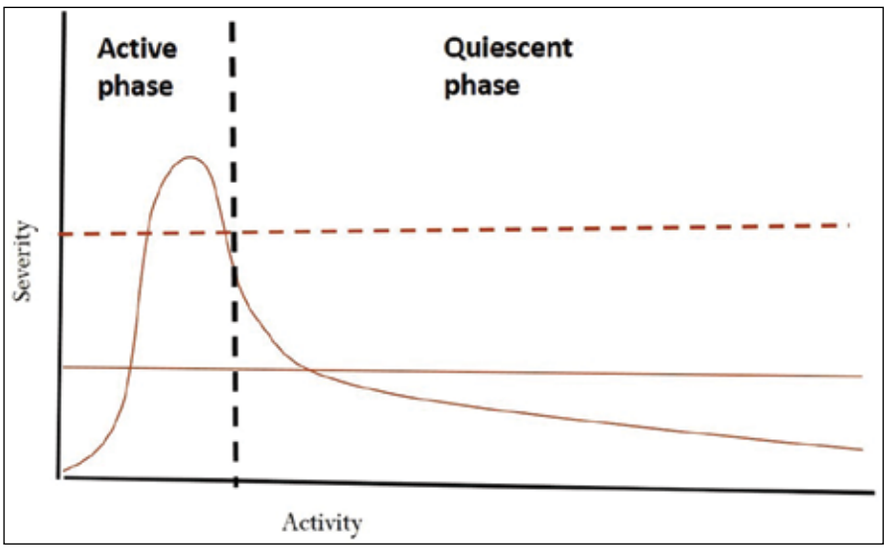

Figure 3: *Rundle's curve showing the natural course of progression and severity of thyroid eye disease.*

established role in the pathogenesis of TED[12]. Also TSHR expression is higher in patients with active disease as compared to inactive disease and the level of this autoantibody can correlate with disease activity and severity[13].

IGF-1R is another potential autoantigen in the pathogenesis of TED and is overexpressed in TED orbital fibroblasts as compared to controls[14]. Activated fibroblasts also secrete potential T-cell chemoattractants, IL16 and CCL5, facilitating the recruitment of T lymphocytes into the orbit which activates orbital fibroblasts leading to secretion of prostaglandins and proinflammatory cytokines[15]. These T – cell mediated events eventually results in soft tissue remodelling and inflammatory events in TED. Long lasting edema leads to atrophy, fibrosis and sclerosis of the extraocular muscles subsequently leading to restrictive strabismus.

Risk factors for development and progression of TED

These risk factors for development of TED have been summarised in (Figure 2)[16].

Clinical course of thyroid eye disease: the Rundle's curve

A unique feature of TED as compared to other autoimmune disease is that it is a self-limiting disease. TED follows a biphasic course. There is a progressive or active phase lasting 6–18 months followed by a stable or inactive phase. This pattern was first described by Rundle, who plotted a graph of orbital disease severity against time (Rundle's curve) and can be plotted graphically for all patients[17]. The steepness of the graph in the active phase reflects the acuity of progression, with a steeper slope often leading to more severe disease (Figure 3).

Clinical features of thyroid eye disease

Appearance and exposure

- Upper eyelid retraction:

Upper lid retraction is present when the lid margin lies at the superior limbus or higher, exposing the sclera. The retraction is more marked laterally causing the characteristic lateral flare and may fluctuate with emotion or fixation (Dalrymple's sign) giving the patient an angry look (Figure 4a). It is associated with lid lag on downgaze (Von Graefe's sign), apparent spasmodic lid overaction on upgaze (Kocher's sign) and incomplete lid closure while asleep (Figure 4b).

Proposed mechanisms of eyelid retraction include increased circulating catecholamines, overaction of the levator palpebrae superioris and superior rectus muscles to compensate for inferior rectus restriction, or inflammation and scarring of the levator complex.

Lower eyelid retraction

Lower lid retraction is present when sclera is visible inferiorly and occurs more frequently in patients of Asian origin[18].

Proptosis

This is the second most common finding in GO following eyelid retraction (Figure 4c). Expansion of the orbital fat and/or muscles limited anteriorly by the relatively tight eyelid tarsoligamentous diaphragm causes the proptosis.

Corneal exposure

Corneal exposure can occur secondary to lid retraction and proptosis and can manifest as irritation, photophobia, watering and blurred vision, corneal punctate epithelial erosions, and frank abrasions or in severe cases, ulcerations and corneal perforation.

Periorbital soft tissue inflammation and congestion:

Symptoms and signs of periorbital soft tissue inflammation include orbital ache at rest or with movement, conjunctival and caruncular injection and oedema, eyelid redness and oedema, and diurnal variation (worse with the head dependent after sleep). These are considered a clinical indicator of disease activity (Figure 4d).

Ocular motility disruption and strabismus

Although the levator muscle is commonly involved in GO resulting in upper lid retraction in over 80% of patients, the extraocular muscles become clinically involved in only 25%, often in older population. Inferior rectus is the most common muscle involved. During the active inflammatory phase, progressive restriction of motility develops, initially intermittently or with gaze. In the post-inflammatory phase, muscle atrophy, fibrosis or sclerosis may result in disabling diplopia in the primary gaze, which may be constant.

Dysthyroid optic neuropathy (DON)

Potentially reversible optic nerve dysfunction seen in 4-8% of all cases of GO caused by direct compression of the nerve by swollen muscles at the orbital apex, presumably impairing axoplasmic flow[19]. Symptoms include colour desaturation and blurring of central vision. An afferent pupillary defect is a specific sign of DON but is not detected in 35% of patients, often because of symmetric loss of vision[20]. Likewise, disc oedema is a specific sign when present, but is absent in over 50% of patients with DON. Visual evoked potential is the most significant clinical test for detection of DON and has been found abnormal in 18% of patients with DON who had good visual acuity. Patients who develop DON are more likely to be male, older, and diabetic compared with their non-DON counterparts. DON may occur in the absence of significant proptosis, usually in Asian population due to a shallower orbital cavity.

Assessment of the clinical features and grading of the disease:

Several classification systems have been used to assess the severity and activity of the clinical manifestations of TED.

1. NOSPECS Classification:

In 1969, Werner reported the NOSPECS Classification (No physical signs or symptoms, Only signs, Soft tissue involvement, Proptosis, Extraocular muscle signs, Corneal involvement, and Sight loss) which he modified and published again in 1977[21,22] (Table 1). It only graded the severity of disease and did not distinguish active and inactive disease. Hence, the indication for treatments used to be based exclusively on the severity of symptoms instead of the activity.

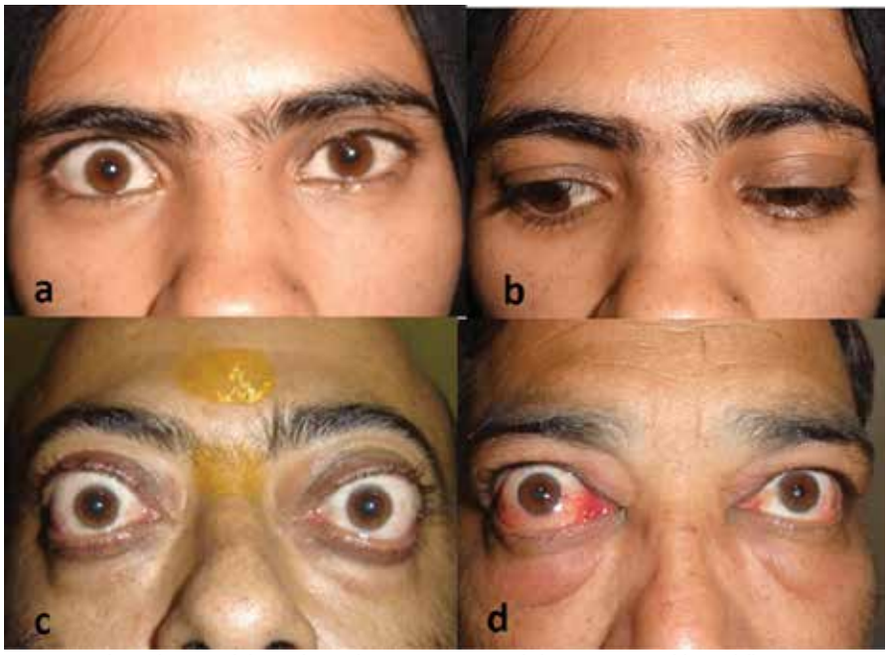

Figure 4: *Clinical signs of thyroid eye disease. Upper eyelid retraction and lateral flare (Figure 4a) and lid lad on downgaze (Figure 4b) are the pathognomic findings in TED. Other findings include proptosis (Figure 4c) and signs of inflammation (Figure 4d) in patients with active disease.*

Table 1: Modified NOSPECS classification of Thyroid Eye disease

NO SPECS modified classification	
Class	**Suggestions for grading**
0	No physical signs or symptoms
I	Only signs
II	Soft tissue involvement 0 Absent (a) Minimal (b) Moderate (c) Marked
III	Proptosis (3 mm or more of normal upper limits with or without symptoms) 0 Absent (a) 3 or 4 mm over upper normal (b) 5 to 7 mm increase (c) 8 mm increase
IV	Extraocular muscle involvement (usually with diplopia) 0 Absent (a) Limitation of motion at extremes of gaze (b) Evident restriction of motion (c) Fixation of a globe or globes
V	Corneal involvement (primarily due to lagophthalmos) 0 Absent (a) Stippling of cornea (b) Ulceration (c) Clouding, necrosis, and perforation
VI	Sight loss (due to optic nerve involvement) 0 Absent (a) Disc pallor or choking, or visual field defect, vision 20/20–20/60 (b) The same, but vision 20/70–20/200 (c) Blindness, vision less than 20/200

2. Clinical Activity Score (CAS)

In 1989, Mourits et al. described the Clinical Activity Score (CAS), which was modified in 1997[23,24]. The scoring system is based on the classical signs of acute inflammation (pain, redness, swelling,

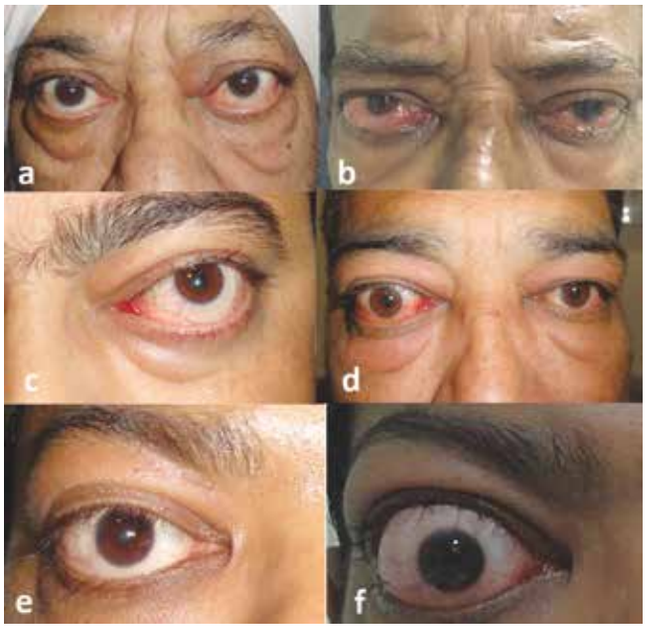

Figure 5: *Signs of clinical activity. Conjunctival congestion (Figure 5a), conjunctival chemosis (Figure 5b), eyelid edema (Figure 5c), eyelid erythema and caruncular inflammation (Figure 5d), increase in proptosis by more than 2mm during the follow up evaluation (Figure 5e,5f).*

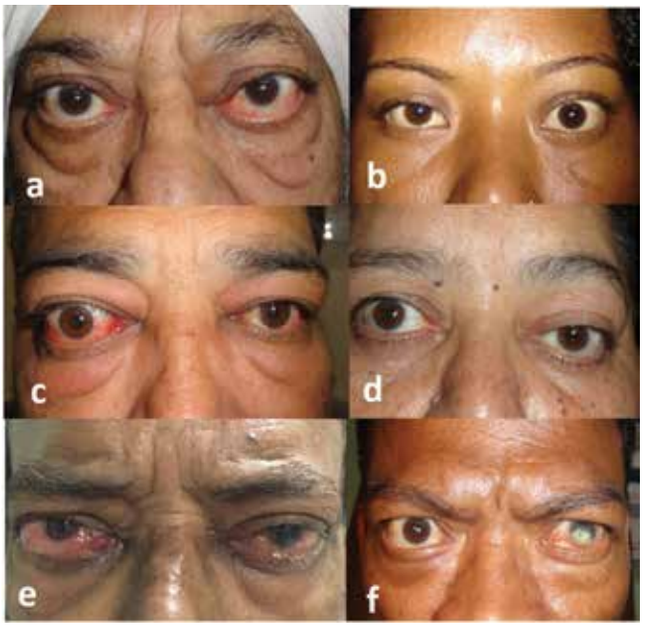

Figure 6: *Grading of the severity of the disease in the active and inactive phase. Mild disease with minimal signs of activity (Figure 6a) and mild eyelid retraction (Figure 6b) and no activity. Moderate to severe disease with signs of activity (Figure 6c) and no signs of activity (Figure 6d). Sight threatening disease with compressive optic neuropathy in active phase (Figure 6e) and corneal exposure due to severe lagophthalmos in inactive phase (Figure 6f).*

and impaired function). This classification system attempts to differentiate active from quiet disease. One point is given for the presence of each of the parameters assessed (Figure 5). The sum of all points defines clinical activity: active ophthalmopathy if the score is above 3/7 at the first examination or above 4/10 in successive examinations (Table 2).

The currently used grading systems used for the assessment of TED are:

- VISA Classification (vision, inflammation, strabismus, and appearance)
- European Group of Graves' Orbitopathy (EUGOGO) Classification

Both the grading systems are based on the NOSPECS and CAS classification system and uses indicators to assess the signs of activity and the degree of severity. Importantly, they allow the clinician to plan and assess the treatment response of patients with GO. The classification systems are not interchangeable and only one classification system should be followed for an individual patient.

Table 2: Clinical activity scoring system

CLINICAL ACTIVITY SCORE (CAS) (EUGOGO)
For initial CAS, only score items 1–7 1. Spontaneous orbital pain 2. Gaze evoked orbital pain 3. Eyelid swelling that is considered to be due to active GO 4. Eyelid erythema 5. Conjunctival redness that is considered to be due to active GO 6. Chemosis 7. Inflammation of caruncle OR plica **Patients assessed after follow-up (1–3 months) can be scored out of 10 by including items 8–10** 8. Increase of >2mm in proptosis 9. Decrease in uniocular ocular excursion in any one direction of >8∘ 10. Decrease of acuity equivalent to 1 Snellen line

VISA classification

The VISA classification system is based on four severity parameters, vision (V), inflammation (I), strabismus(S) and appearance (A) and is graded independently 25. A global severity score is the sum of score of each parametres graded independently. The score for each parameter is as follows, vision: 1 point, inflammatory score: 10 points, strabismus and diplopia: 6 points and appearance: 3 points (Table 3). The maximum score can be 20. The first visit and follow up assessment forms for patients with GO has been designed based on the VISA classification and has been adopted by International thyroid eye disease society (ITEDS, www.thyroideyedisease.org).

EUGOGO classification

The EUGOGO classification is also based on the disease activity and severity parameters[26]. For grading activity, the modified clinical activity scoring system is used. The severity parameters are graded based on comparison with an image atlas that has been developed by the group. A classification system to guide the management of patients with GO have also has been developed based on the impact of the disease on the quality of life and the risk of vision loss and helps in deciding on the management strategy[27] (Table 4) (Figure 6).

Approach to diagnosis

The diagnosis of thyroid eye disease is mainly clinical based on assessment of symptoms and signs. Investigations are directed to assess the systemic thyroid

Table 3: VISA INFLAMMATORY INDEX (ITEDS) scoring system

	Symptoms	Clinical evaluation	Scoring
Vision	Blurred vision Colour desaturation	Visual acuity Colour Vision Afferent pupillary defect Optic nerve evaluation Visual field testing Visually evoked potential	DON absent: 0 DON present:1
Inflammation	Caruncular edema Chemosis Conjunctival redness Lid redness Lid edema Retrobulbar ache at rest With Gaze Diurnal variation	External evaluation and slit lamp examination	0: absent 1: present 0: absent 1: conjunctiva lies behind the grey line of the lid 2: conjunctiva extends anterior to the grey line of the lid 0: absent 1: present 0: absent 1: present 0: absent 1: present but without redundant tissues 2: present and causing bulging in the palpebral skin, including lower lid festoon. 0: absent; 1: present 0: absent; 1: present 0: absent; 1: present
Strabismus and diplopia	Diplopia	Ocular motility Cover test Head posture Diplopia charting Field of binocular single vision	0:Absent diplopia 1:Diplopia on horizontal or vertical gazes 2: Intermittent diplopia in straight gaze 3: Constant diplopia in straight gaze.
Appearance	Appearance concern like bulging eyes, lid retraction, fat bags. Exposure symptoms like photophobia, grittiness	External examination Slit lamp evaluation	

Table 4: EUGOGO Classification of the Severity of the Ophthalmopathy

(1) ***Mild:*** Minimum impact on the patient's life.One or more of the following signs:
- Minor lid retraction (<2 mm).
- Mild soft tissue involvement.
- Exophthalmos <3mm(above the normal range for the race and gender).
- Transient or no diplopia.
- Corneal exposure responsive to lubricants.

(2) ***Moderate to severe:*** patients without sight-threatening GO whose eye disease has sufficient impact on daily life to justify the risks of immunosuppression (if active) or surgical intervention (if inactive). Patients usually present one or more of the following signs:
- Lid retraction (>2 mm).
- Moderate or severe soft tissue involvement.
- Exophthalmos ≥3mm(above the normal range for the race and gender).
- Inconstant, or constant diplopia.

(3) ***Sight-threatening GO:*** patients with dysthyroid optic neuropathy or corneal breakdown due to severe exposure. Other infrequent cases are ocular globe subluxation, severe forms of frozen eye, choroidal folds, and postural visual obscuration. This category warrants immediate intervention.

status and CT scan or MRI is indicated in cases of DON and for orbital bone assessment prior to orbital surgery.

INVESTIGATIONS

1. Thyroid function tests (free T3, free T4, and TSH) to know whether the patient is euthyroid, hypothyroid or hyperthyroid
2. Thyroid specific antibodies (anti-thyroglobulin, anti-thyroid peroxidase and anti-TSH receptor) which may support the diagnosis, but may be negative, especially in late disease. The level of thyroid stimulating immunoglobulin (TSI) correlates with the development of ophthalmopathy in patients with Graves' disease.

- Orbital imaging helps in supporting

the diagnosis and may not be indicated if the clinical features are sufficient to arrive at the diagnosis. Non contrast CT Scan is indicated in the following situations (Figure 7):

- To identify orbital apical crowding in cases of optic neuropathy
- To know the status of the surrounding bone and sinuses prior to decompression surgery.
- For measurement of orbital fat, lacrimal gland and individual extraocular muscles when clinical diagnosis is not conclusive.
- Sequential scanning permits an assessment of natural progression or response to therapy.

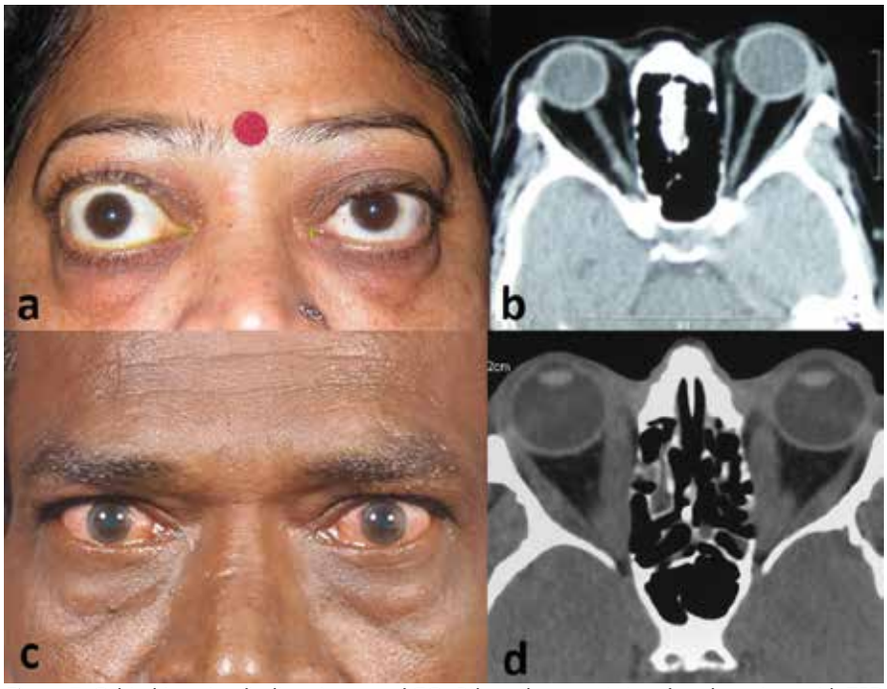

Figure 7: *Orbital imaging findings in case of GO. Bilateral proptosis in a female patient with TED (Figure 7a) and CT scan showing increased orbital fat spaces and stretching of the optic nerve with minimal extraocular muscle thickening suggesting a predominantly fat disease (Figure 7b). Eyelid retraction and severe extraocular movement restriction and optic neuropathy with minimal proptosis in a male patient with TED (Figure 7c) and the corresponding CT scan showing massive enlargement of extraocular muscles causing crowding of the orbital apex in a predominantly muscle disease (Figure 7d).*

MANAGEMENT

The management of thyroid eye disease needs a multidisciplinaray approach. A patient centred approach to treatment is recommended which takes into account the effect of the disease and the treatment on the quality of life and psychosocial well being of the patient.

Treatment of thyroid dysfunction

Since persistence of thyroid dysfunction is a major risk factor for worsening of TED, it is important to make the patient euthyroid. Underlying thyroid dysfunction can be treated by:

- Medical intervention (propylthiouracil, carbimazole, thyroxine, radioactive iodine)
- Surgical intervention (thyroidectomy)
- Radioactive iodine therapy (RAI). Caution must be exercised while advising RAI since it can worsen TED. Hence patients must be screened before starting RAI therapy for the risk factors for worsening of orbitopathy like recent onset hyperthyroidism, severe hyperthyroidism, active Graves's orbitopathy, high serum TSH or TRAb levels or cigarette smoking. Concomitant oral steroids are started few days prior to the RAI therapy if risk factors are present[28].

Risk factor modification

Tobacco smoking is the strongest modifiable risk factor for the progression of orbitopathy. Association between smoking and thyroid eye disease is well established including the following observations:[29,30,31]

i) Smokers tend to have a more severe ophthalmopathy.
ii) Smokers are more likely to show progression or development of ophthalmopathy after RAI therapy for hyperthyroidism
iii) Cessation of smoking is associated with a better outcome of GO.
iv) Smoking delays or worsens the outcome of immunosuppressive therapy for ophthalmopathy

Treatment of Ophthalmopathy:

All patients with TED should be assessed for activity and severity as par the standardised criteria and categorised into active or inactive or mild, moderate to severe or sight threatening ophthalmopathy.

General measures

General Measures advised to all patients with TED should include preservative free topical lubricants, moisture goggles, smoking cessation and nocturnal head elevation. Increased tear osmolality is the main component of dry eye in these patients. Lacrimal gland expresses TSH receptors and circulating TSHR antibodies can bind and contribute to the lacrimal gland impairment, leading to secondary Sjogren's syndrome in long standing cases[32]. Hence, preservative free artificial tears with long retention time like sodium hyaluronate and those with osmoprotective action should be prescribed frequently to protect the ocular surface.

Selenium supplementation

Selenium supplementation have shown to improve the clinical activity score in patients with mild active TED, improve quality of life and slow the disease progression in TED patients with mild GO[33]. However, there is an increased risk of type II diabetes with higher doses.

Specific management

Treatment of TED depends on the activity and severity of the disease. Patients with a VISA score of >4/10 or a CAS score of >3/7 is considered to be active disease. An algorithm for the management of TED is provided in (Figure 8).

Active Phase Disease Management

The aim of the active phase management is to reduce the risk of sight threatening complications and reduce the severe manifestation of the disease until the disease activity dies down. Mild disease can be managed by

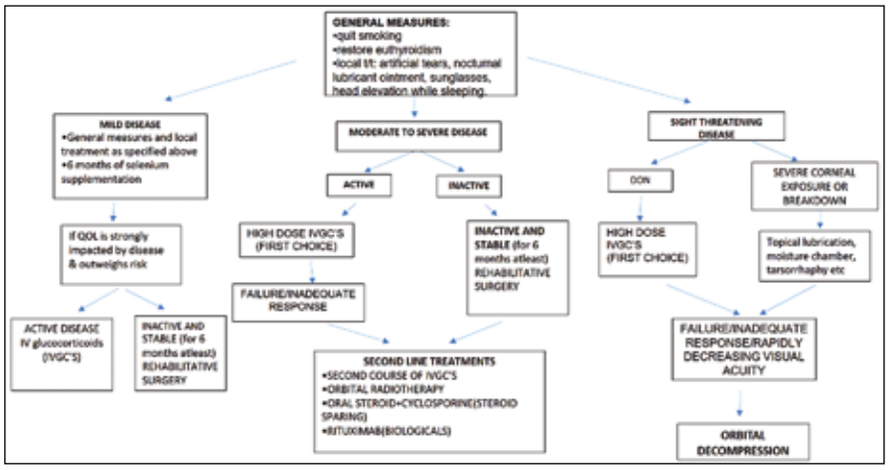

Figure 8: *Algorithm for management of TED.*

supportive care including ocular surface lubrication, moisture goggles, and nocturnal head elevation. Moderate to severe disease requires treatment with anti-inflammatory medications.

1. Anti-inflammatory treatment

a) Corticosteroids:

Corticosteroids remain the first line anti-inflammatory therapy. It can be administered orally, intravenously (IV) or as local steroid injection into orbital soft tissues. Though IV steroids appears to be more efficacious and better tolerated than oral steroids, a recent survey of members of the American society of ophthalmic plastic and reconstructive surgery have found a preference for use of oral corticosteroids[34]. For moderate to severe active disease, IV methylprednisolone is prescribed at a dose of 0.5gm weekly for 6 weeks followed by 0.25g weekly for 6 more weeks for a cumulative total dose of 4.5gm[35]. High dose regimen is reserved for sight threatening GO and the total cumulative dose should not exceed 8gm to avoid the side effects like liver damage[36]. Safety data also suggest that single dose of IVMP should not exceed 0.75gm and consecutive day dosing should be avoided. Concomitant administration of proton pump inhibitors to prevent peptic ulcers and calcium and Vitamin D supplementation is recommended especially in patients who are at high risk for osteoporosis.

b) Orbital glucocorticoid injection:

Periocular or subconjunctival steroid injection is less effective than oral or IV steroids but can be considered when oral or IV steroids are contraindicated. Triamcinolone 20mg injected in the inferotemporal quadrant of the orbit at 4 weekly intervals has shown improvement in diplopia and significant reduction in the thickness of the extraocular muscles[37].

Steroid sparing agents

Steroid-sparing agents such as azathioprine, methotrexate or cyclosporine may be considered if moderate, no improvement has occurred with systemic steroids, or patient is intolerant to steroids. Combined treatment with oral prednisolone and cyclosporine has been found to provide better outcome and lower recurrence rate of moderate-severe and active GO.

c) Biological agents:

Biological agents like Rituximab, Adalimimab and Teprotumumab has been the subject of research for treatment of thyroid eye disease. Biological agents have been focussed on improving the steroid sparing regimen and can target T cells, B cells, IGF-1 receptor, TSH receptor and various inflammatory cytokines[38].

- Rituximab is a monoclonal antibody against CD20, a transmembrane protein present on the B lymphocytes. Preliminary studies have shown promising result in treatment of moderate to severe active TED with a sustained reduction of the clinical activity score. The optimal dosing ranges from 100 to 1000 mg per infusion for 3 to 4 infusions[39]. Conflicting reports on progression of DON has been reported on patients on treatment with rituximab, especially those with longer duration of the disease. Hence, rituximab should be avoided in patients with impending DON or long standing cases.
- Adalimumab is a monoclonal antibody and TNF-alpha antagonist and is approved for the treatment of rheumatoid arthritis, psoriatic arthritis, ankylosing spondylitis etc. Initial studies have shown promising results in treatment of TED with decrease in ophthalmopathy and steroid tapering possible in some patients[40].
- Teprotumumab is a monoclonal antibody and IGF-1 receptor blocker and is undergoing clinical trial as a treatment modality for TED. IGF-1 receptors are highly expressed in the fibrocytes of TED patients and thus teprotumumab might have a role in the reduction or prevention of TED[41].

2. Orbital Radiotherapy:

Orbital fibroblasts and lymphocytes are sensitive to ionizing radiation. Controversy remains over its benefits and its role in management of TED. However studies have shown an average improvement in VISA score in TED patients treated with orbital radiotherapy[42]. Orbital irradiation (OR) can be considered in patients with active disease who have diplopia or restricted motility and who are intolerant to or not responsive to systemic corticosteroids or biological agents. A cumulative dose of 20Gy fractionated over 10 sessions and administered over 2 weeks period is a commonly used regimen.

3. Surgical intervention:

Surgical intervention is usually avoided in the active phase of the disease as manipulation of the orbital tissues can worsen the orbital inflammation. Exposure keratopathy not amenable lubricants might benefit from tarsorrhaphy. Corneal breakdown might require tissue adhesive or corneal transplantation. Prism prescription might help in restoring fusion in patients with primary position diplopia and small angle squint (usually <15 PD) in the active phase of the disease. Orbital decompression in the active phase is reserved for patients intolerant or non responsive to steroids.

CHRONIC INACTIVE PHASE TREATMENT

Rehabilitative surgery for TED is usually performed in the chronic inactive phase of the disease. Surgical management should proceed in the following sequence: orbital decompression, squint surgery, lid lengthening with or followed by blepharoplasty/browplasty.

Orbital decompression

The usual indications for orbital decompression are compressive optic neuropathy, disfiguring exophthalmos, troublesome retroocular pain/discomfort related to orbital congestion, and/or

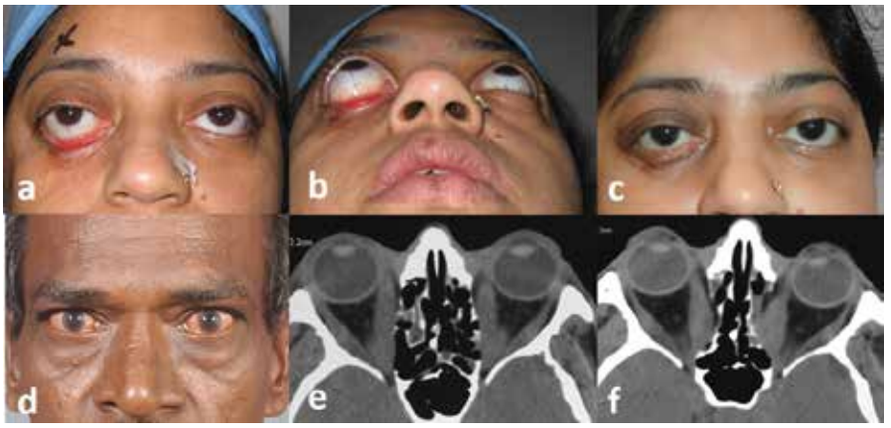

Figure 9: *Orbital decompression for thyroid orbitopathy. Right disfiguring proptosis and lower lid ectropion in the quiescent phase of GO in a female patient (Figure 9a,9b). Appearance following deep lateral orbital and fat decompression (Figure 9c). Compressive optic neuropathy due to apical crowding by bulky extraocular muscles as seen on CT scan in a male smoker with TED (Figure 9d, 9e). Reduction in the apical crowding seen on CT scan following endoscopic medial wall decompression (Figure 9f).*

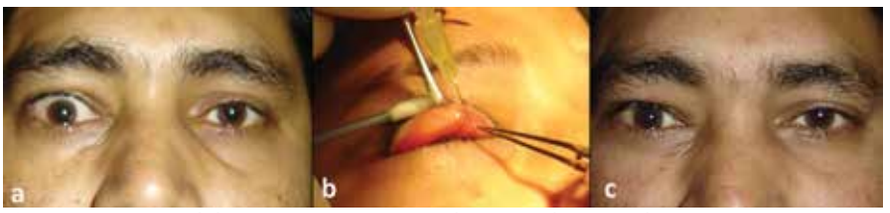

Figure 10: *Upper eyelid retraction in a patient with Graves' disease with recent onset TED (Figure 10a). Transconjunctival injection of 5U of Botox (botulinum toxin) to the levator caused temporary reduction in the retraction with optimal cosmetic appearance (Figure 10b,10c).*

Figure 11: *Mild inactive TED with right upper eyelid retraction (Figure 11a). Transconjunctival graded levator recession was done to correct the retraction (Figure 11b,11c).*

grittiness associated with minor exposure keratopathy not amenable to topical therapies. Orbital decompression for disfiguring exophthalmos is best deferred until the orbitopathy has been inactive for at least 6 months. Decompression can be done either by orbital approach, endoscopic endonasal approach or via transcranial route (Figure 9). Depending upon the amount of proptosis reduction desired, either fat, bone or a combination of fat and bone decompression can be done. Complications associated with orbital decompression include diplopia, ocular motility limitation and can be minimised by doing a 'balanced orbital decompression" which involves simultaneous medial and lateral wall decompression.

Strabismus surgery

Strabismus surgery for correction of diplopia is done after the active phase of the disease is over and orbital decompression if indicated is done with. Strabismus measurement should be stable for at least 6 months before planning any surgical intervention. The aim of strabismus correction is to restore fusion in the primary gaze and downgaze and if that is achieved to correct any other residual incomitance. The extraocular muscles are usually tight and fibrotic in TED; hence recession of the extraocular muscles with adjustable suture technique and retroequitorial myopexy gives best results.

Eyelid retraction

In the active phase of the disease, eyelid retraction can be minimised by botulinum toxin injection to the levator aponeurosis. Injection of 2.5 to 5U of Botox is given either via transcutaneous or transconjunctival route (Figure 10). Severe retraction and exposure keratopathy requires tarsorrhaphy to reduce the lagophthalmos. Conventional surgical management of eyelid retraction is levator recession (Figure 11). Treatment of lower eyelid retraction requires spacer placement in addition to recession of the retractors to provide height and necessary stiffness to support the eyelid against gravity. Injection of fillers in the levator plane can also cause temporary reduction in the eyelid retraction.

CONCLUSION

GO is the most common extrathyroidal manifestation of Graves's disease. The pathophysiology of GO is still not completely understood, however newer studies have elucidated noble mechanisms of the involvement of the orbital tissues in this disease. Introduction of the newer and noble therapeutic modalities like biological agents, IGF-1 receptor antagonists monoclonal antibodies will make it possible to manage this disease better, if not eliminate it. Refinements in the surgical management techniques like minimally invasive and image guided stereotactic decompression have made the surgical management safer and efficacious.

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Role of Immunohistochemistry in Evaluation of Malignant Ocular Tumors

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Abstract: *Immunohistochemistry (IHC) is an important diagnostic and prognostic tool that employs antibodies to identify the cellular components. In the last few decades it has emerged as an important technique to resolve the histomorphological differential diagnosis and helps in establishing the evidence based accurate diagnosis. Hence it contributes in the classification of poorly differentiated tumor and guides in the patient management.*

Key words: *antibody; immunohistochemistry; tumor.*

Ocular malignancies comprises of heterogeneous group of tumor that can involve conjunctiva, orbital soft tissues, eyelid or adnexal structures such as lacrimal gland and lacrimal drainage system. These tumors present with spectrum of clinical symptoms and include several forms of epithelial, stromal, lymphoid, caruncular and secondary tumors[1,4]. Application of immunohistochemistry (IHC) technique in establishing the diagnosis has become an indispensible ancillary study and is crucial in oncologic pathology. The IHC is being used frequently for the prognostic assessment of the tumors. The diagnostic accuracy and prognostic importance of IHC has continuously improved in recent years because of the discovery of the additional tissue specific biomarkers[5,6]. The choroid is the most common site for a neoplasm.

The main Intra-ocular tumors include: Choroidal melanoma and metastasis in adults and retinoblastoma in children. Conjunctival tumors affect adults and include conjunctival carcinoma and melanoma. The commonest orbital malignant tumor in children is Rhabdomyosarcoma, while lymphoma and metastasis are common in adults[3,5].

This review article will focus the utility of IHC in evaluating the malignant ocular tumor and resolving the histomorphological differential diagnosis.

HEMATOLYMPHOID NEOPLASM

Lymphocytic proliferation especially in conjunctiva is at times difficult to compartmentalize as benign or malignant.

Majority of the lymphomas arising in conjunctiva and orbit resembles other Mucosa Associated Lymphoid Tissue (MALT) derived lymphoma and Extranodal Marginal Zone B-cell lymphoma (EMZL) is the most common lymphoma at these sites. The lymphoid tissue of the conjunctiva and probably of the orbit, forms the part of the MALT[3,7]. The great majority of the ocular adnexal lymphomas are Non Hodgkins Lymphoma (NHL) of B-Cell immunophenotype.

EMZL is the most common lymphoma (>50% of the cases) followed by Follicular Lymphomas (FL) and Diffuse large B-cell lymphoma (DLBCL); while Mantle Cell Lymphoma (MCL), Small Lymphocytic Lymphoma (SLL/CLL) collectively comprises of the minority (upto 10%).

EMZL appears as diffuse expansion of marginal zone composed of monomorphic B-Cells, plasmacytoid cells with possible lymphoepithelial lesion and follicular colonization. Monocytoid cells with abundant pale cytoplasm are less frequently observed in ocular sites. The cells are distributed in nodular, diffuse or interfollicular pattern. EMZL cells are positive for CD20, PAX-5, CD79a, BCL-2 and are negative for CD5, CD10, CD23. CD43 expression is less common in ocular EMZL as compared to other sites of EMZL. CD10 and CD21 highlight the partially colonized follicles by marginal zone lymphoma[5,8]. Myeloid cell nuclear differentiation antigen (MNDA) is a recently described IHC marker with strong nuclear expression as a positive interpretation and is positive in 95% of MALT lymphoma and is used to differentiate it from FL in which it is negative[9].

FL is CD20, CD10, BCL2 positive and CD5 negative while CD23 expression is usually negative in the cells but can be seen in few cases. FL needs to be differentiated from reactive follicular hyperplasia and other small cell B-Cell lymphoma especially in trucut/small biopsies in view of diffuse CD20 expression.

Follicular lymphoma will show co-expression of CD20 and BCL-2 (Figure 1) while reactive lymphoid hyperplasia will be CD20 positive and BCL-2 negative. BCL-2 expression will be in the interfollicular in reactive hyperplasia while it will be diffuse in FL (Nodular and internodular area). High grade FL may show loss of CD10 & BCL-2 expression which may pose a diagnostic difficulty.

Stathmin, also known as STMN1 is strongly expressed by GC B cell in a cytoplasmic pattern. STMN1 is strongly positive in most of cases (97%) of FL, even in high-grade lymphoma. Germinal centre B-Cell expressed transcript 1 (GCET-1) expressed in GC B cells and B cells lymphomas arrested at the GC stage of differentiation. Positive expression shows granular cytoplasmic pattern. GCET1 is positive in almost all the cases of FL including these lacking CD10/BCL-2 expression. GCET-1 is negative in SLL/CLL, EMZL and MCL, hence FL a highly specific marker for FL[9-11].

SLL/CLL – These B cell lymphoma comprises of neoplastic lymphoid cells small to intermediate in size with coarse chromatin and scant cytoplasm with interspersed large cell aggregate, representing proliferation centers. The cells are

diffusely CD20+, CD5+, CD23+. Few cases (5%) may be CD5 negative (atypical SLL/CLL) and may cause diagnostic challenge especially to distinguish it from EMZL[10].

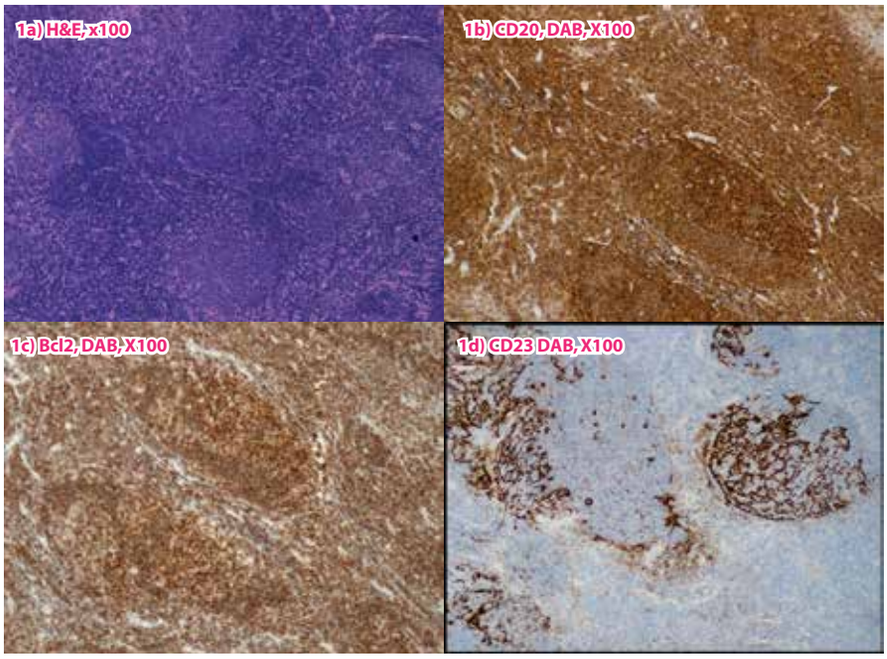
Lymphoid Enhancer Binding-Factor 1 (LEF1) IHC shows nuclear staining as positive interpretation. Strong nuclear LEF1 expression is present in SLL/CLL cells. CD200 is also a valuable IHC marker to distinguish CLL from other small B cell lymphoma. The strong of CD200 is intense membranous which is seen in CLL/SLL while negative in FL/EMZL/MCL[12-14].

Mantle cell lymphoma is a neoplasm derived from B cells of – mantle zone that is mostly composed of a pregerminal center. MCL cells are CD20+, CD5+, Cyclin D1 positive. Few cases (upto 10%) are cyclin D1 negative and many pose diagnostic difficulty especially to differentiate it from CLL/SLL. MCL has a significantly aggressive clinical course and prognosis than CLL/SLL. A new IHC marker (SOX 11) which is overexpressed in MCL giving nuclear staining seen in almost all the cases of MCL including cyclin D1 negative MCL and blastoid variant of MCL. SOX11 is negative in SLL/CLL, FL, EMZL and DLBCL[9,15-17]. Diffuse large B cell lymphomas are diffuse proliferation of large neoplastic B lymphoid cells showing significant mitosis with aggressive clinical course. Histomorphological features are helpful to distinguish it from previously described B cell NHL.

DLBCL are variably positive for CD10, BCL-6, MUM-1 and BCL-2. They may progress from EMZL/FL/CLL or may arise de novo.

Lymphoblastic lymphoma (LBL) is a Lymphoproliferative disorder composed of immature, neoplastic lymphocytes. T-cell. LBL is the second most common subtype of non-Hodgkin lymphoma in children and adolescents, comprising 85-90% of all LBLs. LCA, TdT, CD99 Cd1a and CD34 positivity helps in establishing the diagnosis[18].

Granulocytic sarcoma (GS): Soft tissue infiltration by myelogenous leukemia can present as an orbital mass especially in young adults. Systemic disease is usually present before orbital involvement but rarely orbital mass can be the first manifestation and differential diagnosis includes Malignant Round Cell Tumors (MRCT). On IHC LCA is positive while B-cell markers (CD20, CD79a) and T cell markers (CD3, CD2, CD5) are absent and this immunoprofile is highly suspicious for granulocytic sarcoma. C-kit (CD117), MPO (Myeloperoxidase), CD163 and CD34 are helpful for establishing the diagnosis[19,20].



Case 1) Follicular Lymphoma in 60 year old male as conjunctival mass
Figure 1a: *Neoplastic small to intermediate lymphoid cells in predominantly follicular pattern.*
Figure1b&c: *CD20 and Bcl2 showing diffuse positivity in follicular and interfollicular areas*
Figure 1d: *CD23 highlights the intact and disrupted follicular meshwork*

Langerhan Cell Histiocytosis (LCH) which includes the eosinophilic granuloma can involve the ocular site. CD1a and langerin are highly specific markers besides S100.

EPITHELIAL MALIGNANCIES

Basal cell carcinoma (BCC) is the most common malignant eyelid tumor. Focal squamoid differentiation and keratinization can be seen. Peripheral palisading of tumor cells, stromal retraction artifact and absence of an intraepithelial component favors the diagnosis of BCC. IHC can substantiate the diagnosis. BCC are Ber-EP4 positive while EMA negative while squamous cell carcinoma and sebaceous carcinoma are EMA positive[21].

Squamous cell carcinoma (SCC) is the second most common malignant epithelial tumor of the eyelid. SCC can arise from pre-existing actinic keratosis, Bowen`s disease, Keratoacanthoma, radiation dermatitis or de novo. SCC comprises of infiltrating malignant squamous cells with or without keratinization.

On IHC the tumor cells are diffusely positive for EMA while negative for Ber-EP4, which helps in differentiating it from BCC[22].

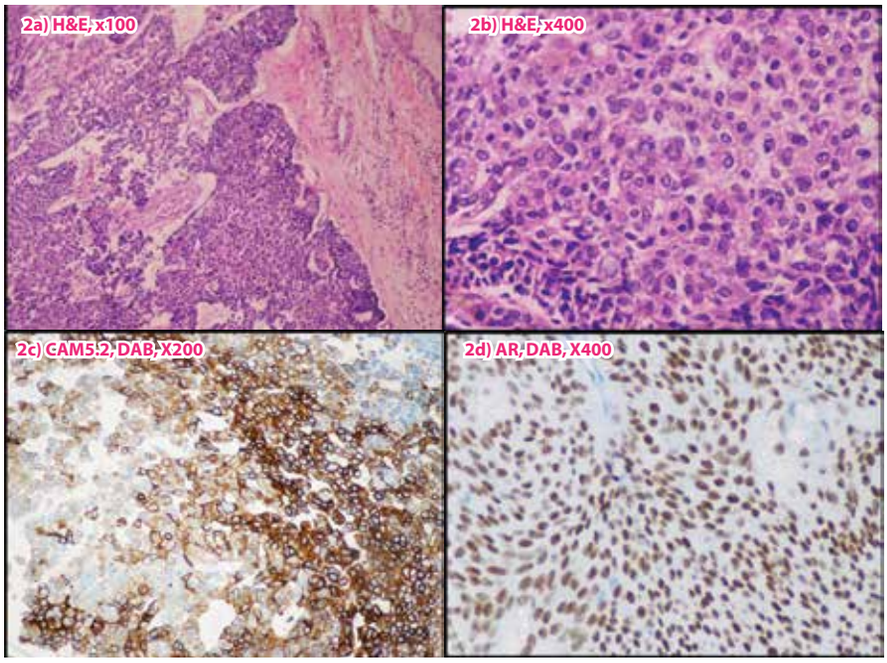
Sebaceous Carcinoma (SEC) is the third most common malignant epithelial tumor and most aggressive due to potential risk of metastasis. The most overlapping differential diagnosis is SCC as both are EMA positive. In many studies Androgen Receptor (AR) & CAM5. has been proved to be an important IHC marker to distinguish SCC and SEC with positive expression seen in great majority of SEC while negative in SCC (Figure 2).

Low grade SEC also needs to be distinguished from sebaceous hyperplasia. P53 expression (nuclear) is seen in majority of SEC with expression directly proportional to the grade, highest expression seen in grade III SEC. Sebaceous hyperplasia show no or insignificant P53 expression[22,23].

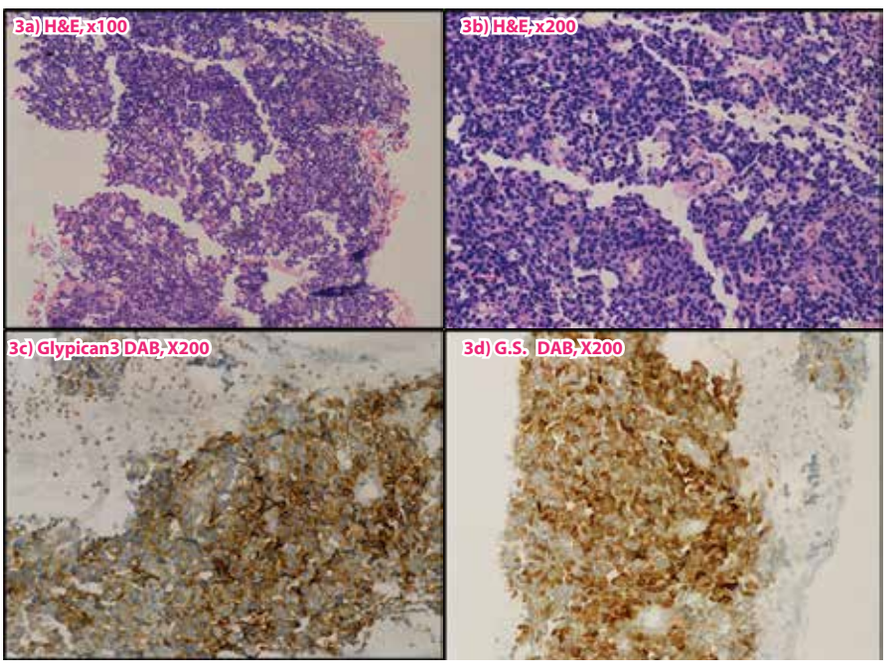
Metastatic epithelial malignancy: Ocular and its adnexal metastasis are uncommon. IHC plays a crucial role when the primary cancer that has metastasized to ocular site is occult. Even in established case of primary malignancy, IHC is essential to rule out a second primary tumor in the eye.

Common ocular metastases are from primary Breast carcinoma followed by lung carcinoma. Other primary cancer sites include colorectal, kidney liver. When a primary tumor metastasized in ocular site is undetected the first IHC panel should comprise of CK7 and CK.

Breast carcinoma, upper GI Tract and lung adenocarcinoma are positive for CK7 while negative for CK20. Breast carcinoma shows ER, GATA-3 and mammaglobin



Case 2: Sebaceous carcinoma of the eye lid
Figure 2a: *Neoplastic epithelial cells in nests with focally infiltrative margins.*
Figure 2b: *Tumor cells exhibiting significant atypia with mitosis(arrow), cytoplasm is pale eosinophilic.* ***Figure 2c:*** *Strong CAM5.2 cytoplasmic positivity.* ***Figure 2d:*** *Strong and diffuse AR nuclear positivity*



Case 3) Metastatic HCC presented as orbital mass
Figure 3a: *Trucut biopsy shows neoplastic cells in diffuse sheets*
Figure3b: *Tumor cells are round with hyperchromatic nuclei and scant eosinophilic cytoplasm.*
Figure 3c&d: *Glypican 3 and Glutamine Synthetase (GS) positivity in tumor cells.*

positivity. Lung adenocarcinoma shows TTF-1 and Napsin Positivity. Colorectal carcinoma shows CK20 and SATB2 expression.

Hepatocellular carcinoma (HCC), Renal cell carcinoma (RCC) and prostatic adenocarcinoma are CK7 and CK20 dual negative. HCC shows expression of Hep-Par-1, glypican 3 and Glutamine synthetase (Figure 3). RCC shows PAX-8, carbonic Anhydrase expression. Prostatic adenocarcinoma show NKX3.1, PSA and PSAP expression.

Neuroendocrine carcinoma is positive for CK (dot like), synaptophysin and chromogranin.

Miscellaneous tumor – Merkel cell carcinoma is a rare malignant primary cutaneous neuroendocrine carcinoma, which can arise on eyelid. They are characteristically positive for CK20 (perinuclear dot like) and show positivity for synaptophysin.

Malignant epithelial lacrimal gland tumors: This category includes adenoid cystic carcinoma (ACC), adenocarcinoma-NOS and mucinous adenocarcinoma. ACC is the most common tumor. The tumor cells shows dual differentiation epithelial cells (luminal), highlighted by CK7 & C-kit while myoepithelial cells (abluminal) highlighted by P40.

OCULAR MELANOMA AND RETINOBLASTOMA

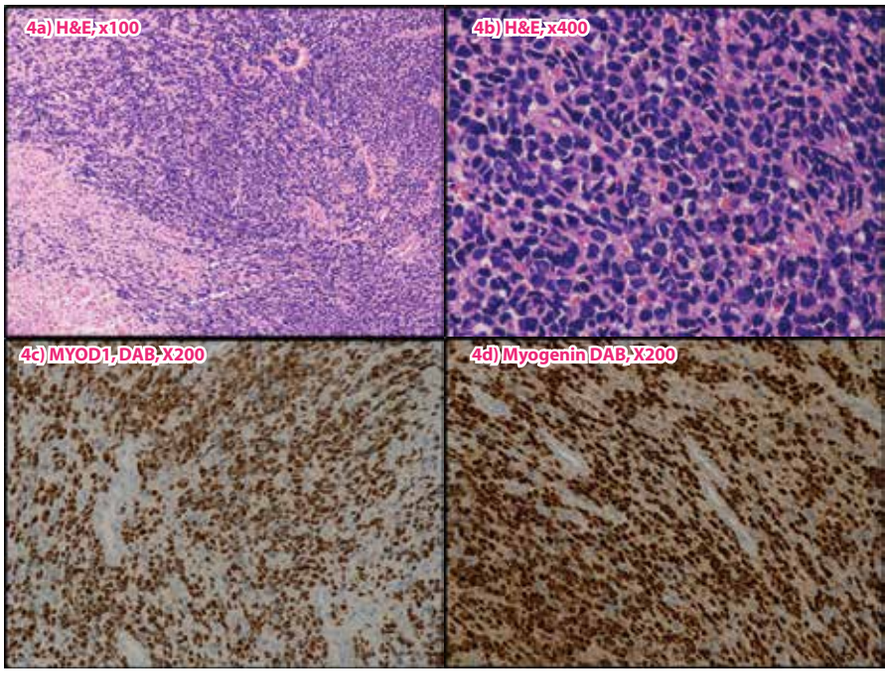
Melanoma of the ciliary body and choroid, collectively called posterior uvea is the most common primary ocular malignant tumor in adults and are highly aggressive. Iris melanomas are less aggressive with a lower evidence of metastasis. Diagnosis is usually made on indirect ophthalmoscopy and ultrasonography while fine needle biopsy has a little role. The tumor cells can be spindle, epithelioid or mixed. The tumor cells are positive for HMB-45, S100, Melan A, SOX-10. Loss of BAP-1 (BRCA associated protein) has been shown to be of prognostic significance and associated with early metastasis and decreased survival[24-26].

To distinguish the benign and malignant melanocytic lesion of conjunctiva, Ki67 and P53 IHC markers have proven a complementary role to histomorphological assessment. Ki67 of > 5% and increased p53 expression has been associated with malignant melanoma and Primary acquired melanosis with atypia[27].

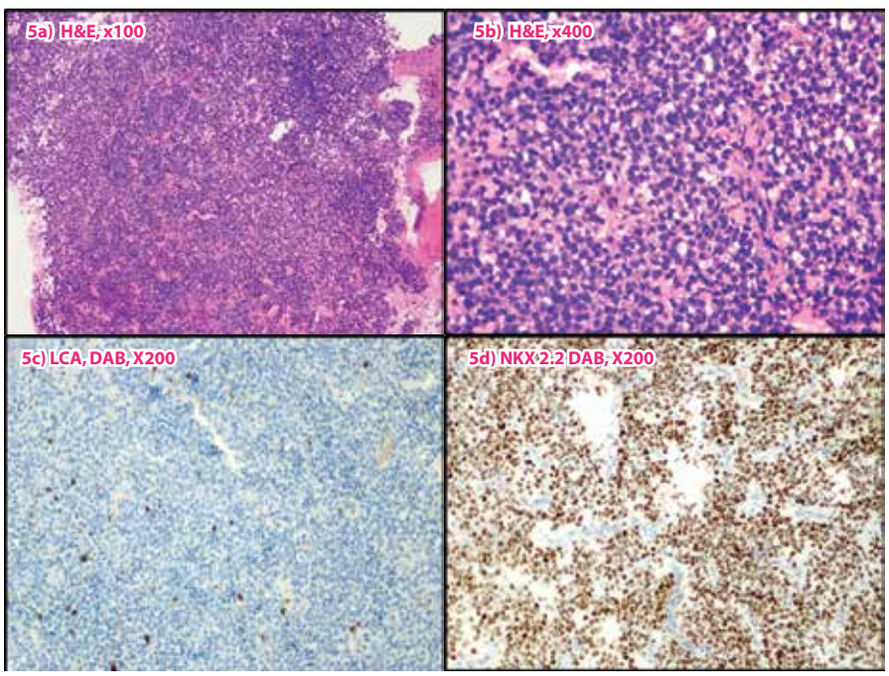
Retinoblastoma is the most common intra-ocular malignancy in the children. Clinical and histomorphological features are highly characteristic of this tumor. These tumors are characteristically CD99 negative. There is no established prognostic role of Ki67 index.

MESENCHYMAL MALIGNANCIES

Rhabdomyosarcoma (RMS) is the most common sarcoma arising in children and adolescent. The tumors present as an orbital mass with round cells to spindle cells morphology in a myxoid background. The differential diagnosis includes Malignant Round Cell Tumors (MRCT), Lymphoma, Ewing sarcoma, myeloid sarcoma. The RMS cells are positive for desmin, myogenin and myoD1 (Figure 4), which are highly sensitive and specific markers.



Case 4) Embryonal RMS in 5 year old child
Figure 4a: *Trucut biopsy shows round neoplastic cells in cords and sheets in a hyalanized stroma*
Figure 4b: *Tumor cells are round with hyperchromatic nuclei and scant eosinophilic cytoplasm.*
Figure 4c&d: *Diffuse MYOD1 and myogenin nuclear positivity in tumor cells.*



Case 5) Ewings sarcoma in 12 year old child as orbital mass
Figure 5a: *Trucut biopsy shows round neoplastic cells in cords and sheets in a hyalanized stroma*
Figure 5b: *Tumor cells are round with hyperchromatic nuclei and moderate amount of vacoulated cytoplasm.* ***Figure 5c:*** *LCA negative in tumor cells and highlights occasional scattered leucocytes in background.* ***Figure 5d:*** *Diffuse NKX2.2 positivity in tumor cells.*

Ewings sarcoma (ES) also present as an orbital mass in children, adolescents or young adults. The differential disease includes MRCT. CD99 is a sensitive marker but specificity is poor as CD99 positivity is well described in lymphoblastic lymphoma. Recently described IHC marker NKX2.2 is highly specific for Ewings sarcoma (Figure 5). CK and Synaptophysin expression can be seen in 20% of the cases[28].

Conclusion: With well-performed and interpreted immunohistochemistry panels, anatomic pathologists can successfully resolve the differentials and establish the accurate diagnosis. It is crucial to understand not only the diagnostic uses of the many available antibodies but also the potential limits and pitfalls. Therefore, a judicious panel of IHC should be ordered in cognizance of sensitivity and specificity and finally, it is important to stress that the selection of an appropriate panel should be targeted at a group of well-constructed differential diagnoses based on careful microscopic examination and clinicopathologic correlation.

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NOTICE FOR GENERAL BODY MEETING

The General Body Meeting of the Delhi Ophthalmological Society will be held during the Annual Conference on Sunday, April 8th, 2018 at 4:30 PM at the Ashok Hotel, Chanakyapuri, New Delhi.

The revised Agenda of the General Body Meeting shall be :

1. Confirmation of the minutes of the last Annual General Body meeting and action taken thereof.
2. Adoption of the annual report of the Executive Committee presented by the Hony-Secretary.
3. Ratification of new members.
4. Report of the Library officer
5. Report of Editor
6. Report from representative to A.I.O.S.
7. Consideration of any other business or resolution that may be laid before the meeting provided that the Hony. Secretary has received due notice at least eight weeks before the meeting for consideration by the Executive before putting it to General body.

7(a).

1. Elections of Various Posts in Winter Conference
2. Election Fund
3. Eligibility Criteria for Election

7(b). Consideration of any other business :

1. Course of Action for denial of bid of DOS to host 2020 AIOS Conference.
2. Complimentary registration Eligibility Criteria
3. Deletion of names of members from voter list, who are neither working nor residing in Delhi
8. Address of the outgoing and incoming president.
9. Election of the Office Bearers and members of Executive Committee and announcement of results
10. Any other matter with the permission of the Chair.

All members are requested to attend.

Thanking you,

Sincerely yours,

Prof. Kamlesh
President, DOS

Prof. Subhash C. Dadeya
Secretary, DOS

Congenital Nasolacrimal Duct Obstruction: Variant based Approach

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Abstract: *Congenital nasolacrimal duct obstruction (CNLDO) is one of the common lacrimal disease but also one of the commonly miss-managed entity. Be it related to method of sac compression, impractical use of eye drops, age of probing or blanket treatment with silicone intubation and balloon dacryoplasty. Different specialties right from pediatricians to general ophthalmologist, pediatric ophthalmologist or oculoplastic surgeon may be involved, therefore the protocol for management of CNLDO is not standard and uniform. For children above 4 years straightforward dacryocystorhinostomy is no more a treatment of choice. Current review aims to highlight the recent advances in understanding of different types of CNLDO and a case based approach.*

Congenital nasolacrimal duct obstruction (CNLDO) is the most common cause of epiphora in infants and children[1]. Actual incidence in newborns is 50% but due to spontaneous perforation of membrane (Hasner valve) at lower nasolacrimal duct (NLD) after birth, clinical symptoms of CNLDO are seen in about 6-30% children[2]. Embryological basis of CNLDO is that the lower NLD is last to canalize during development, and therefore its failure leads to CNLDO. During crying or respiration, spontaneous rupture of membrane may occur over 3-4 weeks after birth but if it doesn't occur, CNLDO symptoms are manifested. This review aims to make you understand about the basics as well recent paradigm shifts in understanding and management of CNLDO. Over the recent years, nasal endoscopy guided (NEG) syringing and probing (S&P) is being accepted as gold standard for the management of CNLDO. NEG is not only needed for repeat or failed cases, which have undergone previous S & P, but also during primary attempt itself. Many cases can be saved from dacryocystorhinostomy (DCR) if done properly through NEG. Even older cases up to 10-12 years of age can be successfully managed with NEG S & P although with decreased success rate. Such older age children should always be given trial of S & P, before considering them for DCR.

Another recent change in the management of CNLDO is the approach based on the type of variant. CNLDO is broadly of two types: simple and complex[3,4]. Complex CNLDO encompasses various variants. Management of these variants not only need NEG but also customized treatment as discussed in this review. It is important to understand that some of the widely practiced methods of primary silicone intubation and balloon dacryoplasty may be entirely unnecessary.

SYMPTOMS/SIGNS OF CNLDO

Symptoms include classic triad of watering, discharge and matting of eyelashes. Three important signs are: positive regurgitation on pressure over lacrimal sac (ROPLAS), increase tear meniscus height and positive Fluorescein dye Retention test (FDRT) (Figure 1). Sometimes acute dacryocystitis may occur which can complicate into orbital cellulitis, orbital abscess or cavernous sinus thrombosis. Dacryocele is a bluish-purple dilated lacrimal sac seen in infants with CNDLO and functional common canalicular obstruction. It is important to carefully follow up and treat Dacryocele early as it is associated with intranasal cyst in NLD which can cause respiratory problems. Other findings may be lacrimal fistula (congenital/acquired), mucocele, incomplete punctal canalization and canalicular stenosis.

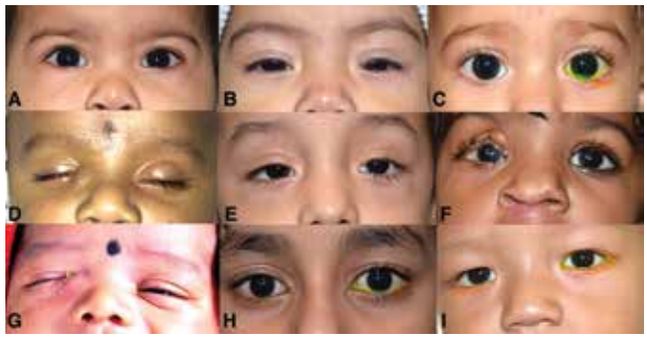

Figure 1: *Panel of photographs showing spectrum of clinical manifestation of CNLDO* ***(A)*** *A 5-month old child presenting with bilateral epiphora* ***(B)*** *A 6-month old child having classic triad of left eye epiphora, discharge and matting of eyelashes* ***(C)*** *Left eye showing increased tear meniscus with positive fluorescein dye retention test (FDRT)* ***(D)*** *A 2-month old child presenting with right dacryocele* ***(E)*** *A child with down syndrome presenting with symptoms of left eye complex CNLDO* ***(F)*** *A 6-month old child presenting with right eye watering and discharge along with Goldenhar syndrome and operated cleft lip/palate suggesting complex CNLDO* ***(G)*** *A 23 days old child presenting with right acute dacryocystitis* ***(H)*** *A 5-year old child presenting with persistent symptoms of CNLDO after twice failed previous attempts of probing suggesting complex CNLDO* ***(I)*** *A 8-months child presenting with recurrent symptoms of watering left eye associated with upper respiratory tract infection suggesting diffuse stenosis of nasolacrimal duct.*

TYPES OF CNLDO (SIMPLE AND COMPLEX)

All cases with soft membranous obstruction at lower NLD with rest of lacrimal system being normal and no intranasal abnormality constitutes simple CNLDO (Figure 2). All others are complex CNLDO. First study comparing simple and complex

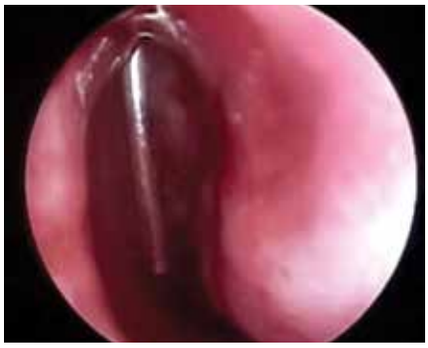

Figure 2: *Nasal endoscopy view of right nostril showing Bowman lacrimal probe coming through nasolacrimal duct opening without any nasal abnormality i.e. Simple CNLDO.*

CNLDO was published by Ali MJ et al reporting demography, clinical features and management outcomes at a tertiary eye care center. Table 1 shows the overview of difference between the two types.

Table 1: Modified table adapted from Ali MJ, Kamal S, Gupta A et al.

Parameter	Simple CNLDO	Complex CNLDO
Average age at presentation	Younger (mean 17.6 months)	Older (mean 45.6 months)
History of prior failed intervention	none	Positive in one-third cases
Intervention needed	Only NEG with S & P	Adjunctive procedures like intubation, balloon catheter needed in one-third cases
Success rate Anatomical Functional	97.8% 94.7%	58% 51%

CNLDO – Congenital nasolacrimal duct obstruction, NEG – Nasal endoscopic guidance, S & P – Syringing and Probing

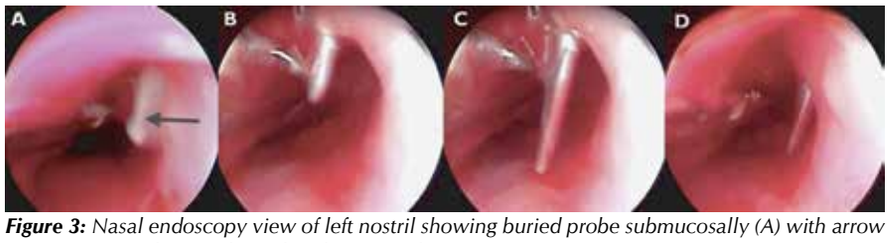

Figure 3: *Nasal endoscopy view of left nostril showing buried probe submucosally (A) with arrow pointing. (B-D) showing buried probe externalization.*

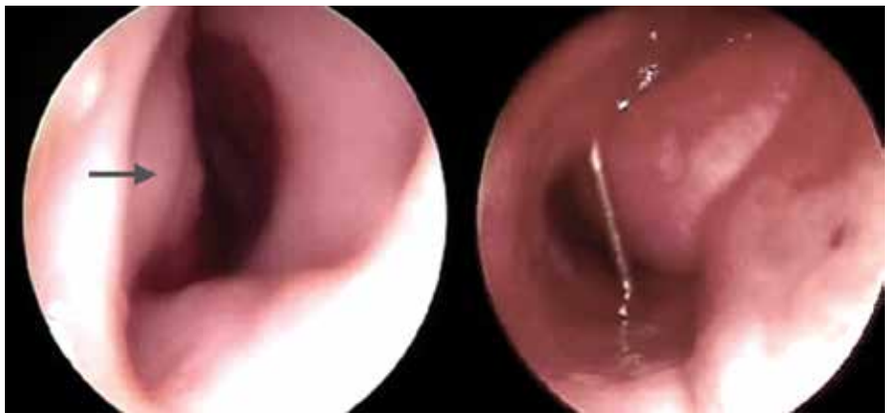

Figure 4: *Nasal endoscopy view of right nostril showing lateralized inferior turbinate (left Picture). Probe seen after medialization of inferior turbinate (right picture). True impaction of inferior turbinate is very rarely seen.*

Complex CNLDO encompasses following

1. *Bony obstruction* – This is due to complete absence of NLD formation or NLD directed into lateral maxillary bone.
2. *Craniofacial syndromes* – Such as Downs syndrome, Crouzon syndrome, Treacher Collins syndrome, Cleft lip/palate, hypertelorism.
3. *Buried probe*[5] – In this variant probe lies sub-mucosally along lateral wall of nose and fails to come out of the NLD opening. It needs tilting of probe to create and enlarge the opening along entire length (Figure 3).
4. *Lateralized/Impacted inferior turbinate* – This means the inferior turbinate lies in close approximation to the lateral nasal wall giving no access to the inferior meatus and visualization of NLD (Figure 4).
5. Dacryocele without intranasal cyst
6. *Anlage duct* – Lacrimal fistula (Anlage duct) may be associated with CNLDO. If congenital it needs fistulectomy.
7. *Multiple blocks* – Stenosis at valvular sites in canaliculus, sac and NLD or sometimes diffuse stenosis of NLD can lead to multiple level NLD blocks.
8. *Dacryocele with intranasal cyst* – Pressure inside Dacryocele can be transmitted to NLD and lead to its dilation and nasal cyst formation (Figure 5).
9. *Atonic sac* – Longstanding CNLDO can lead to atonicity of lacrimal sac.
10. *NLD upto the floor* – Variant in which NLD extends within the bone till nasal floor
11. *NLD into the inferior turbinate* – Caused by misdirection of inferior NLD.
12. *Lateral nasal wall hypoplasia* – Lead to non-differentiation of lateral nasal wall structures such as inferior turbinate, inferior meatus or NLD opening.

NATURAL HISTORY

It is very important to be aware of natural history of CNLDO since the management decision solely rests upon the chances of self resolution at that particular age. Macewen and Young followed children with symptoms of CNLDO till one year of age and calculated the chances of spontaneous resolution (Table 2) and suggested that probing should be performed at one year of age. Subsequently same authors published spontaneous resolution between 12-24 months age and observed resolution rate

Table 2: Predicted spontaneous resolution of CNLDO till one-year age depending upon presenting age (Adapted from Macewen and Young)

Age (months)	Resolution rate
1	96%
3	90%
6	75%
9	36%
12	0%

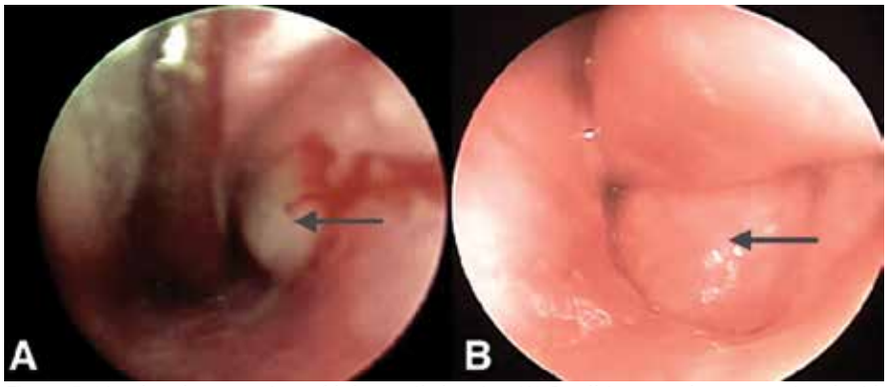

Figure 5: *Nasal endoscopy view of left nostril showing* ***(A)*** *Small intranasal cyst associated with dacryocele* ***(B)*** *A very large intranasal cyst which may cause respiratory difficulties in infants.*

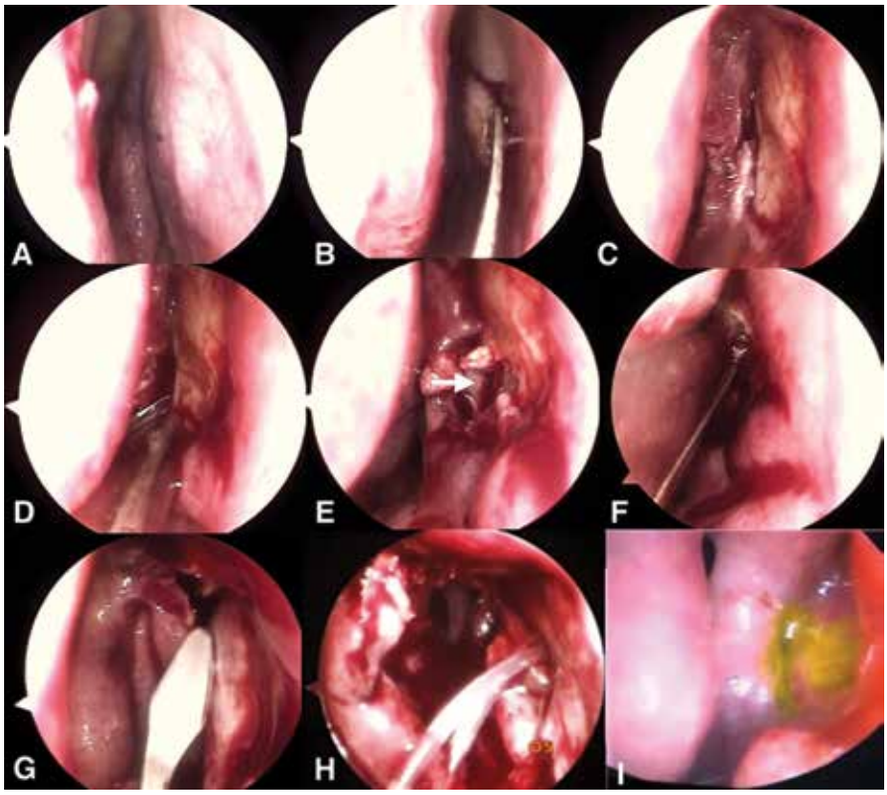

Figure 6: *Panel of intraoperative photographs showing Endoscopic DCR being performed in 4 year-old children with persistent CNLDO with bony obstruction.* ***(A)*** *Left nasal cavity showing septum on left side and lateral maxillary wall on right side* ***(B)*** *Nasal mucosal incision being given with No 15 BP blade* ***(C)*** *Frontal process of maxilla seen after elevation of nasal mucosal flap* ***(D)*** *Initiation of Osteotomy with punch at sac-duct junction* ***(E)*** *Bluish color sac seen after initial osteotomy (arrow)* ***(F)*** *Enlargement of osteotomy To expose lacrimal sac completely* ***(G)*** *Crescent knife for making anterior and posterior lacrimal sac flaps* ***(H)*** *Lacrimal sac opened in book-like fashion with bicanalicular silicone tube in situ* ***(I)*** *Evaluation of ostium at 4 weeks follow up showing positive functional endoscopic dye test (FEDT).*

of 50% between 13-18 months and 23% between 18-24 months age[6]. Appropriate time of probing recommended was 18 months. More recent data from PEDIG (Pediatric eye disease investigator group) observed that 66% of cases are likely to resolve if age is between 6 to less than 10 months[7] Author consider 9-12 months of age appropriate for S & P in non-resolving CNLDO.

MANAGEMENT OF CNLDO

Conservative treatment and Lacrimal sac compression - Compression over the sac increases hydrostatic pressure and can cause membrane rupture at lower NLD. It was described by Criggler in 1923. It can be done 3-4 times a day, 10 times each. It is important to demonstrate and then ask the parents to perform in clinics.

Probing - Timing of probing is controversial. Spontaneous resolution can occur beyond one year age, but success of probing decreases with increasing age. Katowitz and Welsh recommended probing before 13 months of age because they found that probing before 13 months of age was associated with a cure rate of 97%, which dropped to 54.7% after 13 months of age 9. Most important is to explain to the parents about chances of spontaneous resolution versus success rate of probing at particular age. Most clinicians prefer 9-12 months age for first probing.

Balloon catheter dilation and silicone intubation – These measures are not necessary for simple CNDLO but may be needed for complex CNLDO, persistent simple CNLDO, recurrent CNLDO or associated canalicular stenosis. Cost of the procedure is the main factor which has limited its use in recent times.

Nasal endoscopy - Use of rigid nasal endoscope is now becoming gold standard during probing, balloon catheter dilation or silicone intubation. It allows the visualization of NLD opening, helps in proper evaluation and treatment of complex CNLDO, marsupialization of intranasal cyst and correction of associated nasal abnormalities[1]. Figure 2 shows its utilization during management of CNLDO allowing confirmation of probe. Older children should undergo a trial of probing with endoscopy before considering DCR. A rigid 2.7 mm endoscope is used for nasal endoscopy in children.

Dacryocystorhinostomy (DCR) – Is considered for cases of failed probing, absent NLD or failed cases even when adjunctive measures have been used. Both external and endoscopic DCR can be done, and studies have shown that pediatric endoscopic DCR can be performed with good success rate even in children of one-year age. Figure 6 shows primary endoscopy DCR being performed in a 4-year-old child. Early need for endoscopic DCR is required in cases with recurrent acute dacryocystitis with failed probing or need for intraocular surgery.

CONCLUSION

CNLDO is a common cause of epiphora in children. Usually most of the cases tend to resolve spontaneously. For cases persisting beyond 9-12 months of age nasal endoscopic guided irrigation and probing is becoming gold standard. Use of nasal endoscopy not only helps in identification of complex CNLDO but also its management, thus improving the success rate of probing. This is especially important for cases with previously failed probing where endoscopy is not done.

For cases with atonic sac, mucocele, persistent CNLDO and bony obstruction, endonasal endoscopic DCR is considered.

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Contracted Socket

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Abstract: *Management of contracted anophthalmic socket requires understanding of various causative factors, assessment of key areas of tissue deficiency in the socket, a clear mental classification of what needs correction and how much correction is possible. A detailed discussion with the patient about likely benefits or limitations of procedures is very important to keep expectations realistic. The following text details various factors that go into planning any reconstructive procedure for an anophthalmic socket and also elaborates some commonly used surgical techniques.*

Loss of an eye is a big psychological trauma for anyone who has to deal with this unfortunate situation. With facial cosmesis becoming very important in our current social environment, it becomes vital for ophthalmologists to understand proper management of an anophthalmic socket. All anophthalmic sockets undergo some sort of contraction as a result of fibrosis of orbital tissues, which maybe mild and unnoticeable most of the times. However, it usually draws attention when the individual is unable to retain a prosthesis, or there is significant asymmetry compared to the contralateral eye. Contracted socket is best prevented by following proper surgical technique at the time of primary enucleation or evisceration.

Common factors responsible for contraction of an anophthalmic socket are:

1. Poor surgical technique of primary surgery.
2. Fibrosis resulting from multiple surgeries.
3. Implant related complications: migration, exposure etc.
4. Socket left without prosthesis / conformer for a long time.
5. Using inappropriately made / Ill fitting prosthesis.
6. Trauma.
7. Cicatrising conjunctival diseases.
8. Chronic inflammation.
9. Chemical Injury.
10. Radiation.

CLINICAL PRESENTATION

Contraction of soft tissues in an anophthalmic socket presents with shallowing of fornices, irregular fibrosis, atrophy of orbital fat and volume redistribution within the orbit (Figure 1). Together, these factors lead to appearance of a hollow or deep superior sulcus, enophthalmos and eventually an inability of socket to retain an ocular prosthesis in place (Figure 2).

Examination revolves around assessing the principal deficiency in socket, which may be a deficiency of surface only, or compounded by a deficiency in volume of orbit as well. In more severe cases, shortening of palpebral aperture can also be seen (Figure 3).

Evaluation of a contracted socket includes an assessment of:

a) *Size of palpebral fissure:* Both horizontal and vertical fissure height need to be assessed and compared with the normal side. Any pre-existing ptosis must be given due consideration.
b) Tone of Orbicularis, lower lid laxity or ectropion.
c) Depth of fornices.
d) Surface character: Dry or moist, vascularised, healthy or pale.
e) Volume deficiency: Assessment of enophthalmos and deepening of superior sulcus.
f) Presence of obvious cicatricial bands.
g) Extra ocular muscle function.
h) Presence of confounding factors such as concurrent orbital fracture or bony contractures.

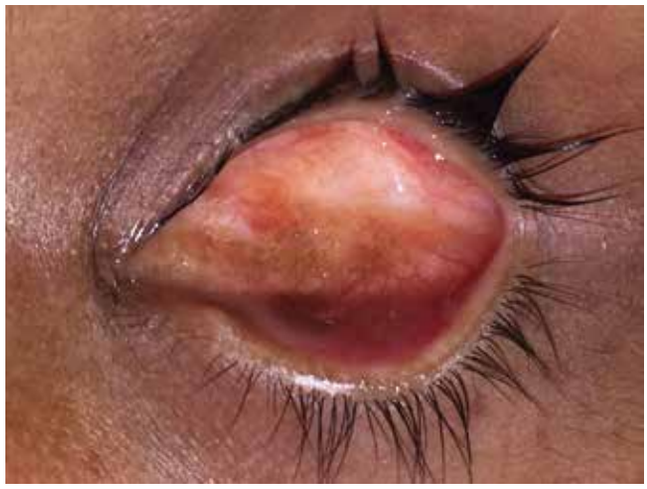

Figure 1: *Shallowing of fornices, irregular fibrosis, atrophy of orbital fat and volume redistribution within the orbit.*

CLASSIFICATION

Gopal Krishna's[1] classification of contracted sockets as mentioned below is probably the most widely accepted one:

Grade-0: Socket is lined with healthy conjunctiva and has deep and well formed fornices.

Grade-I: Socket is characterised by shallow lower fornix, preventing retention of an artificial eye.

Grade-II: Socket is characterised by loss of upper and lower fornices both.

Grade- III: Socket is characterised by loss of the upper, lower, medial and lateral fornices.

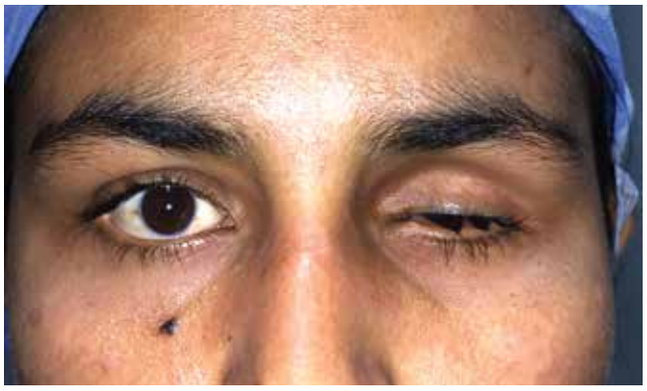

Figure 2: *Hollow, deep superior sulcus and enophthalmic appearance in a case of contracted socket.*

Figure 3: *Shortening of horizontal palpebral aperture in a severely contracted socket.*

Grade-IV: Socket is characterised by loss of all the fornices, and reduction of palpebral aperture in horizontal and vertical dimensions.

Grade-V: Recurrence of contraction of socket after repeated trials of reconstruction.

Another system of classification has been described by Tawfik et al[2] as follows:

Grade 1: Minimal or no actual contraction. Patients complain of inability to retain the prosthesis for a long time. Horizontal lid laxity is often observed in these patients, with subsequent prolapse or retraction of the inferior fornix. Patients with an unusually large or anteriorly displaced implant also fall in this category.

Grade 2: Mild contracture of the inferior and/or the superior fornix. Patient either complains of inability to wear the prosthesis or may complain of a cosmetic disfigurement due to rolling-in of the upper and lower eyelid margin.

Grade 3: More advanced scarring than grade 2. Cicatrisation generally involving the entire upper and lower fornices. Wearing the prosthesis is impossible.

Grade 4: Severe phimosis of the palpebral fissure both vertically and horizontally. Recurrent cases and irradiated sockets are also included in this category.

MANAGEMENT

Primary aim of management is to create a healthy socket which is able to hold a stable ocular prosthesis along with reasonable symmetry of palpebral apertures, canthal angles and superior sulci.

Treatment planning is based on the severity of cicatrisation. Due consideration must be given to the etiology of contraction. As for any cosmetic procedure, a detailed discussion with patient and family is very important regarding the expected outcome and to understand the patient's expectations from the procedure.

Not all cases of socket contraction require surgery, it may just be sufficient to modify the prosthesis in very mild cases. Cases where the fornices are unable to hold prosthesis due to excessive lid laxity require horizontal lid tightening with or without fornix formation sutures.

Fornix formation sutures: are non absorbable double armed sutures that are passed through the respective fornices and exteriorised on the skin side along inferior and superior orbital rims. The sutures are retained for approximately two weeks and an adequate sized conformer is placed to ensure that the socket retains necessary space for holding a prosthesis subsequently.

Patients with shortening of the posterior lamella causing entropion require lengthening of posterior lamella with the help of scleral or cartilage graft in addition to fornix formation[3].

Moderately contracted sockets where there is mainly surface shortening and no volume deficiency with a moist vascularised surface can be treated with either Amniotic membrane graft or mucous membrane graft.

Mucous membrane graft: remains the preferred choice of surgery for moderate contractures. Autologous oral mucosa from lip and cheek can be easily harvested and is generally taken up well by the socket[4] (Figure 4).

An incision is given through the centre of socket from lateral to medial canthus. Blunt dissection is carried out superiorly and inferiorly to release the scar tissue and to allow deepening of fornices. Fornix formation sutures are then passed from superior and inferior fornices. Central area of conjunctival deficiency is measured. Full thickness mucosal graft is harvested from oral cavity either lip or cheek, measuring approximately 40-50% larger than the deficiency in socket to accommodate for subsequent graft contracture. Submucosal tissue is trimmed and graft sutured to free edges of incised conjunctiva with absorbable sutures. An adequate sized conformer is finally inserted to keep the fornices deep. The donor site can be closed by direct suturing or left alone to heal spontaneously.

Secondary Orbital Implant: Situations where the surface tissue is adequate, but there is volume deficiency only because of absence of an orbital implant, can be managed by placing an appropriately sized orbital implant secondarily in the posterior orbit. Choice of implant may vary depending on individual surgeon's experience and preference.

Dermis Fat Graft: More severe cases of contracted socket where there is significant shortening of surface as well as volume of socket are best managed with an autologous dermis fat graft. However, its use should be avoided in patients with severe or recurrent scarring, as insufficient socket vascularity compromises the success of composite grafts[5].

Initial steps of opening up the conjunctiva and deepening of fornices are the same as described above for mucous membrane graft. The upper and outer quadrant of hip is considered safe for harvesting dermis fat graft. The superficial epidermis is removed carefully and an oval shaped block of dermis with fat globules attached to its undersurface is removed from the the hip. The graft is placed in socket carefully, it should neither be too small nor too big for the size of host socket. Muscle stumps of recti, if available can be attached to the edges of dermal component of graft with absorbable sutures. Free edges of

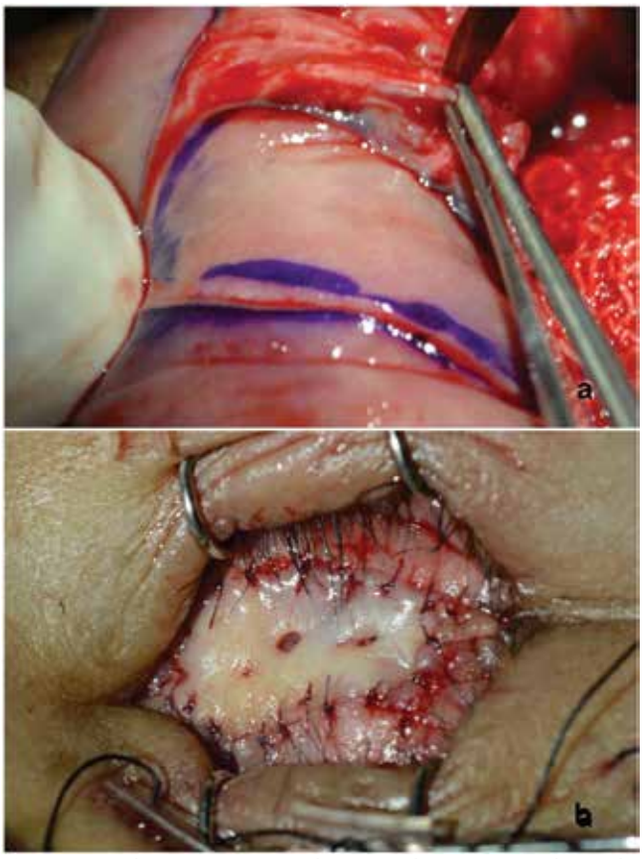

Figure 4: *Oral mucosa is a good source of mucous membrane for reconstructed a contracted socket.* ***(a)*** *Mucosa being harvested from lower lip.* ***(b)*** *Final appearance at the end of surgery with mucous membrane contributing to expansion of surface in a contracted socket.*

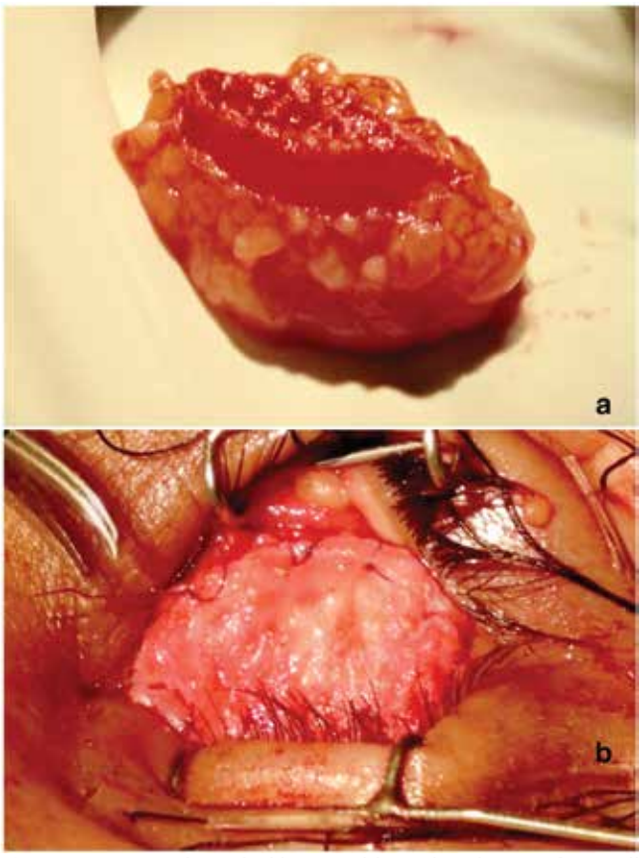

Figure 5(a): *Dermis fat grafted harvested from gluteal region* ***(b)*** *A well fitting dermis fat graft in a contracted socket.*

conjunctiva are also sutured all around to edges of dermis (Figure 5).

Radial forearm free flaps have been described in correcting extreme form of contractures such as those that present in post radiotherapy sockets or multiple failed surgeries[6,7]. Alternately, pedicle temporalis muscle flap can be used to provide volume and also serve as a vascular bed for autologous dermis fat graft[8].

Recalcitrant cases with dry parched surface or post multiple unsuccessful surgical procedures can be managed by spectacle mounted or stick-on orbital prosthesis. Other optical methods to enhance cosmesis include using plus powered lenses to magnify a microphthalmic socket and using prisms to change appearance of malpositioned prosthesis or socket.

Complications that need to be watched for while dealing with flaps / grafts:

1. Poor vascularisation or graft ischemia.
2. Loss of graft (mucous membrane / Dermis Fat) due to necrosis or poor uptake.
3. Cyst formation in the socket.
4. Hair growth and discharge in socket.
5. Erratic growth of fat graft leading to prosthesis extrusion.
6. Donor site complications: Wound infection, poor healing, scar formation.

CONCLUSION

Proper technique of Enucleation/ Evisceration and use of an appropriately sized orbital implant during primary surgery is the best way to prevent the unpleasant sequelae of contraction of an anophthalmic socket. Most useful step in managing contracted sockets is to first evaluate the contracture in terms of surface or volume deficiency or both, and then choose an appropriate surgical technique for best possible outcome.

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Endoscopic Management of Acute Dacryocystitis

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Abstract: *Acute dacryocystitis is a painful condition usually occurs due to underlying nasolacrimal duct obstruction. Acute inflammation of lacrimal sac and peri-sac tissue is classically below the medial canthal tendon in early phases but later may spread to involve the adjacent tissues. Treatment usually consists of antibiotics, analgesics, percutaneous drainage and subsequent external dacryocystorhinostomy (DCR) at later stage. Endoscopic DCR offers several advantages including primary treatment of acute phase with internal drainage of lacrimal abscess. This leads to faster recovery and increase patient comfort. Authors here in describe the clinical manifestations, conservative management and role of endoscopic DCR in the setting of acute dacryocystitis.*

Acute dacryocystitis is the acute inflammation of lacrimal sac usually secondary to microbial infection. It is defined as "A medical urgency clinically characterized by rapid onset of pain, erythema and swelling, classically below the medial canthal tendon with or without pre-existing epiphora mainly resulting from the acute infection of the lacrimal sac and perisac tissues"[1].

Clinical presentation may vary from erythema to lacrimal sac abscess. Standard treatment is conservative and often incision and drainage is performed to relieve the acute episode. External dacryocystorhinostomy (Ex-DCR) is performed at a later stage (3-4 weeks) to avoid spread of infection once superficial tissues normalizes. More often patients may have non-resolving inflammation or recurrent attacks of infection and pain which prolongs the treatment course. With the changing concepts of endoscopic dacryocystorhinostomy (En-DCR), early treatment in acute phase is possible. Moreover it offers several other advantages. The purpose of present review is to the discuss the clinical presentation and endoscopic management of acute dacryocystitis.

CLINICAL PRESENTATION

Acute dacryocystitis is a clinical diagnosis suggested by history of epiphora, discharge, prior similar acute infection. Classical findings of inflammation such as swelling, erythema, pain, tenderness in lacrimal sac area below the medial cathal tendon are seen. Clinically most common finding is swelling (85%), followed by pain (83%), erythema (48%), discharge (40%), fever (6%) and aggravated epiphora (1%). Presentation may be sometimes variable ranging from minimal erythema, and swelling over lacrimal sac area to the abscess formation, spontaneous rupture of abscess with fistula formation or infected mucocele (Figure 1). Rarely it may complicate into persistent abscess or visual threatening orbital cellulitis, orbital abscess, superior ophthalmic vein thrombosis and cavernous sinus thrombosis[2,3].

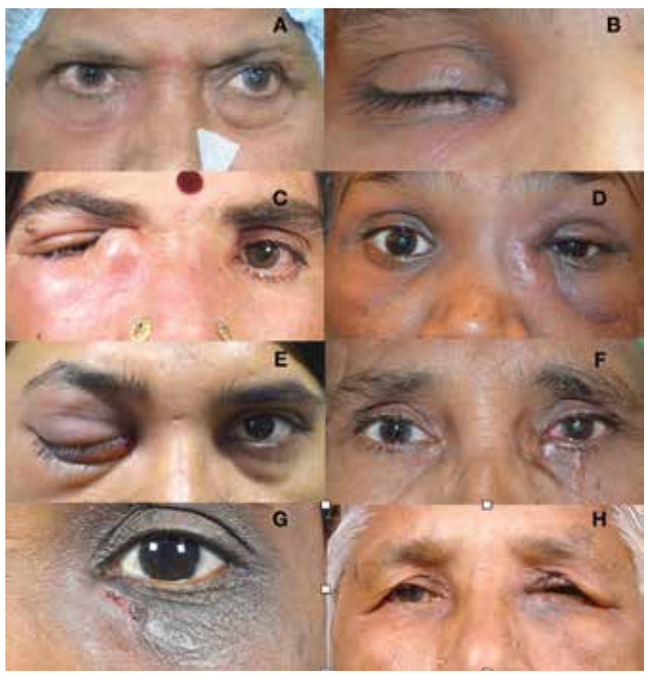

Figure 1: *Panel of photographs demonstrating varied presentation and some of the complications of acute dacryocystitis A. Right eye minimal erythema, swelling and pain suggestive of early acute dacryocystitis in a case with past history of multiple episodes of acute dacryocystitis B. Right eye showing typical signs of acute dacryocystitis with medial canthus discharge C. A case with right side huge non-resolving lacrimal abscess and left eye early acute dacryocystitis D. Left eye acute dacryocystitis with preseptal cellulitis E. Right eye persistent acute dacryocystitis complicating in to orbital cellulitis F. Typical findings of scarred skin with mucocele with chronic congestion of left eye indicating previous multiple episodes of acute dacryocystitis G. Left eye showing acute dacryocystitis with external skin fistula H. Non resolving acute dacryocystitis in spite of repeated course of antibiotics and incision and drainage procedure.*

Clinical course is prolong. The mean days to resolution of acute attack is about 10 days (1-4 weeks). Bilateral acute dacryocystitis may be seen in up to 9% cases. Other complications are progression to lacrimal sac abscess (23%), relapse of acute dacryocystitis (6%), orbital cellulitis (3%) and no response (2%).

MANAGEMENT

Traditional conservative treatment of acute dacryocystitis consists of warm compresses, oral broad spectrum antibiotics and analgesics. Effect is usually seen in 48-72 hours in most cases but complete resolution takes much longer. In case of abscess with pus point, percutaneous incision and drainage (I & D) is done to open sac and drain pus. Although done under local anaesthesia, I & D is very painful due to inadequate effect of anaesthetic agents in inflamed tissues. If done, it is important to break open all the septae/synechiae in and around the sac and place a gauze soaked with antibiotic solution over the incision. Postoperatively cleaning with topical betadine and antibiotic ointment is advised. Procedure may need to be repeated if patient develops recurrent abscess before planned surgery. Sometimes, inflammation may chronically persists in spite of treatment. Causes of non-resolving acute dacryocystitis includes virulent organisms, presence of lacrimal sac abscess, non-penetration of antibiotics in inflamed tissues, antibiotic resistance and associated persistent inflammation.

Ex-DCR is done after 3-4 weeks when inflammation subsides. Some of the challenges faced are scarred soft tissue, fibrosed shrunken sac, fibrosis around common canaliculus leading to common canalicular obstruction and increase bleeding. Skin incision related complications are seen in up to 8% cases and include wound gape, hypertrophic scar and even cicatricial ectropion.

Disadvantages of conservative treatment are prolonged/recurrent infection (which may complicate into orbital cellulitis/cavernous sinus thrombosis), adverse effect of antibiotics, skin scar/fistula formation and failure of subsequent surgery due to scarring and granulation in sac[4].

Endoscopic endonasal DCR can be performed in acute dacryocystitis[5-8]. Advantages include decrease morbidity, shortened duration of antibiotics, faster recovery with acceptable and high success rate. Other advantages compared to external DCR are avoidance of cutaneous scar, less disruption of anatomy and lacrimal pump, decrease intraoperative haemorrhage and concurrent correction of nasal and paranasal sinuses abnormalities. With the advancement of nasal endoscopy equipment, increase experience and better anatomical understanding, success rate of En-DCR now compares favourably with external DCR. Wormald et al stressed the importance of complete sac exposure, mechanical removal of thick frontal process of maxilla and opening of agger nasi cell is needed to clear the fundus of sac. This combined with 360 degree nasal mucosa to sac mucosa approximation results in healing with primary intention around osteotomy[7-9].

PROCEDURE TIMING

En-DCR can be performed in stage of acute dacryocystitis with or without abscess formation. Patients may not be started on oral antibiotics if surgery is planned early. If there is associated orbital cellulitis, orbital abscess, paranasal sinus infection then the prior treatment with intravenous antibiotics is needed. Fistulectomy can be done from cutaneous side if there is long standing fistula formation secondary to repeated attacks of acute dacryocystitis or previous incision and drainage.

SURGICAL TECHNIQUE

Anesthesia: It is best to operate cases under general anesthesia. It is more comfortable for patient and hypotensive anesthesia is maintained which reduces bleeding.

Technique: After induction, nasal endoscopy is performed with a zero degree 4 mm (for adults) or 2.7 mm (for pediatric cases) rigid endoscope. Nasal anatomy and deviated nasal septum if present is noted. Local infiltration is done with 2% lignocaine with 1:80000 adrenaline beneath the nasal mucosa anterior to the axilla of the middle turbinate and maxillary ridge (Figure 2a). Nasal cavity is packed with merocel sponge/sterile gauze pieces soaked in same solution. No.15 Bard-Parker blade or Sickle knife is use to give incision which starts about 8-10 mms above the axilla of middle turbinate and continued anteriorly for 10 mms and then inferiorly till the level of junction of upper two-thirds and lower one-third of middle turbinate (Figure 2b). Nasal mucosal flap is then elevated using either a suction elevator or periosteum elevator to expose the maxillary ridge and frontal process of maxilla (Figure 2c). Nasal mucosal flap can be either removed just in front of uncinate process or left over to protect the middle turbinate and excise later. Lacrimal bone is punctured inferiorly and osteotomy with Kerrison punch is made (Figure 2d). The lacrimal sac is exposed completely from nasolacrimal duct to fundus and agger nasi is opened up (Figure 3e). Bowman lacrimal probe is passed and Crescent knife is used to open the sac in a book like manner to form anterior and posterior flap (Figure 3f). In cases of acute dacryocystitis and lacrimal abscess, purulent discharge can be seen from within the lacrimal sac and wall of the sac may be inflamed and thickened (Figure 3g). It is important to release all the intra-sac synechiae if present. Mitomycin-C (MMC) 0.02% is applied for 5 minutes and circumostial injection (0.1 ml of 0.02% MMC at each site) can be given as described.6 Silicone intubation (Figure 3h) and anterior nasal packing is done.

Marked resolution of symptoms and signs usually occurs within one day (Figure 4). Postoperative systemic antibiotic and analgesics, topical antibiotic eyedrops and nasal decongestants are given for 3-4 weeks. Nasal endoscopy with silicone tube removal and ostium evaluation is done at 4 weeks in OPD. Anatomical and functional success can be demonstrated with the functional endoscopic dye test (FEDT) (Figure 5). FEDT is performed by instilling fluorescein dye in conjunctival sac which is seen flowing through internal common opening and filling up the concavity of ostium through the endoscope.

DISCUSSION

Endoscopic DCR because of its approach from nasal side has many advantages when there is inflammation outside over the sac area. Nasal mucosa and bone is never inflamed in such cases and lacrimal sac marsupialization helps in internal drainage of abscess and purulent discharge thus relieving the symptoms and signs of acute attack. Percutaneous Incision and drainage of lacrimal abscess is a painful procedure and may promote intra-sac scarring and granulation tissue which may lead to failure of subsequent DCR surgery.

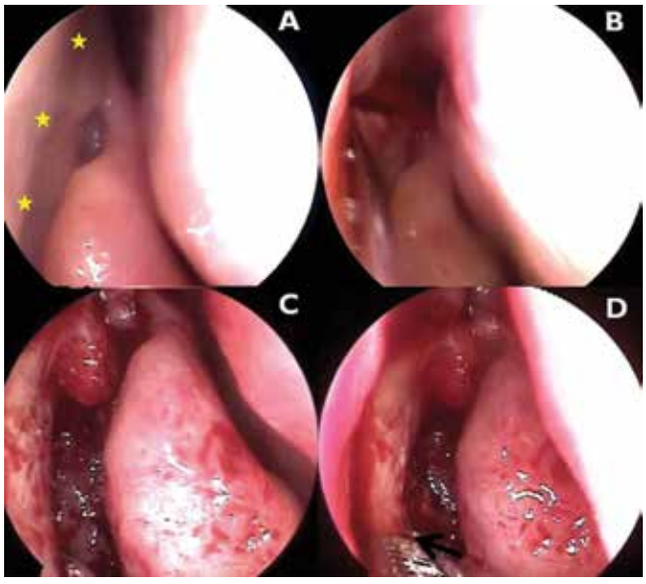

Figure 2A: *Endoscopy view of right nasal cavity showing the markings for local anesthesia infiltration B. Incision of nasal mucosa carried out with sickle knife C. Exposure of the frontal process of maxilla after mucosal flap removal D. Initiation of osteotomy inferiorly.*

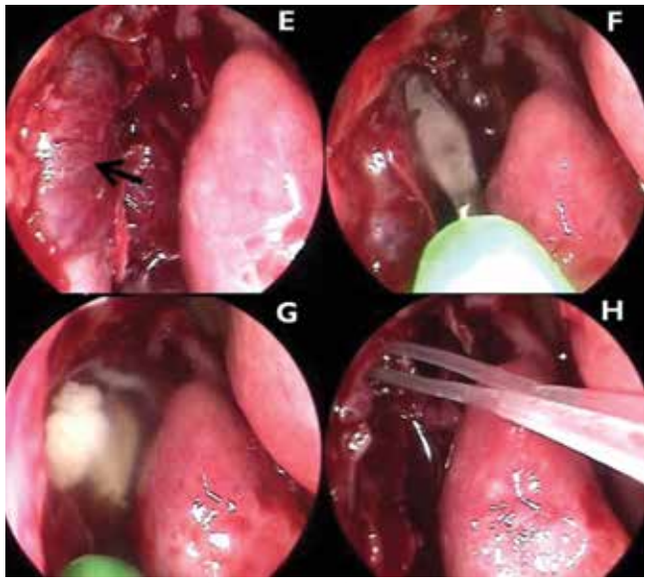

Figure 3E: *Exposure of lacrimal sac after osteotomy (note the inflamed angry looking sac in a case of acute dacryocystitis) F. Crescent knife use to make anterior and posterior flap G. Purulent discharge seen from lacrimal sac in a case of lacrimal abscess H. Appearance at the end of surgery, note the sac is open in book like fashion with silicone tube in situ.*

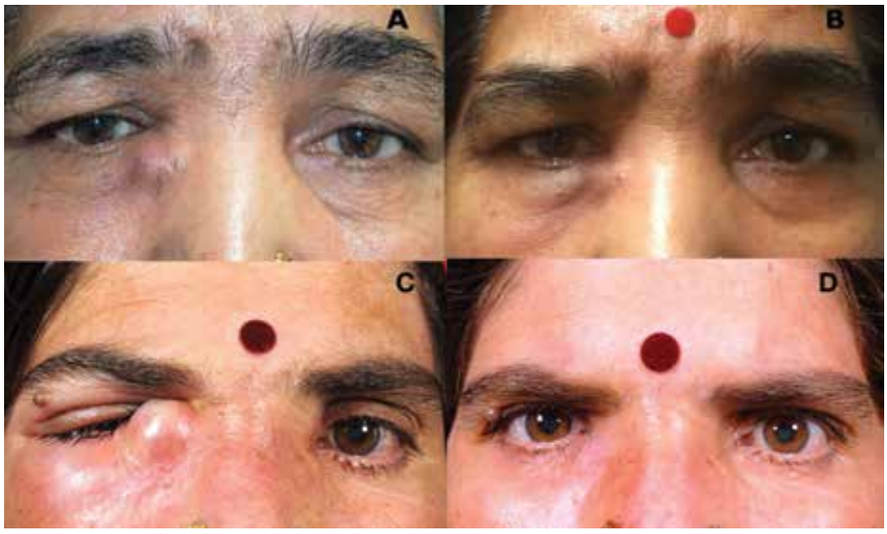

Figure 4: *Clinical photograph of a two cases presenting with acute dacryocystitis where En-DCR was performed A. Preoperative photograph of first case with Right acute dacryocystitis with evolving lacrimal abscess B. Postoperative photograph at first day showing marked resolution of swelling and erythema C. Preoperative photograph of second case presenting with Right non-resolving lacrimal abscess and left evolving acute dacryocystitis. D. Postoperative photograph at 3 days after bilateral simultaneous En-DCR showing resolution of signs bilaterally.*

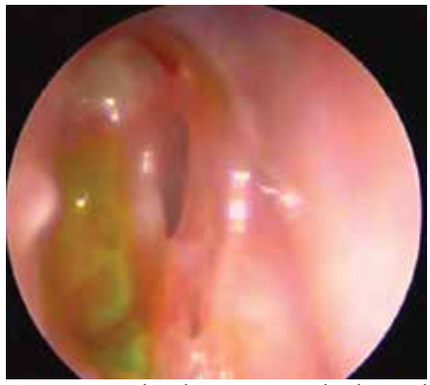

Figure 5: *Nasal endoscopy view of right nasal cavity of first case showing a large, well healed ostium with positive functional endoscopic dye test (Fluorescein seen) at 3 months follow up.*

It is known that episode of acute dacryocystitis is a risk factor for the failure of DCR surgery[10]. In a prospective randomized case series comparing delayed external DCR with early endoscopic endonasal DCR in acute dacryocystitis, success rate of 90% was seen with endoscopic approach compared to 66% with external approach ($p<0.05$). Mean time to the resolution of pain was 1 day for endoscopic DCR compared to 5.5 days for external DCR. Authors concluded that the endoscopic approach achieves higher success rate with minimal tissue manipulation and trauma to the lacrimal system. Functional success rate achieved with endoscopic endonasal DCR in acute dacryocystitis ranges from 90-95% at 6 months follow up and is 81% with long term follow up over one year. With the improved instrumentation such as use of powered drill and diamond burr for superior osteotomy, complete exposure of sac up to the fundus is achieved and adequate clearance around internal common opening is possible. Moreover, endoscopic endonasal approach helps to correct nasal abnormalities such as deviated nasal septum.

Use of adjunctives such as mitomycin-C and silicone intubation in DCR is known to increase the success rate especially in cases with risk of failure. There is no adverse effect of Mitomycin-C such as mucosal necrosis, increase infection or bleeding observed in cases of acute dacryocystitis so far. Similarly silicone intubation appears to be safe without any adverse effects such as increase granulation tissue, nidus for infection or canalicular cheese wiring.

Acute pediatric dacryocystitis needs special mention. Mostly acute dacryocystitis in cases of congenital nasolacrimal duct obstruction (CNLDO) in young infants is managed with systemic antibiotics and irrigation and probing under endoscopic guidance[12]. DCR is indicated in cases with recurrent acute attacks in which probing and adjunctive procedures such as silicone

intubation has failed, and for persistent cases of CNLDO requiring early intraocular surgery. En-DCR because of its advantages over external approach especially in acute dacryocystitis can be done in children. Debate exists over minimal age when it can be performed although it has been performed even up to 8 months to 1 year age[13,14]. There are certain challenges in performing En-DCR in children like narrow nasal cavity, limited working space and anatomical variations, but the results of En-DCR are comparable to external approach.

To conclude En-DCR is safe, effective and appear promising for the primary treatment of acute dacryocystitis with or without lacrimal abscess formation. Furthermore it leads to rapid resolution of symptoms without any recurrence and corrects the underlying nasolacrimal duct obstruction thus relieving epiphora.

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Recent Advancements in Ocularistry

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Abstract: *Ocularistry is an art and science of making custom ocular prosthesis and provides cosmetic rehabilitation to the patients with disfigured eye. Making of prosthesis has become much easier and better with newer techniques and machines. Recent advancements have enabled ocularists to make ultra thin scleral shell which can be worn over a disfigured cornea in pre-phthisical eye, a hollow prosthesis which is light in weight in comparison to the conventional prosthesis, painted conformers to replace the old transparent conformers and digital prosthesis which provides ultimate matching with the natural eye.*

Loss of an eye leaves a huge impact on the cosmetic appearance of a person. As ocularists, it is our responsibility to help restore maximum facial symmetry for these patients by making best possible custom ocular prosthesis. In early days, ready-made eyes were used extensively, but now custom-made eyes are preferred option for good cosmetic rehabilitation. Science and technology helps us to develop in all aspects of life, including custom ocular prosthesis. We have evolved from glass prosthesis to acrylic prosthesis, 2-D prosthesis to 3-D prosthesis and now digital prosthesis. Enhancing our techniques helps us to provide better options to patients with complications like hollow prosthesis for large sockets or painted conformers for the healing period post-operatively. Modern technology has also helped ocularists to provide better cosmetic options for one-eyed patients having phthisis bulbi and who do not want to undergo surgery. Ultra-thin scleral shell is a good option for such cases where asymptomatic eye used to be removed just for cosmetic rehabilitation[1].

Technology has evolved to a great extent and ocularistry has evolved with much better options for the care of one-eyed patients. Following are the recent advancements added in Ocularistry.

ULTRA THIN SCLERAL SHELL

Scleral shell is an ultra-thin prosthetic eye designed to be worn over a discolored and/or disfigured eye. The prosthesis covers the entire surface of the cosmetically blemished eye, restoring its natural appearance.

Case Report

This is a case of 21 years old female who was diagnosed right eye phthisis bulbi post infection (Figure 1). Patient was given two options for cosmetic rehabilitation. Option 1) Trial of scleral shell, and in case of failure then option 2) Evisceration + orbital implant followed by custom prosthesis. Patient wanted to retain her eye and original movements so the plan of making an ultra-thin scleral shell was executed by taking impression of the eye by alginate. This impression was converted into a very thin scleral shell by scleral press method in which a 1 mm thin acrylic sheet is pressed over the impression to make the masterpiece of the shell[2]. This masterpiece was further painted according to the other eye of the patient. This scleral shell after

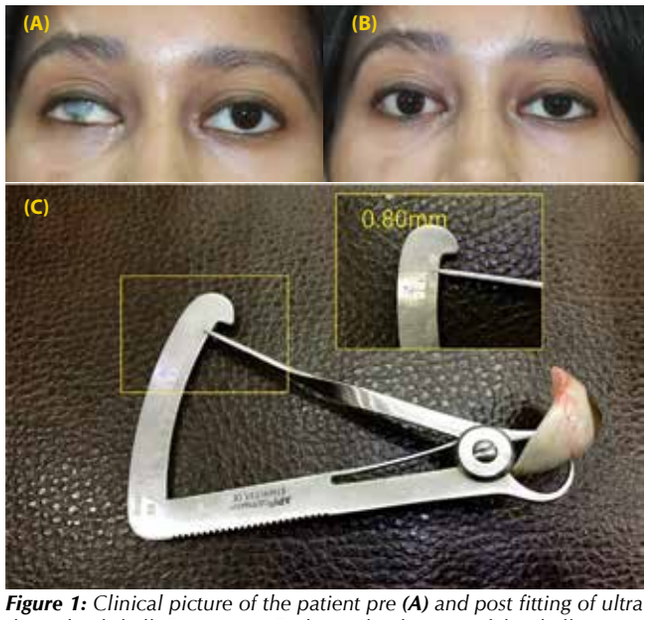

Figure 1: *Clinical picture of the patient pre* ***(A)*** *and post fitting of ultra thin scleral shell* ***(B)****, picture* ***(C)*** *shows the thinness of the shell.*

optical grade polishing was dispensed to the patient. Patient achieved satisfying result without compromising her existing eye. Ultra-thin scleral shell is a good option for phthisis bulbi that provides best movements over the disfigured eye.

HOLLOW OCULAR PROSTHESIS

Hollow prosthesis is thick ocular prosthesis, which is made hollow from inside without compromising its anterior and posterior dimensions.

Sometimes there are cases where there is a large volume loss in the socket and further volume replacement surgeries are contraindicated. Such conditions are like socket without implant, severe injury, and severe phthisis bulbi, lid sparing exenteration etc. The impression of such sockets is usually thick, bi-convex and bulky. Prosthesis made using such an impressions becomes heavy. A heavy prosthesis may cause lower lid laxity, lid sagging and can cause secondary ectropion and shallow lower fornix in future. A hollow prosthesis can be prepared which has same dimensions as conventional prosthesis but hollow from inside, which serves with around 30-45% less weight. A hollow prosthesis can replace the need for surgical volume replacement where another surgery is contraindicated. The major limitation of hollow prosthesis is

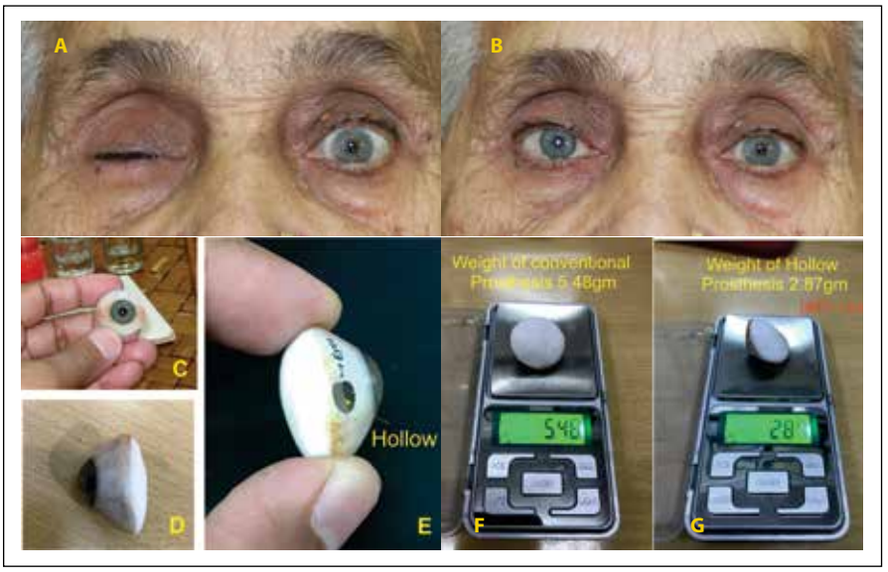

Figure 2: *Clinical picture of the patient pre **(A)** and post **(B)** fitting of Hollow Prosthesis. Picture (**C&D**) shows the conventional prosthesis & (**E**) shows the hollowness of the prosthesis. In picture (**F**) weight of conventional prosthesis 5.48 gram and in (**G**) weight of hollow prosthesis which is 2.87 grams is shown.*

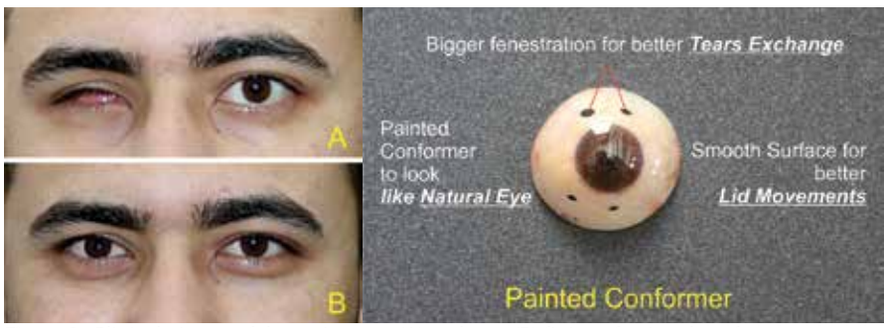

Figure 3: *Clinical picture of the patient pre **(A)** and post **(B)** fitting of painted conformer.*

limitation of movements.

A hollow prosthesis provides similar appearance like conventional prosthesis but less in weight in the socket, which helps in prevention of lower lid complications.

Case Report

This is a case of 67 years old female who was diagnosed right eye anophthalmos post enucleation due to endophthalmitis (Figure 2). There was no orbital implant in her eye and another surgery was not preferred in her case due to her health issues. A custom made ocular prosthesis after taking the impression of socket was made. Weight of the prosthesis was 5.48 grams, which is much higher than the standard range of an ocular prosthesis. Hollow prosthesis (Figure 2E) in which first anterior part was prepared and painted then the posterior part was fused to anterior part without filling the central thickness. This hollow prosthesis has the same dimensions of anterior and posterior surface as that of conventional prosthesis. The weight of this prosthesis was 2.87 grams, which was 48% lighter than the conventional prosthesis (5.48 grams).

Patient was more comfortable with the new lightweight prosthesis. Hollow prosthesis has an advantage in preventing the socket complications in such cases.

PAINTED CONFORMER

Painted conformers are like normal conventional conformers, which are being used in socket, post enucleation or evisceration but they are painted according to the patient's fellow eye. Painted conformers provide cosmetic rehabilitation immediately after the surgery.

Transparent conformers are widely used post eye removal surgeries. It provides benefits such as protection of the wound, minimizing the changes in the socket size and conformation, preventing scar tissue contractures from distorting the socket bed[3] and maintaining the fornix depth[4].

Now days, an ocularist fabricates the painted conformer for the patient which can replace the old transparent conformer. This has several advantages over the transparent conformer by camouflaging the anophthalmic eye immediately after the surgery. With the help of painted conformer, a patient can be socially active after 1 week of surgery or some of them can resume their job and require less leave for surgical healing. Caution- the painted conformers are only for providing cosmetic rehabilitation during the post-surgical healing period. All post-surgical precautions, use of medication, rest and all other activities are same as with normal transparent conformers. Such painted conformers should be changed with the custom ocular prosthesis as per the schedule visit.

Case Report

This is a case of 27 years old male who was diagnosed right eye anophthalmos post evisceration with orbital implantation due to painful blind eye (Figure 3). Patient was delaying his surgery due to his limited leaves. Patient was not willing to join his work with dark glasses, as his work was more in customer dealing. He was suggested to go for a painted conformer after one week of his surgery, which is painted according to the fellow eye. Patient was instructed that this is not a customized prosthesis and doesn't have advantages of a prosthesis that is made after taking the impression of the socket. It is used primarily post-surgery to avoid the adhesion of the conjunctiva during the healing period. Patient was also advised to use protective glasses and to follow all the post-operative instructions given by his surgeon. Painted conformer provides all benefits of conventional conformers over the cosmetically blemished eye and restores its natural appearance during the healing period.

DIGITAL IRIS PROSTHESIS

It is a recently developed ocular prosthesis, which is prepared with the help of digitally printed iris.

The world is inclining towards the digital technology and in last few years ophthalmology has seen the advantages of digital processing and so is ocularistry. Now ocularists are able to make ocular prosthesis with digital technology, which provides as close as natural look of the iris. In this technology a high-resolution picture of the patients' normal eye taken with the help of additional macro lens over the DSLR camera with ring flash.

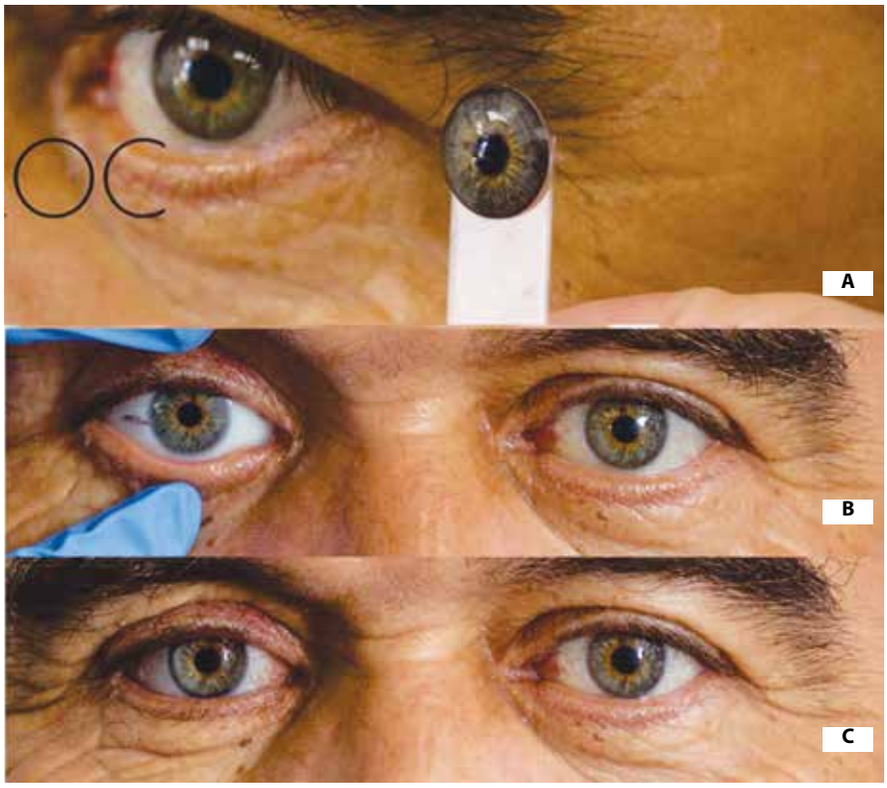

Figure 4(A): *digitally printed iris button matching with the other eye.* ***(B)*** *Shows the master mold and* ***(C)*** *final result with digital iris prosthesis. Image courtesy: Focus Lab, Russia*

This picture is processed in Photoshop to remove the reflections from the pictures and then this image is printed over the photo paper. This photo print is used to make the iris button; it is fused in the prosthesis, which is further painted for scleral tinting. Digital Iris prosthesis is a new technique, which will help ocularists to make prosthesis more accurate and faster in future.

CONCLUSION

Disfigured eye due to any reason causes cosmetic blemish. A carefully made customized ocular prosthesis helps such eyes to look life like. Recent advancements in ocularistry can help in providing better quality service and can reduce the limitation of the management of disfigured eye. With the help of digital prosthesis an ocularist would be able to deliver better and faster service in future.

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Ocular Surface Squamous Neoplasia(OSSN): The Small Malignancy that Looms at Large

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Summary: *The Ocular Surface Squamous Neoplasia (OSSN) is not an uncommon condition. The differential diagnosis of OSSN includes pterygium, pinguecula, papilloma, episcleritis, amelanotic nevus, malignant melanoma. The diagnosis of OSSN is mostly clinical and hence it becomes imperative to differentiate it from the relatively benign conditions. The management of OSSN is dependent on the clinical grading and depth of invasion of the neoplasia. Topical chemotherapy and surgical excision are the two modes of treatment in our armamentarium. Recent studies have shown an excellent response to topical Interferon α2b (IFN- α2b).*

The Ocular Surface Squamous Neoplasia (OSSN) is a broad term including conjunctival intraepithelial neoplastic lesions (CIN) and invasive squamous cell carcinoma (SCC) of conjunctiva and cornea[1]. CIN includes varying grades of dysplasia, ranging from mild, moderate, severe dysplasia to carcinoma in situ[2].Their importance lies in the fact that they can mimic benign lesions like pterygium or even chronic conjunctivitis.

The term Ocular Surface Squamous Neoplasia (OSSN) was coined by LEE and HIRST which has three grades:

I. Benign dysplasia:
   - Papilloma
   - Pseudotheliomatous hyperplasia
   - Benign hereditary intraepithelial dyskeratosis

II. Preinvasive OSSN.
   - Conjunctival/corneal carcinoma in situ

III. Invasive OSSN
   - Squamous carcinoma
   - Mucoepidermoid carcinoma

Morphologically there are three types of lesions gelatinous, nodular or diffuse[3].

CASE PROFILE

CASE 1

(Figure 1A) shows a small OSSN of pigmented variety measuring 3mm x 3mm and abutting the nasal limbus. Patient was started on Topical IFN- α2b (Inj. Relifer on where 0.5ml contains 3 million IU and is mixed with 2.5 ml solution of polyvinyl alcohol eye drops to obtain a final concentration of 1 million IU/ml) 4 times daily and clinical resolution was evident at 4 weeks and complete resolution occurred by 12 weeks (Figure 1B). The Topical IFN- α2b was then tapered off and the patient has remained asymptomatic without evidence of any recurrence since then.

CASE 2

(Figure 2A) shows another small OSSN of pigmented variety measuring 4mm x 4mm on nasal side. Patient was started on same Topical IFN- α2b regimen and clinical resolution was evident at 6 weeks (Figure 2B) with complete resolution by 12 weeks (Figure 2C).

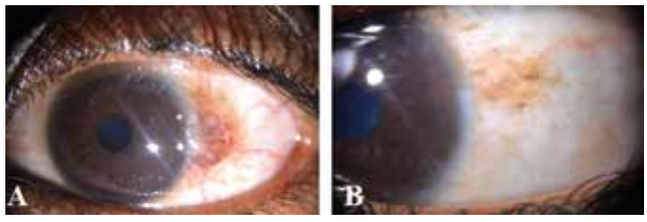

Figure 1A: *Small OSSN of pigmented variety measuring 3mm x 3mm and abutting the nasal limbus.* ***Figure 1B:*** *Complete resolution at 12 weeks.*

Figure 2A: *Small OSSN of pigmented variety measuring 4mm x 4mm on nasal side.****Figure 2B:*** *Clinical resolution was evident at 6 weeks.* ***Figure 2C:*** *Complete resolution by 12 weeks.*

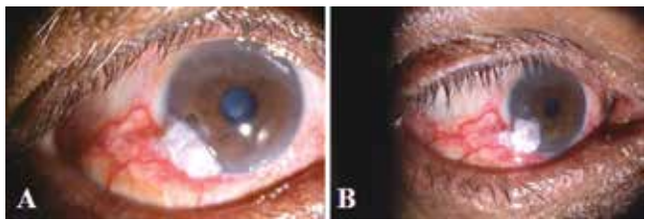

Figure 3A: *Small OSSN measuring 6 mm x 4mm which also involved the cornea.* ***Figure 3B:*** *No improvement even after 12 weeks of topical immunotherapy.*

CASE 3

(Figure 3A) shows a patient who had a small OSSN measuring 6 mm x 4mm which also involved the cornea. Patient was started on Topical IFN- α2b 4 times daily using same regimen. However even after 12 weeks, the improvement was minimal (Figure 3B). It was excised en-bloc using no-touch

technique and histophathology proved it to be microinvasive squamous cell carcinoma. Patient was given adjuvent chemotherapy in the form of topical Mitomycin C and there is no evidence of any recurrence even after 6 months of treatment.

COMMENT

The IFN- α2b is low-molecular weight glycoprotein, produced by leukocytes, has antineoplastic and antiviral properties[4]. IFN- α2b can be administered via topical drops, subconjunctival injections or intralesional injections. Topical IFN- α2b is preferred for OSSN, which are relative thinner, for complete tumour control (immunotherapy) while combination therapy with topical and injection IFN- α2b is used for partial reduction of thicker and extensive OSSN (immunoreduction).

With topical application, clinical resolution usually takes place with a mean treatment time of about 12 weeks. Subconjunctival injection combined with topical IFN- α2b for noninvasive OSSN has a faster time to resolution, about six weeks.

A typical regimen consists of topical IFN- α2b drops with a concentration of 1-3 million IU/mL, applied four times daily; or subconjunctival injections of 3 million IU/0.5 mL, administered weekly. Intralesional IFN- α2b (10 million IU/ml) has been used in giant OSSN (≥6 limbal clock-hours).

Topical IFN- α2b application leads to complete resolution with good prognosis in most Benign and Pre-invasive OSSN i.e. Type I and II. In Invasive i.e Type III or very large pre invasive OSSN a trial of topical chemotherapy should be attempted, which may reduce the size of the lesion making it amenable to complete surgical excision.

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High Flow Arterio-Venous Malformation: Which Imaging Technique is Better?

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Summary: *High flow vascular malformations comprise of arteriovenous malformations and acquired arteriovenous shunts/fistula. They can be quiescient, expansile, complicated or decompensated depending on their clinical stage. In management of high flow malformations, the key is to identify the feeder vessels. Digital Subtraction Angiography (DSA) is the ideal diagnostic technique to identify the feeder vessels prior to endovascular coiling or direct ligation of these vessels.*

Vascular malformations need to be understood differently from the acquired arteriovenous shunts and fistulae in terms of pathogenesis and hemodynamics[1]. They derive either from the arterial system, venous system, lymphatic system or a combination of the above.

The Orbital Society has classified Vascular malformations as[2]:

Type I: NO FLOW comprising of Lymphangiomas and Combined Veno-Lymphatic malformations

Type II: LOW FLOW comprising of predominantly venous malformations encompassing distensible, non-distensible and the combined lymphatic-venous distensible malformations.

Type III: HIGH FLOW comprised of arteriovenous malformations and the acquired arteriovenous shunts.

Based on clinical activity, SCHOBINGER[3] classified "High Flow AV Malformations" as :

I: QUIESCENT: Staining pink blue, Warm to touch, AV shunt on Doppler

II: EXPANSILE: Besides above findings, also show enlargement, pulsatile to touch,

thrill on palpation, bruit audible along with tortuous veins

III: Have further COMPLICATIONS manifested as dystrophic skin, ulceration, persistent pain, bleeding, necrosis and bony lytic lesions.

IV: DECOMPENSATION: The shunt finally leads to congestive cardiac failure due to increased cardiac output and left ventricular hypertrophy.

CASE PROFILE

A 28-year-old female, presented to us with a painless pulsatile mass located at the root of the nose and partially extending onto the medial orbit on either side (Figure 1). Thrill was palpable over the mass and on auscultation a clear bruit was heard. The mass in left orbit and on lateral wall of nose was clearly visible on MRI scans (Figure 2).

Patient underwent pre-op CT-Angiography, which confirmed a high-flow AVM localized to glabella and medial orbit with bilateral facial arteries as possible feeder vessels (Figure 3).

Combined surgery with ENT surgeons was undertaken where both the facial arteries were ligated, right one near the ala of nose and left one near the mandible (Figure 4). There was no perceptible change in the pulsations of the mass. The further surgical intervention was deferred. Post facial artery ligation a repeat CT angiography showed no change in size of AVM (Figure 5). The left facial artery and also vein were ligated again at the level of ala of nose, which led to an increase in the size of the mass and in the thrust of the pulsations. This confirmed that the facial vein is the draining vessel for the high-flow AVM

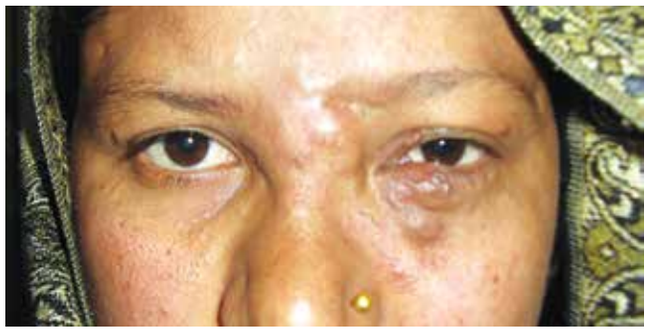

Figure 1: *Pre-op clinical photo showing painless pulsatile mass located at the root of the nose and partially extending onto the medial orbit on either side.*

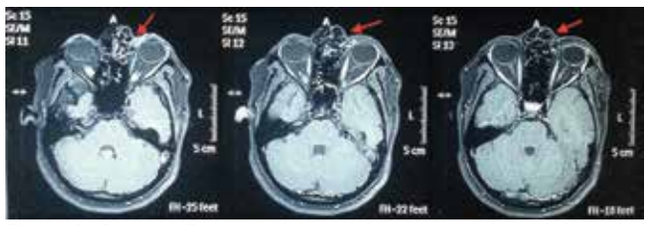

Figure 2: *Pre-op MRI scans showing the mass involving left medial orbit and lateral wall of nose.*

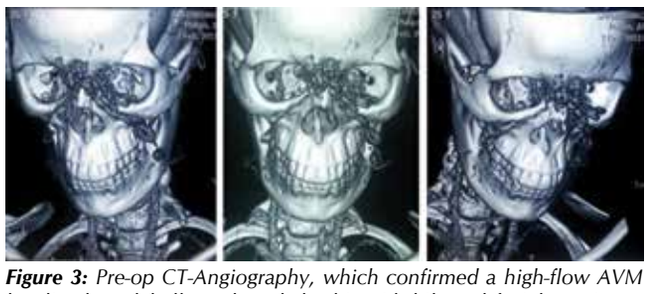

Figure 3: *Pre-op CT-Angiography, which confirmed a high-flow AVM localized to glabella and medial orbit with bilateral facial arteries as possible feeder vessel.*

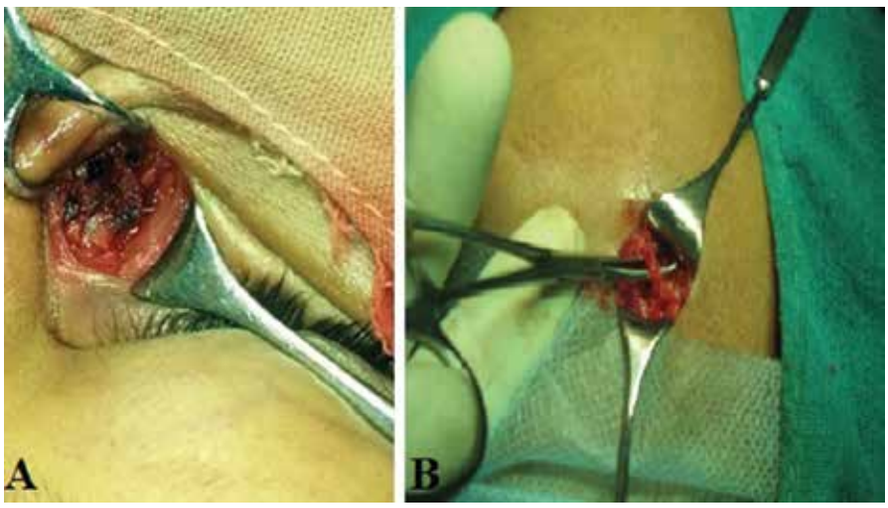

Figure 4: *Intra-op clinical photo of combined surgery with ENT surgeons, where both the facial arteries were ligated.* ***4(A):*** *Right facial artery ligation near the ala of nose.* ***4(B):*** *Left facial artery ligation near the mandible.*

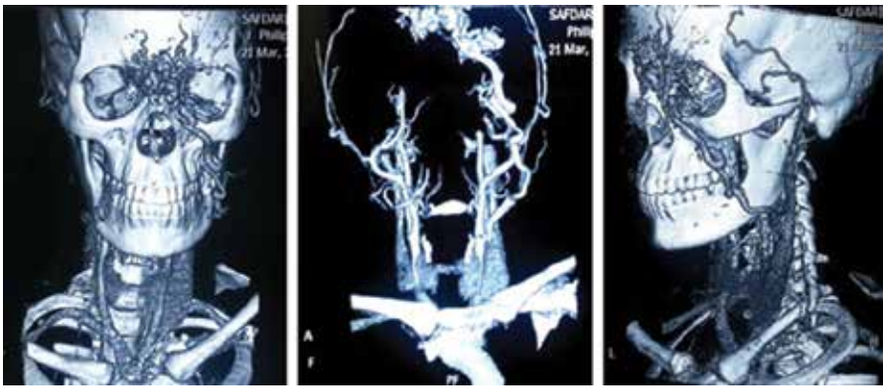

Figure 5: *Post facial artery ligation a repeat CT angiography showed no change in size of AVM.*

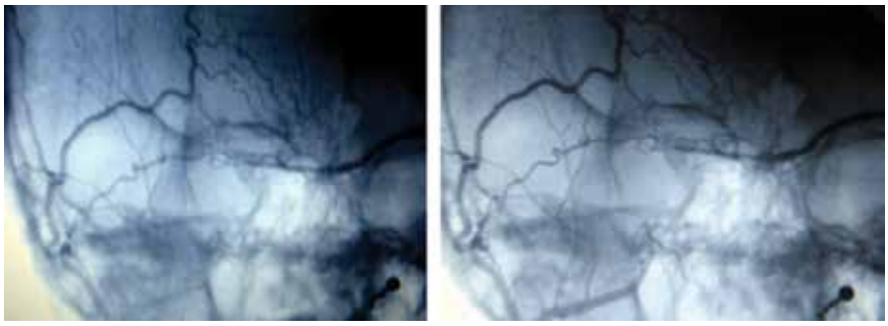

Figure 6: *Digital-Subtraction-Angiography (DSA) clearly indicating aberrant branches from superficial temporal artery and External Carotid Artery as main feeder vessels along with multiple feeder vessels from other branches of External Carotid Artery.*

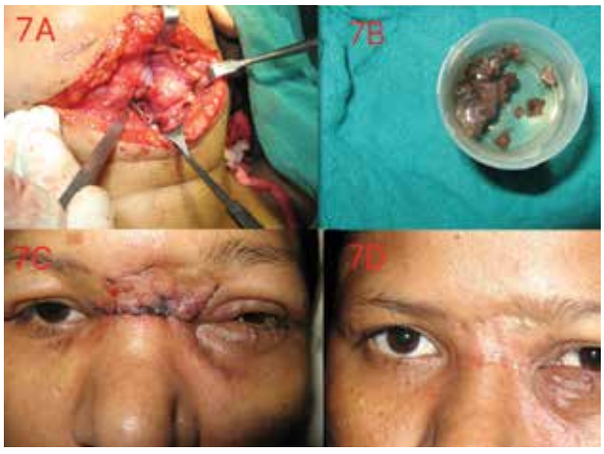

Figure 7: ***(A)*** *Left External Carotid ligation.* ***(B)*** *Excised mass with cyano-acrylate glue* ***(C)*** *Post op day one.* ***(D)*** *Clinical photograph at two months post op.*

and facial artery is not the feeder vessel.

Finally Digital-Subtraction-Angiography (DSA) was done which clearly indicated aberrant branches from Left superficial temporal artery and External Carotid Artery (ECA) as main feeder vessel along with multiple feeder vessels from other branches of External Carotid Artery (Figure 6).

Due to functional difficulties in intravascular coiling of multiple small feeder vessels[4,5], External Carotid Artery was ligated resulting in prompt decrease in size of mass and decrease in strength of pulsations and the mass was excised at the same sitting using Cyano-acrylate glue (Figure 7).

COMMENT

Since CT-Angiography and MR-Angiography may not be able to accurately delineate feeder vessels in a case of High Flow AV Malformation, it could be imperative to perform Digital Subtraction Angiography (DSA), which will be able to identify feeder vessels much more accurately.

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Orbital Apex Syndrome

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Abstract: *Tuberculosis is a significant health problem in India. While presentations of tuberculosis can be variable, orbital involvement is uncommon. There are certain inflammatory syndromes in orbit that present with specific clinical features. Because of the crowding of important neural structures in orbit, it is almost always impossible to do a invasive biopsy and one has to rely on clinical and radiological findings to make a diagnosis. The authors hereby take this opportunity to describe a case of orbital apex syndrome secondary to tuberculosis in which timely diagnosis and medical treatment completely restored vision and movements.*

Tuberculosis both pulmonary and extra pulmonary constitutes a significant health problem across the world especially in developing countries. While pulmonary tuberculosis is the most common presentation, the disease can present with involvement of virtually any body tissue and therefore a variety of presentations are possible. While pulmonary tuberculosis can be diagnosed easily with the help of imaging and microscopy, diagnosing extra pulmonary tuberculosis can pose a challenge to the treating physician. Although the most common ophthalmic presentation of tuberculosis is choroiditis or uveitis, a myriad of presentations can be seen. This often places the ophthalmologist at the diagnostic forefront to diagnose systemic tuberculosis based on ocular findings. One of the rare ocular presentation of tuberculosis is with multiple cranial nerve palsy in which timely diagnosis and management can prevent vision loss or restore vision.

CASE REPORT

A 32 year old hindu married male patient presented with left sided frontal headache since two months. It was followed by binocular diplopia and drooping of left upper lid after a gap of one month. Twenty days later diminution of vision developed in left eye. There was no history of fever, weight loss, anorexia, tuberculosis or contact with a tuberculosis patient, or trauma. There was however a history of a painless swelling with discharging sinuses at angle of right jaw seventeen years back that subsided with treatment for one year. Details of treatment received were not available. General physical examination was unremarkable except for the presence of lymphadenopathy. Firm, non tender, matted lymph nodes were present in bilateral submandibular, right supraclavicular, axillary and inguinal regions. Ocular examination revealed visual acuity of hand movements in left eye with total external ophthalmoplegia and complete ptosis of upper lid (Figure 1). There was complete limitation of monocular as well as binocular movements in all cardinal position of gaze (Figure 2). There was also a grade three RAPD in left eye. Fundus examination showed a hyperaemic and edematous optic disc with peripapillary haemorrhages and blurring of all optic disc margins. Corneal sensation were reduced in left eye, so were the sensations in the forehead and upper lid on left side while sensations in the maxillary and mandibular division of trigeminal nerve were intact. Hertels exophthalmometry was 21 mm and 23 mm in right and left eye respectively at base of 110 mm. Examination of right eye was normal.

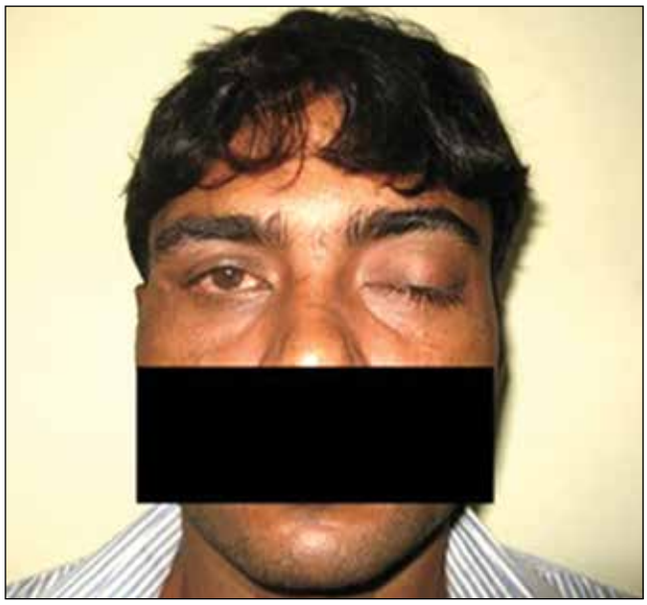

Figure 1: *Clinical photo showing complete ptosis of left eye upper lid.*

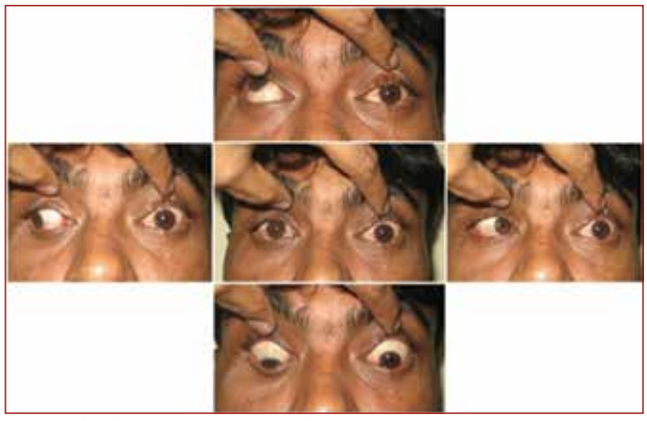

Figure 2: *Clinical photo showing cardinal gazes to demonstrate complete limitation of extraocular movements in left eye.*

Blood investigations showed a lymphoctytosis on differential leucocyte counts and a raised ESR levels. Patient was tested negative for HIV, HbsAg, c-ANCA and p-ANCA antibodies. Ultrasonography of orbit and x-ray chest were normal. Montoux

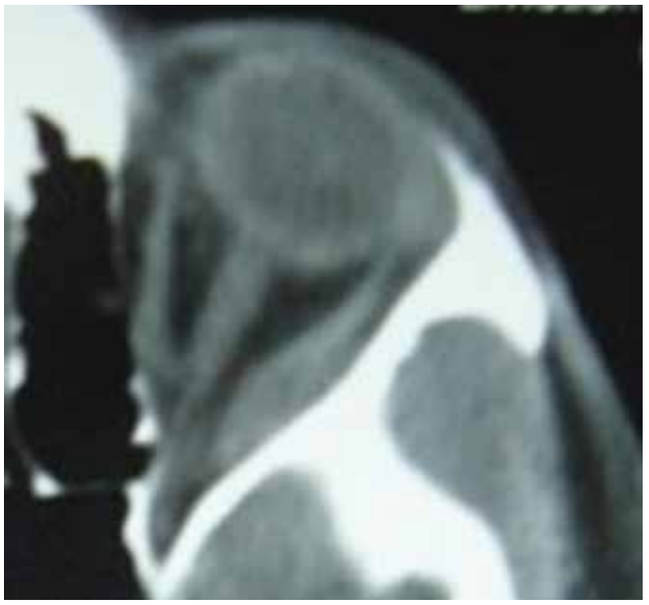

Figure 3: *CT scan in axial view showing bulky left lateral rectus muscle.*

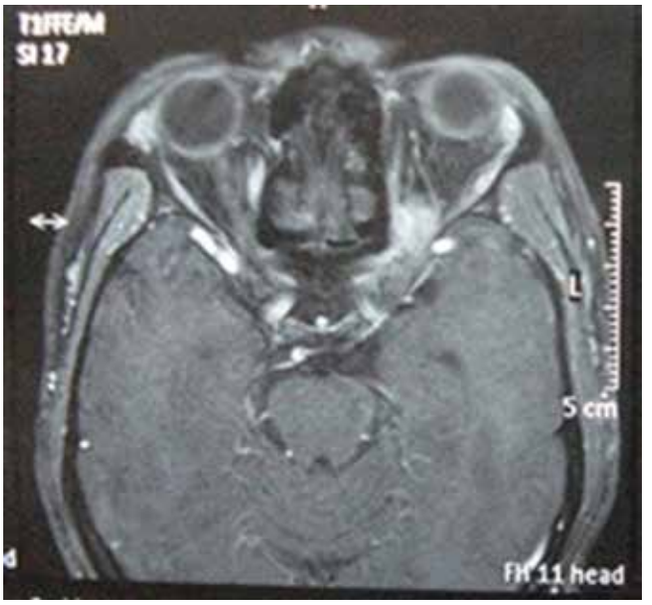

Figure 4: *MRI axial scan showing soft tissue shadow at orbital apex extending upto cavernous sinus with thickened optic nerve sheath.*

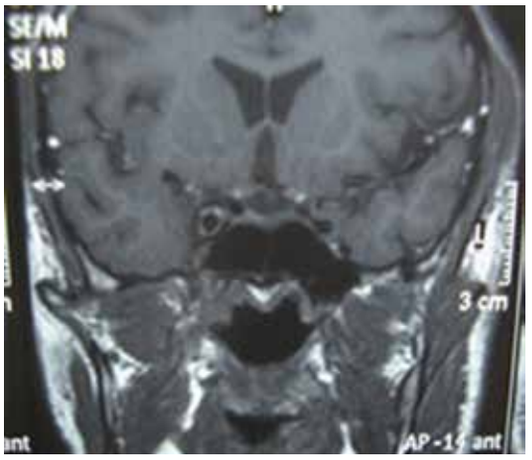

Figure 5: *MRI coronal section showing compression of internal carotid artery.*

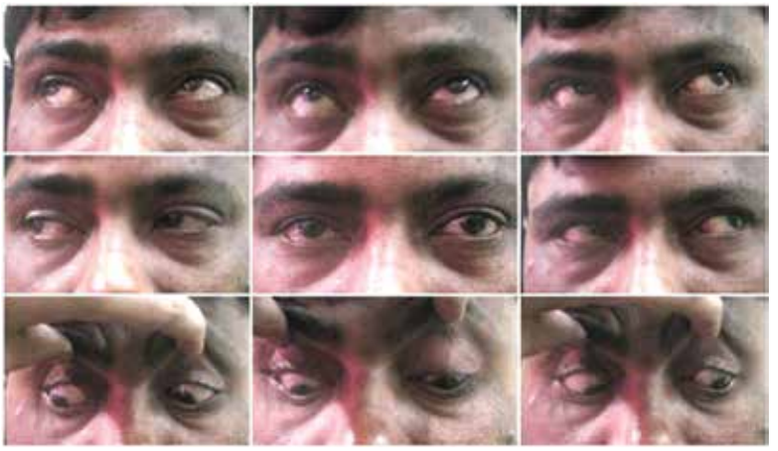

Figure 6: *No limitation of movements in left eye 6 months post treatment.*

test was strongly positive with reading of 35 mm by 30 mm after 48 hours of injection. Computed tomography scan showed an enlarged lateral rectus muscle and no other significant anomaly (Figure 3). Magnetic resonance imaging of orbits and brain was done that showed soft tissue thickening in the left orbital apex region extending up to the left cavernous sinus with diffuse thickening of the optic nerve sheath and a bulky lateral rectus muscle (Figure 4). Coronal section through the middle cavernous sinus area showed compression and decrease in the calibre of internal carotid artery by the soft tissue thickening on left side as compared to right side (Figure 5). A lymph node biopsy from right supraclavicular region showed caseous necrosis and calcification on histopathological examination. A diagnosis of tubercular orbital apex syndrome was made and the patient was started on category one anti tubercular therapy (2HRZE+4HR) and oral prednisolone 1 mg/kg body weight/ day. One month after treatment visual acuity improved to 6/9 on snellens chart, colour vision and contrast sensitivity were normal, however there was partial improvement in ptosis and extraocular movements. The AKT was continued for six months with gradual tapering of oral steroids over next one month at 10mg per week. Follow up at six months post treatment showed return of visual acuity to 6/6 on snellens, normal extraocular movements and absence of ptosis (Figure 6). Repeat MRI scanning showed complete resolution of the soft tissue swelling from orbital apex and cavernous sinus and return of internal carotid calibre to normal (Figure 7). Lymph node biopsy was repeated that showed absence of caseation and was replaced by fibrofatty tissue.

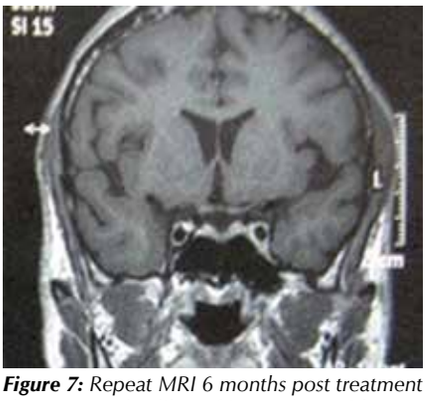

Figure 7: *Repeat MRI 6 months post treatment shows normal calibre of internal carotid artery within cavernous sinus.*

DISCUSSION

Any patient who presents with multiple cranial nerve involvement needs to be evaluated carefully to direct appropriate investigations and determine appropriate intervention. There are very few locations where

Table 1: Algorithm to locate the site of lesion in case of multiple cranial nerve palsy

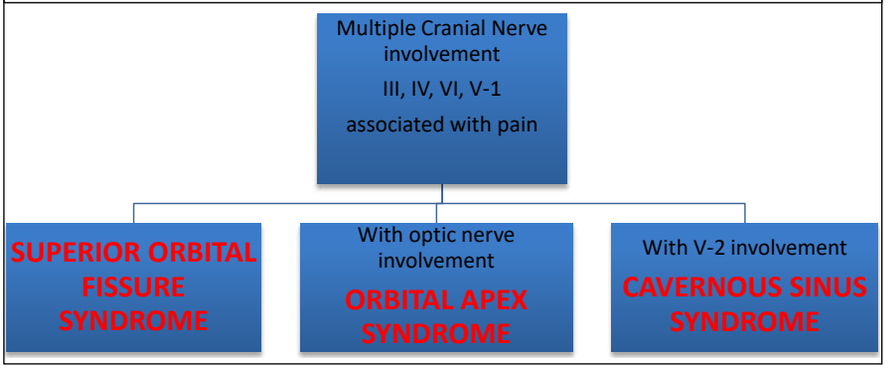

Table 2: Causes of orbital apex syndrome

INFLAMMATORY	Sarcoidosis Systemic lupus erythematosus Churg–Strauss syndrome Wegener granulomatosis THS Giant cell arteritis Orbital inflammatory pseudotumor Thyroid orbitopathy
INFECTIOUS	Fungi- Aspergillosis, Mucormycosis Bacteria- Mycobacterium tuberculosis
NEOPLASTIC	Head and neck tumors Neural tumors Non-Hodgkin lymphoma, leukemia Metastatic lesions
VASCULAR	Carotid cavernous fistula Cavernous sinus thrombosis
IATROGENIC/ TRAUMATIC	Fractures

3rd to 6th cranial nerves are present in proximity to each other and therefore can be simultaneously involved. Three important clinical syndromes that can present with these cranial nerve involvement are superior orbital fissure syndrome (SOFS), orbital apex syndrome (OAS) and cavernous sinus syndrome (CSS). Although each syndrome refers to a particular anatomical site, they form a continuous spectrum and any one condition can lead to other two. There are however subtle clinical signs that can help the clinician in determining the exact anatomical site of involvement and to direct the investigation or imaging. Table 1 represents a simple algorithm to differentiate between the three syndromes[1]

Causes for orbital apex syndrome may be inflammatory, infectious, vascular, neoplastic, iatrogenic (Table 2)[2]. Tuberculosis being a chronic granulomatous infection remains an important cause especially in Indian population owing to its high prevalence. Since these anatomical sites do not allow for biopsy owing to close proximity of vital structures, one has to rely on secondary clinical features and imaging to make the diagnosis. Causal association with tuberculosis therefore remains presumptive. Imaging modality of choice is magnetic resonance imaging. CT is inferior to MRI in imaging of orbital apex and cavernous sinus region and is done only when MRI is contraindicated. MRI characteristically shows enlarged optic nerve sheath and extraocular muscles, hyperintense soft tissue shadows at apex or cavernous sinus, narrowing of internal carotid artery within the sinus[3].

Treatment is aimed at reducing the inflammation with the help of systemic corticosteroids. In cases suspected to be due to tuberculosis steroids should be started under cover of anti tubercular therapy.

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When Benign Turns Fierce

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Abstract: Purpose: *We report a case of Orbital Schwannoma in a middle aged man who underwent sudden expansion in a short span of 15 days leading to globe subluxation, corneo-scleral melt and loss of light perception in the affected eye. The tumor otherwise was a very slow growing one and the patient was practically symptomless for 2 years except for a noticeable upper lid mass of right eye. Schwannomas are known to be slowly growing, painless masses which may undergo rapid expansion of size due to hemorrhage or degeneration. An explosive increase in size in this short a duration is rare. Complete removal of tumor was achieved surgically .The enucleated socket was restored cosmetically using a PMMA orbital implant and custom made ocular prosthesis.*

Key words: *Orbital Tumor, Giant Schwannoma, globe subluxation*

Schwannomas constitute approximately 1% of orbital tumours[1,2]. They are known to be slow growing, painless, benign tumors of the peripheral nerves. Most schwannomas arise from branches of either the supraorbital or supratrochlear nerves and hence produce downward displacement of the globe. With a wide range of clinical presentations, histopathology helps in confirming the diagnosis.

CASE REPORT

A 37 year old male patient, farmer by occupation, resident of Uttar Pradesh, India, presented to us in December 2015 with severe pain, diminution of vision, redness and protrusion of right eye of 15 days duration. He further revealed that he noticed slight protrusion of right eye, which was gradually progressive and painless since 2 years with worsening of symptoms over last 15 days. On examination his right globe was subluxated outwards and downwards with severe conjunctival congestion, full thickness corneal abscess, and a diffuse reddish mass prolapsing through the superomedial fornix from the orbit (Figure 1a). The boundaries of the mass could not be palpated and the superior portion of the mass was flush with superior orbital rim. The forniceal conjunctival vessels over the mass were dilated (Figure 1b). His right eye had no light perception. Left eye visual acuity was 6/6 and both anterior and posterior segment were within normal limits. Patient was started on Intravenous Cefotaxim 1 gm and Amikacin 500 mg 12 hourly for 5 days. Topical Moxifloxacin (0.5%) and Natamycin (5%) eye drop hourly and Itraconazole (1%) and Carboxy methylcellulose (1%) eye drops 6 times/ day. A Contrast enhanced CT Orbit and head was ordered.

Figure 1: *Supero-medial orbital mass prolapsing through the fornix with globe subluxation and corneo-scleral melt.*

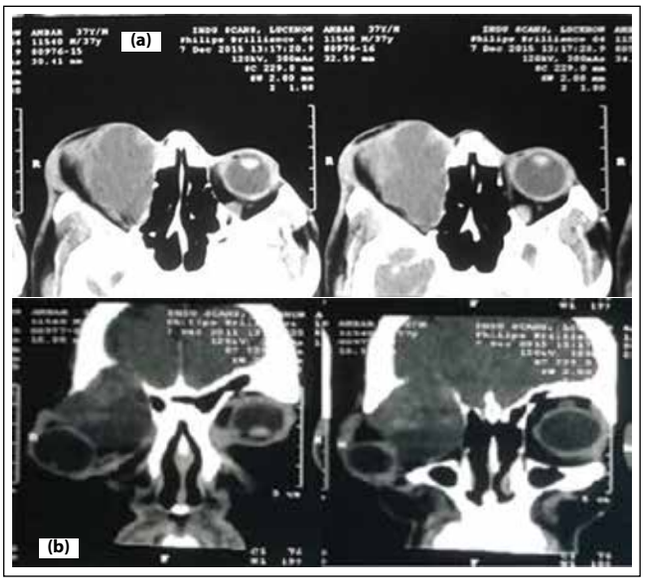

Figure 2(a): *Axial sections* ***(b)*** *Coronal sections showing well-defined, moderately enhancing, ovoid mass occupying the entire medial two thirds of superior and medial extraconal orbital compartment with areas of hypodensity within it.*

The CT images revealed a well defined moderately enhancing ovoid mass occupying the entire medial two thirds of superior and medial extraconal orbital compartment with areas of hypodensity within it (Figure 2a,b).

A medial orbitotomy with tumor excision and enucleation with implant was planned and nil visual prognosis explained to patient. The tumor was approached via a lid split orbitotomy and was found to be well encapsulated and extraconal (Figure 3 a,b). The eye was enucleated using Myoconjunctival technique and tumor excised by blunt dissection from surrounding tissue. The lid incision was closed in three layers. There was excessive oozing from orbital apex. The socket was packed with gauze. On first postoperative day there was no active oozing from socket and a 18mm PMMA implant was placed posterior to tenons and incision closed in two layers. On gross examination the mass was conical in shape and well encapsulated (Figure 4a,b). The histopathology report showed typical Antoni A and B areas, Verocay Bodies along with cholesterol clefts, Foreign Body Giant cells and Foamy histiocytes suggesting a Schwannoma with degeneration and inflammation (Figure

Figure 3(a): *Lid split anterior orbitotomy with exposed tumor* ***(b)*** *The mass after enucleation.*

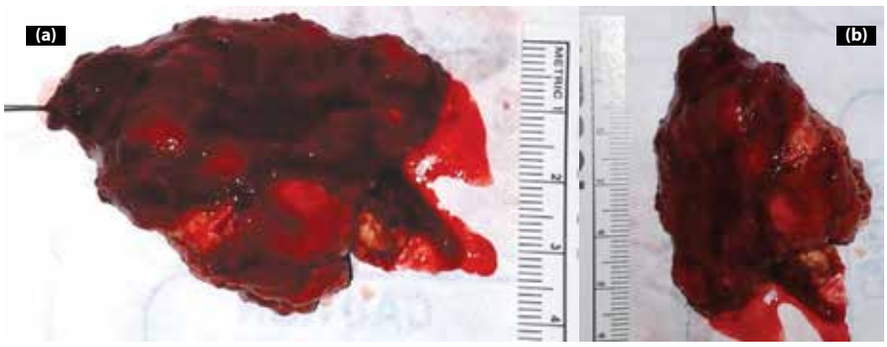

Figure 4: *Excised mass measuring 40mm x 60mm.*

and benign. Previous such reports have mentioned cystic degenerations with exacerbation of signs in long standing tumors[3]. A 15 days short history with sudden expansion due to degeneration with globe subluxation is a rare finding and can often end up as a clinical diagnosis of malignancy. Immunohistochemistry aids identification when inflammation and hemorrhage etc. distort the Histopathology picture[4].

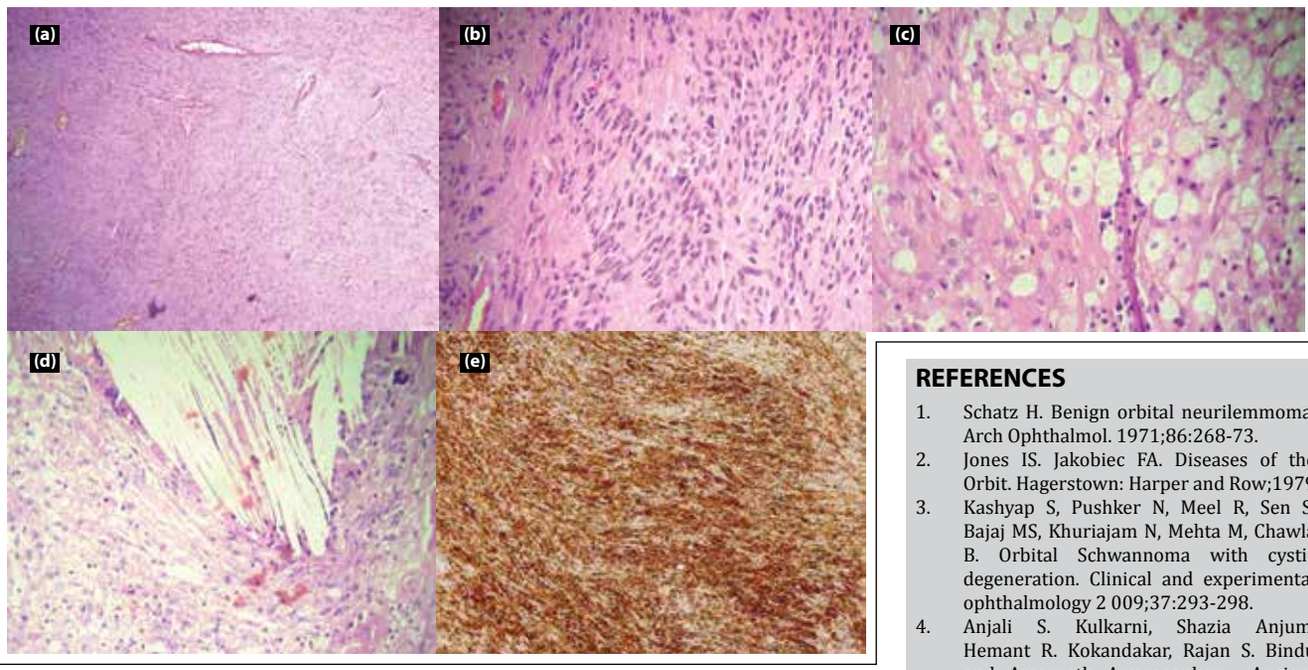

Figure 5: *Histopathology report showing typical* ***(a)*** *Antoni A bodies* ***(b)*** *Antoni B bodies* ***(c)*** *Foreign Body Giant cells and Foamy histiocytes* ***(d)*** *Verocay Bodies along with cholesterol clefts* ***(e)*** *IHC positive for S100.*

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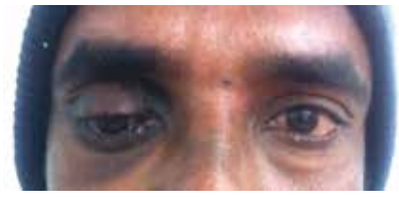

Figure 6: *Follow up at 6 weeks post surgery with wax trial model of custom made ocular prosthesis*

5a to d). Immunohistochemistry was positive for S 100 confirming the mass to be schwannoma (Figure 5e). A custom made ocular prosthesis (COP) was placed at 6 weeks post operative follow up by the ocularist (Figure 6).

DISCUSSION

Schwannomas are known to be gradually progressive, well encapsulated

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DOS Times Quiz 2017-18

Episode-4

Last date: Completed responses to reach the DOS Office by e-mail or mail before 5 pm on 25th April, 2018

Q1. Pagetoid spread is feature of
A) Squamous cell carcinoma
B) Sebaceous gland carcinoma
C) Basal cell carcinoma
D) Xanthogranuloma

Q2. PHOX 2A gene is implicated in which type of Congenital Fibrosis of Extra Ocular Muscles (CFEOM)
A) Type 1
B) Type 2
C) Type 3
D) Tukel Syndrome

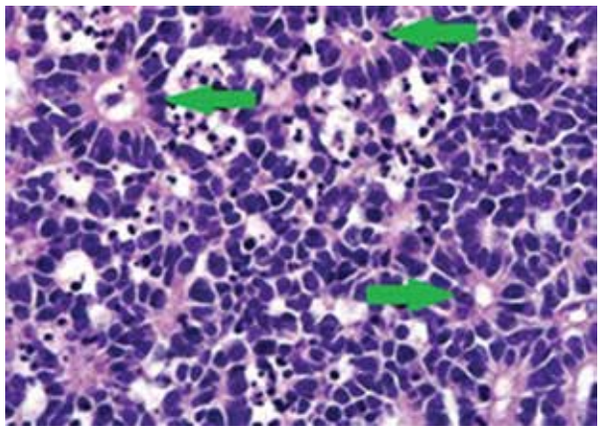
Q3. Chemotherapy for OSSN include all of the following except-
A) IFN α
B) IFN β
C) Mitomycin C
D) 5-Fluorouracil

Q4. Botulinum toxin serotype used for therapeutic purposes
A) Type A
B) Type D
C) Type C1
D) Type C2

Q5. Size of optic canal is reduced in-
A) Ophthalmic artery aneurysm
B) Optic nerve glioma
C) Optic nerve menigioma
D) Sphenoidal meningioma

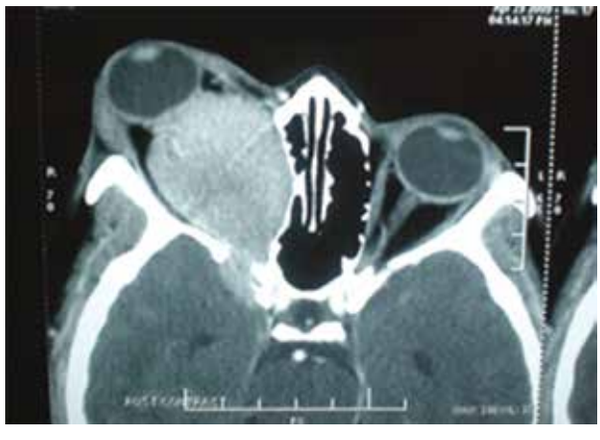
Q6. Which of the following dermal fillers is a collagen stimulator-
A) Hyaluronic acid
B) Bovine collagen
C) Poly L-lactic acid
D) Polyacrylamide

Q7. Identify the histological feature in the slide below-
A) Flexner Wintersteiner rosettes
B) Fleurettes
C) Homer Wright rosettes
D) Pseudorosette

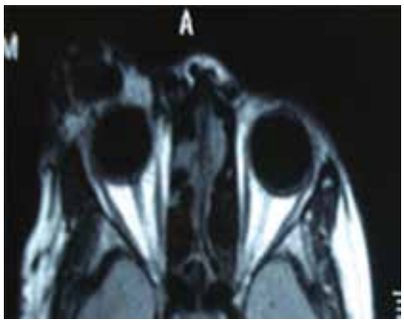


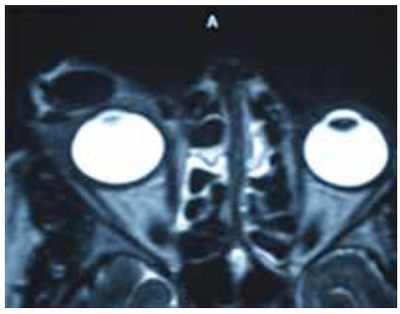
Q8. Which of the childhood tumors is associated with opsoclonus-
A) Retinocytoma
B) Neuroblastoma
C) Ewing sarcoma
D) Rhabdomyosarcoma

Q9. 7 year old child presented with unilateral proptosis and CT scan as shown in figure below. What is the most probable diagnosis-



Q10. T1 and T2 enhanced MRI images of a 40 year old female with upper lid mass are shown below. Identify the lesion-





Compiled by:

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Dr. Komal Saluja

DOS Times Quiz Rules

1. DOS Times Quiz will now feature as **5 Episodes** (Episode 1: July-August, Episode 2: September – October, Episode 3: November – December, Episode 4: January – February, Episode 5: March – April). Entries will have to be emailed before the last date mentioned in the contest questions form. Late entries will not be entertained.
2. Please email (as scanned PDF Only) completed responses for the quiz along with details of the contestant **filled in and signed** to dostimes10@gmail.com (with cc to dosrecords@gmail.com) or mail to DOS Times Quiz, Dr. Subhash Dadeya, Room No. 205, 2nd Floor, OPD Block, Guru Nanak Eye Centre, Maharaja Ranjit Singh Marg, New Delhi.
3. Nonmembers may also send in their entries but will be required to send along with their completed entries, the completed membership application (with the required documents) to enroll as member. Failing this their entries into the contest will not be considered.
4. Contestants are requested to attempt all the 5 episodes of the Quiz contest and send in their applications within the date specified. No entries will be entertained after the last date. The scores of each contestant for all 5 episodes together will be compiled at the end of episode 5 and the winner will be announced in the DOS Annual Conference in April 2018. In the event of more than one winning contestants, a draw of lots will decide the winner. Winner of each episode will also be published in the next episode along with the previous episode answers.
5. Please write to dostimes10@gmail.com or dosrecords@gmail.com for further clarifications if any.

Q. No. Completed Responses for DOS Times Quiz: Episode 4

1. _____ 6. _____
2. _____ 7. _____
3. _____ 8. _____
4. _____ 9. _____
5. _____ 10. _____

Contestant Details

Name: _____ Degree: _____

Designation:_____ Address:_____

_____ State _____ Pin _____

Mobile No: _____ DOS Membership no: _____

Email ID: _____Signature: _____

DOS Crossword

Episode-4

Correspondence to:
Dr. Komal Saluja
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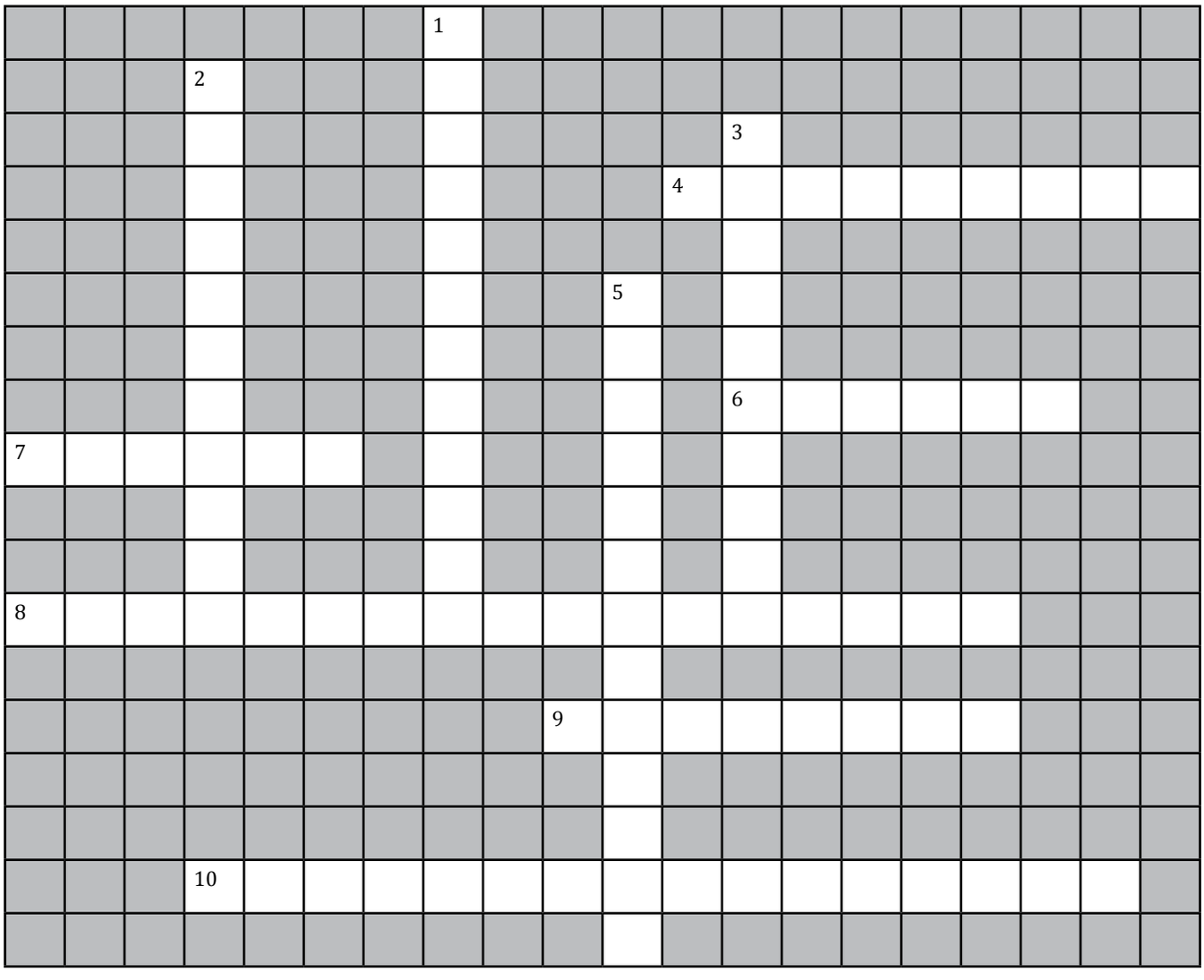

DOWN

1. No Flow arteriovenous malformation(12)
2. Nevus Flammeus(6,5)
3. Tram track sign(10)
5. Bilateral symmetrical enlargement of palpebral aperture(13)

ACROSS

4. Intra-arterial chemotherapy for retinoblastoma(9)
6. Most common cause of Unilateral proptosis in adults(6)
7. X-ray view for orbital floor(6)
8. Collar stud appearance(9,8)
9. Classification for orbital cellulitis(8)
10. Hemorrhagic cystitis(16)

Sclerosing and Embolisation agents used in Ophthalmology

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INTRODUCTION

There are many treatment modalities reported in the literature for hemangiomas, such as intralesional and systemic corticosteroid treatment, surgical excision, thermocauterization, laser photocoagulation, and sclerotherapy. Each of the treatment modalities has its own risks and advantages.

Advantages of sclerotherapy to other hemangioma treatment modalities include it being very simple and safe to apply, affordable, and readily available, with most of this being due to not requiring special equipment for application and having no need for hospitalization of the patient.

SCLEROSING AGENTS

Agent	Dosage	Adverse effects	Contraindications
Sodium morrhuate 5%	0.2 -2.1 ml intralesional	Aching or burning sensation at injection site with discolouration, redness, swelling or ulceration; allergic reactions. Pulmonary embolism, anaphylaxis	Allergy, DVT, vasculitis, uncontrolled DM, thyrotoxicosis, TB, neoplasms, asthma, sepsis, blood dyscrasias, acute respiratory or skin disease, bedridden patients, pregnant/lactatating mothers.
Sodium tetradecyl sulphate (sortradecol) 0.1-3%	1.5 -2 ml	Nausea, vomiting, cough, shortness of breath, pulmonary embolism, pruritus, conjunctival redness, injection site problems (hyperpigmentation, ulcer or necrosis following extravasation), hypersensitivity reaction and anaphylaxis	hypersensitivity, pregnancy, thrombophlebitis, hyperthyroidism. Acute infections, TB, prolonged recumbency, cardiac insuffi ciency, uncontrolled diabetes, arterial disease and asthma.
Polidocanol 1%	1-6 ml	Injection site problems (hematoma, irritation, discolouration, pain, pruritus, thrombosis, ischemia), anaphylaxis, cardiac arrest.	Hypersensitivity and acute thromboembolic disease.
Bleomycin	0.5 mg/kg body wt, not exceeding 10 units at a time. (1 mg = 1 unit) 1 U/ ml of normal saline with 2% lignocaine.	Pain, redness, at injection site, fever , chills, hypersensitivity reaction	Hypersensitivity.
Picibanil (OK-432)	0.01-0.02mg/ ml normal saline. 1-2 ml of solution after aspiration of contents from the lesion.	Fever, injection site inflammation, hypersensitivity reactions.	Hypersensitivity
Ethanol	95% / 98% dehydrated, Denatures proteins and causes thrombosis in stagnant channels. Maximum dose 1ml/kg or 60 ml in one procedure.	Local neurolysis, tissue necrosis, Systemic- CNS depression, hypoglycaemia, arrhythmia	

EMBOLISATION AGENTS			
Poly Vinyl Alcohol	Particles of size 50-2000 microns mixed with saline in suspension form	Direct mechanical occlusion and induction of foreign body reaction and granulation tissue formation thereby obstructing inflow.	Tendency to flocculate, Non uniform delivery leading to occlusion of delivery catheter or more proximal vascular occlusion then intended.
Embospheres	Hydrophilic acrylic based spheres in sterile prefilled syringe.	More compressible, Less catheter clogging, Better penetration into vascular bed and more uniform embolisation.	Higher per vial cost
N- Butyl Cyanoacrylate (NBCA)	Adhesive embolic agent	Polymerisation on contact with ionic environment. Mechanical effect plus chronic inflammatory response	Occasionaly distal embolisation and ischaemia may occur especially in high flow lesions.
Coils	2-30 mm in diameter made of stainless steel and platinum. Dacron fibres increase the thrombogenicity. Platinum coils are MRI compatible.	Mechanical occlusion plus Dacron fibres acting as nidus for thrombus formation.	Sizing of coils is crucial. Smaller coils can embolize distally. Larger coils may recoil into proximal larger vessel.
Balloons	Silicon balloons	Highly efficacious for high flow lesions. Cause mechanical occlusion of vessel with thrombus formation	Balloons may deflate over time bus vessels remain occluded due to associated thrombosis.

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