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.*

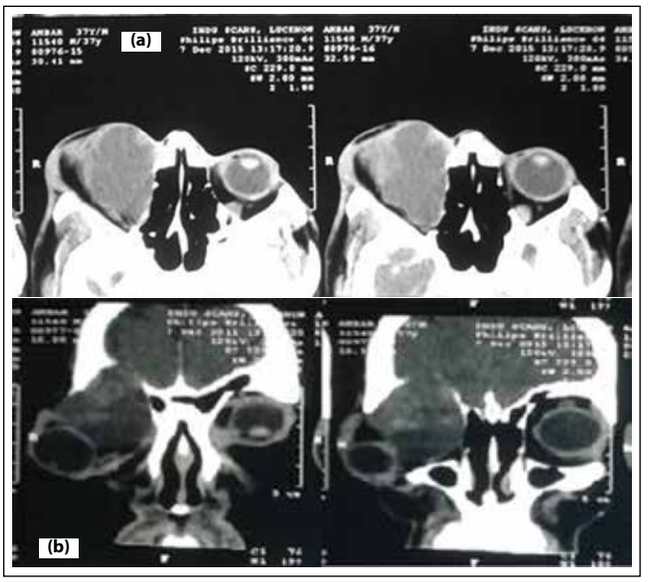

Figure 2(a): *Axial sections* ***(b)*** *Coronal sections showing well-defined, moderately enhancing, ovoid mass occupying the entire medial two thirds of superior and medial extraconal orbital compartment with areas of hypodensity within it.*

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.*

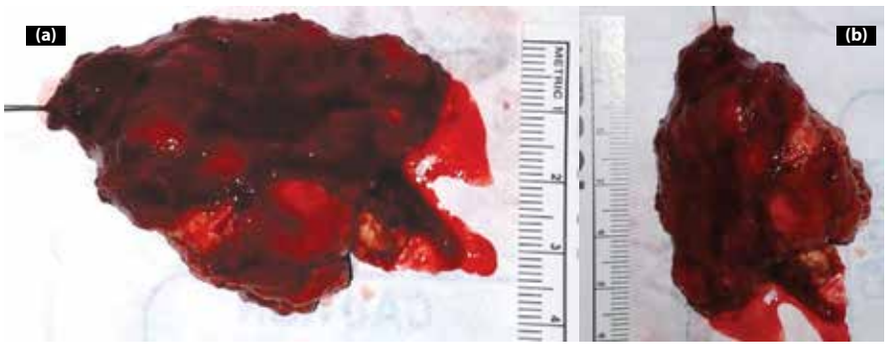

Figure 4: *Excised mass measuring 40mm x 60mm.*

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].

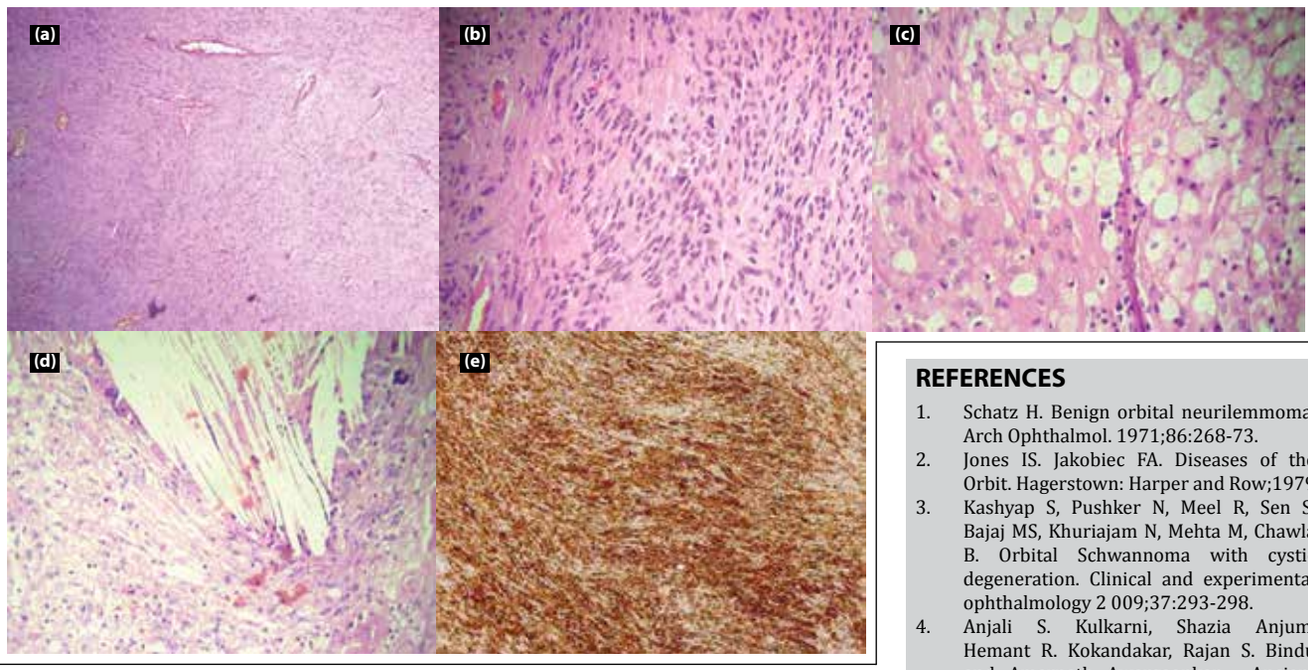

Figure 5: *Histopathology report showing typical* ***(a)*** *Antoni A bodies* ***(b)*** *Antoni B bodies* ***(c)*** *Foreign Body Giant cells and Foamy histiocytes* ***(d)*** *Verocay Bodies along with cholesterol clefts* ***(e)*** *IHC positive for S100.*

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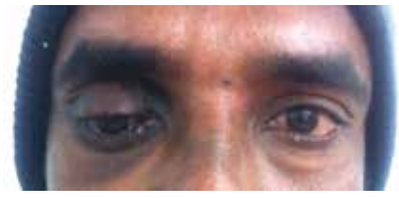

Figure 6: *Follow up at 6 weeks post surgery with wax trial model of custom made ocular prosthesis*

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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