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.

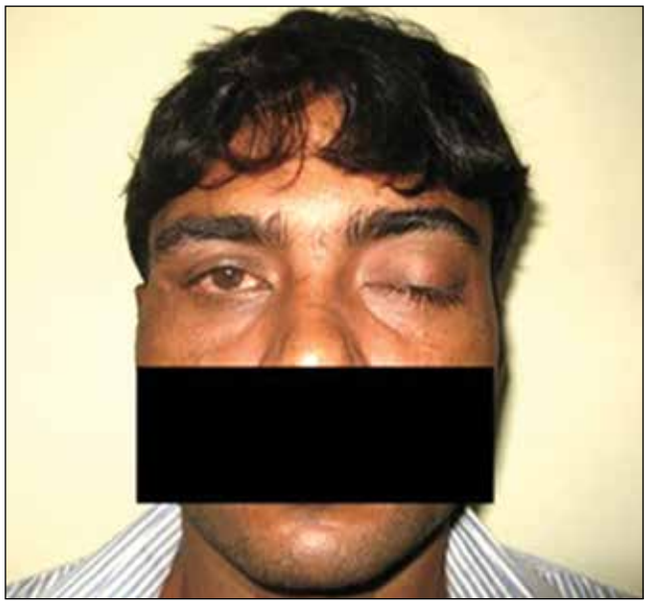

Figure 1: *Clinical photo showing complete ptosis of left eye upper lid.*

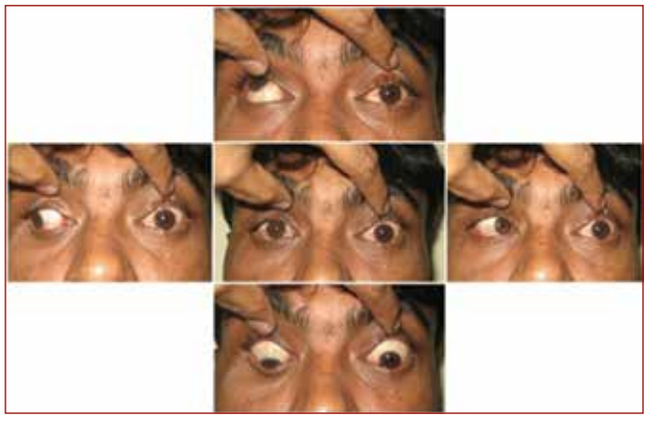

Figure 2: *Clinical photo showing cardinal gazes to demonstrate complete limitation of extraocular movements in left eye.*

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

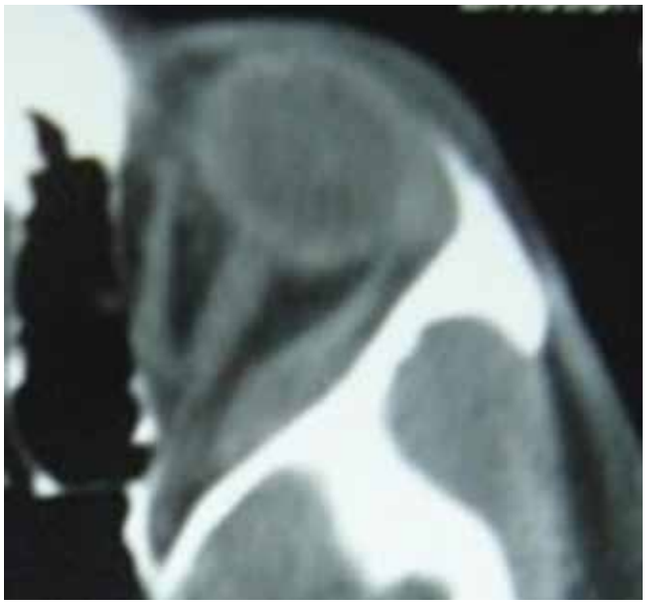

Figure 3: *CT scan in axial view showing bulky left lateral rectus muscle.*

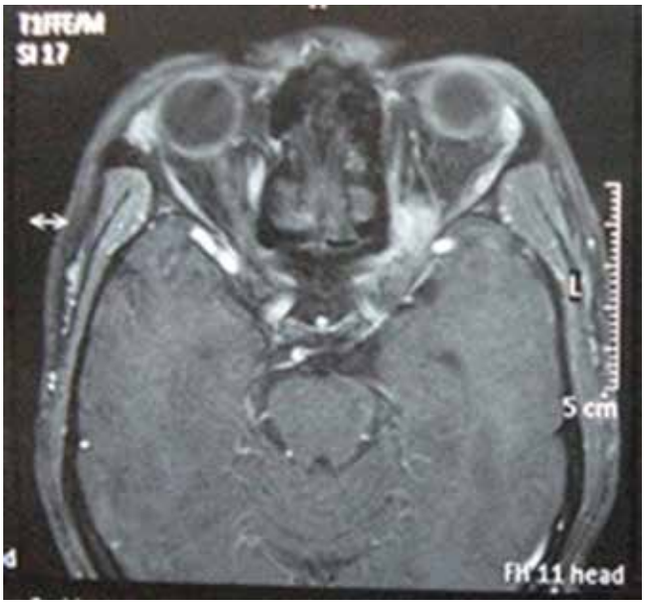

Figure 4: *MRI axial scan showing soft tissue shadow at orbital apex extending upto cavernous sinus with thickened optic nerve sheath.*

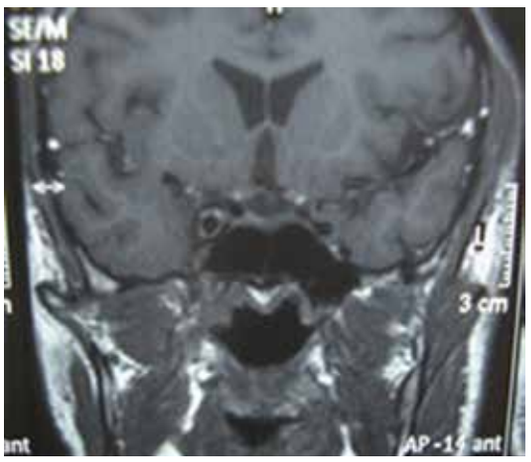

Figure 5: *MRI coronal section showing compression of internal carotid artery.*

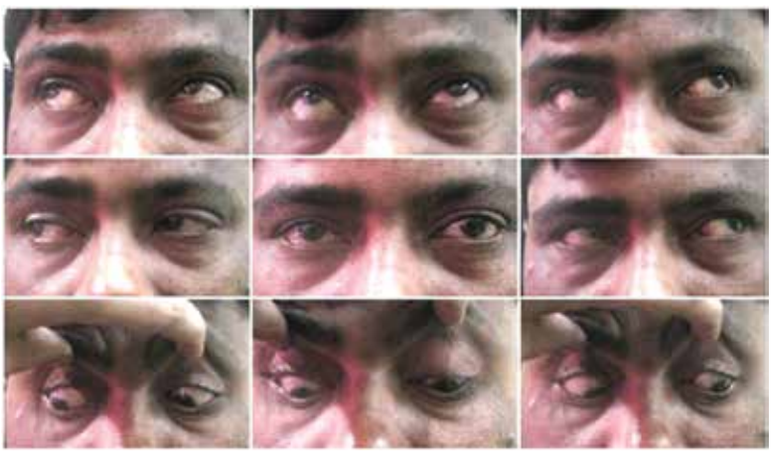

Figure 6: *No limitation of movements in left eye 6 months post treatment.*

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.

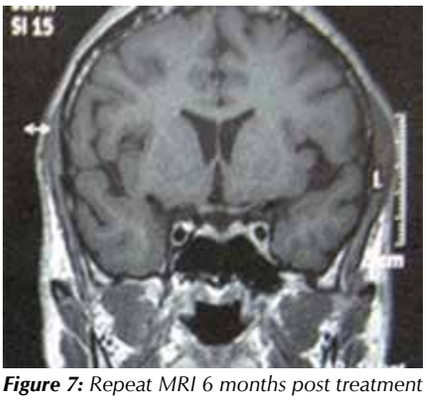

Figure 7: *Repeat MRI 6 months post treatment shows normal calibre of internal carotid artery within cavernous sinus.*

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

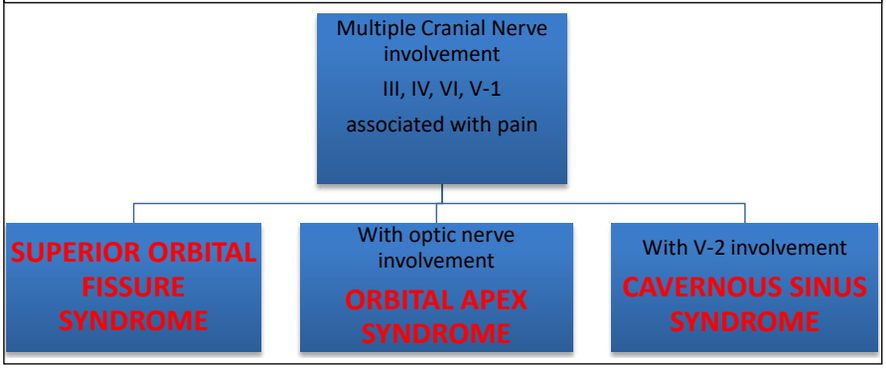

Table 2: Causes of orbital apex syndrome

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.

REFERENCES

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