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CASE REPORT

Orbital apex syndrome in a child

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Abstract A 8-year-old male presented with visual loss, diplopia, ptosis, pain behind the left eye, facial numbness and vomiting of one week duration. The ophthalmological, neurological and radiological examination showed a lesion of the left orbital apex with extension into the cavernous sinus. Examination of the nose and paranasal sinuses did not reveal any abnormality. Transnasal Endoscopic orbital decompression was performed and inflamed granulation tissue found in the orbital apex was removed. Microbiology showed fungal elements which on culture grew Aspergillosis flavus. Antifungal therapy with new generation oral drug (voriconazole) resulted in complete resolution of symptoms. Relevant literature is reviewed and discussed.

Keywords Aspergillosis · Orbital apex syndrome · Mycoses · Cavernous sinus thrombosis

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Introduction

Orbital Apex Syndrome consists of paralysis of all the three nerves [3, 4, 6] supplying the extra ocular muscles and a sensory deficit in the distribution of the first division of the trigeminal nerve, combined with an optic nerve lesion [1]. Fungal lesions involving the orbital apex are the least common cause of this syndrome. Atleast 4 forms of fungal infection of the sinonasal tract have been recognized: Allergic, Non-invasive, Invasive and Fulminating. Infection usually spreads from the paranasal sinuses to the orbital Apex. Aspergillus is a fungus found in soil and organic debris. It usually presents as a localized disease of the lungs and the paranasal sinuses and mainly affects immunocompromised individuals. But the invasive form can also affect normal or mildly immunocompromised individuals. Orbital Aspergillosis is a rare condition.

We report a case of *Aspergillus Flavus* infection, which caused an orbital apex syndrome with cavernous sinus thrombosis in a child.

Case

A 8-year-old boy presented to the neurosurgery dept. in Nov 2004 with complaints of blurring of vision in left eye following a head injury sustained 2 weeks earlier. He was evaluated and diagnosed to have a left carotico-cavernous fistula for which he underwent embolization with stenting. Patient improved symptomatically with no blurring of vision and was discharged from the hospital. In April 2005, he was readmitted in neurosurgery with history of progressive loss of vision with diplopia of 1 week duration. At that time his vision was restricted to perception of light alone. MRI Brain and Paranasal sinuses revealed an enhancing lesion involving the left orbital apex in T2 weighted images. However all the paranasal sinuses were found to be normal. In consultation with the ophthalmologist a diagnosis of ?

granulomatous lesion was made and the patient treated with corticosteroids and antibiotics for a period of one month. Following this, his vision partially improved and he was discharged from the hospital. In August, 2005 he was admitted with complaints of progressive worsening of vision in the left eye with diplopia, ptosis, retro-ocular pain, facial numbness and vomiting of 1 week duration. ENT opinion was sought for the same. On clinical examination the patient was conscious and oriented. He had proptosis of left eye and it was pushed outwards and downwards. There was left 3, 4 and 6 cranial nerve palsies with loss of sensation over the left forehead region (Fig. 1). There was only perception of light in the superior quadrant in the nasal line in the left eye. MRI Brain and Paranasal sinuses revealed an enhancing lesion in T2 weighted images occupying the left orbital apex with extension to the cavernous sinus (Figs. 2, 3). All the Paranasal sinuses were however found to be normal and free of disease. The size of the lesion was found to have increased when compared to the earlier

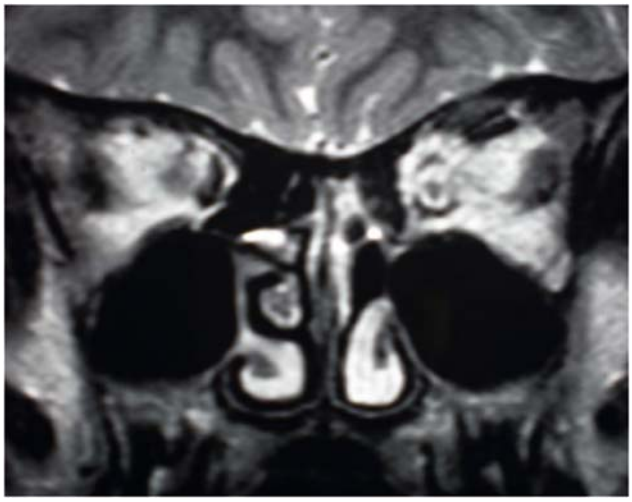

Fig. 3 Preoperative coronal CT scans showing fungal lesion in the orbital apex

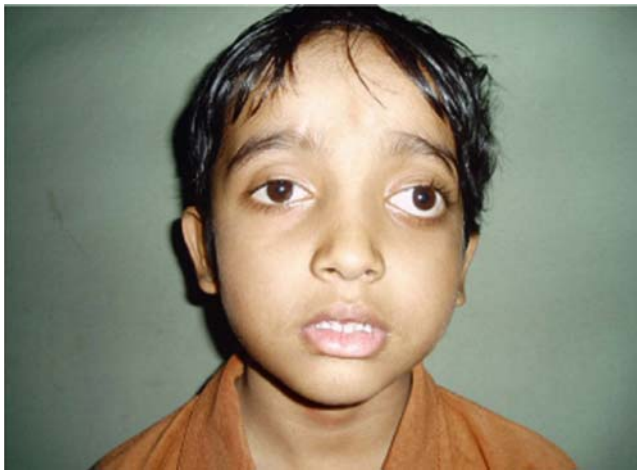

Fig. 1 Preoperative photograph showing left proptosis

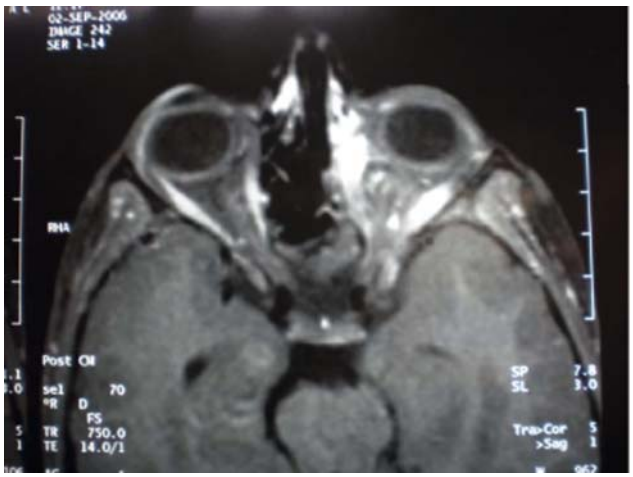

Fig. 2 Preoperative axial CT scan showing fungal lesion in the orbital apex

scan done in April, 2005. A Doppler Scan of the left eye revealed superior ophthalmic vein thrombosis. A diagnostic nasal endoscopy could not be done due to the severe pain. All the haematological investigations were within normal limits except for the erythrocyte sedimentation rate which was found to be elevated. Endoscopic Biopsy with Orbital Decompression was done under general anaesthesia. Anterior to posterior approach was used. Uncinectomy with infundibulotomy and middle meatal antrostomy done on the left side. The anterior and posterior ethmoid air cells were excised. Sphenoidotomy was done. A bulge was visualized in the posterior aspect of the lamina papyracea at level of posterior ethmoids. The lamina was opened all the way from the region posterior to the maxillary ostium to the orbital apex. Bulging medial rectus muscle was identified and soft friable tissue was visualized lateral to the medial rectus extending posteriorly. This tissue was removed and sent for histopathological and fungal studies. A complete medial and inferior orbital decompression was done.

The nasal pack was removed on the first post operative day. The proptosis had subsided with partial recovery of the eye movements. The pain had completely subsided. However there was no improvement in vision. The child was on antibiotic coverage in the post operative period. Microbiological studies grew *Aspergillus flavus* on Sabouraud's dextrose agar and Lactophenol cotton blue mount revealed septate hyphae with spores and vesicles. The patient was started on oral voriconazole (150 mg BD) for a period of three months. The patient was reviewed at the end of three months. He had full range of eye movements without any ptosis or proptosis (Fig. 4). Visual Acuity had improved from perception of light to finger counting at a distance of two feet. Nasal endoscopy showed well healed mucosa with no evidence of recurrence. A repeat MRI

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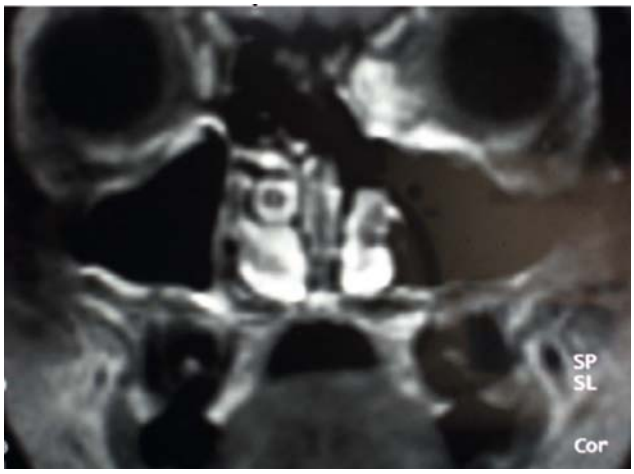

Fig. 4 Post operative coronal CT scan showing regression of the lesion in the orbital apex

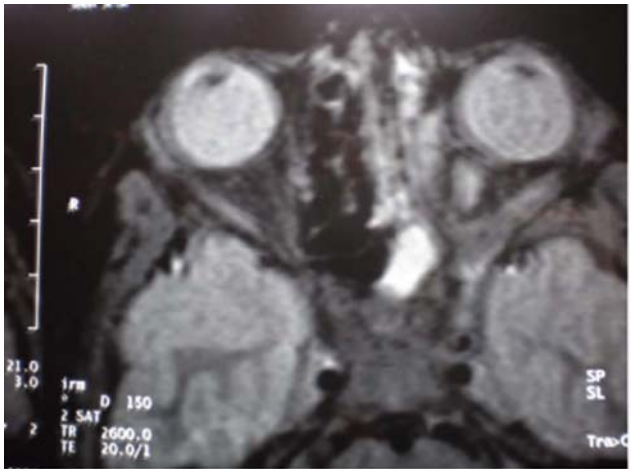

Fig. 5 Post operative axial CT scan showing regression of the lesion in the orbital apex

Brain and Paranasal sinuses showed no evidence of lesion in the region of the left orbital apex. However there was some evidence of disease persisting in the region of the cavernous sinus (Fig. 5). He had been advised to continue voriconazole for a further period of 12 months and he was followed up after one year with no evidence of recurrence.

Discussion

The orbital apex is defined as the region between the posterior ethmoidal foramen and openings of the optic canal and the superior orbital fissure [2]. The apex of the orbit is the entry portal for all the nerves and vessels of the eye and the site of origin of all extraocular muscles except the inferior oblique [3]. Orbital Apex syndrome can be due to a variety of causes like optic nerve glioma, Infraclinoid aneurysm of the internal carotid artery, trauma, orbital tumors, Paget's disease and fungal infections. Fungal infections are the least common. Only rarely is Aspergillosis the causative pathology. In humans, almost all the invasive Aspergillus infections are caused by *Aspergillus fumigatus*. In invasive human infections *A. flavus, A. glaucus, A. niger, A. restrictus, A. terreus* and *A. versicolor* have been found as causative agents [4]. *Aspergillus fumigatus* is the most common species isolated in human infections, followed by *A. flavus* [5]. Invasive fungal infection is an opportunistic infection. Predisposing causes include alcoholism, low dose prednisolone therapy and diabetis mellitus. The route of infection is frequently by inhalation of Aspergillus spores and or airborne metabolites of aspergillus, causing at first allergic aspergillosis.

The main routes of central nervous system contamination are hematogenous dissemination from a distant primary source, mainly lung, and contiguous spread from an adjacent focus such as orbit or paranasal sinus [6]. It may manifest as single solid granuloma, multiple abscesses, necrotic lesions or meningitis. CNS aspergillosis is a dramatic disease with a mortality rate of over 90% in immunocompromised patients. Orbital aspergillosis is a rare condition that may mimic non-specific orbital inflammatory disease and hence it goes undiagnosed. Once the infection extends intracranially the morbidity is high inspite of aggressive management.

Invasive fungal infections can be detected by imaging procedures such as X-ray, CT scan, MRI and sonography, but laboratory confirmation is of great importance. Respiratory secretions, bronchoalveolar lavage fluid as well as other clinical specimens from infected sites should be examined both under the microscope and in cultures.

Systemic aspergillosis is a life threatening opportunistic infection that requires specific antimycotic therapy. Amphotericin B has been the first line drug for systemic therapy inspite of nephrotoxicity. In case of aspergillomata treatment includes aggressive surgical debridement and antifungal therapy with Amphotericin B, Itraconazole, Voriconazole [7 , 8]. Voriconazole is a second generation triazole antifungal drug which is very effective against Aspergillus species. It has been reported to be superior to Amphotericin B in treatment of Aspergillus infections of Paranasal sinuses.

Conclusion

This case report highlights the occurence of fungal infection in a immunocompetent child. The mode of infection is not clear as all the paranasal sinuses were uninvolved. Probably the fungus entered the orbit at the time of head injury or during the interventional radiological procedure (embolization with stenting).The dramatic improvement after surgical decompression of the orbit and antifungal therapy with voriconazole is noteworthy. It is also emphasized that such lesions need to be biopsied before starting steroid therapy as the fungal lesions mimic granulomatous lesions.

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