Volume-10 / Issue-I/June 2022/ISSN No.-2321-3450
DOI: httpp://doi.org/10.36611/upjohns/volume10/Issue1/9
Date received: 17.5.2022 / Date published: 30.6.2022

CASE REPORT

ORBITAL APEX SYNDROME

Authors: Eshani Dutta (1) Rajat Jain* (2) Veerendra Verma (3) Sunil Kumar (4) Abhishek Bahadur Singh (5)

Authors Affiliations: (1) Senior Resident (2) Assistant Professor (3) Professor (4) Additional Professor (5) Associate Professor, Department of Otorhinolaryngology and Head-Neck Surgery, King George Medical University, UP, India

ABSTRACT

The orbital apex syndrome is a relatively uncommon entity, characterized by retrobulbar pain, supraorbital nerve hypoesthesia, palpebral ptosis, loss of vision, ophthalmoplegia and fixed mydriasis caused by involvement of the structures within the orbital apex, namely, second (optic), third (oculomotor), fourth (trochlear), fifth (Opthalmic divison of trigeminal) and sixth (abducens) cranial nerves. Presenting a case of 62 year diabetic male with complaints of loss of vision, ptosis. Management of orbital apex syndrome mainly depends upon localization of the lesion, identification of the etiology followed by full course of antifungal therapy. Clinical diagnosis was supplemented with imaging modalities, like contrast enhanced computed tomography (CECT) of the orbits and paranasal sinuses, as well as MRI PNS with orbits, along with lab investigations. In this article, we attempt to explain in detail the symptomatology, clinical and radiological findings and management of the case.

INTRODUCTION

The superior orbital fissure is the communication between the orbit and middle cranial fossa, lying at the back of the orbit between the lesser and greater wings of sphenoid and curves downward and medially, widening at the orbital apex. It is in close proximity to the cavernous sinus, which extends posteriorly from the superior orbital fissure to the dorsum sella. The optic foramen is located medially on the lesser wing of the sphenoid bone and is narrow in caliber– measuring around 2 mm in the superior narrow aspect and about 8 mm at the basal broader aspect, being about 22 mm in length [1,2]. The contents of the superior orbital fissure include the superior and inferior ophthalmic veins, the ophthalmic division of the trigeminal nerve and its branches (lacrimal, frontal and nasociliary) along with nerves supplying the extra-ocular muscles. Orbital apex syndrome is clinically characterized by paralysis of extra-ocular muscles, sensory impairment of ophthalmic division of trigeminal nerve with extension to the optic nerve, Ophthalmoplegia, ptosis, decreased corneal sensation and occasionally visual loss caused by mechanical compression of the optic nerve. The palpebral ptosis can be explained by the involvement of the superior branch of oculomotor nerve. Mydriasis is secondary to trauma to the parasympathetic fibers that course along the oculomotor nerve. Hypoesthesia over the frontal region and superior eyelid along with retro-orbital pain is due impairment of the lacrimal and ophthalmic branches of trigeminal nerve. Ophthalmoplegia can be attributed to the involvement of oculomotor, trochlear and abducens nerve. Presence of proptosis with swelling of the eyelids and chemosis are suggestive of significant mass extension within the orbits.

CASE REPORT

A 62 year old male presented to the OPD with complaints of right sided headache and facial pain, drooping of right upper eyelid and gradual diminution of vision of right eye, progressing over a span of 20 days. The headache was insidious in onset, unilateral with localization to the right hemicranium and retro-orbital region, severe in intensity causing disturbances with sleep, lancinating type, and not associated with changes in position of the head. Drooping of right upper eyelid had progressed to complete inability to lift the eyelid over 20 days. Reduction in vision of the right eye was sudden in onset and progressed from blurring to perception of light over 20 days

Review Process: Double blind Peer Reviewed by Reviewer Board Members

and was associated with inability to move the right eye. Patient had no complaints of nasal obstruction, nasal discharge or bleeding, alterations in olfaction, swelling or redness of skin over the face, fever or any history of trauma. Patient was a known diabetic taking oral hypoglycemic agents since last 5 years, with poorly controlled sugar levels. On examination, patient did not have any signs of systemic toxicity. Examination of the right eye revealed palpebral ptosis with restriction of extra-ocular movements in all gazes(figure 1), absent perception of light, positive relative afferent papillary defect, and absent corneal reflex but normal fundoscopic findings.

Crude touch sensations were intact and equal over both sides of the face. Remainder of the cranial nerves were normal on examination. There was no para-nasal sinus tenderness or significant finding on rhinoscopy.

Figure 1:

Fig.1 – Restricted extra-ocular movements demonstrated in different gazes
A - Restricted upper eye movement
B - Restricted lower eye movements

C - Restricted medial eye movements
D - Restricted lateral eye movements

Neuroimaging done for the patient included CT of the orbits and PNS which showed soft tissue attenuation in the orbital apex (**figure 2**), while MRI orbits and PNS shows iso- to hypo intense images on T1, T2 weighted MR images and with minimal enhancement on T1 contrast enhanced image (**figure 3**).

Figure 2:

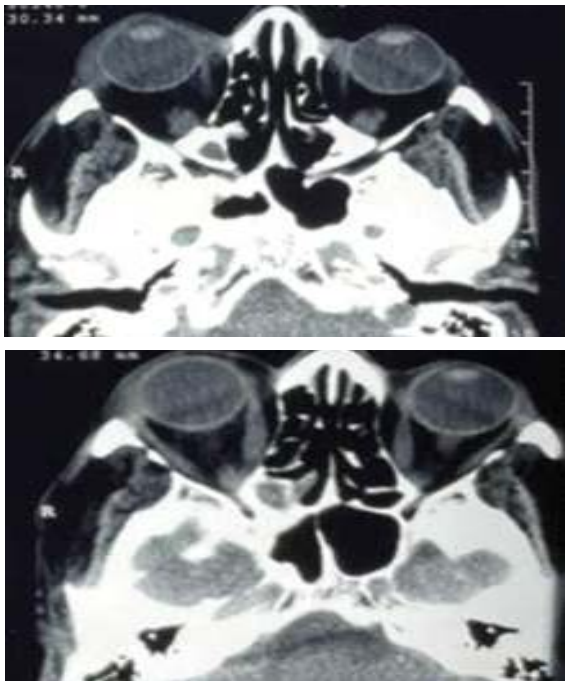
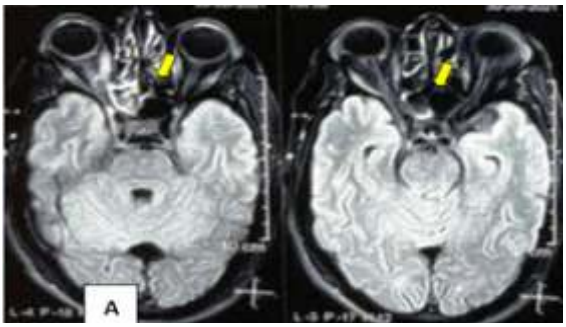


Fig.2 – Axial NCCT of the orbits and PNS showing soft tissue attenuation in the orbital apex

Figure 3:



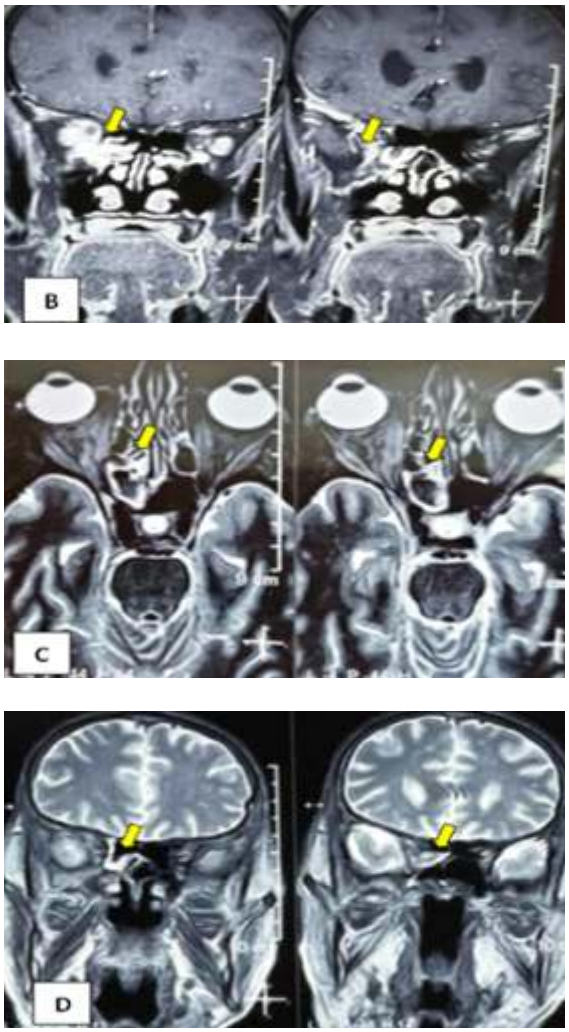


Fig.3 – MRI orbits and PNS shows iso- to hypo intense images on T1, T2 weighted MR images and with minimal enhancement on T1 contrast enhanced image

A – T1 contrast axial view
B – T1 contrast coronal view
C – T2 axial view
D – T2 coronal view

Lab investigations revealed a raised glycosylated hemoglobin level of 13.7%, with deranged sugar profile along with raised serum CRP and galactomannan levels.Patient was subjected an endoscopy-guided biopsy, the histopathological examination of which revealed septate fungal hyphae in the setting of chronic granulomatous inflammation (on the 12th day)(figure 4), suggestive of infection with Aspergillus species.

Figure 4:

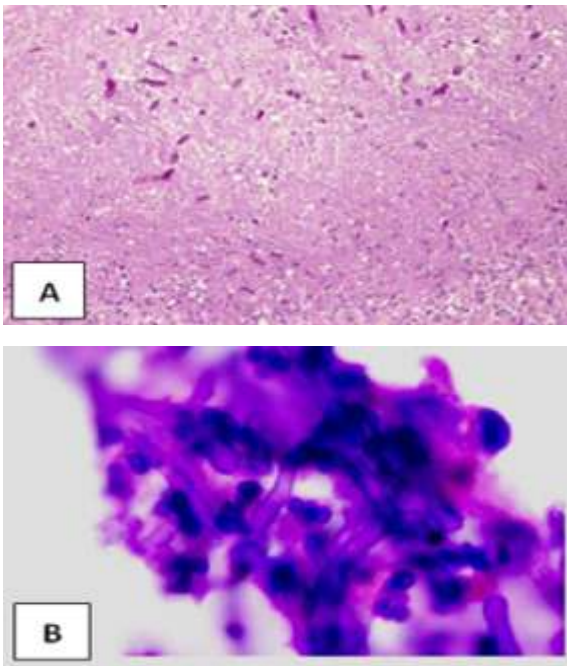


Fig. 4 – Histopathological images showing Aspergillosis : [A] Hematoxylin and eosin staining showing tissue-invasive Aspergilluswithout granuloma formation. [B] PAS staining showing branching of fungal hyphae at 45 degree angle.

Patient was started on oral voriconazole and broad spectrum antibiotics and advised strict glycemic control along with regular transnasal irrigation with saline. Patient showed significant improvement in the palpebral ptosis and ophthalmoplegia after 1month of treatment **(figure 5)**.

Figure: 5

Fig.5 – Improvement in ophthalmoplegia demonstrated after 1 month of antifungal treatment

A - Upper eye movement
B - Lateral eye movements
C- Medial eye movements
D - Lower eye movements

DISCUSSION

Orbit is an anatomically complex region due to the intricate interactions between various bony, vascular and neural structures. Orbital Apex Syndrome is a syndrome characterized by the involvement of optic, oculomotor, trochlear, first division of trigeminal and abducens nerves.

A similar presentation is seen in two other syndromes, namely cavernous sinus syndrome and superior orbital fissure syndrome. Cavernous sinus syndrome results from the combination of orbital apex syndrome and affection of the sympathetic fibers and the maxillary division of the trigeminal nerve. The superior orbital fissure syndrome, or Rochon -Duvigneaud syndrome, occurs secondary to lesions anterior to the orbital apex, close to the annulus of Zinn. It characteristically spares the optic nerve.

Orbital apex syndrome is characterized by vision loss and painful ophthalmoplegia, as well as localizing symptoms like pain around the orbit or the skin around the orbit (involvement of the ophthalmic division of trigeminal nerve) and facial pain (involvement of maxillary division of the trigeminal nerve). Ophthalmoplegia results from involvement of the 3rd, 4th and 6th cranial nerves and can present as vertical, horizontal or torsional diplopia with or without compensatory abnormal head postures. Other signs include abnormal or absent pupillary reflex, absent corneal reflex and proptosis.

The pathology behind orbital apex syndrome can be broadly classified as inflammatory, infectious, or neoplastic in origin. Inflammatory lesions show rapid onset, progression over days,painful ophthalmoplegia and signs of optic neuropathy. Causes include Wegener's Granulomatosis, Churg- Strauss syndrome and Tolosa Hunt syndrome. Other causes include neoplasms (head and neck tumors,hematologic cancers and metastatic lesions), accidental and surgical trauma (cranio-maxillo-facial trauma and surgeries of the orbit and sinuses causing direct compression by displaced bony fragments, foreign bodies or hematomas or inflammatory edema), vascular causes, namely carotid cavernous fistulas, carotid cavernous aneurysms and carotid sinus thrombosis.

Infectious causes can be attributed to viral, bacterial, fungal and parasitic infections which result from contiguous infections of the paranasal sinuses or other surrounding structures. Fungal orbital apex syndrome is a rare entity. Fungal infections are most commonly due to Aspergillus and Mucor. The source of infection is usually the paranasal sinuses and vertebral aspergillosis infections spreading to the orbitalapex via the Batson's plexus have also been reported.[3] Voriconazole is a second generation triazole antifungal, which is reported to be more effective than Amphotericin B in the treatment of Aspergillus infection of the paranasal sinuses.

The primary focus for management should be the localization of the underlying lesion, identification of etiology and early mangement. Neuro-imaging is mandatory in patients with orbital apex syndrome, with CEMRI of the orbit and PNS being the imaging modality of choice. Orbital apex syndrome secondary to acute invasive sinusitis caused by Aspergillus spp shows soft-tissue thickening in the posterior ethmoid air cells, destroying the medial wall of the orbit and extension into the retro-orbital soft tissues on CT **(figure 2)**. In cases of OAS secondary to acute

invasive fungal sinusitis caused by Zygomycosis, non-contrast CT shows hypoattenuating mucosal thickening orsoft-tissue attenuation within the nasal cavity, affected paranasal sinus and orbital apex with predilectionfor unilateral ethmoid andsphenoid sinus involvement and aggressive destruction of bony confines of the sinus **(figure 6).**

Figure 6:

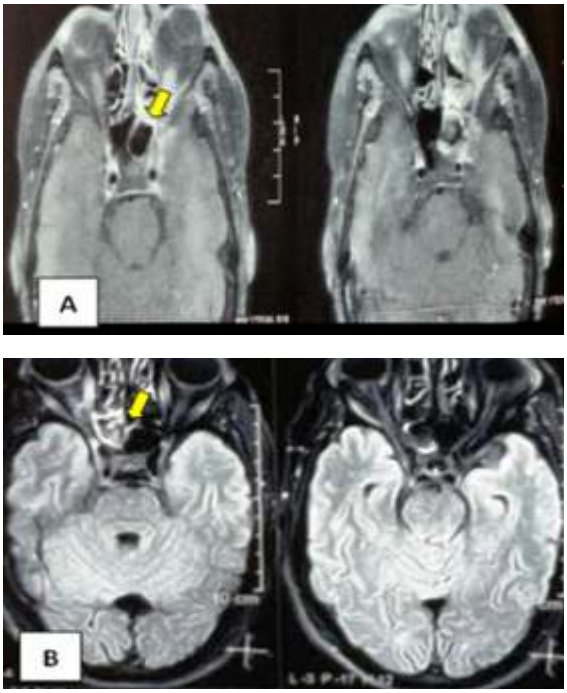


Fig.6 - MRI orbits and PNS – T1 contrast axial images showing a case of [A] Rhino-orbital mucormycosis with no contrast uptake; [B] Invasive aspergillosis with minimal contrast enhancement in orbital apex region.

MRI shows inflammatory changes in the orbital fat and extraocular muscles **(figure 3)**. Hematological work up should include a complete blood count and investigations directed against a particular infectious/fungal agent if suspected. The key to dealing with orbital apex syndrome is that treatment is directed towards the underlying etiology. The most crucial distinction is between an inflammatory cause and an infection, especially fungal infections, since they have a tendency to worsen rapidly and be fatal if steroid treatment is initiated for the latter. The initial step in management in any case of invasive fungal rhinosinusitis (acute or chronic) is the reversal of the immuno compromised state of the individual. In our case, the initiation of empirical antifungal therapy with voriconazole (400mg twice as loading dose on day 1 followed by 200mg twice daily as maintenance dose for 6 months)[4]with strict glycemic control brought about a drastic improvement in the patient's symptoms, along with gradual resolution of the pupillary defect and ophthalmoplegia, thereby negating the need for surgical exploration. Patient was monitored using liver function tests and serial CECT PNS during the course of treatment.

CONCLUSION

Orbital apex syndrome secondary to invasive fungal lesions is a rare entity and may be associated with poor prognosis. Long term voriconazole is found effective in the management of invasive aspergillosis causing orbital apex syndrome.

REFERENCES

1. Yeh S, Foroozan R. Orbital apex syndrome. Curr Opin Ophthalmol.2004;15(6):490–498. doi:10.1097/01.icu.0000144387.12739.9c
2. Jiang N, Zhao G, Yang S, et al. A retrospective analysis of eleven cases of invasive rhino-orbito-cerebral mucormycosis presented with orbital apex syndrome initially. BMC Ophthalmol. 2016;16:10. doi:10.1186/s12886-016-0189-1
3. Camargo JF, Seriburi V, Tenner M, El Khoury MY. Aspergillus osteomyelitis of the lumbar spine complicated with orbital apex syndrome: a potential role of the Batson's plexus in disease propagation. Med Mycol Case Rep. 2012;1(1):9–12. doi:10.1016/j.mmcr.2012.02.004
4. Beatriz Peral-Cagigal , Luis-Miguel Redondo-González , Alberto Verrier-Hernández. Invasive maxillary sinus aspergillosis: A case report successfully treated withvoriconazole

and surgical debridement
5. Mormont E, Laloux P, Vauthier J, Ossemann M. Radiotherapy in a case of Tolosa-Hunt syndrome. Cephalalgia. 2000;20(10):931–933. doi:10.1046/j.1468-2982.2000.00147.J Clin Exp Dent. 2014;6(4):e448-51.
6. Smith JR, Rosenbaum JT. A role for methotrexate in the management of non-infectious orbital inflammatory disease. Br J Ophthalmol. 2001;85(10):1220–1224. doi:10.1136/bjo.85.10.1220
7. Badakere and Patil-Chhablani. Orbital Apex Syndrome: A Review. Dove Press journal:Eye and Brain 2019:11
8. Dong Hyun Kim, MD, Jin Uk Jeong, MD, Seul Kim, MD, Seon Tae Kim, MD , and Gyu Cheol Han, MD. Bilateral Orbital Apex Syndrome Related to Sphenoid Fungal Sinusitis. Ear, Nose & Throat Journal 1–3

***Corresponding Author:**
Dr Rajat Jain
Assistant Professor
ENT department
KGMU, Chowk , Lucknow, UP-226003
Mob.: 7754915828

How to Cite the article
Eshani Dutta etal etal, Orbital Apex Syndrome, UPJOHNS June 2022; 10(1)- 50-55 DOI: httpp://doi.org/10.36611/upjohns/volume10/Issue 1/9