

Orbital Apex Syndrome: An Uncommon Complication caused by a Common Nasal Commensal

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ABSTRACT

Introduction

Orbital apex syndrome, an uncommon condition seen in ENT practice, is characterised by multiple cranial neuropathies, presenting as visual loss, ophthalmoplegia, ptosis and hypoesthesia of forehead due to involvement of Cranial nerves III, IV, VI and VI.

Case Report

This is a case of an 80-year-old female patient, with poorly controlled Type 2 Diabetes mellitus and Hypertension, who presented with right sided headache of 10 days duration, drooping of right eyelid and decreased vision in right eye for 3 days. Clinically, there were features suggestive of right orbital apex syndrome. MRI Brain showed abnormal enhancement in right orbital apex with subtle enhancement of optic nerve in optic canal and intense enhancement of mucosal thickening in sphenoid sinus. Aerobic culture report of the purulent nasal discharge revealed *Staphylococcus epidermidis*.

Discussion

Aetiology of this condition is varied. Rarely known to occur following bacterial sinusitis, it is most commonly seen secondary to fungal sinusitis or orbital cellulitis involving orbital apex. Here we report an unusual case occurring secondary to *Staphylococcus epidermidis* sinusitis. In conclusion, although acute orbital apex syndrome is usually associated with invasive fungal sinusitis, clinicians must be aware that bacterial sinusitis may also present with a similar aggressive pattern.

Keywords

Staphylococcus epidermidis; Sphenoid sinusitis; Bacterial infections; Sinusitis; Complications

Orbital apex syndrome, a condition characterised by multiple cranial neuropathies, is rarely known to occur following bacterial sinusitis without orbital cellulitis, and is most commonly seen secondary to fungal sinusitis or orbital cellulitis involving the orbital apex¹ *Staphylococcus epidermidis*, previously regarded only as a commensal microorganism, is now also seen as an important opportunistic pathogen.²

Case Report

Here we report a case of an 80-year-old female patient, with poorly controlled Type 2 Diabetes mellitus (HbA1c of 13%) and Hypertension, who presented with history of right sided headache of 10 days duration, drooping of right eyelid and decreased vision in right eye for 3

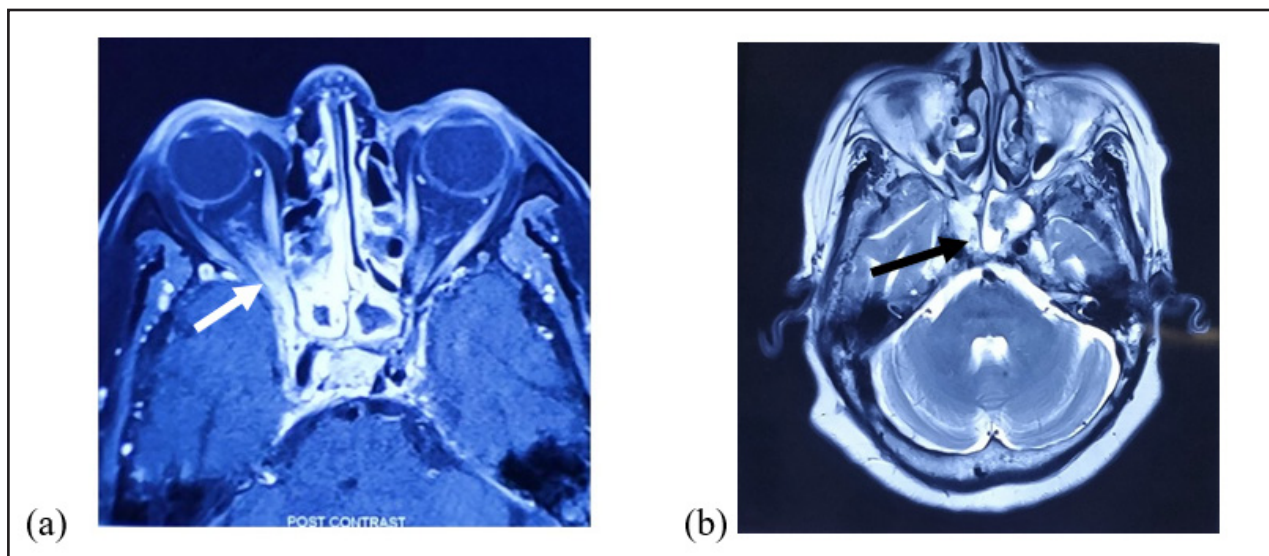
days, which was gradually progressive. On examination of the Right eye: there was complete ptosis, vision was limited to perception of light, there was relative afferent pupillary defect (RAPD), all extraocular movements were restricted and corneal sensation was absent on lateral half. All these findings were suggestive of Right Orbital apex syndrome.

Diagnostic nasal endoscopy revealed deviated nasal septum to left and purulent discharge was noted in bilateral sphenothmoidal recess and in the

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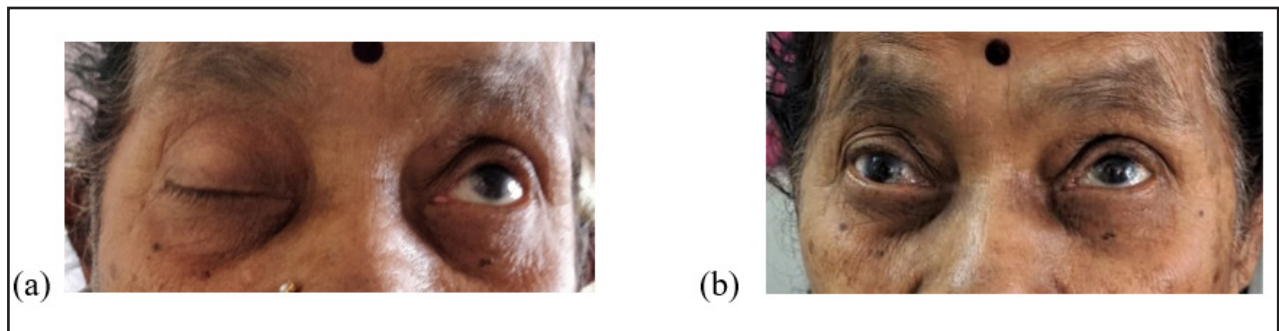
**Fig.1. MRI brain showing (a) abnormal enhancement (white arrow) in right orbital apex with the subtle enhancement of optic nerve in the optic canal
(b) intense enhancement (black arrow) of the mucosal thickening in bilateral sphenoid sinus**

nasopharynx. Swab of the purulent discharge was taken for aerobic culture sensitivity, which was later reported as *Staphylococcus epidermidis*, sensitive to gentamycin, linezolid, daptomycin, Trimethoprim/Sulfamethoxazole, Tigecycline, Teicoplanin, Vancomycin and resistant to Penicillin, Clindamycin and Erythromycin. On investigating further, MRI Brain showed abnormal enhancement in right orbital apex with the subtle enhancement of optic nerve in the optic canal (Fig.1a) and intense enhancement of the mucosal thickening in sphenoid sinus (Right more than left) (Fig.1b). As advised by the Neurologist, further investigations were done to rule out other causes for orbital apex syndrome, such as TB-PCR (Tuberculosis-Polymerase Chain Reaction) which was negative, ANA (Antinuclear antibody) profile which was negative and Serum ACE (Angiotensin Converting Enzyme) level which was normal.

In view of features suggestive of sphenoid sinusitis, the patient underwent Functional endoscopic sinus surgery under General anaesthesia. Intraoperatively, under endoscopic guidance, the polypoidal tissue noted obstructing the right sphenoid ostium was cleared and both ostia were widened. Thickened oedematous mucosa was seen extending to opticocarotid recess and optic

nerve prominence, with thinning of bone near the optic nerve prominence, along with purulent discharge in the right sphenoid sinus, which was cleared completely. Oedematous mucosa noted in left sphenoid sinus was removed. The polypoidal tissue and oedematous mucosa which were removed were sent for KOH (potassium hydroxide) mount and histopathological examination. Simultaneously, the patient was started on Intravenous antibiotics namely Piperacillin-tazobactam 4.5 grams thrice daily for 10 days and injection Vancomycin 500 milligrams twice daily for one week, based on culture sensitivity report, and appropriate anti-diabetic and anti-hypertensive medications and other supportive treatment were given. Postoperatively, on POD (post-operative day) 1, nasal packs were removed. On POD- 4, right eye ptosis improved (Fig.2b). On POD- 6, minimal superior and inferior extraocular movements were observed and on POD-7, patient appreciated perception of light in the right eye. The KOH mount was reported negative for fungal elements. And histopathology revealed focal collection of macrophages, epitheloid cells and multinucleated giant cells, suggestive of granulomatous inflammation, with no caseation, no fungal elements and no atypia.

On 1month follow-up, patient was symptomatically



**Fig.2. (a) Preoperative picture showing right eye ptosis
(b) Picture on Postoperative day 4 showing resolved ptosis of right eye**

better, there was mild restriction of right lateral extraocular movement, perception of light was present and there was no ptosis.

Discussion

Orbital apex syndrome is an uncommon disorder characterized by visual loss, ophthalmoplegia, ptosis and hypoesthesia of forehead. It may occur secondary to a variety of inflammatory, infectious, neoplastic and vascular conditions that cause damage to the superior orbital fissure (causing Cranial Nerves III, IV, VI, VI palsies) and optic canal (causing Cranial Nerve II dysfunction).³

This condition can be life threatening if there is invasion through ophthalmic vessels or bone fissures, leading to intra-cranial involvement. Hence timely clinical suspicion and recognition of the condition is crucial for better outcome.

As our patient had uncontrolled diabetes and rapid progression of symptoms, with cranial nerves II to VI involvement, and an MRI suggestive of right orbital apex involvement, subtle enhancement of right optic nerve and mucosal thickening in sphenoid sinus, our initial suspicion was invasive fungal sinusitis as a possible cause. But KOH was negative for fungal elements. Aerobic culture sensitivity of the purulent discharge showed staphylococcus epidermidis.

Very few cases have been reported in literature about bacterial sinusitis resulting in orbital apex syndrome without orbital cellulitis. And even in those observed cases, Staphylococcus aureus and Pseudomonas

aeruginosa were the common bacteria which were isolated.

Staphylococcus epidermidis, a gram- positive bacterium, often regarded as a culture contaminant, has in recent years been recognised as a pathogen, especially in immunocompromised patients.⁴ In a study by Janusz et al, Staphylococcus epidermidis was the most commonly isolated bacteria, especially in ethmoid and sphenoid sinuses. It was also found to be present in all biofilm- positive mucosal specimens, indicating its pivotal role in pathogenesis of severe chronic infections.⁵ Hence, in this case, we attributed the patient's orbital apex syndrome and optic neuropathy to sphenoid bacterial sinusitis without orbital cellulitis. The patient's immunocompromised state, would have probably contributed to rapid progression of the infection from sinus to orbital apex.

Early endoscopic sinus surgery and appropriate culture sensitive antibiotics, seemed to have helped in reversing some of the cranial nerve damage and the patient's extraocular movements and ptosis improved. The vision was restricted to perception of light, and though there was no improvement noted in vision, it prevented the further deterioration of vision and also the extension of infection intracranially.

In conclusion, although we usually associate invasive fungal sinusitis with acute orbital apex syndrome, clinicians must be aware that bacterial sinusitis may also present with a similar aggressive pattern. Timely diagnosis and early surgical intervention along with appropriate antibiotic coverage has shown to prevent further spread of the disease.

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Conflicts of interest: None

Compliant with ethical standards.

Written informed consent was obtained for publication of data and photographs.

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