

## Invasive Fungal Sinusitis and Optic Neuropathy

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### Abstract

Early determination of a definitive underlying aetiology in a young immunocompetent individual presenting with sudden onset visual loss mimicking optic neuritis presents as a challenge to the treating ophthalmologist. Presence of a relative afferent pupillary defect usually triggers initiation of high doses of systemic steroid therapy which may have unpredictable and surprising neurophthalmic sequelae. This case report elucidates an atypical presentation of optic neuropathy with its tumultuous course in a young male who presented with severe unilateral visual loss and was eventually diagnosed with invasive fungal sinusitis accounting for atypical manifestations.

**Keywords:** Optic Neuropathy; Invasive Aspergillosis; Ethmoid Sinus and Orbital Cellulitis; Optic Neuritis with Fungal Sinusitis

### Abbreviations

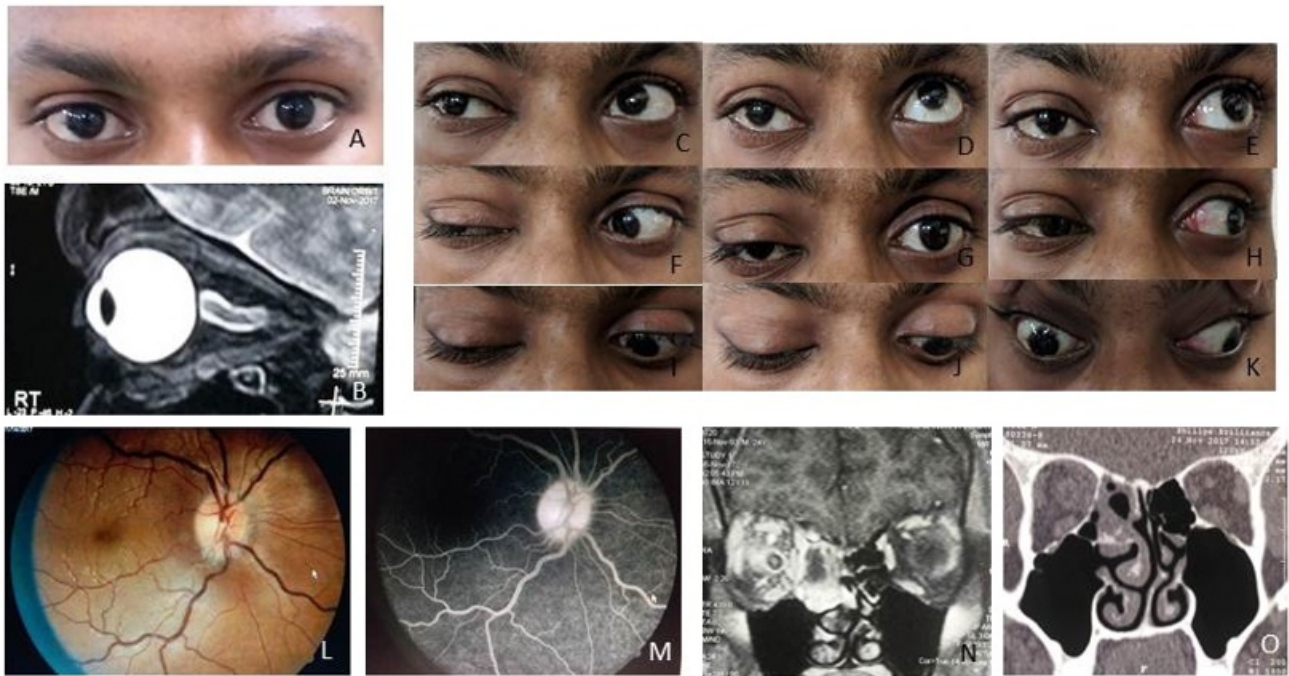
RE: Right Eye; LE: Left Eye; VA: Visual Acuity; RAPD: Relative Afferent Pupillary Defect; ONTT: Optic Neuritis Treatment Trial; I.V.M.P: Intravenous methylprednisolone; OD: Once a Day; VDRL: Venereal Disease Research Laboratory; BCVA: Best Corrected Visual Acuity; CT: Computed Tomography; PNS: Paranasal Sinus

### Introduction

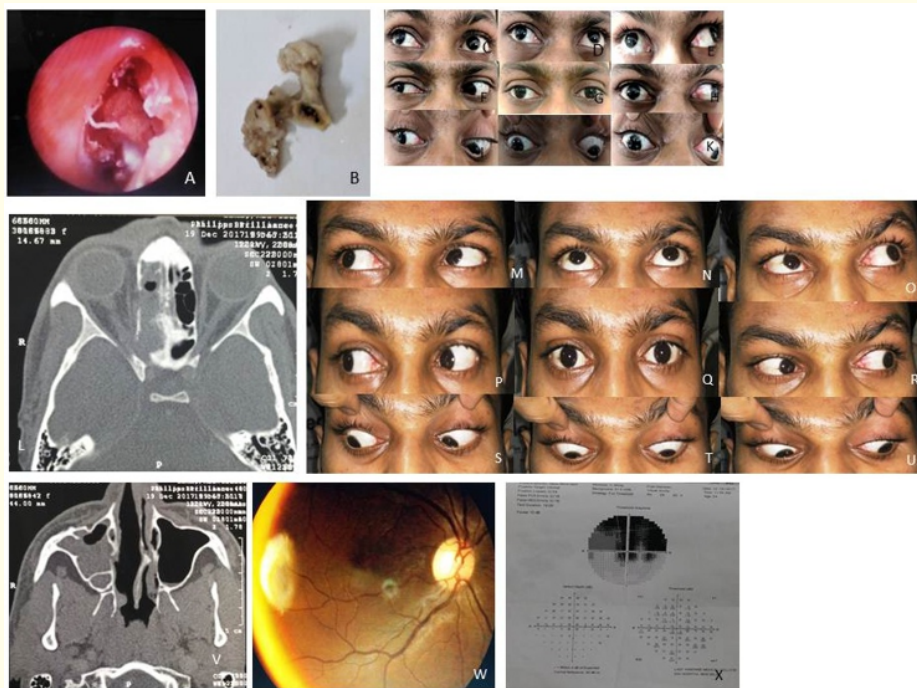
Loss of vision associated with the disease of the orbit or sinuses is either due to trauma, a neoplasm or due to an underlying infectious etiology. The structural closeness of the orbit, paranasal sinuses and intracranial cavity results in transfer of the pathological process from one structure to the other. *Aspergillus* is usually a saprophytic fungus which becomes a pathogenic fungus in the paranasal sinuses due to anaerobic conditions. Unilateral painless visual loss as a result of sino-orbital aspergilloma has been reported in two isolated case reports in patients with steroid responsive optic neuropathy and non-specific optic neuritis [1,2]. Orbital aspergillosis in immunocompetent patients can have variable presentation and can mimic malignancy, optic neuritis, orbital apex syndrome, and typical bacterial cellulitis or orbital abscess [3]. Increase in the usage of steroids, new antibiotics and immunosuppressants has led to an increase in the incidence of aspergillosis of the paranasal sinus. The contiguous spread of the fungal infection can take place through the thin bones of the orbital wall or as a result of the inflammation of the veins communicating between the sinus and the orbit.

## Case History

A 24 year male presented with sudden painless loss of vision in his right eye (RE) since ten days. Left eye (LE) was normal at presentation and throughout the course of disease (Figure 1A). He was a chronic tobacco chewer and smoker for last four years. Vitals, general physical and systemic examination including the central nervous system was normal except for the second cranial nerve examination. The visual acuity (VA) on presentation was finger counting close to face and 6/6 in the RE and LE respectively with accurate projection of rays and insignificant refractive error in both eyes (BE). Near vision was less than N36 in RE with a relative afferent pupillary defect (RAPD) and the patient could not grossly identify red and green colours. The orbit, eyeball, eyelids, anterior segment, intraocular pressure and fundus examination were normal. A working clinical diagnosis of retrobulbar neuritis/optic neuritis was presumed. Magnetic resonance imaging (MRI) of the brain and orbits revealed subtle swelling and oedema of right optic nerve (Figure 1B) in its intra-orbital and intracanalicular segments. No parenchymal lesion in the brain was seen. The patient was admitted and as per the recommendations of the optic neuritis treatment trial (ONTT) [4], was started on a high dose (one gram per day) of intravenous methylprednisolone (I.V.MP) for three days and discharged on 60 mg of oral prednisolone (as per body weight) once a day (OD) with a VA of 6/6 - RE and a normal colour vision. Three days later he presented with a drop in VA to 6/36 which improved to 6/12 with pin hole, reappearance of RAPD, reduction in contrast sensitivity [0.9% (13.1%)], defective colour vision but with a normal appearing optic disc. He was restarted on I.V. MP for three days and discharged after five days on 60 mg OD of oral steroids with a best corrected VA (BCVA) of 6/12. Visual fields in the LE till this stage was normal. Surprisingly, he presented the next day with sudden onset moderate intensity pain, swelling and diminution of vision in RE to an extent that he had doubtful perception of light accompanied with periorbital edema, proptosis of 2 mm, partial ophthalmoplegia (Figure 1C-1K) and blurring of the inferonasal right optic disc margins (Figure 1L). Investigations revealed haemoglobin of 12.3 gm%, a total leucocyte count of 19800/mm<sup>3</sup> with normal liver and kidney function, platelet count, glycosylated haemoglobin (4.8%), fasting blood glucose and serum angiotensin converting enzyme levels. There was no growth on blood and urine culture. Antinuclear antibody, C reactive protein, cytoplasmic and perinuclear anti-neutrophilic cytoplasmic antibody, human immunodeficiency virus, hepatitis B surface antigen, hepatitis C and venereal disease research laboratory (VDRL) were non-reactive. Urgent computed tomography (CT) was done to rule out cavernous sinus thrombosis and demonstrated ethmoidal sinusitis. Injection imipenem was started after a neurology consultation. Fundus fluorescein angiography was suggestive of optic neuritis (Figure 1M). Meanwhile otorhinolaryngological opinion revealed no obvious abnormality on endoscopic examination. MRI brain, orbit and whole spine screening was done and demonstrated right sided optic nerve inflammation, intraconal fat stranding, extra-ocular muscle (superior oblique, medial and inferior rectus) inflammation and thickening, bulky inflamed lacrimal gland pointing to right sided orbital cellulitis (Figure 1N). In view of non-recovery of visual functions, he was started on I.V MP for 5 days after 5 days of I.V antibiotics but the patient lost all vision in his RE on the sixth day and could not perceive light. Non-contrast CT paranasal sinus (PNS) done subsequently confirmed right ethmoid and sphenoid sinusitis with optic nerve thickening and inflammatory changes in retro-orbital fat (Figure 1O). He underwent functional endoscopic sinus surgery and yellowish debris was visualized in ethmoidal sinus area however other sinuses were clear (Figure 2A). The mass (Figure 2B) was sent for histopathology and *Aspergillus flavus* was obtained on culture. He was started on injection Amphotericin B and oral steroids were continued under antifungal cover, both for 44 days. After 23 days of IV antifungals (Figure 2C-2K), BCVA improved to 6/12 in RE. Contrast enhanced CT Brain and PNS revealed erosion involving right lamina papyracea (Figure 2L), anterior cranial fossa and right lateral wall of sphenoid sinus (Figure 2V) adjacent to optic canal leading to extension of soft tissue into right orbit abutting the optic nerve suggestive of a suspicious fungal etiology. He was maintained on oral voriconazole for three months and had complete amelioration of ophthalmoplegia (Figure 2M-2U), BCVA stabilised to 6/12 (RE). Fundus examination revealed optic disc pallor (Figure 2W) and visual field charting demonstrated altitudinal type of field defect (Figure 2X). However colour vision and contrast sensitivity remained defective.



**Figure 1A-10:** Sequential patient photographs during the course of the disease. First presentation (1A). MRI suggested optic nerve edema (1B). Third presentation with ophthalmoplegia (1C-1K). Fundus and FFA revealed optic neuritis (1L, 1M). MRI (1N) and CT scan (1O) confirmed orbital cellulitis and sinusitis.



**Figure 2A-2X:** Sequential patient photographs during the course of the disease with recovery. Intraoperative yellowish debris in ethmoid sinus (2A) was excised (2B). Ophthalmoplegia (2C-2K) persisted. CECT revealed erosion of right lamina papyracea (2L) and lateral sphenoid sinus wall (2V). Three months later (2M-2U), optic disc pallor (2W) and altitudinal field defect persisted (2X).

### Discussion

Disturbance in the visual functions represent one of the commonest symptom as a presenting feature of invasive fungal sinusitis with a possible origin in the sphenoid sinus. Optic nerve involvement in these cases is responsible for this deterioration in visual functions. Although ethmoid sinuses are primarily responsible for the transfer of infection and subsequent manifestation as orbital cellulitis in the paediatric age group however their involvement must also be considered in adults presenting with orbital cellulitis. Altitudinal field defect have been sparsely reported in literature occurring as a result of invasive fungal sinusitis. A study conducted by A Das., *et al.* has reported *A. flavus* to be the predominant aetiological agent in the causation of chronic invasive granulomatous fungal rhinosinusitis [5,6]. Most cases of invasive fungal sinusitis also have sterile cultures as the background fibrosis and paucity of fungal elements that are present withing the giant cells preclude contact of the fungus with the culture medium. Neuroimaging is indispensable in situations where clinical examination does not provide definitive evidence of the underlying etiology. Accumulation of calcium in the fungal colony along-with bony erosion results in easy localisation of the invasive mycotic destruction of the surrounding structures on a CT scan. Invasive *Aspergillus* infection is a consequence of the spread of the fungus to the wall of the sinuses and the surrounding tissues. The etiopathogenesis of this saprophytic fungus culminating into an invasive infection is unknown. However it is commonly seen in immunosuppressed individuals. Surgical debridement and systemic antifungals can achieve successful outcome in sino-orbital aspergillosis [1]. Neuroimaging along-with visual field examination helps to rule out an impending or an early ischaemic/compressive optic neuropathy or orbital apex syndrome [7].

### Conclusion

Aspergillosis may mimic as optic neuritis and has been documented to involve orbital and sino-orbital structures. Fungal infections must be ruled out before initiation of steroid therapy and neuroimaging maybe helpful in these cases. A multidisciplinary intervention may avert potentially blinding sequelae.

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### Conflicting Interest

None.

### Informed Consent

Informed consent was taken from the patient for inclusion of clinical photographs in the case report.

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