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CASE REPORT

A RARE PRESENTATION OF GRANULOMATOUS FUNGAL SINUSITIS WITH OPTIC NERVE INVOLVEMENT- A CASE REPORT

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ABSTRACT

A 30 year old male presented with a history of gradual onset diminution of vision on right eye along with a painless swelling over right forehead with right nasal obstruction and discharge for 4 months. Contrast CT of paranasal sinuses was done and endoscopic sinus surgery was performed. Post operative biopsy showed granulomatous inflammation with hyaline septate hyphae suggestive of Aspergillus. After 1gm of Amphotericin B, patient was discharged on oral voriconazole of which he wasn't compliant. He presented after 3 months with total loss of vision on right eye and temporal field loss of vision on left eye. MRI brain was done which showed nodular thickening of right optic nerve upto optic chiasma. Revision surgery was performed and patient received oral anti fungal for 6 months post operatively and did not show any evidence of recurrence or disease progression.

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INTRODUCTION

Fungal rhinosinusitis is a common condition known widely by its varied presentation. Though there are a wide range of fungal organisms affecting humans, Aspergillus and Mucor species cause significant morbidity. The pathogenesis of fungal infection depends on the immune status of the individual. In immunocompetent individuals, hypersensitivity reactions such as type 1 in case of Allergic fungal sinusitis and type 4 in case of granulomatous fungal sinusitis occur. In immunocompromised persons, pathology involves direct fungal invasion of tissues and blood vessels. We present a case of chronic granulomatous fungal sinusitis in a young male involving optic nerve with rare MRI finding and with clinically unique visual field loss. We also enumerate themorbidity caused by this silent condition with the need for prolonged antifungal therapy to prevent disease progression and recurrence.

CASE

A 30 year old male presented with a history of swelling over right side forehead with headache and right facial pain for 4 month associated with gradual diminution of vision in right eye for 4 months and purulent nasal discharge with nasal obstruction on right side for 3 months. There was no history of nasal bleed, anosmia, fever, seizures, altered sensorium or no history allergic symptoms or comorbidities like diabetes mellitus, hypertension or asthma. On examination, vitals were stable, there was purulent discharge and brownish crusts filling right nasal cavity and visual acuity was 3/60 on right and 6/6 on left eye with no restriction of extra ocular movements or proptosis. A 2 *2 cm hard non tender swelling was noted over right forehead just above medial end of the eye brow with smooth surface and overlying skin was normal. On oral cavity examination, there was no loss of sensation over hard palate, loosening of teeth or ulcer noted. Except optic nerve involvement, all other cranial nerves examination was normal. Diagnostic nasal endoscopy showed thick purulent discharge from the middle meatus, no polyp or mass was noted. Contrast enhanced computed tomography showed heterogeneously enhancing soft tissue density with areas of hyper attenuation filling right maxillary sinus, anterior and posterior ethmoids, frontal and sphenoid sinus with erosion of lamina papyracea, lateral wall of sphenoid sinus and anterior table of frontal sinus. Crusts on KOH mount showed hyaline septate hyphae suggestive of aspergillus species. Patient underwent endoscopic sinus surgery -middle meatal antrostomy, anterior and posterior ethmoidectomy followed by sphenoidotomy and frontal sinusotomy was done and all the necrotic material was debrided.Postoperative biopsy showed granulomatous inflammation with morphology suggestive of Aspergillus. Patient was started on Intravenous Amphotericin 1mg/kg day with strict monitoring of electrolytes, he received 1gm in total during 1 month of hospital stay and discharged on oral Voriconazole 200mg BD for 6months with regular follow up nasal endoscopy. Our patient defaulted follow up and

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voriconazole intake after the first month. Six months later he presented with total loss of vision on Right eye and right sided headache and loss of temporal field of vision on the left side. On examination he had no light perception on right eye and temporal hemianopia on left side. CT showed heterogenous soft tissue density in frontal, sphenoid and ethmoids with extensive bone erosion with no extension into orbit. MRI brain showed enhancing nodular thickening along the right optic nerve from the orbital apex and extending to the optic chiasma and proximal part of optic tract and involving left optic nerve. There was no dural breach or intracranial extension. Poor prognosis regarding vision in right eye was explained to the patient. He underwent revision sinus surgery and DRAF 3. Tissue bits sent for histopathology turned out to be granulomatous fungal infection with morphology suggestive of Mucor and Aspergillus species. Patient received intravenous Liposomal Amphotericin B of 2 gm and discharged with oral voriconazole 200mg BD for 6 months. Ophthalmologic evaluation and diagnostic nasal endoscopy was done at 3 months and at the end of 6 months showing no worsening of vision in left eye and healthy nasal mucosa with no evidence of recurrence on nasendoscopy.

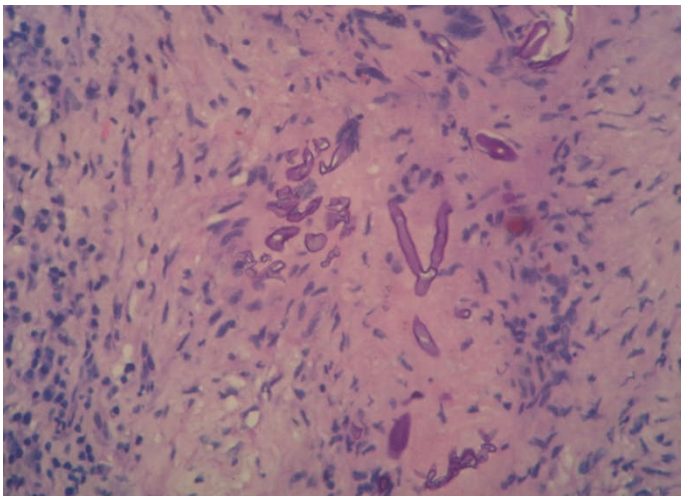

Figure 1. Biopsy slide showing granulomatous inflammation with fungal hyphae

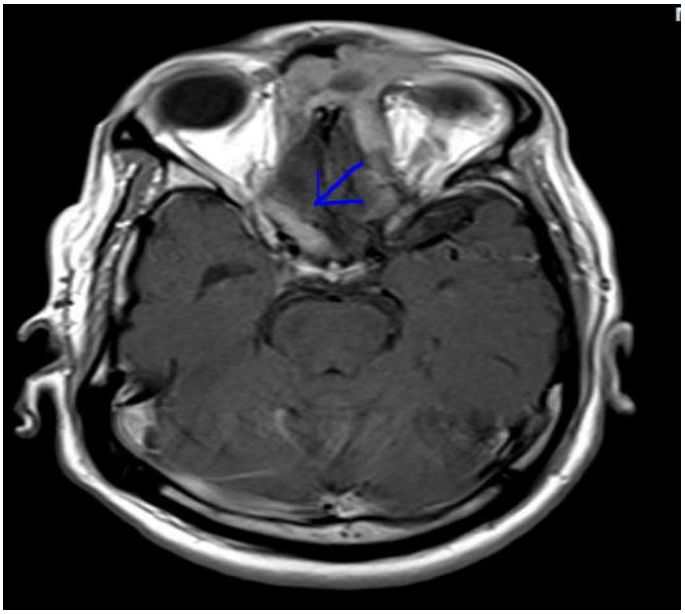

Figure 2: MRI brain showing thickening of right optic nerve (blue arrow)

DISCUSSION

Fungal rhinosinusitis is classified as noninvasive and invasive. Invasive fungal rhinosinusitis is further classified into 3 types: acute invasive (fulminant) fungal rhinosinusitis, chronic invasive fungal rhinosinusitis and granulomatous invasive fungal rhinosinusitis (Chakrabarti *et al.,* 2009). Acute invasive fungal rhinosinusitis usually occurs in immunocompromised patients with rapid progression with time course less than 4 weeks. Fungi belonging to the family Mucoraceae is chiefly responsible for this condition. Chronic invasive fungal rhinosinusitis and granulomatous invasive fungal rhinosinusitis have a time course exceeding 12 weeks. The former usually develops in immunocompromised patients who are infected with Aspergillus fumigatus, whereas the latter is associated with Aspergillus flavus infection (Chakrabarti *et al.,* 2009; Deshazo, 2009). Histologically Chronic invasive fungal rhinosinusitis is characterized by the dense accumulation of the hyphae with occasional invasion of the blood vessels whereas granulomatous form is characterized by noncaseating granuloma with foreign body or Langhans-type giant cells, occasional vasculitis, vascular proliferation, and perivascular fibrosis (Figure 1). Though hyphal forms can be seen, in the center of an area of necrosis, they are faintly seen in hematoxylin and eosin stained sections, they may go unnoticed unless selective special stains like methanamine silver are employed. They appear as septate hyphae, with branching at 45° angles and are about 2–4 mm in diameter (Somdutta Mitra *et al.,* 2018). Condiospores are also seen. This fungus differentiated from mucor specieswhich shows broader non-septate hyphae with dichomatous branching at 90° angles (Sapp *et al.,* 2004; Regezi *et al.,* 2003). Both these features were evident in our patient. Granulomatous invasive fungal rhinosinusitisis mainly described from middle eastern countries like Sudan, Saudi Arabia and also from Pakistan and India. It is also known as primary paranasal granuloma (Chakrabarti *et al.,* 2009; Deshazo, 2009). The characteristics of this condition is that it can produce an extensive disease in an immunocompetent patient leading to morbidity like blindness and the need for prolonged therapy to prevent the disease progression. The disease usually presents with gradual onset headache, facial pain and proptosis. Radiologically granulomatous fungal sinusitis shows hyperattenuating soft tissue mass within the sinuses with calcifications and destructive bone erosions and involvement of adjacent neurovascular structures. MRI provides better soft tissue resolution which shows hypointense in T1 and extremely hypointense in T2 weighted images. In contrast malignancy and bacterial infection shows hyperintense T2 weighted images. In our patient similar findings were noted within the sinuses along with enchancement or nodular thickening along the optic nerve extending into optic tract (Mukherjee *et al.,* 2016; Aribandi *et al.,* 2007) (Figure 2). Ophthalmological examination of the fundus during the initial presentation was showing normal fundus on both eyes. But during second presentation, it showed complete disc pallor on right and pallor in the temporal quadrant on left. Our patient, though he had an extensive sinus disease when he presented initially with minimal involvement of only right optic nerve at an early stage of the disease, the symptoms lasting for 4 months were not disabling. Management in such an early stage involved limited surgical debridement of the involved mucosa without any surgical complication but the post-operative medical management with anti-fungal became pivotal since when the patient defaulted therapy after 1 month the disease progressed

to involve the right optic nerve, optic chiasma and left optic nerve which was not the case the second time when he continued treatment for 6 months with oral anti-fungal leading to cessation of disease progress. Unlike other types of invasive fungal rhinosinusitis, granulomatous invasive fungal rhinosinusitis can be treated by surgery alone without the use of antifungal agents (Milosev *et al.,* 1969). Although the duration of antifungal agent administration for the treatment of granulomatous invasive fungal rhinosinusitis is arguable, Stringer et al. (2000) recommended that patients should receive amphotericin B for 6 weeks and should additionally receive amphotericin B or itraconazole for a little longer to control the disease. In our case, the patient received antifungal agents for 6 months and displayed no clinical symptoms or recurrence 3 months after the surgery. Therefore,we agree that antifungal agents should be administered to patients with granulomatous invasive fungal rhinosinusitis in addition to surgical debridement for more than 3 months. The prognosis of invasive fungal rhinosinusitis is generally poor. Although granulomatous invasive fungal rhinosinusitis has a good prognosis in comparison to the other types of invasive fungal rhinosinusitis, it tends to have a high relapse rate [dyananda] (Somdutta Mitra *et al.,* 2018). Gumaaetal (1992) reported that itraconazole effectively reduces the high relapse rate of granulomatous invasive fungal rhinosinusitis after surgery. Thus our study highlights a rare presentation of granulomatous fungal rhinosinusitis with optic nerve involvement leading to total loss of vision in one eye and temporal field loss of vision in the other eye, the need for surgical debridement followed by prolonged antifungal therapy for 3 to 6 months with Amphotericin B or Voriconazole or Itraconazole. We also place emphasis on the need for patient education about the morbidity this condition can produce if not taken proper care.

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