

Case Report

FUNGAL PANSINUSITIS LEADING TO ORBITAL COMPLICATIONS IN POORLY CONTROLLED DIABETES

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ABSTRACT

Mucormycosis is a rare infection caused by fungus belonging to the order mucorales. They are known opportunistic organisms, which potentially invade and infect a host with compromised immunity. Mucormycosis can present in different clinical forms. The common clinical types include rhinocerebral, pulmonary, gastrointestinal, disseminated and miscellaneous forms. Rhinocerebral mucormycosis is the most characteristic form and frequently involves the structures of the head and neck. It usually occurs in diabetic patients, especially when the diabetes is poorly controlled. The infection usually begins in the paranasal sinus or palate and then invades the adjacent sinuses and retroorbital region. It may extend through the apex of the orbit into the brain. This progression of the lesion gives a characteristic picture, with headache, swelling of the face, intranasal necrosis and serosanguinous discharge, marked proptosis, ptosis and ophthalmoplegia and loss of vision. All these are typically unilateral and are associated with radiologic abnormalities in the paranasal sinuses. Here we report a case of an uncontrolled diabetic male with orbital complications caused by a fungal pan-sinusitis. Early recognition and treatment with urgent surgical debridement and systemic antifungal therapy lead to rapid recovery. Therefore, clinicians should be familiar with the signs and symptoms of the disease so that morbidity and mortality can be reduced.

Keywords: Mucormycosis, Fungal Pansinusitis, Uncontrolled Diabetes, Orbital Apex Syndrome

INTRODUCTION

Rhino-orbital mucormycosis is a rare, opportunistic disease which first affects the para nasal sinuses and nasal cavity. It can lead to high morbidity and mortality. The early signs and symptoms of rhino-orbital mucormycosis are nonspecific that's why early diagnose is really difficult. Its onset is typically acute. Nasal discharge of blood may be present and nasal examination may reveal a black, crusty material. The most frequent signs of this disease are represented by: headache, facial pain, lethargy, diminution of vision affecting one eye, unilateral ophthalmoplegia, impaired colour vision and visual field. The infection spreads, invading the nerves, blood vessels, cartilages, bones, perineural space, inducing thrombosis of the cavernous sinus, the carotid artery and jugular vein and also nervous dysfunction.

Mucormycosis must be suspected in all diabetic patients, particularly those in ketoacidosis, and any debilitated or immune-compromised individual with multiple cranial nerve palsies with or without proptosis. It requires immediate hospitalization because this is a rapidly progressive and possibly life-threatening disease, and the treatment is complex both medical and surgical and must be conducted by a multidisciplinary team.

Fungi are known opportunistic organisms, which potentially invade and infect a host with compromised immunity. Fungal pan-sinusitis complicated with orbital apex syndrome or cavernous sinus syndrome has been described in immunosuppressed patients. Saprophytic fungi responsible for pan-sinusitis include *aspergillus* spp. And those of the mucorales order such as *rhizopus*. The typical presentation of rhino-orbital fungal infection is that of anterior orbital inflammation, severe visual loss, external ophthalmoplegia and fever. We describe a case with atypical orbital apex and cavernous sinus syndrome secondary to fungal pan-sinusitis.

CASES

A 48 year male patient presented to Ophthalmology OPD with blurring of vision right eye for 15 days, right sided severe retro-orbital pain for 15 days, inability to move right eye for 10 days. Patient was also

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having complains of headache, nasal sero-sanguinous discharge for last 15 days with swelling over right side of face. He was a known case of non insulin dependent diabetes mellitus for last 6 years. He was having poor glycemic control over last few months with blood sugar levels between 450 to 570 mg/dl. On ophthalmic examination best corrected visual acuity of right was 6/12 and that of left eye was 6/6. Intraocular pressure was 14 mm Hg both eyes, with upper lid edema with mild ptosis of right side. There was painful 3 mm proptosis of right eye with limitation of all extraocular movements along with miosed pupil of the same side.



Figure 1: Right external ophthalmoplegia due to orbital apex syndrome in case of fungal pansinusitis

Corneal sensation was reduced on right side. On the other hand there were normal extraocular movements, corneal sensation and normal sized normally reacting pupil on left side. The anterior pole of the right eye examined with a biomicroscope showed moderate conjunctival edema (chemosis) and normal in left eye. Fundus examination with a Volk lens revealed a hyperaemic disc with blurring of all disc margins, suggestive of papillitis. The left eye fundus was normal without any evidence of diabetic retinopathy in either eye.

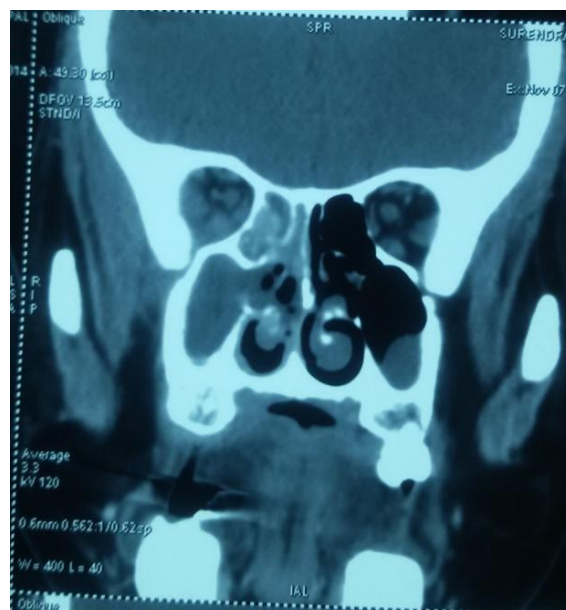


Figure 2: CT scan suggestive of right pansinusitis and right fungal orbital abscess

The CT scan finding was suggestive of a hypodense soft tissue mass expanding and opacifying the right half of nasal cavity with thickening of the turbinates and obstruction to right osteomeatal unit with right pansinusitis. Extraconal extension was seen into the right orbit medial to the medial rectus with a focal hypodense collection in that region. MRI of the brain and orbits confirmed right pansinusitis with involvement of the right orbital apex and increased signal intensities of the right extraocular muscles on

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T2- weighted images. The ophthalmic vein had no vascular anomalies; the right cavernous sinus appeared enlarged but with no signs of thrombophlebitis.

The histopathological exams were performed after prelevation of the biological material from the sinus's mucous membrane during the surgery. The results of the histopathological exams revealed enlarged necrosis with rare PMN and mitotic colonies, abundant inflammatory infiltrate with many PMN and fibrosis, periodic acid-Schiff (PAS) staining identified fungus from tissue debris, that's typical for mucormycosis. So, finally diagnosis of right rhinosinus mucormycosis, right orbital fungal abscess with right orbital apex syndrome and uncontrolled non-insulin diabetes mellitus was confirmed. Because of this was a life threatening and sight threatening situation, urgent surgery was planned by Otorhinology team. First of all, the general status and the glycemic control of the patient were improved with the help of endocrinologists and physicians. Surgical debridement of the necrotic tissue was done till the healthy tissue was reached. Patient was given systemic Augmentin and Amfotericine B for 2 weeks. During the first treatment, with Amfotericine the hepatic and renal functions and potassium levels were monitored. The patient responded to the treatment very well and his diabetic status was controlled. After 6 weeks of treatment the patient regained the ocular motility of his right eye and visual acuity improved to 6/6 on snellen's chart.

DISCUSSION

Fungi are known opportunistic organisms, which potentially invade and infect a host with depressed immunity. Fungal pan-sinusitis complicated with orbital apex syndrome or cavernous sinus syndrome has been described in immunosuppressed patients (Balch *et al.*, 1997; Lee and Sullivan, 1995; Mauriello *et al.*, 1995; Bikhazi and Sloan, 1998). A common complication of pan-sinusitis is orbital and /or cavernous sinus involvement(s) because of the close relationships between the orbit, cavernous sinus and adjacent paranasal sinuses. While the cavernous sinuses are anatomically related to the sphenoid sinuses, the optic canal and orbital apex are related to both the posterior ethmoid (Yeoh and Tan, 1994) and sphenoid sinuses (DeLano *et al.*, 1996; Sapci *et al.*, 2004). Involvement of the cavernous sinus or orbital apex is potentially debilitating, due to the close proximity of numerous cranial nerves within these confined spaces.

We report a case with fungal pan-sinusitis in the presence of poorly controlled diabetes with an orbital apex syndrome. The clinical picture was that of external ophthalmoplegia, orbital fungal abscess, Horner's syndrome, corneal hypoesthesia and papillitis.

Fungal pan-sinusitis commonly occurs in immunosuppressed patients. Cases of rhino-orbital mucormycosis and aspergillosis have been associated with poorly controlled diabetes mellitus, human immunodeficiency virus infection, Hodgkin's disease and chronic corticosteroid therapy (Balch *et al.*, 1997; Lee and Sullivan, 1995; Mauriello *et al.*, 1995; Bikhazi and Sloan, 1998). Early diagnosis with urgent surgical debridement and systemic antifungal therapy is the key to the management of this rhino-orbital infection.

However, a diagnostic dilemma leading to delayed treatment is not uncommon, either because of the absence of previous sinus disease, delayed progression of clinical signs or due to the atypical presentation of fungal infections with the lack of visible orbital inflammation. Sanborn *et al.*, (1984) described cases of sinusitis-related optic neuritis, which were initially misdiagnosed as idiopathic papillitis or demyelinating optic neuritis, leading to consequent mistreatment with systemic corticosteroids in the presence of an underlying sinus infection. Appropriate radiological investigations in the presence of an immunosuppressed patient with acute visual loss and ophthalmoplegia are strongly recommended. A normal sinus X-ray does not exclude an underlying sinusitis. This is due to sub-optimal visualisation of the sinuses, especially that of the ethmoid sinuses, as a result of radiological overlapping of the nasal bone and turbinate. A CT of the paranasal sinuses, orbits and brain is essential to confirm the diagnosis of pansinusitis and evaluate the extent of orbital and brain involvement. Prompt surgical drainage with tissue culture and histology is vital to the clearance of primary infection as well as in the guidance of subsequent anti-microbial therapy.

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Prognosis of such cases is variable. Although there have been cases of reported reversible visual loss following aggressive sinus treatment (Awerbach *et al.*, 1989; Moorman *et al.*, 1999) optic neuropathy can be permanent. On the other hand, ocular motor nerve palsies tend to recover. Lee *et al.*, (2004) evaluated the outcome of isolated sphenoid sinus disease post endoscopic sinus surgery and found a significant improvement in the function of ocular motor nerves compared to visual acuity, with a mean recovery time of 5.1 months. The poor outcome of the optic neuropathy is usually due to ischaemic vasculitis, where certain types of fungi like *Mucor* spp. have the propensity of invading blood vessels with consequent thrombosis and ischaemia. In conclusion, fungal pan-sinusitis with orbital or cavernous sinus involvement can present insidiously, resulting in an irreversible optic neuropathy. Diagnostic imaging of the anterior visual pathway and the paranasal sinuses is indispensable and the importance of prompt surgical debridement and drainage cannot be overemphasised.



Figure 3: Resolution of ocular findings after surgical and medical intervention

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