

Rhino-Orbital Mucormycosis in an Immunocompetent Adult

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Abstract

Mucormycosis is a rare life threatening opportunistic fungal infection caused by fungi of Mycorales order belonging to the Phycomycetes class. Mucormycosis usually develops in the patients with diabetes, prolonged corticosteroid use, hematological malignancies, chronic renal failure and other immunocompromised states. Incidentally we here by report a case of young immunocompetent adult without any obvious risk factors for Mucormycosis presenting with rhinoorbital swelling.

Key Words

Mucormycosis, Immunocompromised, Opportunistic Fungal Infection

Introduction

Mucormycosis is a rare life threatening opportunistic fungal infection caused by fungi of Mycorales order belonging to the Phycomycetes class. *Rhizopus oryzae* species accounts for 60% of all forms of mucormycosis and 90% of rhino-orbito-cerebral cases (1). These pathogens are ubiquitous spore forming saprophytes that invade host tissues especially blood vessels, causing thrombosis, infarction and eventual necrosis (2). Mucormycosis usually develops in the patients with diabetes, prolonged corticosteroid use, hematological malignancies, chronic renal failure and other immunocompromised states. It is rare even in high risk patients, representing 8.3 - 13% of all fungal infections. (3). Mucormycosis follows a rapid progressive course and if left untreated it is rapidly fatal.

This case report is significant because, although a diagnosis of Orbital mucormycosis is rare in adult immunocompetent individuals, it should remain as a differential in patients presenting with orbital cellulitis.

Case Report

A 42 years old male was admitted to the hospital with 7 days history of high grade fever, and initially developed a pea-size swelling near the left medial canthus with slight pain and tenderness. Within a period of 7 days the swelling involved whole of the left eye with protruding eye ball and periorbital swelling with maxillary swelling (Images 1,2). At the time of presentation to the hospital he had large swelling of the left eyeball and periorbital region and maxillary area, with complete occlusion of the left eye ball (*Fig-1*) and he complained of sudden diminution of vision of left eye. Ocular examination revealed no perception of light and pupil was dilated. Direct reaction to light was absent but consensual reflex was present. In addition he had full-blown left orbital apex syndrome (cranial nerves II, III, IV, V-1, 2), VI N palsy. Fundus showed occlusion of the central retinal artery. The left preauricular lymph nodes were enlarged and tender. A small ulcer with cheesy greyish escher was seen on the septum of nose on left side. The patient was investigated

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Fig. 1 Orbital Swelling with Proptosis



Fig.2 Complete Resolution of the Orbital Swelling

with a haemoglobin of 12 gm/dl. Leucocyte count of 14000/cumm, Differential count N84L12E4 with normal renal & liver functions. The conjunctival smear and scrapings from septum of nose and nasopharynx were taken. A fresh 10% KOH mount smear from the nose, showed aseptate right angled hyphae, typical of mucormycosis. Culture of scrapings from the septum of nose showed rhizopus growth after 7 days, in Sabouroud's medium. Histopathology was suggestive of mucormycosis.

The patient was subjected to CECT brain with orbital cuts, which revealed an abnormal heterogenous soft tissue swelling with postcontrast enhancement involving the right ethmoidal gallery with extra and intraconal orbital extension. Immediate treatment with Intravenous liposomal Amphotericin-B was started with a dose of 0.25 mg/kg body weight (in 500 cc of 5% dextrose over 6 hours) which was increased daily by 0.25/kg body weight till a maximum dose of 1 mg/kg body weight/day was reached (total cumulative dose of 2.5 gm) with monitoring of serum electrolytes and renal functions. The patient was simultaneously started on IV Meropenem 1 gm BD

After 2 weeks of hospitalization, patient started responding to the treatment and subsequently the whole of the orbital swelling regressed and patient had normal vision with no residual extraocular muscle weakness and was subsequently discharged after 4 weeks (*Fig 2*).

Discussion

Mucormycosis caused by Phycomycetes, is a fatal but rare infection that generally affect patients who are metabolically or immunologically compromised (4). In our study the patient was a young immunocompetent adult without any risk factors for Mucormycosis. A study reporting the epidemiology of Mucormycosis showed that the mean age of patients affected was 38.8 years with 65% being males. The most common types of infection affected the sinuses (39%), pulmonary organs (24%), cutaneous (19%), and disseminated developed in 23% of cases (5).

Orbital involvement results from invasion of lacrimal duct spreading through the thin medial orbital wall (6). The mucorales hyphae have a predilection for growth into arteries and the lymphatic system (7). The angioinvasion by hyphae produce a fibrin reaction and formation of "mucor thrombi" which occlude the arteries leading to ischemia, infarction and consequent formation of black necrotic eschar of the skin and mucosa that is characteristic of rhino-orbital mucormycosis (8).

Initial symptoms of rhino-orbito-cerebral Mucormycosis include fever (44% of cases), followed by nasal ulceration or necrosis, periorbital or facial swelling (33%) and decreased vision (33% of cases) (9). Other signs and symptoms include proptosis, chemosis, extraocular muscle paresis, perinasal cellulitis,

parasthesia, periorbital edema, mucopurulent rhinorrhea and nasal crusting (10,11). In our case the patient presented with similar signs and symptoms as described above. When the clinical picture includes the presence of sinusitis with black discoloration in the nose and palate in addition to a predisposing factor, a diagnosis of rhino-orbital mucormycosis should be highly suspected (11). Diagnosis of mucormycosis require a preoperative contrast enhanced CT to define the extent of disease which would show edematous mucosa, fluid filling the ethmoid sinuses, and destruction of periorbital tissue and bone margins. MR imaging is useful in identifying intradural and intracranial extent of the infection and also demonstrate the perineural spread (12). However biopsy is necessary to confirm the diagnosis. Invasive hyphae can be seen as ribbon-like, 10-20 micron wide, haphazardly branched organisms with little or no septation (13). Early diagnosis of mucormycosis and initiation of appropriate therapy within 5 days has a survival rate of 83% compared with a 43% survival rate at greater than 5 days (14).

Treatment of mucormycosis involves a combination of surgical and medical modalities in addition to correction of underlying medical disorder if possible. The standard medical therapy for mucormycosis is Amphotericin B in a dosage of 1.0 - 1.5 mg/kg/day for a period of several weeks to months, depending upon the clinical response and degree of drug's side effects (9,12,14). Other promising modalities worth mention include hyperbaric oxygen therapy and nasally nebulised amphotericin B (14). Most studies have showed that surgical debridement is necessary for complete cure (12) however in our case patient was completely cured with medical management only.

The prognosis of mucormycosis appears to depend primarily on two factors: early diagnosis and resolution of the predisposing condition. survival has been positively correlated with the time of diagnosis and initiation of treatment.

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