Paranasal mucormycosis: Unusual presentation in otherwise healthy child

V. P. Venkatachalam · Nitin Anand

Abstract

Mucormycosis is the fungal infection usually involving immunocompromized individuals. In this article we present a case of mucormycosis affecting a 12-year-old healthy boy who was treated by endoscopic surgical technique. Postoperatively patient was put on Amphotericin B nasal douching and oral Itraconazole. There was no recurrence till the last follow up at six month postoperatively.

Keywords Mucormycosis · Paranasal sinuses · Healthy child

Introduction

Mucormycosis is an infection caused by fungus, belonging to the order mucorales of class zygomycetes. These are opportunistic infections, reported to occur in association with Diabetes mellitus [1, 2, 6], leukemias and lymphomas [3], chronic renal failure, and HIV. It is seen to have varied clinical presentations, ranging from limited nose and paranasal [4] to rhino-orbital [5], rhino-cerebral [6], cutaneous, pulmonary, disseminated [7] or intestinal involvement. With the improved diagnostic tools and effective treatment strategies the prognosis has improved in this otherwise fatal infection [8]. The diagnosis is made on the basis of histological ground, which shows invasion of the surrounding tissues and/or vessels. Culture is required to identify the fungal species. Treatment preferred in such invasive disease is the surgical debridement of non-viable tissue with systemic administration of antifungal drugs. Role of endoscopic sinus surgery has also been demonstrated as a conservative approach to mucormycosis [5]. Here we are presenting a rare case of an immunocompetent child who was diagnosed to have mucormycosis of paranasal sinuses.

Case report

A 12 years old male child presented to outpatient department with complaints of nasal obstruction and rhinorrhoea. In spite of antibiotic courses the symptoms did not improve. Patient underwent routine hematological investigations and a Noncontrast Computed Tomograph (NCCT) of nose and paranasal sinuses. Both the maxillary sinuses, ethmoidal and frontal sinuses were found filled with mucosal hypertrophy. Patient underwent Endoscopic sinus surgery under general anesthesia. The histological report showed hypertrophied mucosa. Patient was discharged on antibiotics and steroid nasal spray. Patient was lost to follow-up.

After one year, patient returned with similar complaints of nasal obstruction and rhinorrhoea, patient also gave history of occasional episodes of epistaxis. Diagnostic nasal

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endoscopy revealed multiple polyps in left middle meatus and spheno-ethmoidal recess. A repeat NCCT of nose and paranasal sinuses revealed soft tissue opacification in left maxillary, ethmoidal, frontal and sphenoid sinuses with areas of calcification. Erosion of cribriform plate was also seen on C.T. scan (Fig. 1). Hematological investigations like complete blood count, serum glucose level were found to be within normal range. Chest X-ray was normal. There was no evidence of any systemic disease. Patient was taken up for revision endoscopic sinus surgery. Polypoidal tissue was removed from sinuses. In the core of polypoidal tissue was seen the necrotic tissue – brownish black in colour. Tissue was sent for fungal staining, fungal culture and histopathological examination (Fig. 2). Microscopic examination revealed the presence of fungal hyphae consistent with mucor. Postoperatively patient was started on nasal douching with Amphotericin – B and oral Itracon-

azole (100mg once daily). Patient is under follow up by regular diagnostic nasal endoscopy. Patient is doing well six months postoperatively.

Discussion

Mucormycosis is the term for fungal infection by mucoraceae family of order mucorales. Rhizopus, Mucor, Absidia and Rhizomucor are members of this family. Rhizopus oryzae is the pathogenic species accounting for 60% of all forms of mucormycosis and 90% of rhino cerebral cases of mucormycosis [2]. These pathogens are basically saprophytes growing on dead or decaying tissues and are known to cause an opportunistic infection in clinical setting. Most common predisposing factor reported has been diabetes [9]. Cases of mucormycosis without any predisposing factors have also been reported in the literature [10, 11].

They are known to invade the host tissues especially small and large arteries causing thrombosis and eventually infarction of tissue. With the varied presentation from infection limited to paranasal sinuses to dissemination throughout the body, the prognosis depends upon the early diagnosis and adequate treatment. As the disease progress beyond the limits of sinuses to orbit, brain, skin or lungs the prognosis becomes worse. The mucormycosis may take up a slower indolent invasive course as opposed to the usual acute fulminant course [12]. Cases have been reported where mucor presented as a fungal ball in sinuses [13]. Goldstein MF et al also reported in 1994 a rare case of allergic fungal sinusitis associated with mucor species [14].

Clinically it may present with fever, nasal ulceration or necrosis, facial swelling, diplopia, decreased vision or hemiplegia [5, 8]. All depending upon the extent of the disease involvement. Studies shows anesthesia of the affected area points towards the invasive mucormycosis [2].

For diagnosis, though the broad, irregularly shaped, nonseptate hyphae with right angle branching may be demonstrated on direct microscopy of KOH preparations the histopathology gives appropriate diagnosis of tissue invasion [10]. The culture of fungus is required to specify the species of mucorales involved in the infection. The treatment of mucormycosis should start with the aggressive control of any associated systemic disease causing immunosuppression. Medical management in form of intravenous Amphotericin B has been widely used with success in treating this condition [8, 9]. Cases had been reported where mucor presenting as fungal ball had been treated without intravenous Amphotericin B also [8]. Amphotericin B has also been recommended for local application as it allows increased concentration of agent at the source of active infection [5, 9, 15].

Along with the medical management, the surgical management in the form of debridement of nonviable tissue constitutes the treatment strategies on these cases. Controversy arises with the question of extent of debridement the patient

Fig. 1 CT Scan showing involvement of nasal cavity, ethmoids, sphenoid and maxillary sinuses with areas of hyper attenuation

Fig. 2 Culture bottles showing the mucor growth

requires. Cases have been reported in literature where in spite of clinical symptoms or radiological signs suggestive of orbital involvement, the conservative surgical approach has been resorted to [5, 16]. Endoscopes have opened up a new approach to the management of limited mucormycosis. They provide an effective means of diagnosis and management of these patients. Postoperatively it is used for periodic inspection of the sinuses and nasal cavity. It also helps in monitoring the response to the treatment.

In our case though the histopathology showed invasion of the mucosa of sinuses, it was limited to the sinuses. Endoscopic surgical removal of the disease was followed by Amphotericin B nasal douching and oral Itraconazole. The patient is followed up regularly at monthly interval and there has been no recurrence till the last follow up at six months.

We feel that use of endoscope is an effective tool in not only removing the disease precisely but also monitoring the response to treatment and detecting any early recurrence.

Summary

Mucormycosis is an invasive fungal infection seen usually in the immunocompromized patients. But in rare cases it may be seen as fungal ball in otherwise healthy patients. The prognosis depends on the extent of disease at the time of presentation. Patients presenting with fever, cerebral or systemic involvement carry poor prognosis. Histopathological demonstration of fungal hyphae invading the tissue is must for diagnosis. Fungal culture gives the idea of exact species involved in the infection. Treatment consists of correction of immunocompromised status, surgical debridement of nonviable tissue and systemic antifungal agents.

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