Sinonasal Mucormycosis in a Diabetic - Case Report

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Abstract

Background : Mucormycosis is one of the rapidly progressing and lethal form of fungal infection which involves the nose and paranasal sinuses of the head and the neck regions. It manifests as rhinocerebral, sinonasal, pulmonary, gastrointestinal, cutaneous or disseminated form. The underlying conditions can influence clinical presentation and often delay diagnosis, with resultant poor outcomes.

Objective: To present and discuss the case of a diabetic patient with sinonasal mucormycosis presented with facial pain and discolouration of right side of nose and face due to mucormycosis.

Case report: A 23 years old female patient presented to emergency with uncontrolled blood sugars, pain and swelling over right side of face and blackish discolouration of right side of nose. On examination, there was bluish black discolouration of skin over right side of dorsum of nose and right ala of nose and right nasal vestibule. Sinus CT scan showed features suggestive of fungal sinusitis. Mucormycosis was suspected, confirmed with biopsy and surgical debridement and medial maxillectomy done. Intravenous amphotericin B was administered for 21 days. The patient was followed up regularly, the disease appearing to be resolved.

Conclusion: Mucormycosis is a rapidly progressive fatal infection mostly reported in immune-compromised individuals. Emergency multidisciplinary treatment for control of blood sugars, surgical debridement and effective antifungal medications gives better clinical outcome.

Key words: Mucormycosis, Invasive fungal sinusitis, Medial maxillectomy, Amphotericin B

Introduction

Sinonasal mucormycosis is an acute invasive fungal infection which is rare, opportunistic and potentially fatal, that mostly occurs in immune-compromised patients caused by saprophytic and opportunistic fungi of class Phycomycetes, order mucorales, family mucoraceae belonging to genus mucor and Rhizopus[1]. Following inhalation of spores, the fungi grow and invade neural and vascular structures, leading to vessel thrombosis and resultant mucosal necrosis. The fungi then extend beyond the affected sinus through a combination of bony destruction, perineural and perivascular spread. A high degree of clinical suspicion is required in order to correctly and promptly diagnose and manage this condition[2].

Patients with diabetes have decreased granulocyte phagocytic ability with altered polymorphonuclear leucocyte response, consequently are more susceptible. Diabetes mellitus is associated with 40% of mucormycosis cases overall[3]. The disease is rapidly progressive, so prompt diagnosis and aggressive therapy are essential for successful containment of disease.

Diagnosis of mucormycosis was established on strong clinical suspicion with presence of greyish black crusting on nasal endoscopy which is confirmed by histopathology examination. Immediate correction of underlying immune-compromised status and debridement with intravenous liposomal amphotericin B is required. Early detection and aggressive multidisciplinary management are must for the successful treatment of mucormycosis[4].

The aim of this case report is to present a patient with sinonasal mucormycosis in order to draw attention to its existence in our environment and to emphasize the need for high index of suspicion.

Case report

A 23 years old female patient presented to emergency with pain over right side of face since 8 days with subsequent swelling of right side of face and blackish discolouration of right side of nose and surrounding maxillary area (Fig 1). She was a known case of type 1 diabetes mellitus since 3 years and was on regular

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insulin thrice a day with poorly controlled blood sugars. There was no history of difficulty in breathing or blurring of vision. On examination, there was bluish black discolouration of skin over right side of dorsum of nose and right ala of nose and right nasal vestibule. There was tenderness of right ethmoidal and maxillary sinuses with periorbital oedema of right eye. The examination of oral cavity revealed normal findings with no signs of impending perforation of hard palate.

(Consent was taken from patient regarding using of photographs for article writing.)

Figure 1 (a,b) Patient at the time of presentation.

Investigations at admission showed the following values: RBS-254mg/di. HbAIC- 15. Serum creatinine-2.5mg/d, serum potassium-2.5mg/dl, Hb- 10.5g/dl, total count-7200 cells/cumm, neutrophils-68%, lymphocytes- 30%.

CT of nose and paranasal sinuses showed severe mucosal thickening of right maxillary, ethmoidal, frontal sinus with near complete luminal occlusion with features suggestive of fungal sinusitis (Figure 2,3). No intracranial involvement seen in CT scan.

Based on the history and clinical presentation, provisional diagnosis mucormycosis, ecthyma gangrenosum and lethal midline granuloma were made.

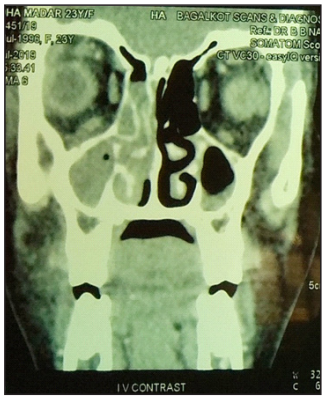

Figure 2- CT Nose and PNS (coronal section)

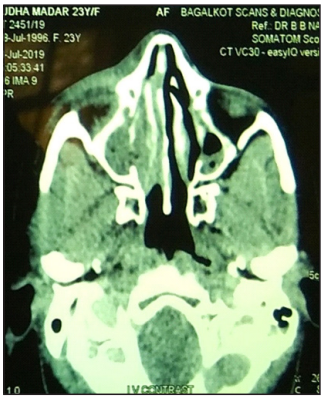

Figure 3- CT Nose and PNS (axial section)

Patient was started on broad spectrum antibiotics, intravenous fluids and injection Amphotericin B deoxycholate 50mg per day with monitoring of serum potassium and serum creatinine levels. Blood sugar levels were corrected with regular insulin injections according to sliding scale. Biopsy was taken at the site of discolouration of mucosa of right nasal vestibule. Histopathological report revealed features suggestive of mucormycosis, KOH mount showed fungal elements and on fungal culture, mucor species were isolated.

Under general anaesthesia, the necrosed part of nose and adjacent face were excised upto bone depth which were found to be totally necrotic. Underlying bone of anterior wall of maxilla was also found to be devitalized and necrotic. Medial maxillectomy done and necrotic anterior wall of maxilla was removed (Figure 4). The necrotic mucosa of sinus was completely taken out. Posterior bony wall of sinus was intact. All the bony edges were saucerized using microdebrider. The surgical excision left behind large defect on the face (Figure 5).

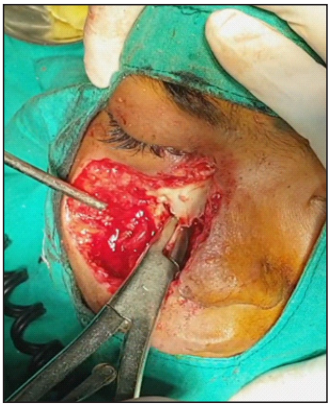

Figure 4 - Maxillectomy

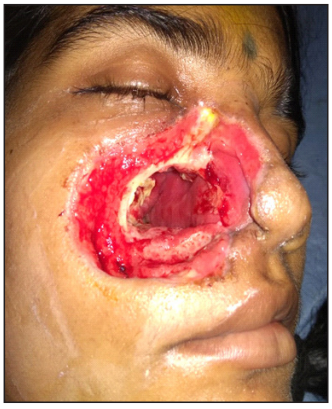

Figure 5 - Facial defect after surgery

Following the debridement and with continuation of injection amphotericin B deoxycholate and blood sugar control, she made a slow but steady progress and her wound became clean with healthy granulation tissue (Figure 6). An obturator was constructed for the patient for facial defect on right side (Figure 7). Injection amphotericin B deoxycholate was given for 21 days and advised tablet voriconazole 200mg BD for 3 months. Free flap surgical reconstruction was planned by plastic surgeon but postponed due to uncontrolled blood sugar levels.

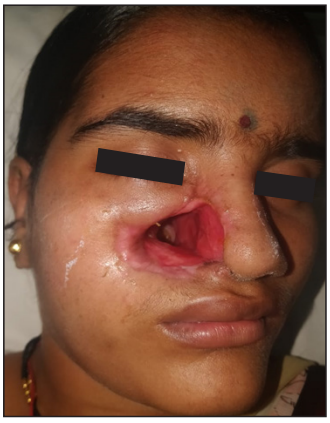

Figure 6 - Healing facial defect

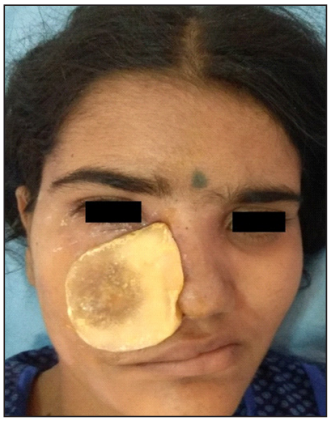

Figure 7 - Obturator for facial defect

Discussion

Mucormycoses are a group of invasive infections caused by filamentous fungi of the Mucoraceae family. Most human infections result from inhalation of fungal sporangiospores that have been released in the air or direct inoculation of organisms into disrupted skin or mucosa. Mucormycosis is primarily seen in patients with chronic conditions, particularly uncontrolled diabetes mellitus and haematological malignancies, because these patients are immunocompromised. Patients with extensive burn injuries, renal failure, prolonged corticosteroid use and deferoxamine treatment have also been reported to have mucormycosis[5].

Diabetic patients are particularly at risk from Zygomycetes (Rhizopus, Mucor) as these organisms have an active ketone reductase system and thrive in high glucose acidotic conditions[1]. In diabetic patients, especially with elevated blood sugar levels, the spores germinate, hyphae develop and fungi begin an inexorable march through lung tissues, invading blood vessels and surrounding tissues. As blood vessels become involved, thrombosis occurs resulting in tissue necrosis and fungi continue to grow in this devitalised tissue[6].

Most patients require both surgical and medical treatments. The line of treatment for such cases requires aggressive surgical debridement of the infected area. Medications also play an important role. Two main goals are sought at the same time: antifungal medications to slow or halt fungal spread and medications to treat any debilitating underlying diseases. AmphotericinB (initially intravenous) is the usual drug of choice for antifungal treatment. Patients with underlying diseases like diabetes need their blood sugar levels optimally controlled. Patients may need additional surgeries and usually need antifungal therapy for an extended time period (weeks to months) depending on the severity of the disease[7]. The prognosis of mucormycosis is usually fair to poor; the prognosis depends on the overall health of the patient, the speed of diagnosis and treatment, the patient's ability to respond to treatments and the complete debridement of the infected body area. In this case, patient withstood surgery and amphotericin B for 21 days and wound healed with regular dressing.

Although amphotericin B deoxycholate (AMP) remains the licensed antifungal agent for mucormycosis, lipid formulations of amphotericin B are considered a safe and efficient alternative. Recommended starting doses for the lipid formulation of amphotericinare 5-7.5 mg/kg/day with higher dosages (up to 10 mg/kg/day) recommended for CNS involvement.

Iron chelation therapy and posaconazole could be considered in cases of refractory infection or polyene intolerance. Itraconazole has known in vitro activity against the Mucorales order; however in vivo its effectiveness is limited to the Absidia species thus limiting its clinical use[8]. When diagnosed early, mucormycosis may be cured by a combination of surgical debridement of the infected area and systemic administration of amphotericin B for up to 3 months[9]. Proper management of the underlying disorder is an important aspect affecting the final outcome of treatment. Survival rates among groups of patients with invasive sinus disease without cerebral involvement may be as high as 50-80%; if infection spreads to the brain, case fatality ratios exceed 80%[10]. Prognosis involves high morbidity and mortality and may improve with rapid diagnosis, early management and reversible underlying risk factors.

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