

CASE REPORT

Mucormycosis Maxilla: Behavior of Disease in an Immunocompetent Young Male Patient

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ABSTRACT

Mucormycosis is an opportunistic fulminant fungal infection usually noted in immunocompromised individuals like diabetic and AIDS patients, patients receiving systemic corticosteroid therapy, cancer chemotherapy, and organ transplant patients. The disease is very rare to affect healthy individuals. Here we report a case of a 21-year-old immunocompetent young male presented with cheek swelling and hard palate ulcer. Clinically, radiologically, and histologically, the diagnosis of maxillary mucormycosis was made.

Keywords: Diabetic, Fungi, Immunocompetent, Mucormycosis, Necrosis.

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INTRODUCTION

Mucormycosis is an opportunistic fulminant fungal infection of class Phycomycetes (Zygomycetes), order Mucorales, family Mucoraceae.¹⁻⁵ Mucormycosis was first described by Paultauf in 1885.⁵ It is one of the most rapidly progressive lethal infection in human being having mortality of 70 to 100%.⁶ The infection manifests as rhinocerebral, pulmonary, gastrointestinal, cutaneous, or disseminated form.¹ The predisposing factors are uncontrolled diabetes, renal failure, tuberculosis, organ transplant, long-term corticosteroids, immunosuppressive therapy, cirrhosis, burns, protein energy malnutrition, leukemias, lymphomas, and AIDS.¹ Mucormycosis has been rarely reported in apparently normal, immunocompetent young individuals⁷ and may create a diag-

nosis as well as therapeutic dilemma. We present a rare case of mucormycosis of maxilla in a 21-year-old young, immunocompetent male patient. This is an ever youngest case of mucormycosis with osteomyelitis of maxilla in an immunocompetent patient on reviewing the literature.

CASE REPORT

A 21-year-old male patient presented to the outpatient Department of Otorhinolaryngology and Head and Neck Surgery, Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh with the left cheek swelling, pain, and left eye swelling for past 1½ years, insidious in onset with ulcer and fistula on left side of hard palate since 6 months. He also complained of blurring of vision since 5 months. There was no history of visual loss or bleeding from the nose. He had no history of diabetes mellitus or HIV infection or prolonged corticosteroid therapy. The patient had a history of left endoscopic clearance and orbital decompression at private hospital, 12 months back. Postoperative biopsy was suggestive of granulomatous lesion only. On examination, intraorally, erosion of left side of hard palate with blackish discoloration around was noted with halitosis. There was a 3×3 cm swelling present over left cheek. There was left conjunctival chemosis. Previous computed tomography (CT) nose and paranasal sinuses coronal and axial views showed heterogenous soft tissue involving left nasal cavity, ethmoids, maxillary sinus, and medial and retrobulbar orbit (Fig. 1). Latest CT showed hard palate fistula left side and soft tissue density left maxillary sinus and orbit medially (Fig. 2A). The fungal smear from hard palate ulcer was aseptate hyphae. Based on clinical examination and fungal smear, patient was taken up for left total maxillectomy by external approach (Weber Ferguson incision). Postoperative histopathology was reported as mucormycosis with osteomyelitic maxillary bone (Fig. 2B). Hematoxylin and eosin (H&E) section showed bland necrosis of intertrabecular marrow spaces with mucor ×200 and broad aseptate ribbon-like foldable on itself fungal profile conforming to the morphology of mucormycosis ×400 (Fig. 3A) and right angle branching (Fig. 3B). Periodic acid–Schiff (PAS) stain highlighted similar fungal profiles within giant cells surrounded by fibrosis and chronic lymphoplasmacytic infiltrate ×200 (Fig. 4A). Gomori

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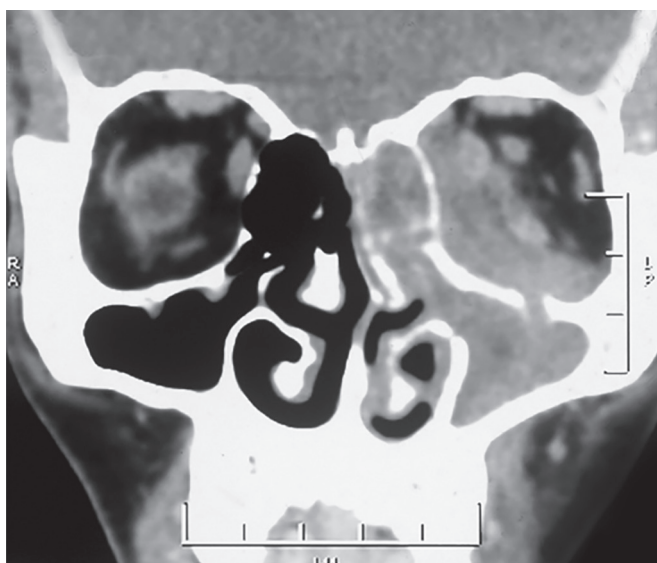
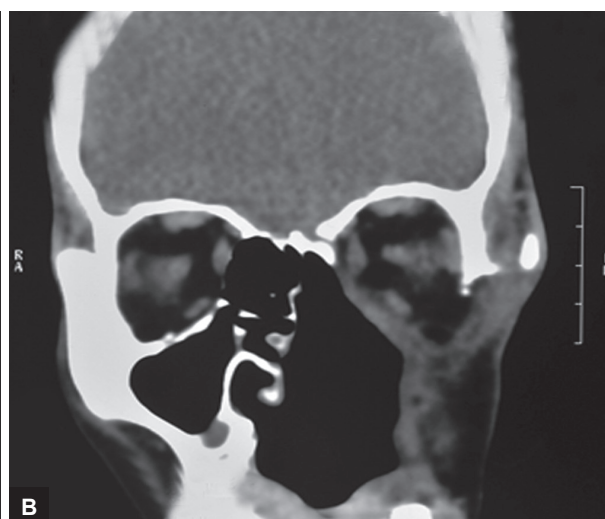
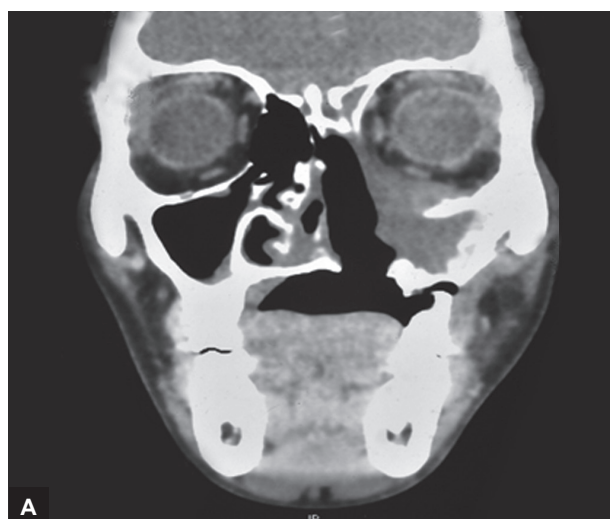


Fig. 1: Computed tomography PNS coronal and axial views showing heterogenous soft tissue involving left nasal cavity, ethmoids, maxillary sinus, and medial and retrobulbar orbit

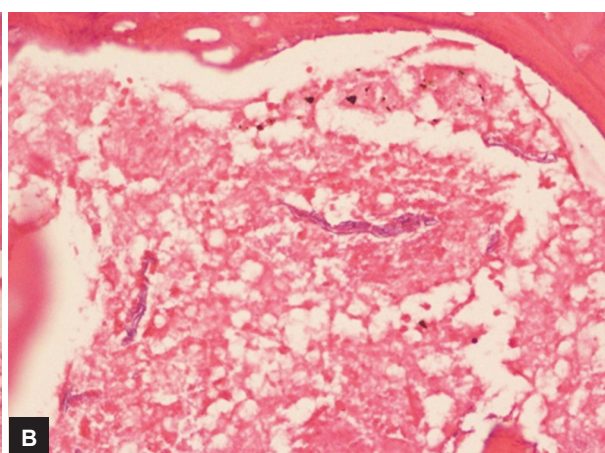
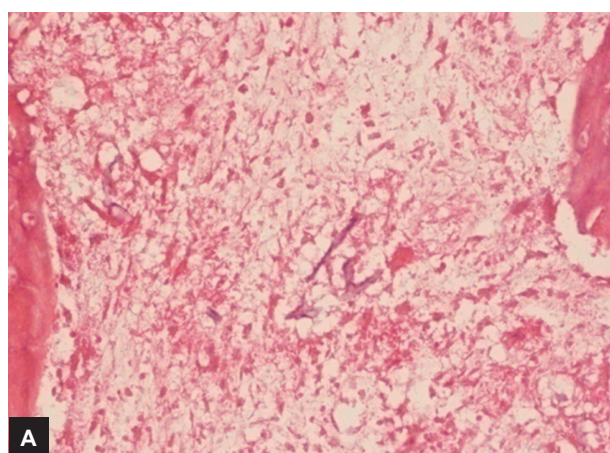
methenamine silver (GMS) stain highlighted similar black-colored argyrophilic fungal profiles $\times 400$ (Fig. 4B). Immediately postoperatively, liposomal Amphotericin B was started intravenously and cumulative dose of 3 gm was given. At 8th postoperative week, patient had well-healed Weber Fergusson incision scar. Plain CT scan axial and coronal was done at 8th postoperative week after total maxillectomy and 3 gm of intravenous liposomal Amphotericin B which showed well-healed cavity with no residual disease (Fig. 5A). Nasal endoscopy performed also showed well-healed cavity (Fig. 5B). The patient was given dental prosthesis for the cavity later on and he is alright with a follow-up of 1 year.

DISCUSSION

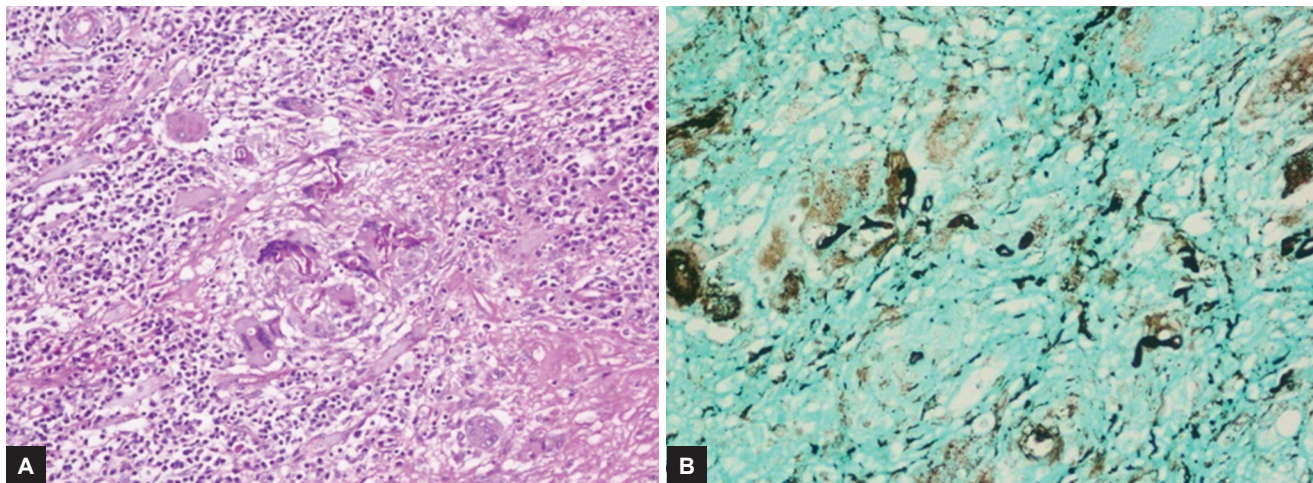
Zygomycosis usually occurs in patients with an underlying factor, such as diabetes mellitus, immunosuppressive therapy, solid organ and hematopoietic cell transplantation,



Figs 2A and B: (A) Computed tomography PNS showed hard palate fistula left side and soft tissue density left maxillary sinus and orbit medially; and (B) postoperative disease free cavity



Figs 3A and B: (A) H&E section showing broad aseptate ribbon-like foldable on itself fungal profile conforming to the morphology of mucor $400\times$; and (B) H&E section showing right angle branching fungal profile conforming to the morphology of mucor $400\times$



Figs 4A and B: (A) Periodic acid–schiff stain highlights similar fungal profiles within giant cells surrounded by fibrosis and chronic lymphoplasmacytic infiltrate 200 \times ; and (B) and GMS stain highlights similar black-colored argyrophilic fungal profiles 400 \times



Figs 5A and B: Clinical: (A) Nasal endoscopic; and (B) patient showing well-healed Weber Fergusson incision at 8th postoperative week

leukemia, lymphoma, burns, glomerulonephritis, gastroenteritis, broad-spectrum antibiotic use, hemodialysis, or deferoxamine therapy.⁸⁻¹⁴ Our patient was young immunocompetent with no comorbidity.

Rhinocerebral mucormycosis is the most common form of the infection, accounting one-third of all cases of mucormycosis.^{11,15} Nasal obstruction or congestion with noisy breathing, headache, odontalgia, maxillary pain, and hyposmia or anosmia may be seen. Necrotic eschars in the nasal cavity, the turbinates or the palate, and necrotic facial lesions correspond to aggressive angioinvasive infections.^{15,16}

Diagnosis requires concurrent use of clinical, mycological, pathological, and radiological methods.¹⁷ We diagnosed our patient by clinical history, examination, and positive fungal smear with aseptate hyphae. Histopathology demonstrated aseptate hyphae branching at right angle.

The treatment of zygomycosis comprises three main factors: Surgical debridement, antifungal therapy, and

relieving underlying metabolic problems or improving immune status.¹⁵

In our patient, surgical debridement of left maxilla was done in the form of total maxillectomy with preservation of orbit. The antifungal treatment with cumulative dose of intravenous 3 gm of liposomal Amphotericin B was given.

Khiste et al¹⁸ reported a case of a 50-year-old immunocompetent female with mucormycosis of maxilla masquerading as neoplasm. But our patient was quite young to develop this acute invasive disease.

Retrospective analysis for 10 years by Chakrabarti et al¹⁹ in India found that rhino-orbito-cerebral type (44.2%) was the commonest, followed by cutaneous (15.5%) and renal (14.0%). Considerable number of patients, 22 (22.9%) out of 129 patients were apparently healthy hosts in this series. But none of them was as younger like the one in our study. Although other reports²⁰ like isolated mucormycosis orbit in a young child of 7 years has

been mentioned, but our patient had presentation in nose, paranasal sinuses, and orbit also. Our patient was treated outside with endoscopic sinus surgery and no antifungal was given at that time but the patient survived and resisted the disease probably because of his younger age and no associated comorbidities. The clinical symptoms, the patient presented with, did not had acute fulminant course.

Mucosal and cutaneous epithelium and endothelium act as a barrier to tissue invasion and angioinvasion. Therefore, invasive fungal infection in immunocompetent patient is rare. Mucor infection in such patients may be due to ability of mucor sporangiospores to attack epithelium previously damaged by infection, direct trauma, or due to toxins or proteases secreted by sporangiospores that may directly destroy endothelial cells.²¹

CONCLUSION

In immunocompetent/otherwise healthy individuals, mucormycosis infection has a worldwide distribution. What might be the real predisposing factors involved in its pathogenesis in such patients and the real causes of this peculiar geographic distribution still remains unknown. It is likely that, a chronic insult of a well-defined and localized body area might have resulted in a local Immunocompromised state, as in our youngest immunocompetent patient, thus leading to the development of an invasive fungal infection.

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