

Necrosis of maxilla, nasal, and frontal bone secondary to extensive rhino-cerebral mucormycosis

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ABSTRACT

Mucormycosis is an opportunistic fulminant fungal infection caused by zygomycetes. This fungus can cause a variety of infections in human beings, particularly in the uncontrolled diabetes mellitus. Zygomycetes impinge into the vascular network, resulting in thrombosis and necrosis of the surrounding hard and soft tissues. The infection begins in the nose and paranasal sinuses due to inhalation of fungal spores and spread to orbital and intracranial structures either by direct invasion or through the blood vessels. Sinus mucormycosis is often accompanied by a poor prognosis and a high mortality rate. Hence, aggressive surgical intervention with antifungal therapy is usually necessary. Early diagnosis and prompt treatment can reduce the mortality and morbidity of this lethal fungal infection. We report a case of aggressive rhino-cerebral mucormycosis in a 58-year-old female patient with uncontrolled diabetes mellitus.

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INTRODUCTION

Mucormycosis is one of the most rapidly progressing and fatal form of fungal infection in human beings.^[1] It is caused by zygomycetes, which can be found on decaying vegetation and in the soil. This fungus invades the arteries, forms thrombi within the blood vessels and cause necrosis of the surrounding hard and soft tissues.^[1-3] The infection begins in the nose and paranasal sinuses and can spread to orbital and intracranial structures either by direct invasion or through the blood vessels.^[4] Usually, mucormycosis manifests as rhinocerebral, pulmonary, gastrointestinal, cutaneous, or disseminated form.^[1,3,5] The predisposing factors are depressed immune system, diabetes mellitus,

hematopoietic stem cell transplantation, hematological malignancies, or long-term steroid use.^[3,5,6]

The first case of human mucormycosis infection was reported by Paltauf in 1885.^[7] The mortality rate ranges from 10 to 100% depending on the place of infection and underlying diseases. Sinus mucormycosis is often accompanied by a poor prognosis and a high mortality rate (46%).^[5] Successful treatment of mucormycosis depends upon the early diagnosis, reversal of underlying predisposing risk factors, surgical debridement, and prompt antifungal therapy.^[3,8]

CASE REPORT

A female patient of 58 years reported to the Department of Oral and Maxillofacial Surgery with chief complaint of pain in the left maxillary posterior region and difficulty in eating. Intra-oral examination presented total denudation of the entire palate with extensive necrosis of overlying mucosa [Figure 1]. The condition became worse with ptosis, diplopia, ophthalmoplegia, cutaneous pustules, and left facial numbness [Figure 2]. Pain was moderate

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in nature, aggravated on bending the head. Patient had history of intermittent fever, purulent discharge from cutaneous pustules, paraesthesia, or foul odor. Patient was known case of diabetes. Our provisional diagnosis was rhino-cerebral mucormycosis, osteomyelitis with purulent oronasal communication, and with suspicious palatal tumor. Paranasal sinus radiograph showed haziness of left maxillary sinus with erosion of lateral sinus wall.

Biochemical investigations revealed fasting blood sugar level of 264 mg/dl and postprandial blood sugar level of 396 mg/dl. Incisional biopsy of hard tissue along with the adjacent soft tissue from the palate was taken under local anesthesia. Histopathological examination showed the presence of 'rhizopus oryzae' confirming the diagnosis of rhino-cerebral mucormycosis.

Aggressive medical therapy was started in consultation with Department of Medicine to control blood sugar and

to control further spread of fungus. Amphotericin B was started after testing in the dose of 0.5 mg/kg on alternate days, a total of 3,000 mg. Blood urea and creatinine levels were monitored as the drug can cause renal toxicity. Patient responded well to the treatment with prompt cessation of acute signs and symptoms. Computed tomography (CT) was advised which revealed erosion of bone from virtually whole of the facial skeleton particularly palate, left maxillary sinuses, and frontal sinus [Figure 3].

Surgical debridement of frontal and maxillary sinus and sequestrectomy was planned. The initial sequestrectomy removed the palate, maxilla, and left maxillary sinus membrane. Drainage was established through frontal sinus. Outer and inner tables were removed [Figure 4]. Histopathological examination of excised necrotic bone confirmed extensive rhino-cerebral mucormycosis. Patient was discharged after 15 days in good general condition and anti-fungal therapy (Amphotericin B) was continued for 6 months.



Figure 1: Intra-oral photograph showing total denudation of the entire palate with extensive necrosis of overlying mucosa



Figure 2: Profile photograph showing ptosis of the left eye and cutaneous pustules on the cheek

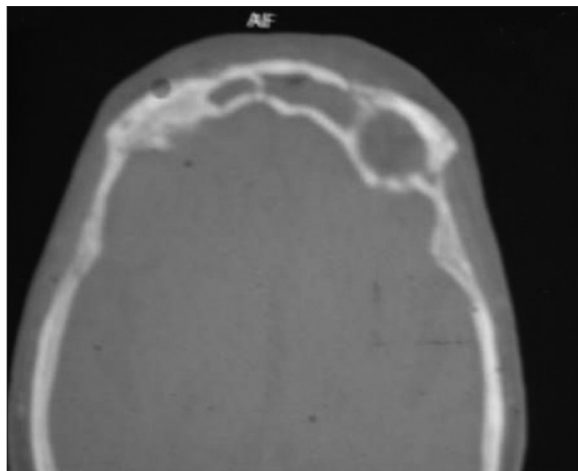


Figure 3: Axial section of CT scan showing erosion of the frontal bone and expansion of the frontal sinus



Figure 4: The outer and inner tables of frontal sinus removal through bilateral subfrontal approach

Patient was seen after 1 year in fairly good health. Until now, no recurrent infection signs or symptoms have been noted clinically.

DISCUSSION

Mucormycosis (Zygomycosis, phycomycosis) is an acute opportunistic infection caused by a saprophytic fungus. In 90% of the cases of rhino-cerebral mucormycosis, rhizopus is the main pathogen. This microbe may be cultured from the oral cavity, nasal passages, throat, and stool of healthy patients without clinical signs of infection. Predisposing factors for mucormycosis are uncontrolled diabetes mellitus, renal failure, cirrhosis, malignancies, long-term corticosteroid and immunosuppressive therapy, and AIDS.^[1,2,4] Our patient also had uncontrolled diabetes.

Diabetes decreases granulocyte phagocytic ability with altered polymorphonuclear leukocyte response of the patient. Peripheral vascular disease in diabetic patients also causes local tissue ischemia and increased susceptibility to infections; therefore, thrombosis of the internal maxillary artery or descending palatine artery caused by mucormycotic infection results in necrosis of the maxilla.^[4,9]

This fungal infection usually originates from the paranasal sinuses. Once fungal hyphae enter into the blood stream, they can disseminate to other organs such as cerebrum or lungs. Mucor hyphae form thrombi within the blood vessels that reduce vascularity to the tissues and cause necrosis.^[2,4]

The most common clinical presentation of mucormycosis is rhino-orbital-cerebral infection. The infection usually presents as acute sinusitis with fever, nasal congestion, purulent nasal discharge, and headache. The most common presenting features are ophthalmologic, including ptosis, proptosis, visual loss, and ophthalmoplegia.^[10] Our patient also presented with the same features.

The successful treatment of mucormycosis requires four steps: (1) early diagnosis; (2) reversal of underlying predisposing risk factors like control of diabetes; (3) surgical debridement of necrotic bone; and (4) prompt antifungal therapy.^[3] A multidisciplinary approach is required for management of such patients.

Initiation of antifungal therapy within 5 days after diagnosis is associated with improvement in survival (83 vs. 49% survival).^[11] Intravenous amphotericin-B is the drug of choice. The usual starting dose is 5 mg/kg daily, and the dosage sometimes will increase up to as high as 10 mg/kg daily in an attempt

to control the infection.^[3,8] Antifungal therapy should continue until all signs of infection have been resolved, and often extends for months.

Debridement of necrotic tissues may be critical for complete eradication of mucormycosis. Combination of surgical management with antifungal therapy has a favorable outcome.^[8] The mortality rate varies depending on its form and severity. A review study has mentioned the mortality rate 10% (localized cutaneous type) to 100% (generalized disseminated type) depending on the infected place and underlying diseases, and it is 46% in sinus mucormycosis.^[5]

Mucormycosis is a fatal infection with poor prognosis but rhino-cerebral mucormycosis has a higher survival rate, because it frequently can be diagnosed earlier and the most common underlying cause, diabetic ketoacidosis, can be treated readily. Survival rates now exceed to 80% with early medical and surgical management.^[12]

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