Fatal Rhino Orbital Zygomyosis (mucormycosis) mimicking as Deep neck abscess in a HIV patient – A Rare case report

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Abstract

The case reports of zygomycosis in the literature are plenty. Also there are reports indicating an increase in the atypical presentations in a HIV/AIDS patient. This case report assumes importance due to the confusing picture of co-existing deep neck space involvement of mucormycosis in a HIV patient with normal CD4+ cell count.

Keywords: AIDS/HIV, Mucormycosis, Neck abscess

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Introduction

Zygomycosis is one of the deep mycoses in a HIV patient, with others being histoplasmosis, cryptococcosis and aspergillosis. Rhizopus oryzae (arrhizus) is the most common cause of infection. This species belongs to family of Mucoracea under the order Mucorales and the class Zygomycetes. The other order Entomophthorales, under the class Zygomycetes is less common mycoses affecting mostly the immunocompetent host, whereas the species of mucoraceae are known mostly to occur in immunocompromised hosts. The sequence of these infections in such patients is nearly lethal; although the disease may become indolent once the risk factors are reversed¹. We present an eighteen year female patient, positive for the retrovirus, who succumbed to the rhino-orbital form of these deep mycoses. There are very few reports of zygomycosis in a HIV patient ²-¹⁴.

Case Report

An Eighteen year old female patient presented to the outpatient Ophthalmology
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Department with sudden onset of left eye proptosis and diminution of vision in the left eye for past two days. Patient gives history of left cheek swelling, with low-grade fever, difficulty in mouth opening and difficulty in swallowing. There was no respiratory difficulty or voice change. Patient was a known case of HIV I infection for past six months with her recent CD4+ cell count of 600 cells/cu mm. On examination, the patient revealed left eye axial proptosis, upper and lower lid edema and conjunctival chemosis. Extra-ocular movements were restricted in all range of gaze, vision reduced to finger counting and pupils being non-reactive to light and consensual light reflex was present. Patient had diffuse swelling of the left cheek and left neck extending down to the body of the mandible. Mouth opening was restricted to two-finger breadth, and a bulge was present in the lateral pharyngeal wall with tonsils pushed medially. Emergency contrast enhanced computed tomogram revealed soft-tissue density, filling and expanding the left maxillary (Figure 1) frontal and ethmoidal sinus (Figure 2), left nasal cavity, nasopharynx obliterating the fossa of Rosenmuller and Eustachian tube opening. Maxillary sinus ostium and infundibulum on the left side were widened (Figure 1). Left eye proptosis was present and soft-tissue density was seen in the medial wall of the orbit (Figure 3) and preseptal space. Retro-orbital fat appeared hazy. Soft tissue density with an air fluid level was also seen in left temporal, infratemporal and left masticator space (Figure 4).

Figure: 1 CECT showing soft-tissue density, filling and expanding the left maxillary widening the Maxillary sinus ostium and infundibulum
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Figure: 2 CECT showing soft-tissue densities in ethmoidal sinus

Figure: 3 CECT showing Left eye proptosis and soft-tissue density was seen in the medial wall of the orbit

Figure 4: CT scan showing Soft tissue density with an air fluid level in left temporal, infratemporal and left masticator space.

Patient was taken up for emergency decompression by incision and drainage, which revealed purulent serosanguinous collection in the above spaces. Orbital decompression was also accomplished via external route. Culture of the purulent
specimen revealed no growth. Patient was started on Amphotericin-B at 1 mg/kg/day with intravenous antibiotics. Though the patient showed improvement in ocular symptoms the patient eventually succumbed to the secondary acquired pneumonia.

Histopathology showed invasive zygomycosis with PAS/Gram stain positive for fungus confirming the diagnosis (Figure 5) and patient had CD4+ count of 580/cu mm.

Figure: 5 Histopathology showing invasive zygomycosis with PAS/Gram stain positive for fungus

Discussion

Fungal mycoses are common in immunocompromised hosts like diabetes, hematological malignancies like leukemia, lymphoma, multiple myeloma, septicemia, and hepatitis and chemotherapy patients. The other minor risk factors are malnutrition, increased serum iron, burns and intravenous drug abuse. Mucoraceae is ubiquitous and less virulent saprophytic fungae that spread by inhalation, ingestion and direct contact. The organism exists as molds in the environment and manifest as hyphae in the infected tissues. Spores, on inhalation get deposited in the nasal mucosa, then grows and invades the blood vessels (thrombosing vasculitis) leading to tissue necrosis. Neutrophils, being the major host defense mechanism, patients with neutropenia and diabetic ketoacidosis usually succumb to the disease. There are very few case reports in literature of
zygomycosis in HIV patients, thus raising the issues of the role of cell-mediated immunity in this infection. Because of the aerobic nature of fungi, the rhino cerebral form is the most frequent (55%), followed by pulmonary localization (30%)\textsuperscript{15}.

However, this element is rare in HIV patients, probably because AIDS patient primarily has a deficiency of T cell dependent immunity and only rarely have a bone marrow suppression leading to a severe neutropenia as observed in a patient with hematological malignancies (5). Transient episodes of neutropenia occurring within four months before presentation may be a risk factor for this disease (9). So, for the above reason our patient with HIV infection and normal CD4+ counts presenting with deep mycoses becomes a rare clinical presentation. Moreover, such a patient with co-existing picture of deep neck space abscess confounds the diagnosis of zygomycosis. The other opportunistic infections such as tuberculosis, aspergillosis and also neoplasia, like sarcoma were considered in the initial diagnosis.

The CECT scan had an important role in the diagnosis of the disease. Pterygopalatine fossa involvement usually suggests that the disease is extensively spread in the patient\textsuperscript{16}. Pterygopatine fossa was involved in the patient. This space acts as a conduit for the spread of infection to other spaces like orbit, infratemporal fossa, parapharyngeal space and retropharyngeal space. Parapharyngeal space involvement in this patient made the initial diagnosis mimicking the deep neck abscess. Infra-temporal space involvement has led to trismus in this patient. Patient also had orbital apex involvement and orbital abscess, which led to the rapid onset of proptosis and deterioration of vision. Fungal invasion in HIV positive patient occurs mainly at counts less than 150 cells/cu mm. However, this patient had CD4+ count of >500 cell/cu mm raised the issues in the role of cell-mediated immunity in this patient. Histopathology also showed invasive zygomycosis with PAS/Gram stain positive for fungus.

**Conclusion**

HIV and mucormycosis is a fatal and rare combination. Mucormycosis can occur in patient even with CD4+ counts >500 cu.mm. Deep neck space involvement may confound the picture. Because of the rapidity with which this disease progresses, prompt and aggressive therapy is essential.
References:


