Mucor Mycosis maxilla with palatal destruction An Interesting Case Report with Literature Review

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Abstract:

Mucor mycosis of maxilla is caused by filamentous fungi belonging to mucoracea family. This fungus is ubiquitous and can be found throughout the world on bread, air, soil etc. Rhinocerebral form of mucor mycosis is rather common. In this case report a detailed description of a patient with mucor mycosis involving maxilla with associated palatal destruction is given, along with a review of published literature on this topic. Rhinocerebral mucor mycosis is common in diabetics and immuno compromised individuals. The sheer inefficiency of phagocytic mechanism in diabetics help these organism to invade and proliferate. This condition is rather sinister because of the ability of mucor to invade other areas via peri vascular spread.

Introduction:

Mucor mycosis is the term used to describe infections caused by nonseptate fungi belonging to class zygomycetes and family mucoracea. The first histological description of this condition was by Paltauf in 1885. Mucor mycosis involving maxillary sinus is common in patients with uncontrolled / poorly controlled diabetes mellitus / in patients during immunocompromised states. Mucor mycosis involving maxillary sinus commonly present with orbital cellulitis. This condition is not quite frequent, and hence a great degree of awareness is necessary to suspect these lesions. This condition is rather common in patients with diabetes ketoacidosis. In patients with diabetic ketoacidosis infection is commonly caused by Rhizopus oryzae, because these organisms are known to produce enzyme ketoreductase which helps these organism to utilize the ketone bodies generated by the patient. Mucor is angio invasive in nature. That is the reason for the high mortality rates of
these patients. Angio invasion by these organisms cause thrombosis of blood vessels and tissue necrosis. This would mean that medications administered systemically will not reach adequate concentrations to neutralize the infection. Hence medical management alone is not sufficient in these patients. It should also be borne in mind that Mucorales are ferrophilic fungi. Acidosis present in these patients prevent binding of iron to transferrin making it available to the proliferating mucorales.

Case Report:

38 years old female patient reported with:

1. Foul smelling discharge right nasal cavity – 3 years
2. Swelling over right palate – 2 years
3. Loss of upper right premolars – 6 months (? devitalization)

On examination:

Nasal cavity: Whitish mass could be seen inside right nasal cavity. This mass was insensitive to touch and looked cheezy in appearance.

Oral cavity: Slough covered lesion seen on the right side of the palate. On probing there was no bone felt under the slough.

Intra oral view of the lesion
Imaging:

CT scan of nose and paranasal sinuses showed heterodense mass occupying right maxillary sinus with destruction of medial wall. The mass could be seen involving the right nasal cavity with destruction of palate.

![CT scan coronal cut of nose and sinuses](image)

Biopsy:

Biopsy from the lesion showed large number of aseptate ribbon like hyphae with right / obtuse angle branching in necrotic tissue. Consistent with mucor mycosis.

Management:

This patient was managed by strict glycemic control followed by endoscopic debridement of the lesion.
Discussion:

Mucor mycosis is the most suddenly occurring fatal infection involving nose and sinuses. This is commonly seen in diabetics and immune compromised individuals. It is commonly caused by Rhizopus, Rhinzomucor, and cunninghamella genera. These fungi have broad aseptate hyphae with uneven diameters with long sporangiophores. These fungi are saprophytic, ferrophilic and ubiquitous in nature. Two main types of mucormycosis occur in humans. They are superficial and visceral.

Superficial mucormycosis – Involves external ear, skin and nails commonly.

The visceral form could involve gastrointestinal, rhinocerebral, pulmonary or disseminated. Among the viseral types rhinocerebral is the most common.

Classification of rhinocerebral mucormycosis:

Type I : Rhino-orbito-cerebral. This form is highly fatal. Mucormycosis in this type involves nose, orbit and brain.

Type II: Rhino-maxillary form. This form is not fatal. The case discussed here belongs to type II category.

Features of mucormycosis:

1. Mucor infections have strong predilection to blood vessels, nerves and lymphatics. If artery is involved it causes avascular necrosis of that area.

2. Mucor thrives well in acidic glucose rich medium. This is commonly seen in diabetics. Hyperglycemia also impairs cell mediated immunity thereby facilitating spread of mucormycosis.

3. Increased availability of micronutrients like iron facilitates proliferation of fungi.
Pathophysiology of mucormycosis maxilla

1. Mucor spores
   - Ingestion/Inhalation
     - Nasal/oral Mucosa
       - Diminished phagocytic Response
         - Gemination of spores
           - Invasion of Blood vessels
             - Stasis Thrombosis
               - Ischemic necrosis
       - microangiopathy
         - Cerebral Ischemia
           - Impaired healing
             - Cavernous Sinus spread
               - Spread to Orbital apex
                 - Spread to Ethmoid artery
                   - Osteomyelitis maxilla
Early diagnosis of rhinocerebral mucor mycosis really paves the way for appropriate and timely treatment. Central nervous system involvement is nearly fatal.

Locoregional spread of mucor mycosis has not been described extensively in the literature. On the contrary angioinvasion finds extensive mention. Hosseini et al regard pterygopalatine fossa as a reservoir of the disease from where it can spread to adjacent areas, like orbit and infratemporal fossa.

References: