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Case Report

Indolent Mucormycosis of Nasopharynx – A Case Report

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Keywords

- Mucormycosis
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- Indolent course Abbreviations
- MCA: Middle cerebral artery
- DKA- Diabetic ketoacidosis
- HPE- Histopathology
- PCR- Polymerase Chain Reaction

Abstract

We present a rare case of indolent mucormycosis of nasopharynx. Mucormycosis of nasopharynx usually presents as a fulminant disease with rapid progression to death if not treated early in the course of the disease (1, 2). The indolent course of the disease was what made our case different. The diagnosis was made on histopathological examination of the biopsy from nasopharynx. This case emphasizes the fact that indolent mucormycosis needs to be treated even if the patient is clinically asymptomatic.

INTRODUCTION

Mucormycosis is a rare, but highly fatal fungal infection. It belongs to the class *zygomycetes.* Rhinocerebral mucormycosis is the most common variety, presenting with usually underlying Diabetes Mellitus or Diabetic Ketoacidosis (DKA). Initial symptoms include acute sinusitis, fever, congestion, purulent nasal discharge and unilateral headache. The infection spreads contiguously through palate, orbit, and brain with periorbital edema, proptosis, blindness, facial numbness and cranial nerve palsies. Cavernous sinus thrombosis and invasion of carotid artery are seen .Usually the infection has a very rapid course. Mucor is found worldwide in soil and decaying organic matter. Modes of transmission are inhalation, ingestion and cutaneous exposure. Incidence is approximately 1.7 cases/million. The number of cases is increasing with increase in the number of cases of immunosuppression. The spores become hyphae and are fatal as they spread through paranasal sinuses and invade the brain and the orbit. Direct soft tissue invasion and formation of cerebral abscesses are frequent. Extension along blood vessel is followed by invasion and thrombosis [1-4]. In addition to classical fulminant form of mucormycosis, a chronic form also exits.

CASE PRESENTATION

A 65year old male presented with stroke with right sided ophthalmoplegia and left sided hemiplegia due to MCA territory infarct. He was a Known Case of Diabetes Mellitus on irregular treatment. Along the course in the hospital, patient developed a crust on the palate. The crust was biopsied. Biopsy was reported as Mucormycosis. Clinically, patient did not have signs and symptoms of mucormycosis. Later, the patient developed yellowish black escher filling up his nasopharynx. This was biopsied and sent for HPE and culture. The HPE was reported as Mucormycosis without invasion. Culture, however showed no growth.

Patient still did not exhibit any symptoms of Classical mucormycosis. Due to this clinical picture, the diagnosis of indolent mucormycosis was considered. Radical Debridement of nasopharynx was done. Patient was started on I V Amphotericin B. The patient improved clinically, the crusted area on the palate healed well. There was complete mucosalisation of the palate. Nasal douching of the nasopharynx was done post operatively. The nasopharynx healed well.

On follow up, after 1.5 years, the nasopharynx healed completely, the exenterated sinuses are well mucosalised and patient is doing well.

DISCUSSION

Mucormycosis is a rare, but highly fatal fungal infection. It belongs to the class *zygomycetes.* Zycomycosis is an umbrella term used for diseases caused by many non septate filamentous fungal species classified under order Mucorales and Entomophthorales.

HISTORY

In 1855, Kurchenmeister described a case of zygomycosis in a patient of neoplastic lung on the basis of its histopathology which was probably first authentic human case. Furbringer in 1876 first described pulmonary mucormycosis caused by Absidia. At that time, most of pathogenic zygomycetes were originally classified as members of genus Mucor. These organisms were later assigned different genera and families within order Mucorales. Platauf coined term "Mycosis Mucorina" and first described a well documented case of systemic infection with gastric and rhinocerebral involvement in 1885. In 1943, Gregory and collegues, in a series of three fatal cases with diabetic ketoacidosis, reported typical findings of advanced Rhinocerebral zygomycosis, ptosis and ophthalmoplegia [4].

Mucorales

The fungal species belonging to order Mucorales are ubiquitously found prevalent in the environment. They are found in food items, soil, air and frequently encountered as laboratory contaminants. The virulence of these fungi is clearly of low order and sporadic infections occur throughout world particularly in severely debilitated patients. Being an opportunistic infection, zygomycosis is produced by contaminant fungi in a host whose immunological defence mechanisms are weakened by endogenous causes like malignancy, leukemia, diabetes mellitus or exogenous causes like immunosuppressive therapy [4].

Entomophthorales

The fungal species belonging to order Entomophthorales are essentially arthropod parasites. They cause uncommon clinical entity called as entomophthoromycosis, which is chronic granulomatous type of subcutaneous, mucocutaneous and visceral infections.

Epidemiology

Mucor is found worldwide in soil and decaying organic matter. Modes of transmission are inhalation, ingestion, cutaneous exposure. Incidence is approximately 1.7 cases/million. The number of cases is increasing with more immunosuppression. The risk factors for mucormycosis are as follows-

Diabetes

DKA

Deferoxamine therapy

Iron overload

Immunosuppression: Neutropenia, steroids, bone marrow transplant

Trauma/burns

IV drug use, malnourishment

Pathogenesis

Once the spores of mucormycosis are inhaled, in healthy people, the nasal cilia transport it to pharynx and it gets cleared through GI tract. In susceptible people, the infection usually begins in nasal turbinates and spreads from there.

Role of Iron: Fungal hyphae produce a substance called rhizoferrin, which binds iron avidly. The iron-rhizoferrin complex is then taken back into the fungus, and the iron becomes available for vital intracellular processes. The body is protected from fungal infection partly by limiting availability of iron in normal patients. But the serum from patients in DKA stimulates growth of fungus. Also, the Deferoxamine-iron chelate is a siderophore for Rhizopus (binds iron and transports it into the cell) which causes increased iron uptake by fungus and stimulates its growth. The decreased neutrophil count and abnormal phagocytic function is also induced by DKA

Clinical Features: Mucormycosis usually presents in one of the following forms-

Rhinocerebral Infection:

PULMONARY

Cutaneous

Rhinocerebral Mucormycosis is the most common form, presenting with usually underlying DM or DKA. Initial symptoms include acute sinusitis, fever, congestion, purulent nasal discharge and unilateral headache. The infection spreads contiguously through palate, orbit, and brain with periorbital edema, proptosis, blindness, facial numbness and cranial nerve palsies. Cavernous sinus thrombosis and invasion (Figure 1). of carotid artery are seen. Usually the infection has a very rapid course.

Pulmonary mucormycosis is caused through the inhalation of sporangiospores. The patients are severely immunocompromised by virtue of an absolute lack of circulating neutrophils, secondary to hematologic malignancy like leukemia, lymphoma, profound immunosuppression or bone marrow transplantation. The lesions may be focal or diffuse. The invasion of blood vessels may result in destruction of lung parenchyma. The usual course of disease takes about 1 to 4 weeks from onset to terminal stages of illness. The clinical manifestations are non- specific and may include chest pain, dyspnoea and hemoptypsis [4].

The cutaneous type of zygomycosis can be either primary infection or secondary to the disseminated form. It is relatively benign form of disease in comparison to the other clinical types. Necrotizing fasciitis may be secondary to cutaneous or subcutaneous zygomycosis and has a very high mortality rate.

The presentation of mucormycosis in most of the cases is acute, with rapid progression of symptoms and death of the patient if not treated. But, the indolent or chronic presentation of nasopharyngeal mucormycosis is extremely rare. Early diagnosis and prompt initiation of treatment can help in salvaging the patient from further progression.

Our patient presented with very subtle symptoms with a chronic course. The diagnosis was picked up by histopathological examination. Patient was then treated with Amphotericin B and he recovered well.

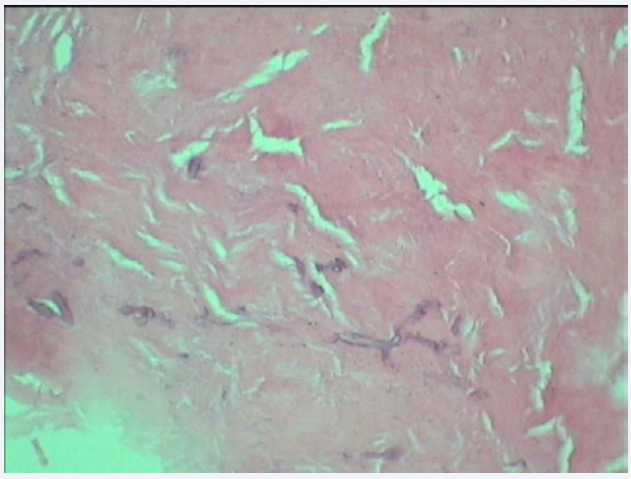

Figure 1 Photomicrograph of aseptate broad hyphae of mucor on histopathology.

Diagnosis

The diagnosis rests on high index of suspicion as a delay of even twelve hours in diagnosis may be fatal.

Lab diagnosis: The diagnosis is by biopsy suspicious areas with histopathological examination and Culture which is considered to be the gold standard. Nested PCR is also available for earlier diagnosis.

On H and E stained slides, mucor is seen as broad, nonseptate hyphae with right-angle branching.

Treatment: Early aggressive surgical intervention and Amphotericin B 1.5mg/kg/day for at least 6-8 weeks, probably longer will be needed. Voriconazole and caspofungin are not effective.

Other co morbidities need to be managed such as correcting acidosis, stopping the immunosuppressant and steroids.

Take home message: Even though the entity of indolent mucormycosis is rare, one has to think about this in case of non healing ulcers of nasopharynx, especially with underlying immunosuppressive conditions.

Early debridement and Amphotericin B therapy can limit the progression of the disease.

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