Case Report

Rhino-Sino-Orbito-Cerebral Mucormycosis, a Case Report and Literature Review

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ABSTRACT

Mucormycosis is rare, life threatening infection, especially in diabetic with ketoacidosis. The infection is caused by a group of saprophytic fungi of class phycomicetes rhizomucor, rhizopus and mucor are more common fungi. The diagnosis is based on high clinical suspicion, CT / MRI imaging and histopathological confirmation. We present a 50 years, diabetic female, with history of mainly left sided facial pain, swelling is recent decreased left eye movement as well. High suspicion of Mucormycosis on CT which confirmed on histopathology later on.

Patient responded well clinically with amphotericine B posaconazole & endoscopic debridement. Patient was discharged 10 days later with followed up MRI and antifungal treatment for 6 month.

Key Words: Mucormycosis, Diabetes, Paranasal sinuses, Orbits Brain, Immunosuppression, Computerized Tomography (CT), Magnetic Resonance Imaging (MRI).

INTRODUCTION

Mucormycosis, also called Zygomycosis or phycomyocosis, was described by Paull Tauf in 1885 [(1)]. Paranasal mycosis usually Mucor or Rhizopus [(2)], manifests as two distinct enteties, a benign or non invasive infection and a relatively serious, invasive infection which usually occurs in immunocompromised individuals[(3)]. Invasive form causes vascular thrombosis and tissue necrosis with signs and symptoms due to involvement of nose, PNS, orbits or central nervous system. The risk factors include ketoacidosis and hematologic malignancies. The prognosis is poor, especially in presence of severe sequalae resulting in high mortality even in patients with prompt diagnosis and correct treatment [(4)].

CASE REPORT

We present 50yrs old house wife, came in medical OPD in King Abdul Aziz Hospital Makkah (Saudi Arabia) for nasal swelling, pain and swelling of left maxilla with recent onset of restricted left eye movement & ptosis. Patient was known diabetic, but not ketoacidotic with poor oral antidiabetic control, on external examination showed left sided nasal / cheek skin involvement and ptosis of left eyelid with provisional diagnosis of fascitis, mainly left sided (Fig1). Laboratory findings revealed increased neutrophils (14500 / mm^3), blood glucose level 360g/100ml. normal electrolytes and renal parameters. Liver enzymes and coagulation profiles were in normal limits. Left eye examination revealed minimal painful restriction of movement and redness which aggravated though blood sugar was controlled and antibiotics were given. After two days, suppuration from her left nostril appeared, with some black material in it. So suspicion of Mucormycosis wad made. CT PNS and brain was recommended along with histopathology after incisional biopsy of left nasal cavity mass. Amphoterisin B therapy was started. CT study showed opacification of sinuses, more on left side with medial wall of orbit destruction and intracranial extension in left basifrontal region (Fig 2 a & b). Histopathology also revealed aspetate hyphae with right angle branching, which are typical for mucormycosis (Fig 03). She recovered completely after endoscopic sinus / nasal debridement and antifungal treatment. Patient remained well after two month follow up with no residual left eye damage, this all was due to early diagnosis of mucormycosis with diagnostic support of imaging studies.

DISCUSSION

Mucormycosis is commonly caused by Rhizopus, absidia, Mucor, Rhizomucor species, however different fungi (Cunningamella, Saksenala, Syncephala strum, Cokeromy, Mortiella) can also by found. Few cases reported in medical literature are caused by apophytomyces, usually in healthy individuals with traumatic inoculation [05]. Fungi usually acts as apportunistic infection, may be present in solid, decaying organic debris and bread moulds. The predisposing factors are diabetes mellitus and hematologic malagnancies with neutropenia. The risk factors are chronic steroid intake, organ transplant,

chronic renal insufficiencies, chemotherapy, ferrum intoxicity and tooth extraction.

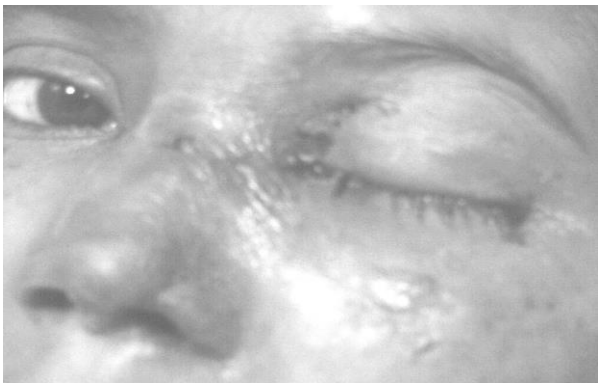

Figure No.1: Nasal / left maxillary swelling with color changes and left eye ptosis

A

B

Figure No.2: A, B CT Coronal images showing involvement of basifrontal, both ethmoids, left maxillary and left orbital regions.

Different forms seen by intalation / inoculation of spores, these include, rhino, orbital, respiratory, gastrointestinal, cutaenous and mixed. Only in advanced disease, symptoms become more specific otherwise in initial stages, may be difficult to detect. Necrosis in present in 20-40% of cases and is considered bad prognostic sign. CNS is affected through orbital apex while orbit is involved by nasoalacrimal duct, medial wall dehiscences or the anterior and posterior ethmoidal orifices. The fungus adheres to blood vessel wall, leading to ischemia and necrosis. Perineural spread through the trigeminal branches has also been described.

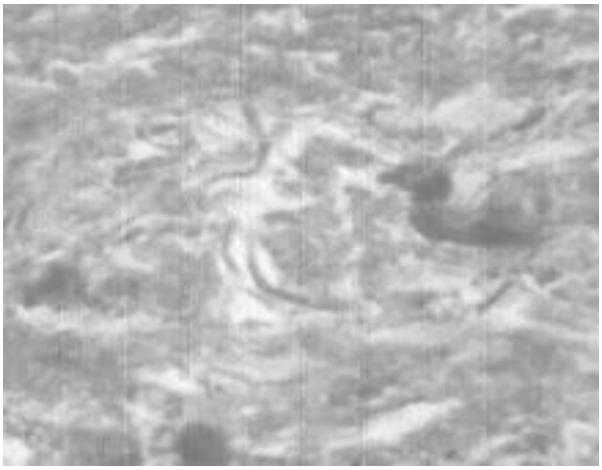

Figure No.3: Aseptat hyphae, branching at right angle, typical form Mucormycosis.

The diagnosis is usually made by clinical suspicion, imaging findings and histopathology. Clinically, fever is the most common early symptom (44% cases) [6], followed by nasal ulceration or necrosis, periorbital or facial swelling and decreased vision (33%cases), 80% may develop necrotic lesions on nasal or oral mucosal later on. Imaging findings may be non specific initially however later on polypridal mucosal thickening and hyperdense foci in affected sinuses / nose are highly suggestive of fungal disease on CT[3]. MRI gives a better visualization of brain, orbits, perineural invasion and vascular obstruction. It is usually preferred choice because of use of nephrotoxic drugs and presence of compromised renal function.

CONCLUSION

In conclusion the early diagnosis is imperative in management and reduction in bad sequalae in invasive Rhino, Orbito, sinocerebral mucormycosis as in this case, The treatment should be focused on proper antifungal (amphotericine B and Posaconazole) and surgical debridement. Follow up is by clinical examination and MRI. The prognosis has improved dramatically, with a mortality rate of 85% in 1960 to 30-35% in 2010 [1].

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