Case Report

Rhino-Orbito-Cerebral Mucormycosis

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ABSTRACT

Mucormycosis represents a group of life-threatening infections caused by fungi of the order Mucorales of the subphylum Mucoromycotina (formerly known as the class Zygomycetes). We report a 28-year-old women who presented with fever and facial pain. She was first time detected as diabetic and was in ketoacidosis. She had right opthalmoplegia with Left sided hemiplegia. Histopathologic examination from maxillary sinus was suggestive of mucormycosis. CT angio showed complete occlusion of Right internal carotid artery and Right cavernous sinus thrombosis. She responded to Amphotericin B Deoxycholate.

Key words : Mucormycosis, Diabetes, Cavernous sinus thrombosis, Amphotericin B

Introduction :

Mucormycosis is an opportunistic and frequently fulminating fungal infection caused by members of the family Mucoraceae, order Mucorales and class Zygomycetes. It is commonly reported in immunocompromised patients such as poorly controlled diabetes mellitus, blood dyscrasias, malnutrition, neutropenia, iron overload, organ transplant, and immunosuppressive therapy. Diagnosis is confirmed by histopathological demonstration of the organism in the affected tissue. Early diagnosis and treatment of mucormycosis is extremely important due to the aggressive course of the disease. The disease may manifest in six different ways as rhinocerebral, pulmonary, cutaneous, gastrointestinal, central nervous system or disseminated forms. Rhinocerebral mucormycosis is the most common type accounting for 30% to 50% of the cases and its extension to the orbit and brain is quite usual making it potentially life-threatening disease.

Case Report :

28 year old woman presented with complaints of fever and pain on face since 5 days and altered sensorium since 1 day. She had no history of headache, convulsions, bleeding tendencies, cough, burning micturition, loose motions, vomiting. She was not a known case of diabetes, hypertension. On examination she was drowsy with a pulse rate of 110/min regular, febrile, no neck stiffness, no external bleeding spots. She had ptosis of right eye with external ophthalmoplegia and absent direct and consensual light reflex. Redness over right cheek region and oral cavity examination showed white ulcerative necrotic oval patch on hard palate ***(Fig. 1 and 2).*** Systemic examination was within normal limit. On investigations, TLC was 13,000/cmm. KFT, LFT and electrolytes were normal. RBS was high and ketones were large by dipstick method. She was started on insulin, I/V fluids and antibiotics. She developed left hemiplegia by 3rd day of IPD stay. Keeping high suspicion of mucormycosis, nasal endoscopy was done. It showed black necrotic inferior and middle turbinates. Debridement and resection was done and sent for histopathological examination ***(Fig. 4).*** CT angio of brain showed complete occlusion of right internal carotid artery by an extensive thrombus and right cavernous sinus thrombosis with infarction of right gangliocapsular region. ENT, Neuro surgery, Oromaxilofacial surgery opinion was taken - and was advised to manage conservatively. Although liposomal Amphotericine B is the drug of choice, due to financial constraints she was started on deoxycholate salt of amphotericin B - IV 1mg/kg/day dose. As her KFT started to derange we shifted her on half of previous dose. In view of cavernous sinus thrombosis, she was also given LMWH & overlapped with warfarin 5 mg OD.

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Fig. 1 : **shows palatal ulcer with erosion**

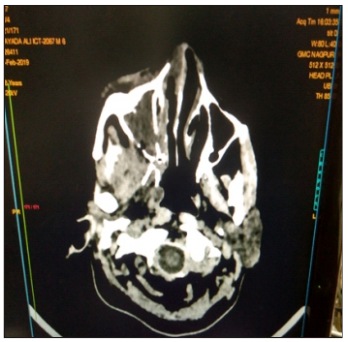

Fig. 3 : **shows soft tissue density in right maxillary sinus.**

Fig. 2 : **shows complete ptosis of right eye**

Fig. 4 : **nasal endoscopy shows black necrotic middle turbinate.**

HPE report came after 7 days as sinonasal mucormycosis. Therapy was given for total 6 weeks with monitoring of vitals and laboratory parameters. She started improving and by time of discharge she was able to stand with support. She was discharged on oral posaconazole and adviced to follow up.

Discussion :

Patients with diabetic ketoacidosis (DKA) are at high risk of developing rhinocerebral mucormycosis. The acidosis causes dissociation of iron from sequestering proteins, resulting in enhanced fungal survival and virulence. High index of suspicion of mucormycosis must be maintained, even in the absence of a known history of diabetes. If hyperglycemia is present, due to upregulation of receptor (GRP78) that binds to Mucorales, it enables tissue penetration. Patients with itraconazole or voriconazole prophylaxis are at increased risk and typically present with disseminated mucormycosis. Breakthrough mucormycosis also has been described in posaconazole or echinocandin prophylaxis.

Rhino-orbital-cerebral mucormycosis : is the most common form of the disease. Symptoms are nonspecific and include eye or facial pain and facial numbness followed by the onset of conjunctival suffusion and blurry vision. Fever may be absent in up to half of cases. White blood cell counts are typically elevated. If untreated, infection usually spreads from the ethmoid sinus to the orbit, resulting in compromise of extraocular muscle function and proptosis, typically with chemosis. From the orbit, spread often takes place via hematogenous or contiguous dissemination to the frontal lobe of the brain and/or via venous drainage to the cavernous sinus. Painful necrotic ulcerations of the hard palate, but this is a late finding that suggests extensive, well-established infection. Definitive diagnosis requires a positive culture from a sterile site or histopathologic evidence of invasive

mucormycosis. Biopsy reveals characteristic wide ($\geq$ 6 - to 30 - m), thick-walled, ribbon-like, aseptate hyphal elements that branch at right angles. Treatment has three steps : (1) early initiation of therapy; (2) rapid reversal of underlying predisposing risk factors, if possible; and (3) surgical debridement. Liposomal amphotericin B is drug of choice. Combination lipid polyene echinocandin therapy was associated with significantly better outcomes than polyene monotherapy in a retrospective clinical study involving patients with rhino-orbital-cerebral mucoromycosis. Isavuconazole, posaconazole, is best reserved for oral step-down therapy in patients whose condition has substantially improved on polyene-based therapy or for salvage therapy in patients who are intolerant of polyene-based regimens or whose infection is refractory to these regimen.

Conclusion :

Clinical suspicion should be maintained for such dangerous infection when patient presents in diabetic ketoacidosis. Due to lethal nature of disease, early initiation of therapy helps in rapid reversal of disease process.

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