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A Case Report of Rhino-Orbital-Cerebral Mucormycosis

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ABSTRACT
Rhino-orbital-cerebral mucormycosis is a rare but life threatening infection that generally occurs in patients with diabetes mellitus and other immune deficiency conditions. Rhino-orbital and Rhino-cerebral are two form of the disease and the mixed presentation in the form of rhino-orbital-cerebra mucormycosis is very rare. As such the condition is a medical emergency. Early recognition and treatment are essential because it may lead to death in few days.
Keywords: *Mucormycosis, multiple cranial nerve palsies, cerebral abscess, diabetes mellitus.*

INTRODUCTION

Mucormycosis is a rare but aggressive opportunistic fungal infection that is commonly caused by members of the family Mucoraceae that include Rhizopus, Rhizomucor, Mucor and Absidia. Mucormycosis can cause severe, sometimes fatal disease in susceptible individuals with uncontrolled diabetic ketoacidosis, neutropenia, chronic glucocorticoid use, hematological malignancy, and chronic malnutrition and burn patients [1].

CASE

A 40 years old newly diagnosed diabetic male patient, came with complaints of vomiting since 10 days, deviation of angle of mouth to left side since5 days, gradual loss of vision in right eye and doping of right eyelid since 5 day. Detailed history revealed that patient was apparently alright 10 days back when he developed vomiting about 5-6 episodes in a day .It was Non projectile, Non foul smelling, Contained water and food particles. There was no associated hematemesis, malena, pain abdomen, abdominal distension and loose motions. Patient also developed deviation of angle of mouth to right side sudden in onset, with drooping of right eyelid sudden in onset & progressive. There was no history of fever, chills, rigors, headache, convulsion, weakness in limbs, altered sensorium, head injury, bowel bladder. There was no significant past and family history. On general physical examination patient was conscious, oriented, afebrile. Pulse rate was 92/min and Blood Pressure was 120/70 mmHg. There was no pallor, cyanosis, clubbing, lymphadenopathy or edema. Systemic examination

revealed, normal higher mental function tests. Cranial nerve examination revealed multiple cranial nerve palsies. Olfactory nerve examination was normal. There was normal visual acuity, visual field, and colour vision in left eye. In right eye there was complete loss of perception of light, hence visual field and colour vision could not be tested. Occular movements in the form of adduction, abduction, elevation, depression, intorsion and extorsion were preserved in left eye, but completely lost in right eye. The pupil on left side was normal sized, with normal direct light reflex and loss of indirect light reflex. But the pupil on right side was dilated and negative for both direct and indirect light reflex. The right eye also had ptosis. Corneal and conjunctival reflex as lost on right side, with reduced facial sensation on right side suggesting involvement of trigeminal nerve. Patient even had lower motor neuron type facial nerve palsy. Other cranial nerves examination did not revealed any abnormality. The motor system, sensory system, reflex, coordination, and gait were normal. There were no cerebellar signs. Other system examination revealed no abnormalities

Investigations were done and Hemoglobin was 12.2 gm/dl, White blood count was 9700/ul , Platelet count was 2,62,000/ul and Mean Corpuscular Volume was 90.6 fl. Random blood sugar was 500 mg/dl. Fasting blood sugar was 254 mg/dl and post prandial blood sugar was 386 mg/dl. Renal function and liver function test was normal. Urinary examination reveals sugar 2+, and large urinary ketones. Electrocardiogram, ultrasound abdomen-pelvis and chest x-ray was normal. Computed tomography of Para nasal sinuses were done which showed mucosal thickening in right frontal ,maxillary, b/l ethmoid & sphenoid sinuses. Right osteomeatal complex is blocked. Thickening is seen extending into extra conal space of right orbit on medial & inferior aspects s/o sinusitis with orbit extension. Magnetic resonance imaging, plane with contrast of brain with orbit was done which showed acute sinusitis involving right maxillary, ethmoid & sphenoid sinus with right orbital extension causing orbital cellulitis, subperiostel abscess & right optic neuritis, probably of fungal etiology. Otorhinolaryngoloist opinion was taken, sinus debridement was done. The debrided content was sent to culture and was positive for mucormycosis. Ophthalmologist opinion was taken and with patients consent enucleation of right was done. patient was treated with injectable antifungal voriconazole. He was even treated with insulin, and was out of ketosis. Patient general condition improved and was discharged after 1 month. Repeat sinus debridement was done and it was culture negative.

Figure 1: showing right eye ptosis

Figure 2: showing facial nerve palsy-deviation of angle of mouth to left, blunting of right nasolabial fold

Figure 3: showing dilated left pupil

Figure 4: patient at the time of discharge

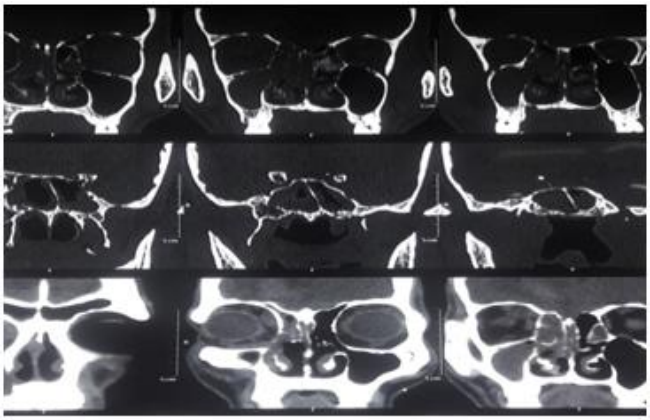

Figure 5: CT PNS showing sinus involvement

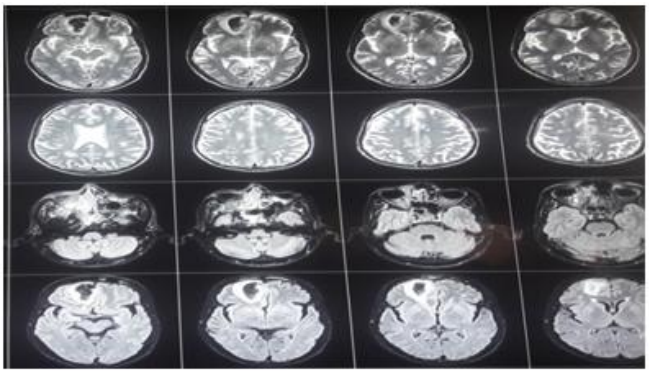

Figure 6: MRI BRAIN WITH ORBIT

DISCUSSION

The most common underlying illnesses of rhino-orbital-cerebral mucormycosis are diabetes mellitus, hematological malignancies, hematopoietic stem cell transplantation, and solid organ transplantation. Sporangiospores are deposited in the nasal turbinates and Para nasal sinuses in immunocompromised patients. [2] Qualitative and quantitative abnormalities of neutrophils, monocytes and macrophages increase the risk for development of mucormycosis. Altered iron metabolism also is a critical factor in the pathogenesis of patients with diabetes mellitus who are at risk for rhino-orbital-cerebral mucormycosis. Angioinvasion with thrombosis and tissue necrosis is a key pathophysiological feature of human Mucorales infection. The ethmoid sinus is a critical site from which sinus mucormycosis may extend through the lamina papyracea into the orbit, extraocular muscles, and optic nerve. The brain may be seeded by invasion of the ethmoidal and orbital veins, which drain into the cavernous sinuses. Diplopia and ophthalmoplegia may be the earliest manifestations of cavernous sinus syndrome before changes are apparent on diagnostic imaging modalities. Negative diagnostic imaging does not exclude cavernous sinus mucormycosis. Mucormycosis of the maxillary sinus has a constellation of clinical features that are different from that of ethmoid sinus mucormycosis [2]. A painful black necrotic ulceration may develop on the hard palate, indicating extension from the maxillary sinus into the oral cavity. Orbital apex syndrome is an ominous complication of mucormycosis of the orbit. Once within the orbital compartment, organisms may extend posteriorly to the optic foramen, where the ophthalmic artery, ophthalmic nerve and optic nerve are threatened by invasion, edema, inflammation and necrosis. Early diagnosis of sinus mucormycosis is critical for prevention of extension to orbital and cerebral tissues. Optimal therapy requires a multidisciplinary approach that relies on prompt institution of appropriate antifungal therapy with amphotericin B, reversal of underlying predisposing conditions, and, where possible, surgical debridement of devitalized tissue. Outcomes are highly dependent upon the degree of immunosuppression, site and extent of infection, timeliness of therapy, and type of treatment provided [2]. Bhansali et al analyzed some of the most frequent ophthalmologic signs and symptoms associated with mucormycosis. In their retrospective review of 35 patients, ophthalmoplegia (89%), proptosis (83%), loss of vision (80%), chemosis (74%), periorbital

swelling (66%), and periorbital pain (43%) were the most frequently cited complaint When looking at nonophthalmic signs and symptoms, sinusitis, nasal discharge/ulceration, facial swelling, cranial nerve VII palsy, and palatable necrosis were the most commonly reported [3]. The therapy of rhinocerebral mucormycosis includes aggressive surgical debridement, administration of high-dose amphotericin B, and control of underlying predisposing conditions, especially diabetes and immunosuppression or immunodeficiency. Hyperbaric oxygen suppresses fungal growth in vitro and has theoretical value in treating mucormycosis because it reduces the tissue hypoxia and acidosis that accompany vascular invasion by the fungus. [4] Patients with poorly controlled diabetes and ketoacidosis are at high risk of developing rhinocerebral mucormycosis, with systemic acidosis creating an ideal environment for the growth of *Rhizopus*. However initial presentation of rhinocerebral mucormycosis infection can often appear non-specific making correct diagnosis extremely difficult until the disease has caused significant morbidity, aggressive fungal invasion of the paranasal sinuses, orbit, hard palate and brain [5]. The basis of mucormycosis treatment remains a combination of extensive surgical debridement and amphotericin B for a protracted period of 4–6 weeks [6]. Although not currently used as first line treatment, the concurrent use of posaconazole, a triazole antifungal drug has been shown to be effective against mucormycosis and use has been increasingly reported when amphotericin B has had to be discontinued due to adverse side effects [7]

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