RHINO – ORBITAL MUCORMYCOSIS

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INTRODUCTION

Mucormycosis is caused by fungi from the genera Mucor, Absidia, and Rhizopus. Rhino orbital mucormycosis (ROM) is a rare, rapidly progressive opportunistic infection [1]. The organism is found in air, soil, vegetable matter, skin, body orifices and bread mold. People with predisposing systemic diseases like Diabetes mellitus with ketoacidosis and immune deficiency disorders are more susceptible to the infection with this organism. The disease was called mucormycosis by Paltauf who described the first case in 1885.

We report a case of rhino – orbital mucormycosis in a diabetic patient, who presented with proptosis, ophthalmoplegia and orbital cellulitis.

CASE REPORT

A 64 Years old female, house wife from Dinga (District Kharian) presented on 4th October, 2006 with complaint of painless drooping of left upper eye lid for the last 15 days. It was followed by pain in left upper molar tooth and painless loss of vision in left eye for last 10 days. Patient was known diabetic for the last 12 years and was on oral hypoglycaemic drugs. Vision in left eye was no perception of light and in right eye, 6/12 improving to 6/9 with glasses. There was left complete ptosis, puffiness of the upper eyelid and facial nerve paresis. On elevating the lid, there was 4 mm proptosis, inferotemporal dystopia and exotropia of 30 prism diopters. There was conjunctival congestion and total ophthalmoplegia in left eye (Fig.1). Rest of anterior segment examination in both eyes was unremarkable. Fundoscopy showed preproliferative diabetic retinopathy in both eyes and disc congestion in left eye. IOP was 14 mm of Hg in right eye and 16 mm of Hg in left eye. Blood complete picture, urine routine examination and chest radiograph - PA view were unremarkable. Blood sugar fasting was 10 mmol/L and random was 16 mmol/L. X ray paranasal sinuses followed by CT Scan showed hazy left maxillary sinus. Tissue biopsy was taken from left maxillary sinus and sent for histopathology which confirmed the diagnosis of mucormycosis. Blood glucose was controlled under supervision of medical specialist. Subtotal exentration of left orbit (sparing the eyelids) along with removal of left maxillary sinus was performed (Fig. 2). Inj. Amphotericin B, 12 mg 6 hourly IV was started. Patient recovered postoperatively but unfortunately expired on 7th postoperative day after remaining in coma for 24 hours.

DISCUSSION

Mucormycosis classically involves the nasal mucosa with invasion of the sinuses, orbit, and brain. The infection can involve the lungs, central nervous system [2], gastrointestinal tract, and skin, but it is probably best known for its rhinocerebral presentation. Most cases of mucormycosis are acute surgical emergencies; however, several cases of subacute or chronic, indolent form have been reported with signs and symptoms developing over 4 weeks. Conditions most commonly associated with mucormycosis include uncontrolled diabetes mellitus, chronic steroid use, metabolic acidosis, organ transplantation, leukemia/lymphoma, treatment with desferoxamine, and AIDS.

The spores of these fungi are ubiquitous and gain entrance to the human body through the mouth and nose. Individuals who are immunocompetent do not develop the disease. In individuals who are immunocompromised, germination of the

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Received: 18 Dec 2007; Accepted 30 April 2008

spores and hyphae formation occurs. The spores attach to the nasal or oral mucosa where massive spore formation occurs, and then the fungus directly invades the blood vessels causing necrotizing vasculitis with thrombosis of the vascular lumina and

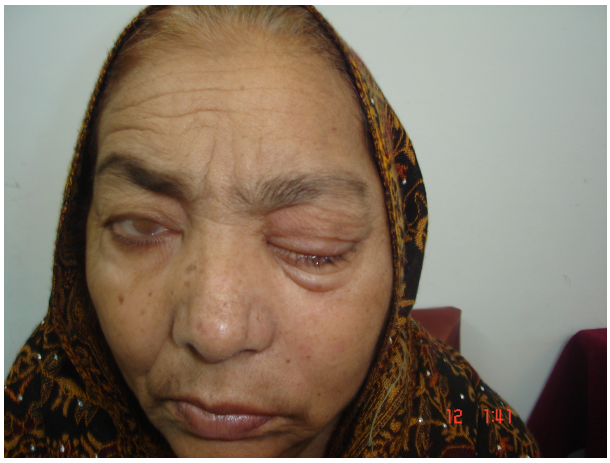

Fig. 1: Left complete ptosis, puffiness of upper eye lid and facial nerve paresis

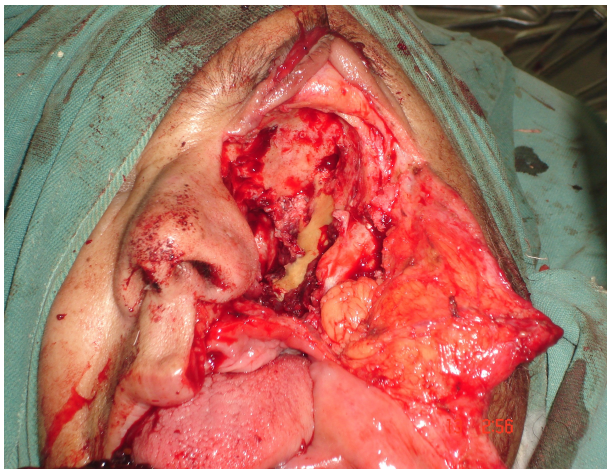

Fig.2: Subtotal exentration of left orbit (sparing the eyelids) along with removal of left maxillary sinus

resultant infarction. Extension to ethmoid sinuses can lead to orbital involvement. Intracranial spread can occur through the ophthalmic artery, superior orbital fissure, or cribriform plate.

The patient usually presents with headache, nausea, fever and lethargy. The nasal symptoms may include purulent discharge, stuffiness, rhinorrhea, epistaxis and nasal hypoaesthesia. Ophthalmic manifestations may be in the form of unilateral orbital apex syndrome [3], including severe pain, visual loss, total ophthalmoplegia, corneal anaesthesia, and multiple cranial nerve palsies. Orbital cellulitis presenting with early visual loss is one of the hallmarks of mucormycosis. Orbital cellulitis per se is not a requisite, but thrombosis of orbital veins, demonstrable by venography, may account for congestive signs and symptoms. Some degree of proptosis and lid swelling is invariably present.

Mucormycosis should be differentiated from inflammatory pseudotumor, contiguous sinusitis, metastatic tumor, lymphoma, nonspecific granulomatous inflammation (Tolosa-Hunt syndrome), nasopharyngeal carcinoma, cavernous sinus thrombosis, diabetic ophthalmoplegia and migrainous ophthalmoplegia.

Of great clinical importance is the early recognition of an acute orbital inflammatory syndrome in the diabetic patient [4], which should immediately suggest an opportunistic fungal infection such as mucormycosis (phycomycosis). Contrary to popular opinion, uncontrolled acidosis need not be present and in fact, orbitocerebral phycomycosis can occur in otherwise healthy patients especially if there is history of injury compromising the cutaneous barrier [5]. Classically, a progressive and often fatal picture of cavernous sinus thrombosis evolves rapidly.

The standard medical therapy for ROM is amphotericin B in a starting dose of 0.25 mg/kg/day and maximum of 2-4 gram for a period of several weeks to several months, depending on the clinical response, tolerance of the patient and laboratory monitoring especially nephrotoxicity. In our case the patient was obese and critically ill so we started with 0.5 mg/kg/day. Drug toxicities can limit the use of amphotericin in some patients and Posaconazole has been

recommended as an alternative [6]. Surgery should be instituted without delay once the condition is diagnosed. Surgical procedures range from debridement of the necrotic mucosa, ethmoidectomy, sphenoidotomy and radical maxillectomy with orbital exenteration. Both endoscopic and open approaches have been described, in both single and multiple stages.

CONCLUSION

To conclude, premorbid diagnosis is dependent on a high index of suspicion, immediate sinus mucosal biopsy followed by rapid correction of the underlying metabolic derangements, intravenous amphotericin B and surgical clearance of all infected tissue. Survival depends on the combined effort of the ophthalmologist, otorhinolaryngologist, mycologist, and internist.

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