Mucormycosis - Genuinely Sight Threatening and Life Threatening

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Introduction

Mucormycosis is a well described but often misdiagnosed and eventually mismanaged complex disease which appears unseemingly benign but is fatally malignant in its behaviour. The nidus of infection is always hidden in the sinus but the manifestation is invariably orbital and hence the need for the ophthalmologist to be aware of this entity.

It is usually described that the infection is most commonly seen in uncontrolled diabetes as acidosis and hyperglycemia provide a rich source of nutrients for growth: but any immunocompromised state can precipitate the disease.

Here are a few cases with varied presentations of mucormycosis.

1) A 36 year old male presented with acute onset proptosis of the right eye of 7 days following an episode of viral fever. On examination his visual acuity in both eyes was 6/6. Except for the minimal axial proptosis of 22 mm in the right eye, extraocular movements were full, anterior segment and fundus of both eyes were normal. On further examination he had diffuse fullness on the right side of face and infraorbital anaesthesia.

Blood investigations revealed leucopenia

ENT examination was done and sinus endoscopy revealed frank sinusitis with slough in the paranasal sinuses. No organism was identified.

The patient rapidly deteriorated with restriction of extraocular movements and acute loss of vision in the right eye within a few days of presentation. His vision dropped to no perception of light. There was a relative afferent papillary defect in the right eye but fundus was normal. A clinical diagnosis of orbital apex syndrome was made.

MRI showed enhancing lesion involving the right anterior orbit and right sphenoidal sinus and extending to the cavernous sinus (Fig. 1)

MRI after endoscopy and sinus lavage with Amphotericin –B and debridement of mucor (Fig 2.)
Within 24 hours, the patient developed a central retinal artery occlusion which gave us a strong suspicion of Orbital Mucormycosis.

On repeat endoscopy, black tarry slough were seen in the sinuses which was sent for biopsy that revealed mucor. Extensive orbital debridement, medial wall decompression & sinus irrigation with lyophilized Amphotericin B was done.

But he developed meningitis and altered sensorium. Disease had spread to the other eye with periorbital edema & proptosis suggestive of Cavernous sinus Thrombosis. Patient ultimately succumbed.

2) A 53 year old female, a known diabetic (uncontrolled) presented with moderate proptosis, complete ptosis, and decrease in vision in the left eye, one week following tooth extraction.

On examination, visual acuity was perception of light. There was complete ptosis, with total ophthalmoplegia and lower motor neuron facial nerve palsy. There was no reaction to light, both to direct and consensual in the left eye. On examination fundus showed a pale retina with cherry red spot. Clinically our first suspicion was mucormycosis.

Endoscopy revealed the presence of black necrotic slough in the paranasal sinuses confirmed by MRI (Fig. 3). She too deteriorated rapidly and developed dysphasia, hemiplegia.

3) 58 year old, a well controlled Diabetic with past history of pulmonary tuberculosis came with sudden loss of vision in the right eye of 2 weeks duration with proptosis (22 mm) and total ophthalmoplegia (Fig. 4a) with numbness on the right side of face. Vision in the right eye was Perception of light and in the left eye was 6/6.

Total ophthalmoplegia was present on the right side. Patient was diagnosed to have Orbital apex syndrome and was treated with steroids. He showed no improvement but developed an ophthalmic artery occlusion. Sinus endoscopy showed tar smeared brawny sinus and fungal growth of rhizopus. In the Right eye limited extenration was done but it did not help. Eventually he presented with cavernous sinus thrombosis and died due to pneumonia (?flare up of pulmonary tuberculosis) MRI showed mucor involving the sphenoidal sinus. (Fig. 4 b-c).

3) 60 year old female, a known diabetic (uncontrolled) presented with right eye periorbital oedema and sinusitis. On examination her visual acuity was 6/6 in both eyes. There was right infraorbital anaesthesia. In consultation with ENT, an endoscopy was done which showed black necrotic areas in the maxillary sinus (Fig. 5). These areas were completely debrided and was sent for histopathological examination which showed broad non-septate hyphae (Fig. 6a).

Patient was immediately treated with Amphotericin – B injection and showed dramatic improvement (Fig. 6b).
lesion is medially situated. Exenteration is done in only severe cases when vision is lost and there is threat to the other eye.

Antifungal therapy without exenteration is currently recommended for initial management of patients with rhinoorbital mucormycosis. Amphotericin-B is the agent of choice given daily and increased gradually to reach a cumulative dose of 2 to 4 grams. Treatment must be continued for weeks to months.

To reduce the nephrotoxicity of conventional amphotericin –B, lipid formulations have been introduced. Lipid formulations allow a significant increase in therapeutic index with decreased toxicities.

Repeated debridement without exenteration and local delivery of amphotericin B have been used in patients in whom the disease was diagnosed early.

The overall mortality is 50-70 % or higher with cerebral involvement Prognosis depends on early diagnosis and treatment as well as resolution of the underlying metabolic disorder. Exentration is unnecessary, however repeated surgical debridement may be necessary. The optimum duration of treatment depends on therapeutic response.

Orbital Mucormycosis should be differentiated from allergic fungal sinusitis which is a Type-1 Hyper sensitivity response to inhalation of fungal spores. This is usually seen in immunocompetent patient with history of allergic asthma/chronic sinusitis. Tissue diagnosis shows allergic mucin,Charcoat Leyden crystals (eosinophilic granules) and fungal hyphae (Fig. 7). But there is no fungal tissue invasion and good response to steroids.

Discussion

The clinical presentation of rhinoorbital mucormycosis can be acutely fulminant or indolent. Disease often presents as cold orbital cellulites with ophthalmoplegia and cranial nerve dysfunction. Sudden loss of vision can be due to toxic optic neuritis, central retinal artery occlusion or cavernous sinus thrombosis. A characeristic black eschar, representing tissue necrosis can be seen on skin, nasal mucosa which is always very late. Vascular invasion and occlusion are hallmark features of mucor but again signs that occur too late in the course of this disease.

Early clinical suspicion is mandatory to save the vision. In our experience, any immunosuppressed individual, not necessarily a diabetic (age no bar), presenting with orbital & periorbital edema, chronic sinusitis, infra orbital anesthesia is a Mucor suspect. One need not necessarily wait for tissue biopsy to demonstrate broad, non septate hyphae but can definitely start on empirical therapy with Amphotericin B than to wait for the hyphae growth.

Pterygopalatine fossa is considered the main reservoir for rhinocerebral mucormycosis and extension into the orbit and facial tissues usually follows this route. After proliferation in the nasal cavity, mucor reaches the pterygopalatine fossa, inferior orbital fissure, retroglobal space of the orbit resulting in ocular signs. Facial soft tissues, palate and inferotemporal fossa can be infected through the connecting pathways from pterygopalatine fossa. So the most definite method of treatment of mucor infection is debridement of pterygopalatine fossa.

In early cases just a sinus lavage with Amphotericin –B and good sinus debridement is sufficient for moderate to severe proptosis. Orbital decompression is helpful if...
MRI shows erosion of frontal bone as the mass erodes into the bone and endoscopy shows the paranasal sinuses filled with greenish material consistent with the allergic mucin.

**Points to Remember……..**

- Mucormycosis is not seen in diabetics alone but can occur in any immunosuppressed individual.
- Its ocular presentations can vary from loss of vision to proptosis, ophthalmoplegia, infraorbital anaesthesia …..
- One must suspect Mucormycosis if there is any evidence of sinusitis with the above clinical features
- Sinus Endoscopy & MRI are two most important investigative parameters that can help in detecting mucormycosis
- If the clinical features and above findings strongly correlate, it is always better to err towards mucormycosis
- A peep into the pterygopalatine fossa during endoscopy will be of help as it is commonly the reservoir of infection
- Lyophilized Amphotericin –B is better than conventional Amphotericin –B
- Exenteration is always reserved as the last choice

**References**