Case Report

Rhino-Orbital-Cerebral Infection by *Syncephalastrum racemosum*

Alice J Mathuram*, Promila Mohanraj**, Mary S Mathews***

Abstract

Invasive rhino-sinusitis infection has been known to be caused by zygomycetes commonly belonging to the genera *Rhizopus, Mucor and Rhizomucor*. We report a middle aged diabetic gentleman who had invasive rhino-orbital-cerebral infection caused by *Syncephalastrum racemosum*. This genera belonging to zygomycetes group of fungi which usually causes skin and soft tissue infection but invasive infection with this fungus is rarely known.

Fungal rhinosinusitis occurs in two forms-the invasive form seen in immunosuppressed individuals and the allergic and chronic non-invasive forms seen in immunocompetent hosts. The common fungal agents causing rhinosinusitis are *Aspergillus flavus, Aspergillus fumigatus*, dematiaceous fungi such as *Curvularia lunata*, *Bipolaris spp* and *Drechslera spp* and Zygomycetes such as *Rhizopus spp, Mucor spp* and *Rhizomucor spp*. This is a case report of invasive rhino-orbital-cerebral infection caused by *Syncephalastrum racemosum* belonging to the Syncphalastraceae family, under the order Mucorales.

A 63 year-old gentleman presented to the emergency department with history of vomiting, irrelevant talking and diminished vision in the left eye for 5 days. He had several episodes of holocranial headache for the last 3 months. He had taken various analgesics for the same without relief of symptoms. He was recently diagnosed with diabetes and was on oral hypoglycemic agents with poor compliance.

On examination he was noted to be drowsy with ptosis, proptosis, chemosis and complete ophthalmoplegia of his left eye. His left pupil measured 3 mm and was not responsive to light, with a grade 4 relative afferent papillary defect. The right pupil measured 2 mm, reacted well to light and extra-ocular movements were normal. Corneal reflex was absent on the left and fundus examination revealed papilloedema. His facial sensations were normal and there was no facial asymmetry. There were no other neurological deficits. Examination of the cardio-vascular, respiratory systems and abdomen was normal. Clinical diagnosis of orbital apex syndrome with raised intracranial pressure was made and invasive fungal infection was considered.

MRI of the brain showed marked oedema of bilateral anterior frontal lobes, involvement of bilateral cavernous sinus by peripherally enhancing soft tissue, expanded ethmoidal air cells with breach of the bone, soft tissue at the left orbital apex and extensive sinusitis along with lateral and third ventricular dilatation. Functional endoscopic sinus surgery and debridement of the sinuses was done. Tissue, when examined on direct microscopy showed broad aseptate fungal hyphae. Culture grew *Syncephalastrum racemosum*, a fungal organism belonging to the family, Syncphalastraceae. The unique distinctive microscopic morphology confirms the identification of this organism.

The patient was started on intravenous amphotericin B 50 mg once daily. Blood sugars were controlled with short-acting insulin. Surgical debridement of the intra-cranial lesions was deferred by neurosurgery because of the poor surgical risk due to extensive intra-cerebral involvement. His sensorium gradually improved and vomiting stopped. 10 days later he developed a central venous catheter related blood stream infection and sensorium deteriorated. He was initiated on appropriate antibiotics but he succumbed.

Discussion

*Syncephalastrum racemosum* has been found in soil in India, Southern United states, Panama and Israel. It has been found to contaminate poultry feeds and has been found in a variety of plants and foodstuffs including oats, wheat, soya, nuts, honeycombs, rice, sugar cane, corn, and barley. It has likewise been cultivated from a variety of water sources, bird guano, and composting plant debris. It was earlier debated whether these organisms caused human disease. *Syncphalastrum* species is now well known to cause skin and soft tissue infection and onychomycosis. It has been implicated in a case of intra-abdominal infection. There is one report in the literature so far of *Syncephalastrum* causing invasive sino-orbital disease in a patient with liver cirrhosis due to Hepatitis B infection with renal failure due to a hepatorenal syndrome. Our patient has type II diabetes mellitus for the past one year. These fungi invade the walls of blood vessels causing mechanical and toxic damage to the intima of blood vessels leading to thrombosis. Infection starts in the nose and para-nasal sinuses and spreads to the eye via the angular and lacrimal vein. It extends intra-cranially by invading the superior orbital fissure, ophthalmic vessels and cribiform plate and occasionally invades the carotid artery. They can cross the dura from the sinuses and affect the frontal lobes causing obtundation, the cavernous sinus leading to thrombosis or carotid-cavernous fistula or affect the temporal lobes causing cerebral infarction. Usually death occurs in these patients within 4 weeks of onset if they are not treated. Our patient presented late to the hospital and had extensive intra-cerebral extension. Debridement of the intra-cranial lesions and sinuses along with amphotericin B would have been the ideal way of managing this patient. Treatment for this infection is the same as mucormycosis. Newer agents such as posaconazole has been found to be effective only in about 60% patients who fail standard...
therapy with amphotericin B. Caspofungin used in combination with amphotericin B is associated with improved outcomes. Deferasirox, an iron chelator is now being tried as an adjunct to liposomal amphotericin B in the treatment of zygomycosis.

In conclusion, high index of clinical suspicion of invasive fungal infection is required in immunosuppressed patients presenting with headache and eye symptoms or signs. *Syncephalastrum* is a species of zygomycetes which very rarely can cause invasive disease.

**References**

8. Van Burik JA; Hare RS; Solomon HF; Corrado ML; Kontoyiannis DP. Posaconazole is effective as salvage therapy in zygomycosis: a retrospective summary of 91 cases. *Clin Infect Dis* 2006;42:e61-5.