



## CASE REPORT

# Rhino-orbital mucormycosis

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### Abstract

Rhino-orbital mucormycosis is a rare but life-threatening infection that generally occurs in patients with immune deficiency. As such the condition is a medical emergency. Early recognition and treatment are essential because it may lead to death within a few days. Fungal infections of the eye are uncommon but are being seen with increasing frequency in patients with immune deficiency.

**Keywords:** Diabetes, Fungus, Mucormycosis, Rhino-orbital mucormycosis

### Introduction

Different levels of vision loss, including blindness, can occur from secondary optic nerve dysfunction.<sup>1, 2</sup> The optic nerve may be involved where there is a breach in the thin bony structure that separates the optic nerve from the paranasal sinuses.

Because of their increased scope of practice, optometrists are constantly confronted with disease despite by bacteria, viruses, fungi and other microorganisms. Among these pathogens, the fungi are receiving ever increasing attention despite the rarity of the disease.<sup>3</sup>

This might be due to a number of factors, particularly those that reduce host resistance and allow opportunistic infections to develop. This case report is about a patient who developed rhino-orbital mucormycosis. The aim of this report is to alert practitioners to the importance of early diagnosis of mucormycosis because eye and/or orbital signs usually are the first clinical manifestation of the disease.

### Report of case

The patient was a 55-year-old male who presented to the private optometric clinic with a one-week history of pain in his right eye with associated redness and swelling. The patient had been examined at the provincial hospital two-days before with a diagnosis of herpes zoster and a prescription of acyclovir was given. His past medical history was significant only for diabetes mellitus and he had been unable to control his blood sugar levels over the past three weeks. Since he started taking the acyclovir, he noticed that the pain and swelling in his right eye had increased. He also reported decreased vision. On physical examination, he could not open the right eye and could perceive only bright lights with the right eye. In addition, the right eye had generalized proptosis. The extraocular muscles of the right eye were restricted in all directions of gaze. The pupil of the right eye was dilated and non-reactive to direct and indirect light. The right optic disc was pale. Visual acuity in the unaffected left eye was 6/6 and the patient did not have any visual deficits in the unaffected left eye. The patient did not have vesicular skin rash that typically characterizes herpes zoster lesions.

The patient was referred back to the provincial hospital and he was hospitalised for tests and treatment. CT scan of the orbits and sinuses demonstrated an air-fluid level in the right maxillary sinus and mucosal thickening of the right anterior ethmoid sinus (see Figure 1). MRI of the head showed enhancement of the intraconal fat and rectus muscles of the right eye (see Figure 2). The patient underwent

surgical sinus debridement and biopsy. Biopsy sample facilitated by periodic acid-Schiff (PAS) showed broad, nonseptate and irregular hyphae (filaments that constitute the vegetative form of the fungus. Fungus resembles a simple plant, in that it has a definite cell wall, is nonmotile and reproduces by means of spores but do not possess stem, roots, leaves or chlorophyll). Treatment with amphotericin B was started. The diagnosis was fungal angitis consistent with mucormycosis called rhino-orbital mucormycosis. The patient underwent additional operation, including exenteration of the right eye. Amphotericin B was the only antifungal agent used. Antibiotics were used to combat secondary infections. He was discharged home in good condition after being hospitalised for three weeks.

### Discussion

The case reported here is mucormycosis of the rhino-orbital form with involvement of the ethmoid sinuses. Despite advances in diagnosis and treatment, a high mortality rate of 30-70% still exists for this disease.<sup>4</sup> Death may occur within two weeks if untreated or unsuccessful treated. Mucormycosis, also known as zygomycosis or phycomycosis was first described by Paultauf in 1885.<sup>5</sup> It is an acute opportunistic infection caused by broad, nonseptate saprophytic fungus found in soil, air, bread mould and rotten fruit and vegetables. It can be cultured from the mouth, nasal tract, throat and faeces of healthy persons.<sup>5, 6</sup>

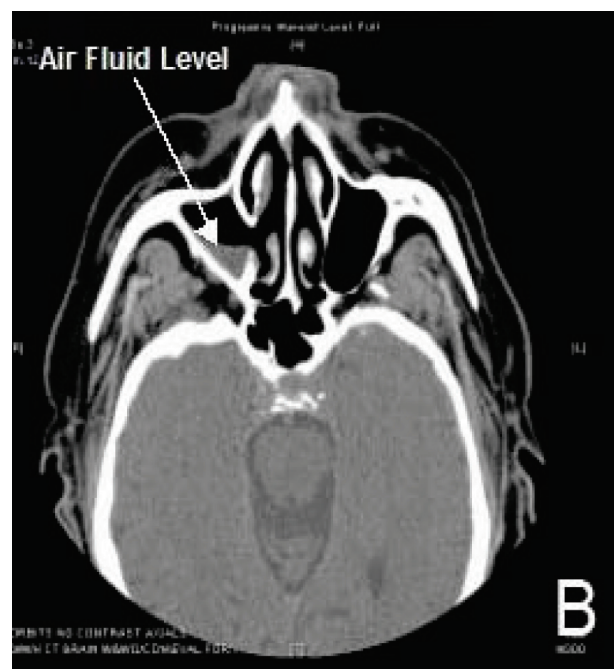
The fungus belongs to the Phycomycetes class, whose most common genera are *Mucor*, *Rhizopus*, *Absidia* and *Basidiobolus*. Contact with the microorganism occurs through the spore inhalation.<sup>7</sup> Infection spreads along vascular and neuronal structures and infiltrates the walls of blood vessels. Infection causes erosion of the bony walls of the ethmoid sinuses and the infection spread into the orbit and retro-orbital area and into the brain (cerebro-rhino-orbital mucormycosis). Death may occur from cerebral abscess.

Infection by this organism usually complicates any underlying chronic disease, such as for the patient in this case. Commonly, mucormycosis attacks people with compromised immune systems. Reduced ability of the serum to bind iron at low pH may be the basic defect in the body defence system.<sup>8</sup> The high iron, glu-

cose-rich, acid milieu facilitates fungal growth. Human resistance to fungal infection rests on the ability to restrict the availability of iron to the



**Figure 1.** Image A of the CT scans of the orbits and sinuses demonstrates an air-fluid level in the right maxillary sinus. Mucosal thickening of the anterior ethmoid sinus and preseptal cellulites are also seen. In affected patients, the diagnostic study of choice is a CT of the orbits and sinuses. (Courtesy of Dr T Segoati, Polokwane Provincial Hospital)



**Figure 2.** Image B of the MRI of the head showing enhancement of the intraconal fat and rectus muscles of the right eye. MRI, if available, can add diagnostic information by showing the extension of the infection into the surrounding blood vessels, orbital fat and intracranial areas. (Courtesy of Dr T Segoati, Polokwane Provincial Hospital)

invading fungus by binding it to proteins such as apotransferrin.<sup>9, 10</sup> Fungal hyphae produce a substance called rhizoferrin, which binds iron avidly. This iron-rhizoferrin complex is then taken up by the fungus and becomes available for vital intracellular processes.

Several predisposing conditions have been reported in the literature.<sup>4, 7</sup> The most common condition is diabetes with ketoacidosis.<sup>11, 12</sup> Diabetic patients are predisposed to the mucormycosis because of the decreased ability of their neutrophils to phagocytise and adhere to endothelial walls.<sup>13-15</sup> Furthermore, the acidosis and hyperglycemia provides an excellent environment for fungus to grow.

The occurrence of mucormycosis in HIV/AIDS patients is rare.<sup>16, 17</sup> HIV status for this patient was negative. Tests for herpes zoster virus were negative. Symptoms of herpes zoster virus include blurred vision, eye pain and red eye which may precede the skin rash. The initial diagnosis of herpes zoster might have been based on eye pain alone.<sup>18</sup>

Orbital and nasal findings are the most common presenting symptoms and signs.<sup>7, 19, 20</sup> Orbital involvement may include loss of function of the second, third, fourth and sixth cranial nerves with proptosis, ptosis, chemosis, diplopia, orbital pain, central retinal artery occlusion, dilated pupil and loss of vision.<sup>7, 19, 20</sup> Diplopia may appear due to paralysis of the motor nerves or intraorbital extension of the infection and involvement of the ocular muscles. Retrobulbar extension of the process may lead to decreased vision or blindness due to optic nerve injury or retinal artery occlusion.<sup>19, 20</sup> Progressive extension of necrosis into the brain can lead to cavernous sinus thrombosis, exophthalmus, complete ophthalmoplegia and papilledema.<sup>19, 20</sup>

## Conclusion

The management of mucormycosis demands a multidisciplinary approach. Nevertheless, the results are not always satisfactory and there are irreversible sequelae with irreparable functional damage. Prognosis may improve with rapid diagnosis, early management and reversible underlying risk factors. Underlying disease is the most important determinant for survival.

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