Orbital mucormycosis with survival

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SUMMARY Orbital mucormycosis is reported in a healthy patient with maturity onset diabetes who was treated with orbital exenteration, amphotericin B, and ketoconazole. A six-year follow-up shows no evidence of recurrence.

Orbital mucormycosis is a rare, fatal condition that usually develops in patients with debilitating disease. Reports of the disease in healthy individuals have been few. We report only the third surviving patient who developed this condition. In most cases mucor invades tissues that have already been altered by disease. Why a usually saprophytic, non-pathogenic fungus becomes invasive is not clear.

The development of mucormycosis has been attributed to decreased host resistance, local lesions incident to primary disease, and disturbance brought about by therapy. Most cases occur in patients with diabetes. However, other predisposing factors are leukaemia, multiple myelomas, carcinoma, anaemia, burns, septicaemia, and hepatitis. The use of antibiotics, steroids, and ionising radiation favours the growth of the organism.

Diagnosis is often made at necropsy, though some cases are recognised during life. The cerebral form is the most easily identified, since ocular manifestations are apparent in 50% of cases.

Since the introduction of amphotericin B treatment in 1958 several patients with mucormycosis have survived. We report such a patient who is alive six years later and shows no sign of recurrence.

Case report

A 61-year-old Caucasian man presented in July 1978 with a six-week history of double vision. He had diabetes of one year’s duration. At examination his visual acuity was 6/6 in both eyes. His right intraocular pressure was 35 mmHg and right proptosis was 21 mm, with loss of right abduction and left depression. The fields were full and colour vision was normal, but the thyroxine level was low; however, a TRH–TSH test gave a normal result. A orbital computerised tomography scan was normal (Fig. 1). He was treated with acetazolamide 500 mg twice daily. His proptosis increased to 26 mm over a two-month period and his symptoms remained. He was treated with prednisolone 60 mg daily and given replacement thyroxine. His proptosis reduced to 22 mm and his steroid therapy was tailed off. In February 1979 he returned with proptosis of 25 mm and intraocular pressure of 40 mmHg. Prednisolone 100 mg daily was recommended and his proptosis fell to 23 mm.

In July 1979 he returned with proptosis of 26 mm. Ethmoidal x-ray showed thickening and an orbital computerised tomography scan (Fig. 2) confirmed an opaque ethmoidal sinus and an enlarged right medial

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Fig. 1 Orbital computerised tomography scan showing a normal right orbit and ethmoidal sinus.
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Fig. 2 Orbital computerised tomography scan showing opaque ethmoidal sinus and thickened medial rectus muscle.

Fig. 3 Appearance consistent with orbital cellulitis.

rectus muscle. Subsequently he developed a right ischaemic optic neuropathy and what appeared clinically to be a right orbital cellulitis (Fig. 3). Antibiotics were tried without benefit. He underwent a right orbital and ethmoidal exploration, and cheesy material was removed for biopsy. Culture and histological examination confirmed Rhizopus (Fig. 4). He was given amphotericin B intravenously in a dosage of 0.25 mg/kg body weight. Surgical exploration revealed extensive involvement of the orbit, ethmoidal sinus, and anterior wall and mucosa of the right maxillary antrum. Furthermore, there was fungal invasion in three places in the dura mater of the anterior cranial fossa. With neurosurgical and otolaryngological assistance, orbital exenteration and ethmoidectomy were performed. In addition the orbital floor and antral mucosa were removed. Almost all of the necrotic and infected tissue was excised.

Two weeks after surgery the amphotericin B was stopped and he was started on ketoconazole 200 mg daily for a further six weeks. At four months there was no evidence of recurrence either clinically or radiologically, and he was fitted with an exenteration prosthesis.

Fig. 4 Orbital tissue showing typical non-septate hyphae. (H and E, ×300).
The patient remained well until November 1984, when he presented with a left central retinal vein occlusion. Examination showed no recurrence of Rhizopus. Six months later his left visual acuity had not improved beyond 6/60, but he remained well.

Discussion

Although many cases of mucormycosis have been reported, true orbital mucormycosis is very rare. Two cases were reported in patients with poorly controlled diabetes and debilitation who survived. At initial presentation our patient had no evidence of the condition, and the most likely presenting diagnosis was dysthyroid eye disease or orbital pseudotumour. However, the use of steroids and the fact that he was mildly diabetic predisposed him to mucormycosis. The infection started in his ethmoidal sinus and spread directly to the orbit. His case illustrates the preference of this organism to invade blood vessels. He was treated with the antifungal agent ketoconazole because it was considered more effective with fewer side effects than other available drugs; this was the first use of ketoconazole in Great Britain. The drug is now known to cause liver damage in one out of every 15,000 patients treated. However, in the case of our patient the combination of exenteration and antifungal therapy saved his life.

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References


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