

Rhinocerebral mucormycosis with extension to the cavernous sinus.

A case report

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Rhinocerebral mucormycosis is a fulminant mycotic infection that occurs primarily in patients with poorly controlled diabetes mellitus. The patient's upper airway is the usual port of entry for this infection, with fungus organisms initially seeding either the nasal mucosa or the hard palate (Hamill et al., 1983). The infection then spreads to the adjacent paranasal sinuses and orbit with associated bone and tissue destruction. There may be evidence of cavernous sinus thrombosis (Soloniuk et al., 1988).

We report the successful treatment of a diabetic boy who was initially seen with rhinocerebral mucormycosis and cavernous sinus thrombosis.

CASE REPORT

A 15 year-old boy was admitted to the hospital with a 20 day history of unilateral ophthalmoplegia associated with proptosis, visual disturbance. The skin of the left side of the face was infiltrated by fungus (Figure 1). He had had poorly controlled diabetes mellitus for about four years. A computed tomographic scan demonstrated a mass involving ethmoidal, maxillary, and sphenoid sinuses, orbit and cavernous sinus on the left side (Figure 2). Immediate orbital, maxillary, ethmoidal and sphenoidal extirpation was performed without taking any biopsy previously. The preoperative clinical diagnosis of mucormycosis was confirmed pathologically (Figure 3). Postoperatively the diabetes was brought under control. Just after the surgical therapy, amphotericin B was initiated with a total dose of 0.3 mg/kg in two divided doses, and continued 1.2 mg/kg over a 2 months period. The patient showed marked clinical improvement, visual acuity and ocular movements increased and the skin lesions returned to normal. The patient was discharged at the end of the second month and unfortunately he has not applied so far.



Figure 1.
15 years-old patient with rhinocerebral mucormycosis.

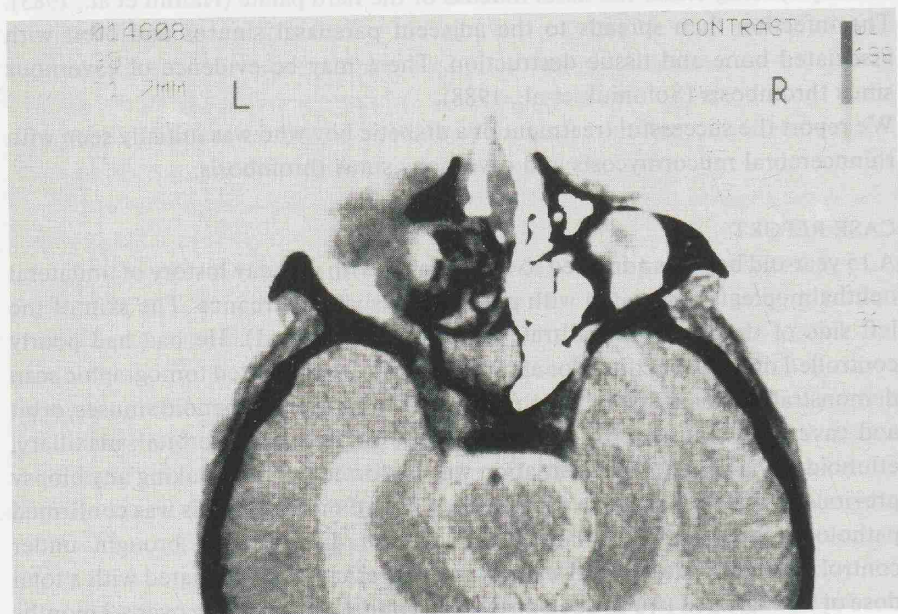


Figure 2. Postcontrast axial scan. The scan obtained from the floor of the orbit shows bone destruction of the ethmoidal sinus walls. A soft tissue lesion in the left ethmoidal and sphenoidal sinuses is seen. The cavernous sinuses are asymmetric (widening of the left cavernous sinus was thought to be due to involvement of the cavernous sinus). The left inferior rectus muscle was enlarged and the sclera showed contrast enhancement.

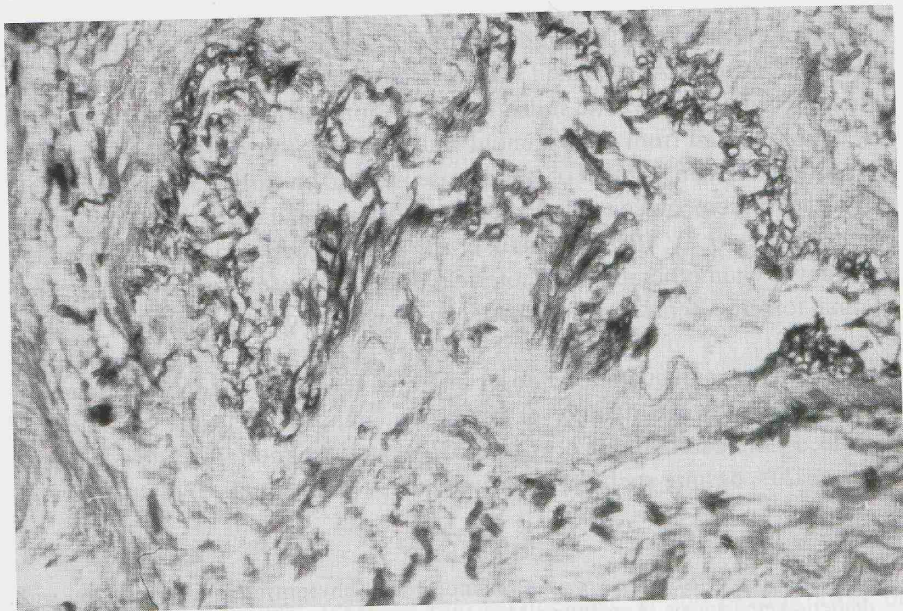


Figure 3. Innumerable hyphae are seen invading a vessel wall in the biopsy section from maxillary sinus. H+E x 230.

DISCUSSION

Rhinocerebral mucormycosis is an opportunistic infection produced by a fungus from the phycomycetes family, generally regarded as the most lethal of the group (Hamill et al., 1983). The onset of the disease is typically marked by local pain and swelling in the involved area which causes a necrotic and bloody discharge. These symptoms are followed by opthalmoplegia, proptosis and central nervous system symptoms as the infection spreads. The infection usually begins in the nose, extends into the adjacent paranasal sinuses. It may then progress through the ethmoid sinus and from there through the retroorbital region. It may also extend into the brain through the apex of the orbit. The organism also has an affinity for the internal elastic lamina of arteries and therefore blood vessel invasion is common (Abramson et al., 1967; Couch et al., 1988). This is why cavernous sinus thrombosis is commonly seen. Cavernous sinus thrombosis was present in our case.

Rhinocerebral mucormycosis should always be considered in sinusitis among diabetics, regardless of blood sugar control, as well as in the immune-compromised patient. The definitive diagnosis rests with histological examination. Large irregularly wide, non-septate hyphae with right angle branching on silver or periodic acid-schiff stain identify mucormycosis (Abramson et al., 1967; Price et al., 1980).

In differential diagnosis, *Aspergillus fumigatus*, other species of *Aspergillus* and species of *Mucor*, *Absidia* and *Rhizopus* should be considered as potential aetiologic agents of mycotic sinusitis. In addition, Actinomycetic osteomyelitis has been reported from the sphenoid sinus with fatal intracranial extension. The successful management of this disease is based on early diagnosis, adequate surgical debridement, control of underlying disease, and therapy with antifungal agents (Couch et al., 1988; Soloniuk et al., 1988). Amphotericin B has been the mainstay of chemotherapy. Amphotericin B should be used with caution because of renal toxicity. Renal function should be monitored during treatment (Battock et al., 1968).

Radical surgery, adequate total doses of amphotericin B, and medical management of diabetes mellitus are of paramount importance in the treatment of advanced rhinocerebral mucormycosis. However, the mortality rate is between 20% and 70% despite such an intensive therapy (Ochs et al., 1988).

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