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Rhinocerebral mucormycosis: An unusual case presentation

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Abstract

Rhinocerebral mucormycosis is a rapidly fatal fungal disease which involves the nose, paranasal sinuses, orbit and central nervous system. The fungal infection is usually secondary to immunosuppression, diabetic acidosis, or antibiotic, steroid or cytotoxic therapy. It can also occur in patients suffering from burns, malignancy and haematological disorders. Current treatment consists of correction of the underlying disorder, repeated debridement of the wound in combination with intravenous amphotericin B.

This paper describes our experience with a case of rhinocerebral mucormycosis. This is an unusual case in which mucormycosis was seen in a young female where no underlying cause was found. She responded to surgical debridement in combination with intravenous amphotericin B.

Introduction

Rhinocerebral mucormycosis is a potentially lethal disease of the nose and paranasal sinuses caused by fungi belonging to class Phycomycetes order Mucorales. The disease rapidly spreads to the orbit and brain and is eventually fatal unless clinical recognition is early and specific treatment is constituted rapidly.

Mucormycosis is an opportunistic infection and its pathogenic nature becomes evident when the patient's general resistance has been altered by metabolic disorders or chemotherapeutic agents. However, in this case we did not find any underlying disorder.

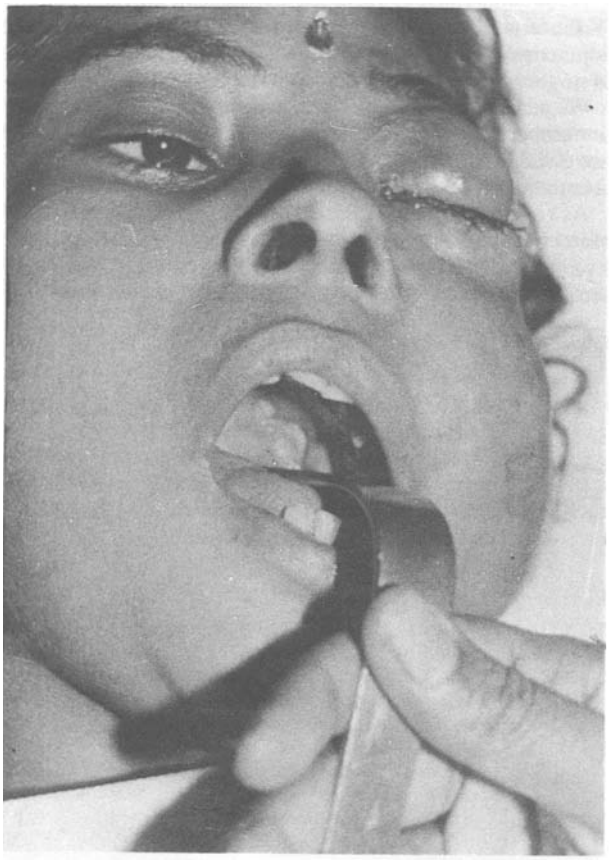

FIG. 1

Clinical photograph of the patient at the time of admission. Ulcers are seen on the soft palate.

Case report

A 24-year-old female presented with swelling of the left eye, left side of the face and left-sided nasal obstruction for 15 days. She had a sudden loss of vision in the left eye and had noticed ulcers on the palate on the day of admission to the ENT ward (Fig. 1). Otolaryngological examination revealed two ulcers on

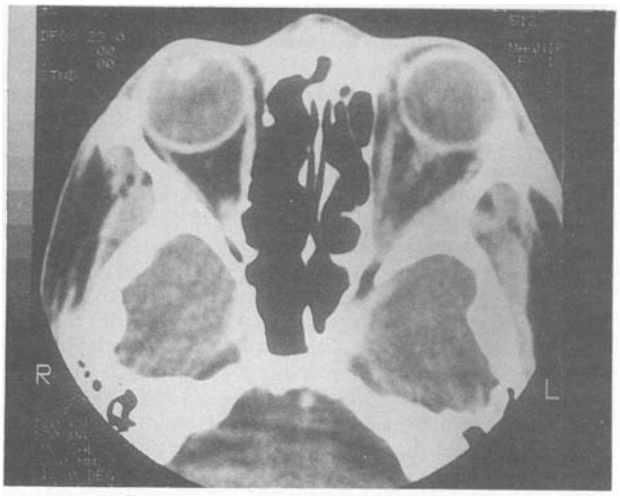

FIG. 2

CT scan showing radio-opaque densities in the left retro-orbital tissues suggestive of orbital cellulitis.

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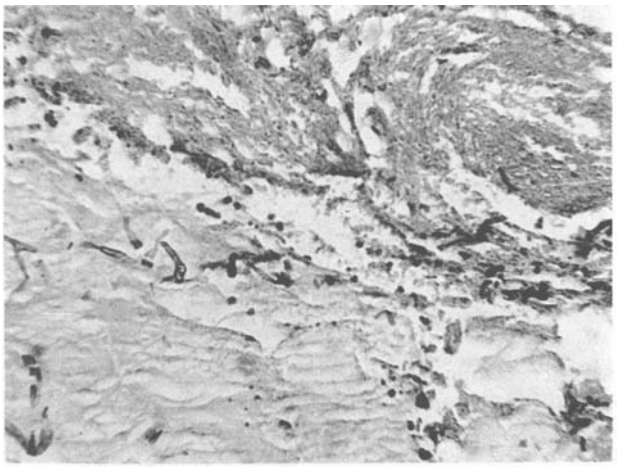

FIG. 3

Photomicrograph showing necrosis, mycotic infiltration of the perivascular tissue. Fungal hyphae are seen invading the blood vessels (HE ×160).

the hard palate on the left side, 2 × 2.5 cm and 3 × 2 cm in size respectively, with necrotic edges and hyperaemic margins. The ulcers were painless, covered with slough and did not bleed on touch. There was no perforation into the nasal cavity or maxillary sinus. Nasal examination showed presence of mucopurulent discharge but no nasal mass was evident. Ophthalmic examination revealed that her left eye had proptosis with ophthalmoplegia. Visual evaluation revealed absent perception of light in the left eye. A CT scan showed evidence of orbital cellulitis, radio-opaque densities were seen in the retro-orbital tissues (Fig. 2). There was no evidence of cavernous sinus thrombosis.

The diagnosis was established from the edge biopsy of the palatal ulcer. Histopathology showed characteristic features of mucormycosis. The sections (Fig. 3) showed extensive areas of necrosis, pinkish hyalinized areas with foci of fungal hyphae all over. The fungus consisted of broad, non-septate, branching hyphae better demonstrated by GMS staining. These fungal hyphae were seen invading blood vessels. There was also mycotic infiltration of the perivascular tissue.

Routine blood tests showed haemoglobin to be 12 g/dl WBC 10,800, polymorphs 53 per cent, lymphocytes 45 per cent, eosinophils 2 per cent. ESR was 27 mm/first hour. The fasting blood sugar was 88 mg and the post prandial blood sugar was 118 mg. VDRL and HIV tests were negative. T cell count and IgG levels were found to be within normal range. A chest X-ray did not reveal any abnormality.

The initial treatment plan was an early wide surgical debridement of the affected tissue. This involved clearance of diseased tissue from the left side of the nose, maxilla, ethmoids and orbit which included exenteration of the eyeball. Four weeks later a repeat debridement was performed in which necrotic bone and soft tissue was cleared from the anterior wall of maxilla, floor of orbit and from the palate. Surgery was followed by administration of intravenous amphotericin B (0.2 mg/kg body weight initially and increased to 0.4 mg/kg body weight). Amphotericin B was given via a subclavian line for a period of six weeks. The patients renal and liver functions were monitored periodically. The effectiveness of the antifungal agent was evidenced by rapid healing of the palatal ulcers.

Three months after completion of amphotericin B treatment, the patient noticed a discharge from the floor of the orbit. A biopsy of the suspicious area confirmed recurrence of mucormycosis. She was again treated with a six week course of intravenous amphotericin B. Repeat biopsies from the orbit, maxilla and ethmoids were all negative after completion of treatment.

The patient is disease free one year after her discharge from the hospital.

Discussion

Rhinocerebral mucormycosis is a potentially lethal disease of the nose and paranasal sinuses caused by fungus belonging to the class Phycomycetes, order Mucorales. The disease rapidly spreads to the orbit and brain and is eventually fatal unless clinical recognition is early and specific treatment is constituted rapidly.

The lesions in rhinocerebral mucormycosis are usually necrotic or haemorrhagic with ulceration of the mucosa. The spread of the fungus is unique in that it has a great affinity for arteries. It penetrates their tough muscular walls, grows within the lumina and stimulates acute arteritis or thrombosis. Later, it invades veins and lymphatics and sets in the characteristic lesions of a combination of infarction and inflammation.

The disease is more frequently associated with diabetic acidosis than with any other disease (De Weese *et al.*, 1965). The long-term suvival rates range from 66 per cent (Bahadur *et al.*, 1983) to 85 per cent (Pillsbury and Newton, 1977).

In this presentation what is noteworthy is the fact that no underlying disorder was found. Whether the cause of the disease in this particular case was due to a virulent fungal strain or not, is debatable.

The treatment consists of correction of the underlying disorder if any, repeated surgical debridement of the wound in combination with intravenous amphotericin B. Bahadur *et al.* (1983) reported good results with oral potassium iodide but this agent was not used in our case.

Acknowledgement

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References

Bahadur, S., Ghosh, P., Chopra, P., Rai, G. (1983) Rhinocerebral phycomycosis. *Journal of Laryngology and Otology,* **97:** 267–270.

De Weese, D. D., Schleuning, A., Robinson, L. B. Mucomycosis of the nose and paranasal sinuses. (1965) *Laryngoscope,* **75:** 1398–1407.

Pillsbury, H. C., Newton, D. F. (1977) Rhinocerebral mucomycosis. *Archives of Otolaryngology,* **103:** 600–604.

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