The Journal of Laryngology and Otology
March 1983. Vol. 97. pp. 267–270

Rhinocerebral phycomycosis

by

S. Bahadur, P. Ghosh, P. Chopra and G. Rai (New Delhi)

Rhinocerebral phycomycosis (mucormycosis) is a rapidly fatal fungal disease which involves the nose, paranasal sinuses, orbit and central nervous system. Diseases in man caused by fungi belonging to the class phycomycetes or zygomycetes can be divided into two groups: mucormycosis and entomphthoramycosis (Clark, 1968).

This paper describes our experience of three cases of rhinocerebral phycomycosis.

Case No.1

S.S., a 10-month-old male child, was admitted to the ENT Ward on 10 January 1980 with complaints of a feeding problem for 15 days and swelling of the right eye for the previous week. There was no other significant history available. General physical examination showed that the infant was malnourished, anaemic and afebrile, and his weight was 5.8 kg. Ophthalmic examination revealed that his right eye had proptosis with ophthalmoplegia. The pupils were dilated and fixed. Fundus examination revealed clouding and blurring of the disc with dilated veins. There was evidence of a flame-shaped haemorrhage. Throat examination showed ulceration of the palate. Nasal examination revealed marked crusting and a large septal perforation. Routine blood tests showed haemoglobin to be 6.8 Gm. per cent, TLC 11,200/cu. mm., polymorphs 43 per cent, lymphocytes 55 per cent, eosinophils 2 per cent, ESR 52 mm./1st hour. Urine test was normal. Radiological examination of the chest did not reveal any abnormality. X-ray of the paranasal sinuses showed opacification of the maxillary antra with erosion of the bony nasal septum. An exploration was carried out under general anaesthesia. There was extensive necrosis of bone involving the hard palate, nasal septum and ethmoids including the cribriform plate. A clinical diagnosis of rhinocerebral mucormycosis was made. Histopathology confirmed mycotic osteomyelitis which was consistent with mucormycosis. Unfortunately, the child died before specific anti-fungal treatment could be commenced.



Fig. 1

Clinical photograph of case No. 1, showing proptosis of right eye and ulceration of the palate.

Case No. 2

V.S., a 1½-year-old male child, was admitted to the ENT Ward on 20 February 1981 with complaints of ulceration of the nose and feeding difficulty for the previous two months. The patient

From the Departments of Otolaryngology and Pathology, All India Institute of Medical Sciences, New Delhi-110 029 (India).

https://doi.org/10.1017/S002221510009410X Published online by Cambridge University Press



FIG. 2

Clinical photograph of case No. 2, showing ulcerative lesion of the nose. Part of bare septum can be seen in the picture.

had earlier suffered from abdominal distension with subacute obstruction. He had been treated elsewhere where a nasogastric tube was passed. The child improved, but on removing the feeding tube it was noticed that there was a small ulceration of the nose. The ulcer progressively increased in size. On physical examination, the child was found to be moderately dehydrated and poorly nourished. There was extensive ulceration of the nose. The ulcer extended from the medial canthus of the right eye to the right ala of the nose, and it measured 5 cm.×3 cm. The floor of the wound was irregular and extended into the nasal cavity where normal anatomy was distorted. The turbinates could not be identified. There was a perforation of the hard palate in the midline 2 cms. behind the upper alvolus. There was congestion in the right eye.

Investigations included a haemogram which showed Hb. to be 9.6 Gm. per cent, TLC was 9,500/cu. mm. of blood, and ESR was 13 mm./1st hour. X-ray chest was normal. X-ray PNS showed haziness and erosion of the right maxillary sinus. Biopsy showed broad, non-septate hyphae compatible with mucor species. Debridement of the wound was done twice under general anaesthesia. The patient was treated with intravenous amphotericin B, in an initial daily dose of 0.2 mg./kg. body weight. A total daily dose of 3 mg. was administered for one week. It was later increased to 5 mg., added to 150 ml. of 5 per cent dextrose. Treatment was continued for three months during which he received a total dose of 180 mg. of amphotericin B. Regular recordings of the serum electrolytes and blood urea were done during this period. The child improved and the lesion regressed completely.

Case No. 3

A 65-year-old male was referred on 20 November 1980 from the renal unit where he had been admitted with acute renal failure. He required a tracheostomy for aspiration of excessive secretions in the chest. There was no history of diabetes. However, the patient developed a pseudomonas infection and was treated with intensive gerramycin therapy. He improved considerably and was detubated. A week later he complained of nasal obstruction and



FIG. 3

X-ray nose and sinuses in case No. 3, showing destruction of the nasal septum and part of the hard palate with opacity of right maxillary antrum.

https://doi.org/10.1017/S002221510009410X Published online by Cambridge University Press

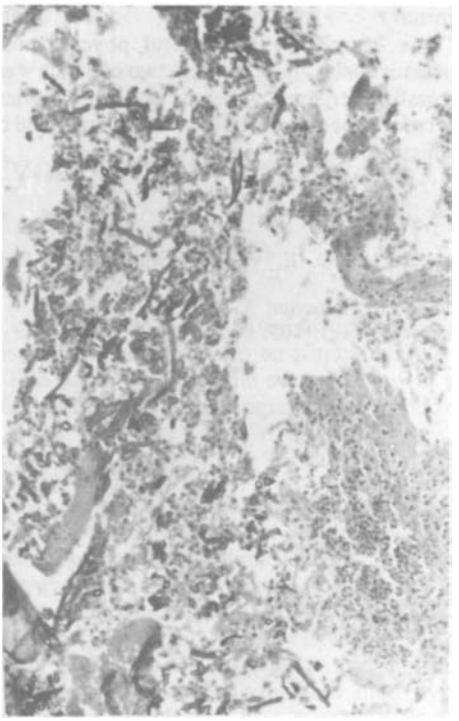


FIG. 4

Photomicrograph showing inflammatory exudate and necrosis in which are embedded numerous broad non-septate hyphae of varying shapes. (H and E ×120).

a foul smell. His general physical examination was normal. Nasal examination revealed the presence of an extensive necrotic slough in both nasal chambers. There was a large septal perforation. Throat examination showed perforation of the palate with necrotic margins. X-ray of the para-nasal sinuses showed haziness of the maxillary antra with obstruction of the nasal septum. He was treated with oral potassium iodide in combination with repeated debridement of the wound. Potassium iodide therapy was continued for a period of six months. His haemogram, serum cholesterol and serum potassium with ECG recordings were done at regular intervals. He has been followed-up for one year and is found to be free of disease. The palatal defect was covered by providing him with a palatal prosthesis.

Discussion

Rhinocerebral mucormycosis is more frequently associated with diabetic acidosis than with any other disease (De Weese *et al.*, 1965). However, the disease can also occur in patients suffering from burns, malignancy, haematological disorders, and other chronic illnesses in immunosuppression; and it has been associated with antibiotics, cortisone and cytotoxic therapy.

Current treatment consists of correction of the underlying disorder and repeated debridement of the wound in combination with intravenous amphotericin B. Pillsbury and Newton (1977) have reported a long-term survival rate in 85 per cent of their 13 cases of rhinocerebral mucormycosis. One of our cases was treated with amphotericin B along with repeated debridement of the wound and has obtained a complete cure. However, another patient has done extremely well with surgical debridment of the wound in combination with oral potassium



FIG. 5

Higher magnification of the above, showing morphologic details of hyphae. Segment of necrotic bone is also seen in the picture. (H and E ×250).

https://doi.org/10.1017/S002221510009410X Published online by Cambridge University Press

iodide therapy, the latter drug being preferred because this patient had previously suffered from renal failure and amphotericin B is known to be toxic to the kidneys. The exact mode of action of potassium iodide is not known. It is believed that it has some fungicidal property.

Summary

Three cases of rhinocerebral phycomycosis (mucormycosis) are presented, two of which were successfully treated, whereas the third patient died before specific antifungal treatment could be commenced.

References

CLARK, B. (1968) *Systemic Mycosis.* J. A. Churchill Ltd., Ed. by G. E. W. Wolstenholme and Ruth Porter, p. 159.

DE-WEESE, D. D., SEBBUNING, A., and ROBINSON, L. B. (1965) *Laryngoscope,* **75:** 1398.

PILLSBURY, R. C., and NEWTON, D. F. (1977) *Archives of Otolaryngology,* **103:** 600.

https://doi.org/10.1017/S002221510009410X Published online by Cambridge University Press