Short report

Cerebrospinal fluid rhinorrhoea from massive osteolysis of the skull

G. VASUDEVA IYER

From the Department of Neurology, Medical College and Hospital, Trivandrum, India

SUMMARY Involvement of the skull by a rare disorder—massive osteolysis—led to CSF rhinorrhoea and meningitis. This is believed to be the first time that such a case has been reported.

Head injuries are by far the most common cause of CSF rhinorrhoea. It has been estimated by Lewin (1954) that about 2% of patients with head injuries develop it. Nontraumatic leaks of CSF may be of the high pressure type, as with intracranial tumours, or result from congenital defects, focal atrophy or, very rarely, from osteomyelitic erosions of bone (Ommaya, 1968). This report concerns a patient who had an unusual calvarial lesion which was diagnosed and reported earlier as massive osteolysis (Iyer and Nayar, 1975). She recently developed CSF rhinorrhoea and meningitis.

Case report

A 58 year old woman was admitted to the Neurology Unit in the Medical College Hospital, Trivandrum in June 1978 complaining of headache, vomiting, and delirium. During the previous six months she had noticed dripping of watery fluid from both nostrils on forward bending of the head. It was intermittent and not associated with headache, vomiting, nose block, or sneezing. One week before admission she developed fever, intense headache, and delirium.

For the past 13 years she had noticed an enlarging depression above her forehead. Starting as a midline dimple in the frontal region, it gradually achieved its present size without causing any pain or discomfort. She was admitted for investigation in 1970 and 1974, and after detailed study including biopsy a diagnosis of massive osteolysis involving the calvarium was made (Iyer and Nayar, 1975).

Physical examination revealed a delirious patient with marked neck stiffness. She had an oblong (120×60 mm) depression in the midline of the skull starting 20 mm above the nasion and extending almost to the vertex (Fig. 1). The skin over the depression appeared normal. The area of the depression was totally devoid of bone, and at the periphery an irregular bony margin could be felt. Pulsation could be detected over this area quite similar to that seen over the open fontanelle

Fig. 1 Photograph of the patient showing the large depression above the forehead.

Address for reprint requests: Dr G. V. Iyer, Department of Neurology Medical College and Hospital, Trivandrum-695011, India.

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of an infant. The fundi were normal, and no neurological abnormalities could be detected. Systemic examination was noncontributory.

Routine blood examination showed a polymorphonuclear leucocytosis. The CSF pressure was increased and the fluid appeared turbid; the protein content was 0.38 g/l and sugar 1.1 mmol/l. Microscopy revealed abundant polymorphs; no organisms were detected with Gram's stain nor isolated in culture. Radiography of the skull (Fig. 2) showed total disappearance of bone under the depression, extending from the coronal suture to the frontal sinus. There was no osteosclerosis at the periphery of the lesion. Special views to visualise the cribriform plate of the ethmoid did not reveal any erosion.

A diagnosis of purulent meningitis secondary to CSF leak was made, and the patient was started on high doses of crystalline penicillin and chloromycetine. There was remarkable improvement within three days. A second lumbar puncture at the end of one week showed a protein level of 0.50 g/l, sugar of 3.3 mmol/l, and only rare polymorphs. While she was convalescing, detailed blood studies (ESR, RBC, WBC, and platelet counts, VDRL, serum protein, calcium, phosphate, alkaline and acid phosphatase) bone marrow studies, and skeletal survey were carried out and were normal. The patient was unwilling to undergo surgery for repair of the CSF leak and, therefore, further investigations to locate the fistula were not undertaken.

Fig. 2 Radiograph of skull taken in 1978, showing extension of the lytic area to the frontal sinus.

Discussion

Serial radiographs showed that this patient had destruction of the frontal bones starting almost 12 years previously and progressing relentlessly with recent extension to the frontal sinus leading to CSF rhinorrhoea. The bone appeared to be totally lysed, and the area was not replaced by pathological tissues as in conditions such as eosinophilic granuloma, plasmacytoma and so on. Repeated aspiration biopsy from different portions near the margin of the lytic area in 1974 and later did not reveal any abnormal tissue. The prolonged course, normal blood chemistry, normal bone marrow picture, and the absence of replacement of the lytic area by pathological tissue suggested the diagnosis of massive osteolysis. Such spontaneous, progressive, and total disappearance of bones has been documented under various names such as phantom bones, disappearing bones. The first report of such a condition concerned a patient with a boneless arm (Anonymous, 1838). Thereafter about 64 cases have been reported, with the involvement of many bones with the exception of the skull. The first reported example of calvarial involvement was the initial report on the present case (Iyer and Nayar, 1975), and three more cases have been reported subsequently (Rajalekshmy *et al.*, 1977).

The radiological appearance is typical and shows dramatic dissolution of bone without evidence of sclerosis. It is a peculiar feature of this disease that it extends across the joint line to affect an entire anatomical region—for example, upper femur with portions of pelvis; clavicle with scapula and the ribs. The condition rarely leads to complications other than pathological fracture. Paraplegia from vertebral involvement (Gorham *et al.*, 1954) and pleuropulmonary complications from involvement of the thoracic cage have been reported (Case Records of Massachusetts General Hospital, 1964). Our patient had no complication at the time of the first report in 1975, but with extension of the lytic process on to the frontal sinus CSF leak and meningitis resulted.

Neither the cause nor the treatment of massive osteolysis is known. It is believed that it may be a form of angiomasia, where a non-neoplastic proliferation of vascular channels occurs leading to active hyperaemia and progressive dissolution of bone (Gorham and Stout, 1955).

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