

Langerhans Cell Histiocytosis: Multifocal Eosinophilic Granuloma - A Case Report

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INTRODUCTION

Localised langerhans cell histiocytosis of bone (eosinophilic granuloma) is a benign tumor like condition with a variable clinical course [1]. Peak incidence is between the ages of 5 and 10 yrs and 75% of cases occur in persons under 20 years of age [2]. We present a case of langerhans cell histiocytosis: multifocal eosinophilic granuloma of skull bone with typical radiologic features.

CASE REPORT

A six years old girl presented with swelling over the right parietal region. There was no history of fever or trauma. On clinical examination there was minimal tenderness over the swelling. The patient was later subjected to radiological and hematological examinations. Hematologic investigations were within normal limits.



Fig. 1: AP Skull Radiograph shows multiple lytic lesions in the parietal and frontal bones.

Anteroposterior and lateral skull radiographs showed multiple lytic lesions on the parasagittal region of the right parietal bone and at fronto-temporal suture line of the right frontal bone [fig. 1,2].

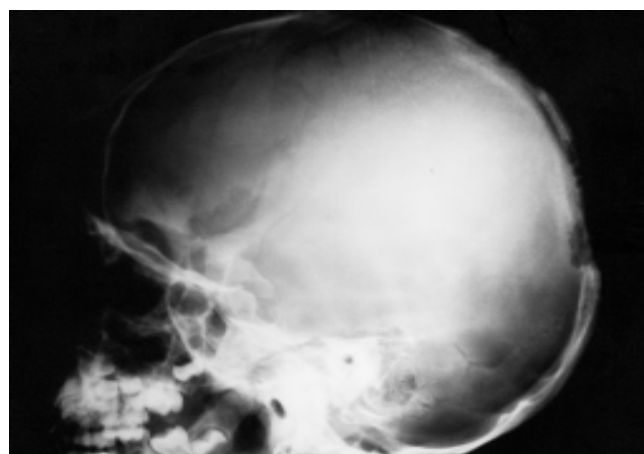


Fig. 2: Lateral Skull Radiograph shows multiple lytic lesions in the parietal and frontal bones.

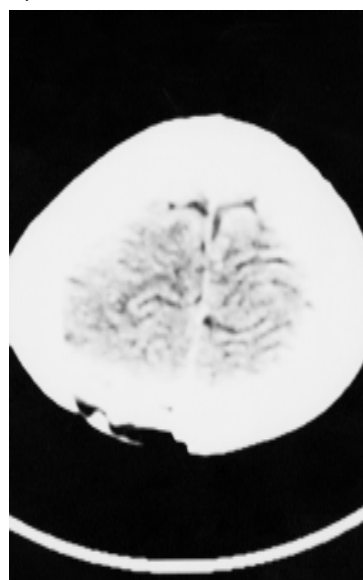


Fig. 3: Axial NECT Brain shows osteolytic erosion of the right parietal bone in its posterior aspect with small intact bony fragment "Button sequestrum".

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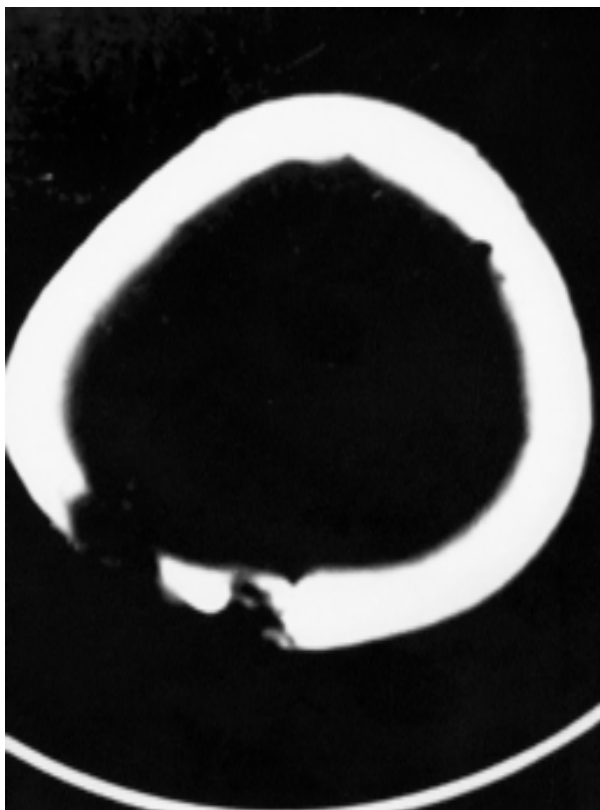


Fig. 4: Axial CT Brain (Bone window) shows osteolytic erosion of the right parietal bone in its posterior aspect with small intact bony fragment "Button sequestrum".

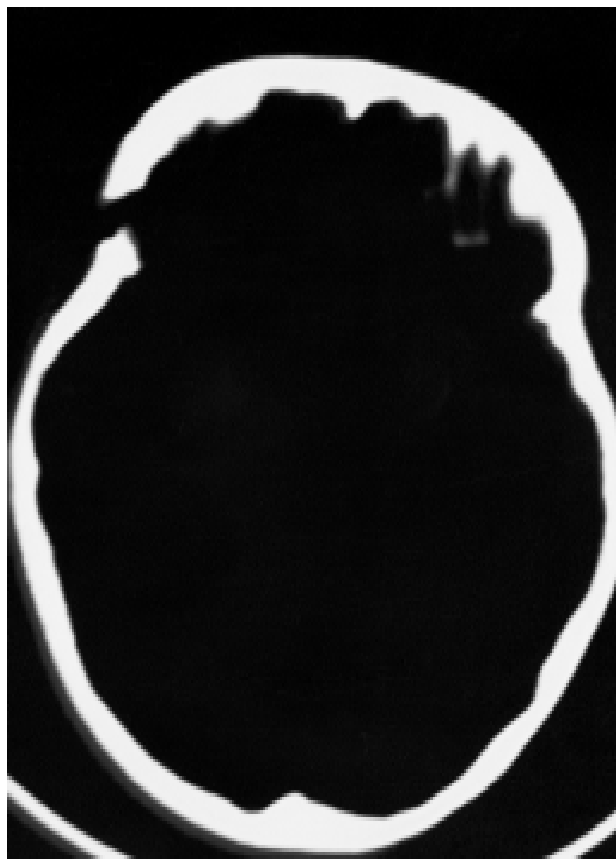


Fig. 5: Axial CT brain (Bone window) shows osteolytic lesion in frontal bone on the right side at fronto-temporal suture line.

Axial CT sections of the cranium showed osteolytic lesion in the high parietal bone in its posterior aspect on the right side, measuring 4.3cm in width with a small intact bony fragment in the centre (button sequestrum). Inner and outer table of the skull were eroded [fig. 3,4]. Another osteolytic lesion measuring 1.4 cm in width was noted in the frontal bone on right side at fronto-temporal suture line [fig. 5].

Chest radiograph posteroanterior view showed no abnormality. Ultrasonography of abdomen was normal.

The radiological features were suggestive of langerhans cell histiocytosis: multifocal eosinophilic granuloma.

DISCUSSION

Langerhans cell histiocytoses are a group of seemingly diverse diseases all characterized by the abnormal accumulation of inflammatory histiocytes. The severity of manifestations and the duration of illness in histiocytoses vary from benign and self-limited to acute, disseminated and fatal. Eosinophilic granuloma was first described by Jaffe and Lichtenstein in 1944 [3]. Lichtenstein observed the similarities between eosinophilic granuloma, Hand-Schuller-Christian disease and Litterer-Siwe disease and proposed that they be considered as variants of a single process collectively known as "Histiocytosis 'x'" [4] also known as Langerhans cell histiocytosis.

Eosinophilic granuloma accounts for upto 60% of all cases of langerhans cell histiocytosis and most frequently affects the skeleton in a monostotic or polyostotic fashion with the solitary lesions prevailing by more than three to one over multiple lesions. Any bone can be involved, but more than 50% of lesions occur in the skull, spine, pelvis, ribs and mandible [2]. Presence of proliferating histiocytes is the histologic hallmark of langerhans cell histiocytosis, including eosinophilic granuloma. Morphologically these disorders are characterized by proliferation of mononuclear and sometimes multinucleated cells with pale, ill defined eosinophilic cytoplasm and lobulated nuclei with longitudinal grooves. Electron microscopy used for definitive diagnosis shows distinctive structures called langerhans or Birbeck granules. These are rod like structures, with a striated core that may have a dilated end, giving them a tennis racket appearance. The Birbeck granules are organelles seen singly or in small clusters in the cytoplasm or as invagination of the plasma membrane and they are believed to be products of internalization of complexes derived from antigen at the cell membrane. Expression of CD1a by immunohistochemistry is also considered diagnostic of Langerhans cell histiocytosis [2, 5].

The radiological features vary considerably depending on

the site of the lesion. In the skull round to oval osteolytic lesion about 1-4 cm in diameter is seen. More than one lesion may be present. The lesion tends to have sharp borders with a punched out appearance. Involvement of both the inner and outer tables results in a double-contour or bevelled edge appearance. The lesion usually causes asymmetric bevelling which can be palpated beneath the scalp. At times a button sequestrum may be present within the osteolytic lesion, representing residual bone. The sequestrum may be seen better on CT scans [2,4], and is considered to be diagnostic hallmark of eosinophilic granuloma.

Radiographic findings of eosinophilic granuloma in skull resemble venous lakes, arachnoid granulations, persistently enlarged parietal foramina (especially when unilateral). Epidermoid cysts or haemangiomas may produce lesions resembling eosinophilic granuloma in the skull [2].

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