CASE REPORT

Solitary Eosinophilic Granuloma in Left Parietal Skull : Case Report and Critical Review of Literature

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Introduction:

Langerhans cell histiocytosis is a term that encompasses a spectrum of clinical conditions ranging from a single, sometimes self limited osteolytic bone lesion to a fulminant, disseminated process that may be fatal. Three clinical subtypes of Langerhans cell histiocytosis are Hand-Schuller-Christian disease, Letterer-Siwe disease and eosinophilic granuloma. Farber recognized a histological similarity among these disparate conditions[1]. and Lichtenstein suggested that although certain differences existed between groups of patients, a reliable distinction could not be made in individual cases on the basis of histopathology alone.[2] Lichtenstein introduced the term histiocytosis X to denote a proliferation of histiocytes, with "X" to convey the uncertain histogenesis of the disease.

The aetiology of this disorder or disorders is unknown. Interestingly, it now appears that each type of Langerhans cell histiocytosis involves the proliferation of a clone of Langerhans cells.[3] suggesting that the disease may be a neoplastic disorder with a variable clinical course.

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The Langerhans cell is a distinct member of the mononuclear phagocytic system, [4] a system that includes tissue histiocytes and macrophages. Monostotic eosinophilic granuloma is the most common form of Langerhans cell histiocytosis. It occurs predominantly in children with 75 percent of cases occurring before the age of 20. The most common sites of involvement are the skull (usually the vault rather than the base), rib, femur, jaw, humerus and vertebral body. It has been estimated that the overall incidence of Langerhans cell histiocytosis is 0.6 case per million children less than 15 years old[5] and most these cases involve monostotic eosinophilic granuloma. A slight male predominance has been noted for monostotic eosinophilic granuloma and it rarely progresses to a disseminated form of diseases.

Case report:

A six years old boy presented with swelling over the left parietal region for two months with the history of trauma. Swelling gradually increased in size instead of reducing though initially treated by antibiotic. There was no history of fever but mother complained of trauma which was a coincident. On clinical examination, there was swelling over left posterior parietal region with minimum tenderness on palpation, no rise of local temperature but bony defect was felt around the

swelling. The patient was later subjected to radiological and hematological examinations. Haematological investigations were within normal limits. Anteroposterior and lateral skull radiographs showed lytic lesions.

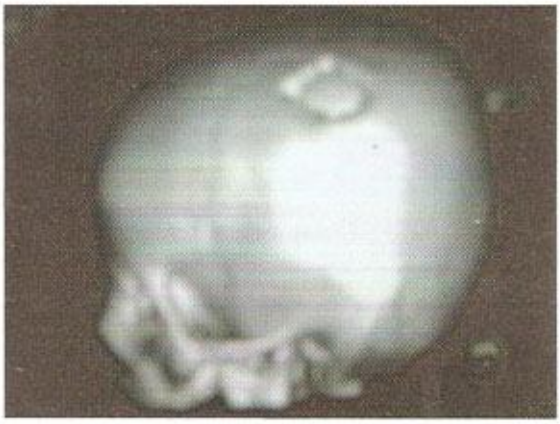

Figure-1: 3-D reconstruction of CT showed a bony defect

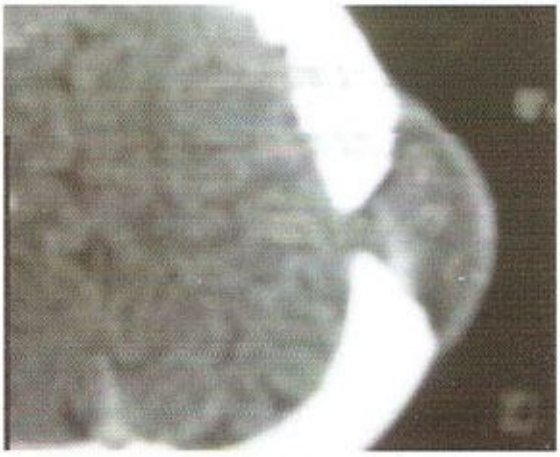

Figure-2: CT of axial section showed a isodense mass filling the bony defect

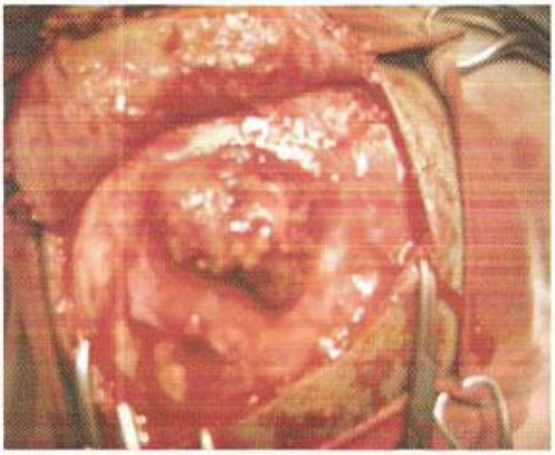

Figure-3: The lesion attached with dura

CT scan of head showed a bony defect with a isodense lesion. As history was confusing, it was planned for excisional biopsy.

A radical excision was done through a parietal craniotomy. The tumor was a gelatinous brownish mass with hematomas. It was protruding thought the bony defect and attached with outer surface of the dura. The lesion was excised totally and the dura was kept free. The bone fragment was replaced and the bone defect filled with bone dust. Excised tumour mass was sent for histological examination which showed many Langerhans cells, eosinophils, neutrophils and lymphocytes. There were necrotic elements too. Diagnosis of eosinophilic granuloma was made. The result of the maneuver was excellent and there were no complications. A new CT of the brain after six months showed no further lesion and the aesthetical result was excellent.

Discussion:

Solitary eosinophilic granuloma of the skull is a rare lesion, the natural history of which has not been defined completely. Langerhans cell histiocytosis (histiocytosis X) is a clonal proliferative disease[6]. Characteristically, parents of the patient noted an enlarging, mild tender posterior parietal skull mass following trauma during the several weeks before the initial assessment. Laboratory finding was normal. Radiographically, the abnormality consisted of a parietal osteolytic defect filled with soft tissue mass. Whole body bone scans or bone surveys used to rule out further lesions. The definitive diagnosis was made by histological examination, which showed the characteristic histiocytes, eosinophils, and multinucleated cells of eosinophilic granuloma. The treatment consisted of a craniotomy and excision of total mass followed by repeated follow-up at interval. The case was referred to paediatric haematooncologist and the patient completed a course of chemotherapy (vincristine). Six months follow-up period showed no recurrence clinically. Based on a review of patient and those reported in the literature, it is stress that a solitary eosinophilic granuloma may foreshadow future disease elsewhere and is not an easily dimissed local disease, especially in children. After the initial treatment, we recommended that careful long term follow-up and treatment of additional lesions with radiation therapy or chemotherapy should be done.

References:

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