

**CASE REPORT**

Solitary Eosinophilic Granuloma of Frontal Bone: A Rare Entity

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Abstract

Histiocytosis is a term applied to a group of rare disorders of the reticuloendothelial system. Langerhan cell histiocytosis (LCH) is characterized by the clonal proliferation of a special type of cell, the Langerhans cell. These are dendritic antigen-presenting cells that are normally distributed in many organs. LCH is presents as one of three clinicopathologic entities: acute disseminated Langerhan cell histiocytosis (Letterer-Siwe syndrome), unifocal eosinophilic granuloma, or multifocal eosinophilic granuloma. Unifocal lesions usually affect the skeletal system and may be asymptomatic, may cause pain and tenderness and in some instances, pathological fractures. This disorder is relatively indolent and may regress spontaneously or needs local excision or irradiation. We present a case of solitary eosinophilic granuloma of frontal bone.

Key Words

Eosinophilic Granuloma, Histiocytosis, Lytic Lesions

Introduction

Eosinophilic granuloma is one of the expressions of basic disorder of proliferation of immature dendritic cells known as langerhan cells, whereas Langerhan cell histiocytosis (LCH) refers to whole cluster of conditions resulting from it. In the past these disorders were referred to as histiocytosis -X and subdivided into three categories: Letterer-Siwe syndrome, Hand-Schüller-Christian disease and eosinophilic granuloma. These three conditions are now believed to represent different expressions of the same basic disorder. Eosinophilic granuloma is the most benign and localized of the three LCH entities and can be solitary or multiple. In most instances, these proliferations are monoclonal and therefore likely to be neoplastic (1). Proliferating Langerhans cells in these disorders do not resemble the normal dendritic cells, but instead have abundant, vacuolated cytoplasm with vesicular nuclei. This appearance is more like that of tissue macrophages (histiocytes), hence the term Langerhan cell histiocytosis. Conventional treatment of LCH is with surgery, radiotherapy, chemotherapy and steroid injections, alone or in combination. Spontaneous regression of localized disease has also been reported (2). We report a case of solitary eosinophilic granuloma of skull which was managed successfully with curettage only.

Case Report

A five year old boy presented to the emergency department of our hospital with moderately painful swelling on the right side of forehead with a gradual increase in size and frequent episodes of headache for 2 months. The swelling was 4x2 cms, globular, well circumscribed smooth, uniformly firm but fixed to the underlying frontal bone. Patient had a normal systemic examination. Radiographs of skull and CT-head (*Fig 1*) demonstrated frontal bone destruction co-incident with site of swelling. On surgery it was a lesion involving the diploic bone, indenting but not infiltrating the dura.

Histopathology of the lesion depicted cellular components of langerhan cells admixed with chronic cellular infiltrate and eosinophils (*Fig 2*). Findings were consistent with eosinophilic granuloma. In addition the routine base line investigations, bone marrow, bone scan and ultrasonography of abdomen were normal except for a raised ESR. Patient was managed with subtotal curettage only with no known recurrence till date.

Discussion

Eosinophilic granuloma is characterised by bone involvement only and it was Otani and Erblich who described the eosinophilic granuloma in children older than two years, with a characteristic bony site(3). Eosinophilic granuloma is actually a focal variety of

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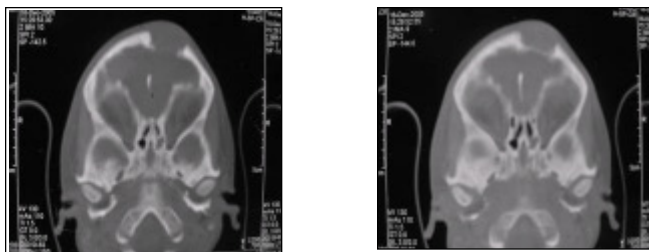


Fig 1. CT Scan of Skull with Evident Frontal Bone Osteolytic Defect Involving Both Tables

Langerhan cell histiocytosis (LCH). It occurs most often in children and young adults, as many as half the patients with solitary lesions are younger than 10 years (4). It can affect almost any bone, but commonly involves mandible. Solitary eosinophilic granuloma of skull is a rare condition, the natural history of which has not been defined completely. Characteristically the patient notices an enlarging tender skull mass for weeks to months; most commonly in parietal and frontal bone areas of skull (5). Our patient had involvement of frontal bone with a gradual increase in size of swelling.

Eosinophilic granuloma of any skull bone can be within diploe may or may not compress brain parenchyma without dural infiltration or it can be projecting into the cranial cavity compressing the brain parenchyma with dural infiltration (6). In our patient the lesion was indenting but not infiltrating the dura. Eosinophilic granuloma of orbit frontal bone usually involves the superior temporal quadrant and presents as rapidly progressive lid edema and erythema, bone pain and tenderness (7).

Like Letterer-Siwe syndrome and Hand-Schüller-Christian disease, langerhan cells of eosinophilic granuloma express HLA-DR, S-100 and CD1a. Unlike normal resident dendritic cells, it is the co-expression of CCR6 and CCR7 which allows the langerhan cells to migrate into the tissues that express the relevant chemokine-CCL20 in skin and bone (ligand for CCR6) and CCL19 and 21 in lymphoid organs (ligands for CCR7).

Langerhan cells can cause osteolysis by elaboration of interleukin-1 and prostaglandin-E2 (8) Histopathological examination of the lesion provides definitive diagnosis of the condition. The lesions cellular components include pathologic langerhan cells, chronic inflammatory cells and eosinophils (9) Radiograph and computed tomography shows extensive frontal bone destruction (7) In our patient CT scan of skull clearly demonstrates bone destruction.

Eosinophilic granuloma of orbital frontal bone has been reported to resolve after low dose irradiation (10) intralesional corticosteroids, (11) simple biopsy, (2) subtotal curettage with or without postoperative low dose radiotherapy or chemotherapy (12) and craniectomy with

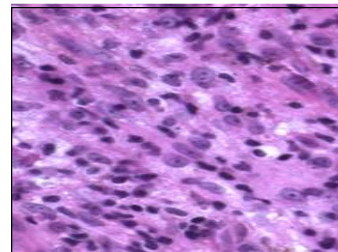


Fig 2. Characteristic Microscopic View of Cells with Vacuolated Cytoplasm

cranioplasty for lesions >4cm (6). Our patient underwent sub total curettage without any adjuvant therapy. Systemic evaluation and long term follow-up after any kind of treatment modality is mandatory in every case.

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