Solitary eosinophilic granuloma of the parietal bone in an adult patient

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ABSTRACT

Eosinophilic granuloma (EG) is a well-recognized benign form of Langerhans-cell histiocytosis. The estimated incidence of EG is 3-4 per million of the population. More patients are children and adolescents between the ages of 1-15 years. In this report, we present an adult patient. A 37-year-old male was admitted with headache and right parietal swelling that was present for 2 months together with epileptic attacks. Cranial CT scan revealed a right parietal osteolytic lesion with large epidural and subcutaneous mass, causing brain compression. Total removal of the mass was carried out with cranioplasty. Histopathological examination disclosed Langerhans' cell histiocytosis and immunohistochemical detection of S-100 antigen. At follow-up, he had no neurological deficits, and control CT was normal. For symptomatic solitary calvarial EG, surgical excision is the main treatment option. Surgical treatment is simple, quick, and allows histological diagnosis of the osteolytic lesions.

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Eosinophilic granuloma (EG) is a benign form of the 3 clinical variants of histiocytosis-X. It is an uncommon, localized granulomatous process, forming solitary or multiple lytic skeletal lesions, and most often presents in childhood. In general, these diseases are summarized under the term Langerhans-cell histiocytosis (LCH). The localized form of LCH, in which the disease is limited to bones, lymphatic nodes, or the lung, is commonly referred to as EG. Surgical excision, radiotherapy, and chemotherapy, either alone or in combination, are the main treatment options.[1-7] The objective of this report is to present a unifocal EG of the parietal bone in an adult patient, which is quite rare, and to review the current management.

Case Report. A 37-year-old male was referred to our clinic with headache and epileptic attacks with soft tissue swelling in the right parietal region that was present for 2 months. On physical examination, there was a solitary mass lesion in the right parietal region of the head with soft tissue swelling. The neurological examination was normal. Skeletal radiographic survey, bone scan, chest radiographs, complete blood cell count, liver function tests, coagulation studies, and electroencephalography were normal. An x-ray of the skull revealed a large, well-defined, irregular, marginated osteolytic lesion involving the right parietal bone (**Figures 1a & 1b**). A CT scan revealed an osteolytic lesion on the right parietal skull with a large epidural and subcutaneous mass (**Figures 2a & 2b**). Total removal of the mass was carried out with cranioplasty. Histopathological examination disclosed Langerhans' cell histiocytosis and immunohistochemical detection of S-100 antigen. The postoperative course was uneventful. The patient received an antiepileptic drug for 2 months (phenytoin 5mg/kg per day). At follow-up 4 years after the operation, he had no neurological deficits, and control CT scan was normal.

Discussion. Eosinophilic granuloma, the most benign form of LCH, accounts for approximately 70% of the cases of LCH and is most commonly found in the flat bones of the cranium, mandible, ribs, and pelvis.[1-9] Solitary cranial EG is thought to be the most common presentation of this disease, the cause of which is still unknown.[2] The estimated incidence of EG is 3-4 per million of the population.[4] The LCH affects all age groups, although it is more common in children. Large clinical series document that 50% or more patients are children and adolescents between the ages of 1-15 years.[6] The etiology and pathogenesis of LCH remains unknown. Analysis of DNA content in histiocyte infiltrates demonstrate both DNA diploidy and aneuploidy. Recent studies, based on molecular analyses, have demonstrated that Langerhans cell

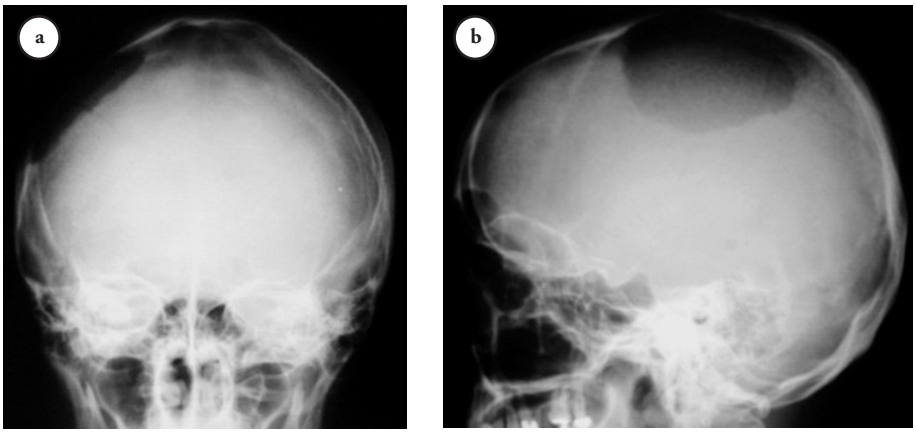

Figure 1 - Antero-posterior **a)** and lateral **b)** x-rays of the skull revealing a large, well-defined, irregular, marginated osteolytic lesion involving the right parietal bone.

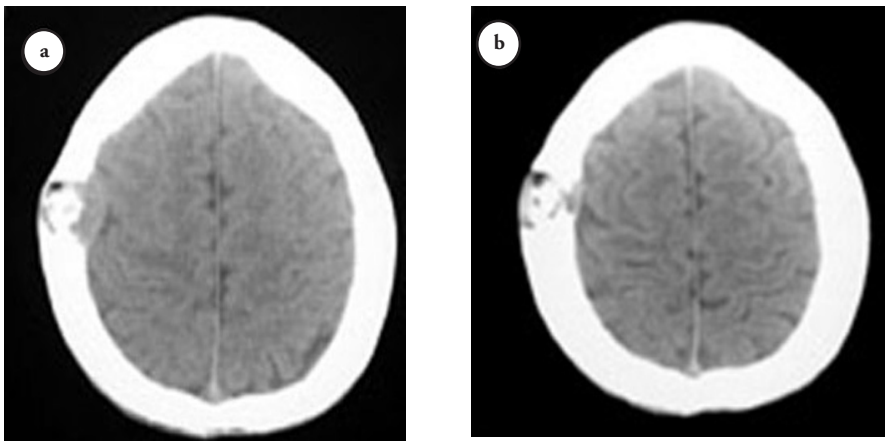

Figure 2 - Computerized tomography scans (**a, b**) revealing an osteolytic lesion on the right parietal skull with a large epidural and subcutaneous mass.

populations within LCH lesions are clonal in origin. These data strongly suggest that somatic genetic changes can contribute to the etiology and pathogenesis of LCH, as documented in neoplastic disorders.[6-8]

The hallmark of LCH is the proliferation and accumulation of a specific histiocyte: the Langerhans' cell. In bone this may cause pain and adjacent soft-tissue swelling and might cause epileptic attacks, as our case, but some lesions are asymptomatic. The LCH can involve any bone, but most lesions occur in the skull especially the calvarium and temporal bones. In the cranium, LCH usually presents as a fixed hemispherical mass.[2,6,9] The role of radiology in the assessment of EG of the bone was first described by Ochner.[2] Imaging diagnosis of the disease in bones is first based on the plain radiographic appearance, which is usually a central destructive, aggressive-looking lesion. In the skull, the lesions develop in the diploic space, are lytic, and their edges may be beveled, scalloped or confluent, or show a "button sequestrum."[1,9,10] The value of plain films is limited and the extent of the disease is better defined by CT scanning. Punched-out radiolucent defect without reactive sclerosis or periosteal thickening is usually detected, which is similar to various osteolytic lesions.[9] An MRI presents low intensity on T1-weighed image and high intensity on T2-weighed image with heterogeneous enhancement.[6,8-10] A CT is better suited for demonstrating bone detail, MR imaging for bone marrow and soft-tissue involvement and to assist with planning a biopsy or a surgical excision. Radionuclide bone scan can also be used to detect the bone involvement as well as to rule out recurrence or regrowth after surgery.[1,6-10] Biopsy of one of the lesions is necessary to confirm the diagnosis. Additional studies should include skeletal radiographic survey, bone scan, chest radiographs, complete blood cell count, liver function tests, coagulation studies, and measurement of urinary osmolarity after a supervised water deprivation and, if indicated, a test dose of vasopressin.[2,6]

Treatment for LCH is usually reserved for patients with symptomatic lesions. Single bone lesions are treated with curettage.[1-9] However, in very rare instances, the spontaneous healing of EG has been reported in recent literature.[11,12] Recurrent lesions are best managed with irradiation or short courses of systemic chemotherapy. More extensive disease involvement requires systemic chemotherapy and, in some cases, marrow transplantation.[1-9] The prognosis for LCH is uncertain. Young age and signs of organ dysfunction predict a poor prognosis.[6] The prognosis of unifocal EG is very good, with successful disease control being achieved in 95% of cases.[3]

In conclusion, for symptomatic solitary calvarial EG, surgical excision is the main treatment option. Surgical treatment is simple, quick, and allows histological diagnosis of the osteolytic lesions.

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