Neurocirugía
2008 19: 453-455

Solitary Langerhans cell histiocytosis orbital lesion: case report and review of the literature

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Summary

Solitary eosinophilic granuloma is a rather benign and localized form of Langerhans's cell histiocytosis. Definitive diagnosis is made by histopathology including immunohistochemical detection of S-100, HLA-DR and CD1a antigens. We report the case of a twenty-five year old boy presented with headache and orbit's pain. A CT scan showed a left supero-lateral orbital mass with evidence of bone erosion. The different options of treatment are discussed and the literature is reviewed.

KEY WORDS: Eosinophilic granuloma. Histiocytosis X. Birbeck granules.

Granuloma eosinófilo de la orbita: caso clínico y revisión de la bibliografía

Resumen

El granuloma solitario eosinófilo es un tumor normalmente benigno y es una forma localizada de la hisitiocitosis de las celulas de Langerhans. La diagnosis definitiva viene de un estudio histopaológico incluida la búsqueda con métodos inmunohistoquímicos de S-100, HLA-DR y de los antígenos CD1a. Proponemos el caso de un joven de venticinco años que presentaba dolor de cabeza y dolor de una órbita. Una TAC muestra una masa en sede orbital supra-lateral izquierda con una evidencia de erosión del hueso. Se discuten las diferentes elecciones de tratamiento y se revisa la bibliografia.

PALABRAS CLAVE: Granuloma eosinófilo. Histiocitosis X. Granule de Birbeck.

Introduction

Histiocytosis X is a disease complex that includes Letterer-Siwe disease, Hand-Schuller-Christian disease and eosinophilic granuloma. Its total incidence is estimated to be 1 in 2 million[1]. Eosinophilic granuloma, the most benign and localized form of Langerhans's cells-histiocytosis, may be solitary or multiple primarily interesting the skull and the facial bones. It is estimated that it occurs in 75% of the cases in children and young adults under 25 years and prevalently males[2,7]. Its etiopathogenesis is still unknown, although it's believed to be a disorder reactive to an infectious, probably viral agent, or an altered regulation of the reticular system with an excessive proliferation of Langerhans cells. Definitive diagnosis of eosinophilic granuloma is based on the histological appearance, which is characterized by a polymorphous, confluent or perivasculary accentuated infiltrate consisting of histiocytes, eosinophilic granulocytes, lymphocytes, and finally the ultrastructural demonstration of Birbeck granules in the proliferating histiocytes[3]. We describe a patient with a solitary granuloma in the orbit's region who had total surgical removal of the lesion followed by any other medical treatment. The literature is reviewed concerning the clinical course of unifocal lesion of histiocytosis X.

Recibido: 12-06-07. Aceptado: 01-02-08

Case report

A twenty-five year old boy presented with complains of headache and orbit's pain on the left side for 2 months. Neurological examination was unremarkable, haematological and biochemical parameters were within the normal limits. Computed Tomography scan showed an osteolytic lesion to level of the external portion of the orbital roof (Fig. 1). Therefore we have decided to resort to the surgery intervention with bicoronal skin incision behind the hairline and curettage of the lesion known. Surgically removed fragments of the lesion showed extensive reparative phenomena of the bone, with fibrosis and a myofibroblastic proliferation simulating a soft tissue tumour. There were interspersed foci of inflammatory cells, sometimes organized in granulomas, constituted by a mixed population of eosinophils, mononucleated and with multinucleated cells (Fig.2). Besides smooth-actin-positive areas of proliferating myofibroblasts, immunohistochemistry documented

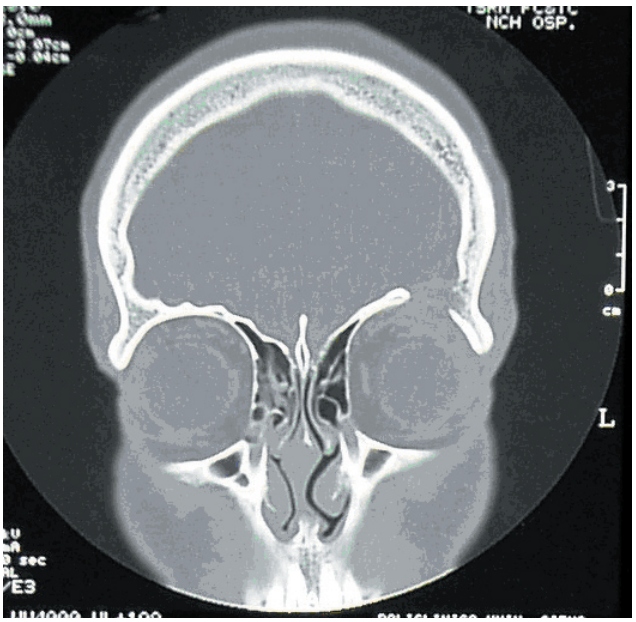

Figure 1. *Coronal CT scan shows the osteolytic area.*

S-100 and CD1a positive cells consistent with Langerhans cells inside the granulomatous foci. A diagnosis of Langerhans cell histiocytosis was performed. Postoperatively the patient experienced no complications; bone scintigraphy and other investigations for thoracic, visceral or vascular manifestation of systemic histiocytosis X disease was done and with negative results. Any other treatment, for the oneness of the lesion, was decided. Follow-up 6 months after curettage of the lesions showed no recurrence or evidence of systemic involvement.

Discussion

Histiocytosis X is a disease complex that includes Letter-Siwe disease, Hand-Schuller-Christian disease and eosinophilic granuloma (EG). The current classification is based only on the number of organs involved and on the number of sites affected in each organ: unifocal eosinophilic granuloma (involving a single organ and a single, generally bony, site), multifocal eosinophilic granuloma (involving multiple sites in a single organ) and acute

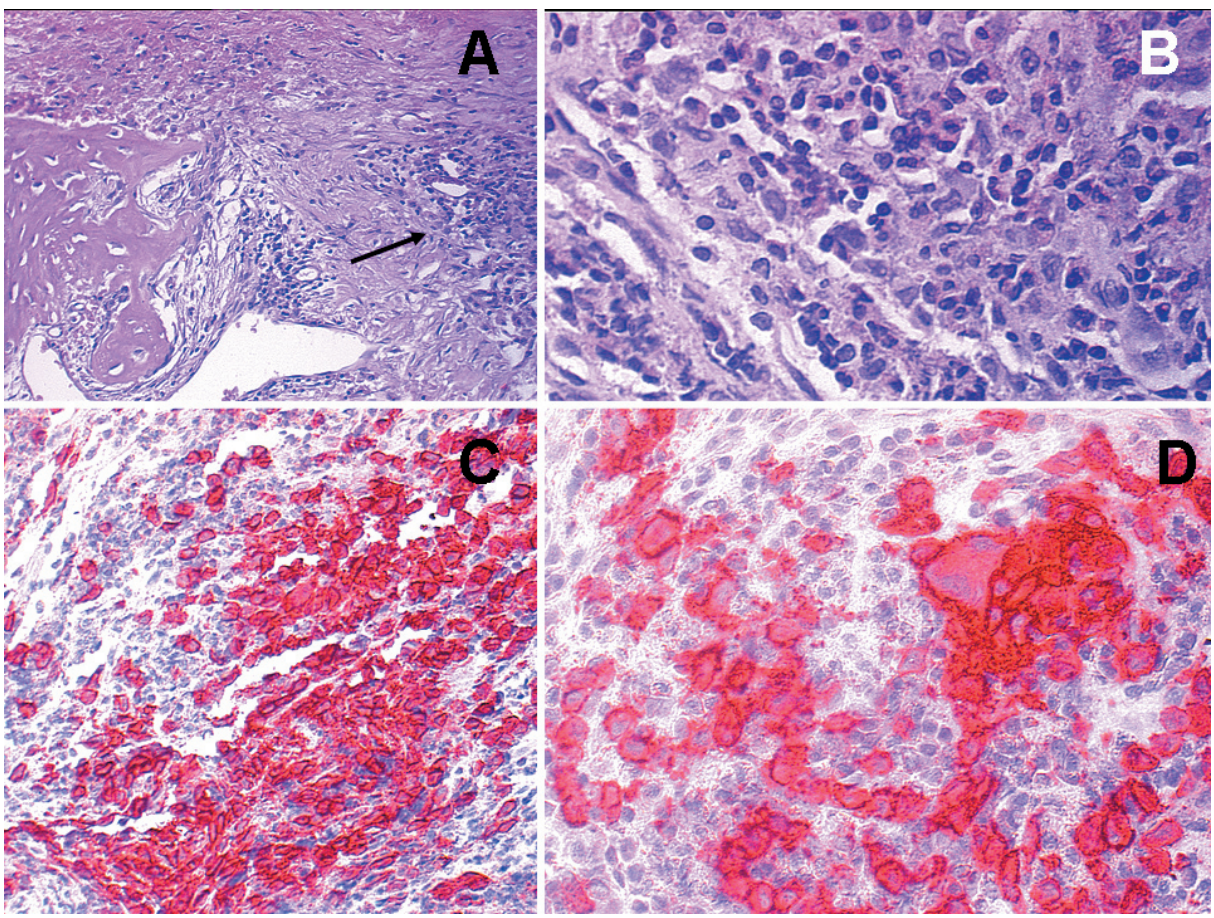

Figure 2. *Bone partly replaced by fibrous tissue, in which granulomas (arrow) are recognizable. Detail of a granuloma (B). S-100 (C), and CD1a (D) positivity of Langerhans cells. (Haematoxylin and eosin, original magnification: A, x 50; B, x 200 Immunohistochemistry, Streptavidin-Biotin method: C, x 100; D, x 400).*

disseminated histiocytosis (involving multiple organs). Although most reported cases describe the orbital frontal bone as the site of origin in the orbit[4], our cases demonstrated that unifocal eosinophilic granuloma may occur in the lateral wall of the orbit.

The etiology of LCH remains unknown, and it has been considered either as a reactive disorder, or a neoplastic process, and an aberrant immune response. Today there is evidence that it is a clonal proliferative disorder of Langerhans cells. Imaging diagnosis of the disease in bone 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 bevelled, scalloped or confluent (geographic), or show a "button sequestrum." There are many accepted forms of treatment for eosinophilic granuloma: these include surgical resection, local irradiation, systemic and local corticoids and systemic chemotherapy[5,8].The prognosis of the unifocal eosinophilic granuloma is usually favourable with control of disease being achieved in about 95% of cases. Therefore a watchful conservative approach is recommended by some authors for selected cases[6]. Patients with a unifocal symptomatic or multifocal progressive disease require treatment. We think the treatment of choice is the complete surgical excision of the lesion and whenever only a partial resection is possible, radiotherapy plays a significative role. Systemic therapy is instead required when soft tissues are involved and in cases of multiple skeleton localisations. Some 50% of patients requiring systemic therapy respond to steroid therapy alone. Chemotherapy seems to be necessary only in patients with systemic disease or multiple organ involvement that do not respond to radiotherapy and the most commonly used drugs, either individually or combined, are vinblastine,methotrexate, chlorambucil and 6-mercaptopurine. The recurrence rate of eosinophilic granuloma after the treatment modalities ranges from 1, 6% to 25%, depending largely on the location and extension of the lesions. The decision for treatment is further complicated by the well documented potential of spontaneous healing of EG.

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Ulivieri S.; Oliveri G.; Filosomi, G.: Solitary Langerhans cell histiocytosis orbital lesion: case report and review of the literature. Neurocirugía 2008; 19: 453-455.

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