Isolated Primary Langerhans' Cell Histiocytosis of Central Nervous System

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Summary

Solitary eosinophilic granuloma that involves the CNS is an uncommon lesion and most frequently affects the hypothalamus. We report a new and rare case of solitary eosinophilic granuloma of the left temporal lobe in a patient without systemic disease. The diagnosis was confirmed by electron microscopy and immunochemical techniques. The treatment of choice is surgical resection. There is a better prognosis in comparison with cases accompanied by systemic disease.

KEY WORDS: Eosinophilic granuloma. Langerhans'cell histiocytosis. Temporal lobe. Histiocytosis "X".

Histiocitosis primaria aislada de las células de Langerhans, del SNC

Resumen

El granuloma eosinófilo aislado del SNC es una entidad rara, siendo su localización más frecuente el hipotálamo. Presentamos un nuevo caso de histiocitosis aislada del lóbulo temporal en un paciente sin afectación sistémica. El diagnóstico se ha confirmado tanto por microscopía electrónica como con las técnicas inmu-no-histoquímicas. El tratamiento de elección es la resección quirúrgica radical, siendo el pronóstico en estos casos es mejor cuando están en el contexto de enfermedad sistémica.

PALABRAS CLAVE: Granuloma eosinófilo. Histiocitosis de células Langerhans. Lóbulo temporal. Histiocitosis "X".

Introduction

It is more than 44 years since Lichtenstein grouped eosinophilic granuloma (EG) of bone, Letterer-Siwie disease and Hand-Schuller-Christian syndrome under the term "histiocytosis X"1, now more correctly denominated Langerhans' cell histiocytosis (LCH). Involvement of the central nervous system (CNS) is frequently observed in LCH either by contiguous spreading from osseous foci or limited to the hypothalamus or posterior pituitary gland. In contrast, isolated LCH of a cerebral hemisphere without evidence of systemic disease is rare, with only a few cases previously reported1,5,7,8. We report a new-case.

Case report

A 36-year-old male with no relevant history (no history of diabetes insipidus, exophthalmos, otitis media, or lymphadenopathy) was admitted to our department in June 1997 for an episode of motor aphasia two weeks earlier. Physical examination was normal and no skin or genital lesions, lymphadenopathy or hepatosplenomegaly, fever or weight loss were noted.

The analytical results were normal. CT scans taken on the same day at the referring hospital showed a left temporal mass with surrounding hypodensity that was enhanced with contrast. (Fig. 1.a). Magnetic resonance imaging (MRI) demonstrated an intraaxial lesion in the left temporal lobe that was intensely enhanced with gadolinium (Fig. 1.b, c). The hypothalamus region appeared normal on all CT and MR scans. Skull and chest X-ray films were normal.

Left temporoparietal craniotomy was performed under general anaesthesia after stereotactic localization, revealing a white-greyish mass of firm consistency that was dissected with ease. The excision was macroscopically complete.

In the postoperative period, the patient continued to suffer episodes of motor dysphasia, which were treated with phenytoin and a 10-day regimen of dexamethasone. The patient is now symptom-free. The histological study with eosin and haematoxylin staining revealed various types of inflammatory cells such as histiocytes, eosinophils and lymphocytes. Immunohistochemical analysis showed strong S-100 protein and vimentin immunoreactivity and negativity for glial fibrillary acid protein. The histiocyte cell population showed positivity for HLA-CD. Electronic microscopic study disclosed a trilaminar rod-shaped structure characteristic of Birbeck granules.

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Discussion

LCH is a disease complex that includes Letterer-Siwe disease, Hand-Schuller-Christian disease and eosinophilic granuloma. Bone is the most commonly affected tissue, although skin, liver, spleen, and the reticuloendothelial system can also be involved. The most common intracranial sites of involvement are the hypothalamus and pituitary. To our knowledge, there have been very few reports of a solitary eosinophilic granuloma in the cerebral hemisphere.

According to international criteria, LCH is demonstrated either by ultrastructural detection of Birbeck granules or immunohistochemical evidence of CD1 expression. Our case fulfills the histological criteria. The histochemical and ultrastructural criteria aid in the differential diagnosis of LCH versus other histocyte-derived lesions.

The CT and MRI images of LCH are not distinctive enough to have diagnostic value.

The origin of LCH is unclear but it has been proposed as an immunological disorder. Most previously reported cases of localized forms of LCH appeared to respond well to surgery alone and only a few cases received additional radiotherapy. In the present case, the patient remains symptom-free after a two-year follow-up and has required no additional treatment, confirming complete surgical resection as the treatment of choice for isolated primary LCH of the CNS.

References


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