Characterization of langerhans cell histiocytosis: analysis of a case series.

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Introduction

Langerhans cell histiocytosis (LCH) has an incidence of 5 x 1,000,000 inhabitants. Its diagnosis is challenging due to its nonspecific clinical manifestations. Bone lesions are the most frequent manifestation, occurring in 75%. Although any bone can be affected, the most frequent locations include the skull, long bones, ribs, pelvis and vertebrae. Otologic involvement occurs in a range of 15-61% of cases, and may manifest as otitis, cerebellar syndrome, facial paralysis and/or hypoacusis. Treatment varies according to its extent. Systemic therapy with corticosteroids and cytostatic agents for at least one year has improved the prognosis of multisystem LCH and is the standard of care.

Objetive

To describe 6 cases of Langerhans cell histiocytosis in pediatric patients, addressing its clinical presentation, evolution and treatment.

Methods and Materials

Review of clinical records of the Instituto de Neurocirugía Dr. Asenjo and Hospital Dr. Luis Calvo Mackenna (2001-2024)

Results

Six cases were reported during the study period, with an equal distribution between females and males. The mean age of the patients was 8 years, with the youngest patient being 3 years old and the oldest being 17 years old at the time of diagnosis. Reasons for consultation varied and included ataxia, facial palsy, headache, otalgia, retroauricular pain, and abducens nerve palsy. The most frequent location of the lesions was the mastoid 33.3% (Figure 1- 4), followed by the sphenoid 16.67%; the petroclival 16.67%; the petrous apex 16.67% and the cranial base 16.67%. Only one patient had a lytic lesion in a secondary location, identified by radiography at the distal end of the humerus (Figure 5). An incisional biopsy was performed in all patients, with 33.33% of biopsies performed by transmastoid approach, 50% by transsphenoidal approach and 16.67% by retrolabyrinthine approach. In one of the cases, intratumoral corticosteroids were administered concomitant to the biopsy. Deferred biopsy allowed the diagnosis of LCH in 100% of patients. Treatment consisted of chemotherapy in 50% of cases, indomethacin in 33.3%, and intratumoral administration of corticosteroids in a second surgical time in 16.67%.

Figures

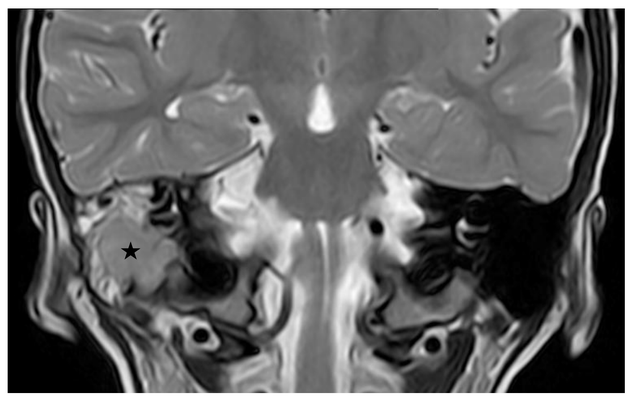

Figure Nº 1. (★) *Right mastoid lesion in T2 sequence in MRI.*

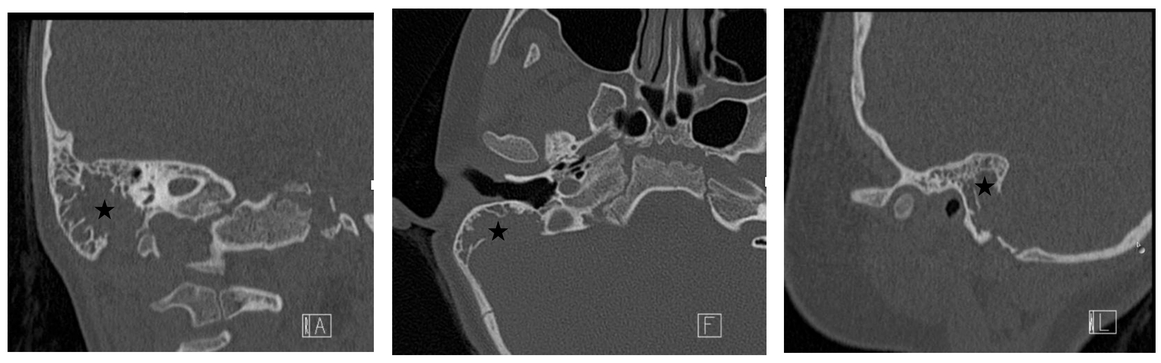

Figure Nº 2, 3 y 4. *Lesion (★) with soft tissue density not detected with contrast medium centered in the right mastoid with destruction of the intercellular septa and the internal table, with extra-axial extension.*

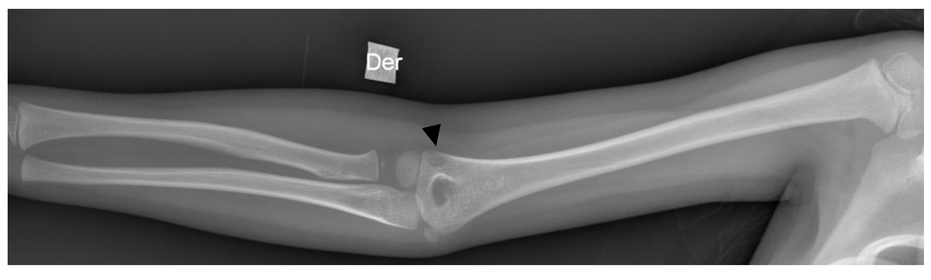

Figure Nº 5. *Lithic lesion (◥) in distal humerus metaphysis.*

The otorhinolaryngologic presentation includes otitis media, cerebellar syndrome, facial paralysis or hypoacusis. It is recommended to perform a complete laboratory and imaging study with computed tomography, magnetic resonance imaging, abdominal ultrasound and PET CT. The definitive diagnosis is made by histology and immunohistochemistry (S-100, CD1a and CD207). Treatment must be individualized, among the alternatives are: expectant management, surgical resolution, radiotherapy, chemotherapy and systemic corticosteroids. Radical resection may be indicated in bone lesions < 2 cm.

Discussion

LCH is a clonal myeloproliferative disease with a variety of clinical presentations and prognosis that originates in immature Langerhans cells. It is associated with genetic changes due to mutation of the BRAF V600E gene.
It affects especially children under 15 years of age. It is predominantly male. The differential diagnosis is broad, complex and requires the exclusion of other malignant or infectious diseases.
The most frequent bony localization corresponds to the skull.

Conclusions

There are no previous descriptions of its incidence in the Chilean literature, so this study provides an approximation to the national reality. Further research including other health centers in the country is required to estimate more accurately the incidence of this pathology in the Chilean population.

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