ISOLATED CONJUNCTIVAL INVOLVEMENT IN LANGERHANS CELL HISTIOCYTOSIS

HISTIOCITOSIS DE CÉLULAS DE LANGERHANS CON AFECTACIÓN CONJUNTIVAL AISLADA

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ABSTRACT

Case report: A 43-year-old woman referred for a hyperemic node in the inferior conjunctival fornix. No improvement was observed after 10 days of antibiotic and anti-inflammatory topical treatment, so biopsy-extirpation was performed. Histopathological and immunohistochemical findings suggested the diagnosis of Langerhans cell histiocytosis (LCH). No findings of extraocular manifestations were shown after an intensive clinical and analytical investigation.

Discussion: LCH is a disease with different organ manifestations whose diagnosis depends on histological findings. We present a patient with isolated conjunctival affection, which to our knowledge is the third case published in the literature (Arch Soc Esp Oftalmol 2009; 84: 217-220).

Key words: Histiocytosis, Langerhans cell, conjunctival node, ocular manifestations.

RESUMEN

Caso clínico: Mujer de 43 años que consulta por un nódulo hiperémico en fondo de saco conjuntival inferior. Tras diez días de tratamiento antibiótico y antiinflamatorio tópico no se observa mejoría, por lo que se realiza una biopsia-extirpación. El resultado anatopatológico e inmunohistoquímico es informado como histiocitosis de células de Langerhans (HCL), procediéndose a realizar un estudio sistemico con diferentes pruebas complementarias, sin hallar afectación a otro nivel.

Discusión: La histiocitosis de células de Langerhans es una enfermedad de afectación muy variada, cuyo diagnóstico se establece mediante hallazgos histológicos. Presentamos un caso de implicación exclusivamente conjuntival, habiéndose publicado sólo tres casos previamente en la literatura médica consultada.

Palabras clave: Histiocitosis, células de Langerhans, nódulo conjuntival, manifestación ocular.
INTRODUCTION

Histiocytoses make up a group of infrequent diseases which affect patients of any age group and with highly variable clinical expressions. According to the latest proposed classification (1), Group 1 includes Langerhans cell histiocytosis (LCH), a term which comprises the diseases classically known as histiocytosis X. Group 2 comprises the non-Langerhans cell histiocytosis, while Group 3 comprises malign histiocytoses. Langerhans cells are a specialized type of macrophages originating in the bone marrow. It has been postulated that the pathological core of LCH is clonal proliferation and the accumulation of this type of cells in various organs (1).

Almost 20 years ago, the Histiocyte Society was established to improve the management of these patients. This society has published three successive versions with updated diagnostic, therapeutic and follow up protocols. The latest version of the LCH III protocol, published in 2001, establishes 4 groups of patients: those who exhibit localized involvement of only one organ, those having multifocal bone disease and those having multi-systemic involvement, differentiating in this group those having vitally important organs affected (central nervous system, hematopoietic system, lungs, liver and spleen) and those who don’t.

The definitive diagnostic of this pathology can only be obtained by a histological study of an affected organ. We present an LCH case with exclusively conjunctival involvement.

CASE REPORT

A 43-year-old woman without relevant history who attended the emergency services due to the presence of a hyperemic nodule in the inferior conjunctival sac (fig. 1). The rest of the ophthalmological exploration gave normal results. Suspecting an inflammatory or infectious process, treatment with terramycine and hydrocortisone in ophthalmic cream was prescribed for 10 days. In the absence of improvements, the nodule was removed. The anatomic-pathologic study revealed a fragment of conjunctival mucosa with an accumulation in the middle area of cells with a broad cytoplasm and slit nuclei, accompanied by eosinophiles and lymphocytes (fig. 2). The sales having the largest sizes showed positive for S-100, CD1a (figs. 3 and 4) and negative for HMB45 antibodies and cytokeratines. These findings were compatible with Langerhans cell histiocytosis.

After the histological diagnostic, a systemic study was performed which included hemogram without cytopeniae, normal kidney and liver functions, absence of thyroid alteration, chest X-ray with slight alterations obtained via high resolution CAT as a pre-tracheal adenopathy smaller than 1 cm, lack-luster glass images in apical segment of right inferior lobe and one nodular lesion in the left inferior lobe. A bronchoscopy with biopsy was made without finding anything related to LCH. The bone series and the SNC imaging tests discarded involvement at that level.
The patient was assessed by the Dermatology Service due to exhibiting lesions in palatine region mucosa which, after a biopsy, were identified as a melanotic hyper-pigmentation which did not suggest histiocytosis.

The check-ups made in the three following years after the excision of the conjunctival node did not find any systemic involvements related to LCH or relapse at the local level.

**DISCUSSION**

The most frequent ophthalmological expression of Langerhans cell histiocytosis is proptosis secondary to the involvement of the orbital bone. The infiltration of other ocular regions is rare (4).

Within the localized forms of LCH, the most frequent involvement referred to the bones with lytic lesions in various locations. Other organs which are affected in an isolated manner include the skin and the lymphatic glands. The exclusively conjunctival involvement is an extremely rare presentation of this disease (4). The medical literature refers only two additional cases in adult patients (3,5).

Saxena et al (3) published the case of a 26 year old patient with a solitary limbar nodule without additional pathology. The diagnostic was made similarly to our diagnostic. Melamud et al (5) described a case with isolated epibulbar involvement without systemic involvement, and finally Monos et al (2) documented a case of congenital histiocytosis in a newborn infant involving the palpebral conjunctiva without other expressions.

The definitive diagnostic of LCH is made with a histopathological study of the injury, with the identification of an infiltrate made up of histiocytary cells together with lymphocytes and eosinophiles being characteristic. Langerhans cells express various markers which, by means of immunohistochemical dyes and electronic microscopy, will lead to a provisional diagnostic (S-100, ATP-ase, α-D-manosidase, soya lecithin) and a definitive diagnostic (CD1a, Birbeck granules) (2). Due to technical reasons, it was not possible to verify in our patient the presence of Birbeck granules, but the positive results for CD1a rendered the diagnostic final.

Once the diagnostic has been reached, it is necessary to carry out a complete protocol-based study to identify the possible involvement of other organs, as this would modify the treatment as well as the prognosis for the patient. In the majority of isolated involvements, the usual procedure is excision or local treatment without requiring systemic chemotherapy (3). However, the latter becomes essential in multi-systemic diseases with different protocolary indications.

The course of the disease is unforeseeable, ranging from spontaneous regression to a rapid and fatal progression. Recurrences and long-term sequels are frequent. For this reason, a long-term follow-up of these patients is essential even when the disease is considered to be under control (2).

Even though it is exceptional, Langerhans cell histiocytosis must be included in the differential
diagnostic of conjunctival nodular injuries and the diagnostic as well as the subsequent management must be multidisciplinary.

REFERENCES


