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Short communication

A case report of orbital Langerhans cell histiocytosis presenting as a orbital cellulitis☆

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ABSTRACT

Clinical case: A 10-year-old girl was seen with a 3-week history of right upper lid swelling and with no other symptoms or fever. There was no recent history of sinusitis, trauma, or previous infection involving the periorbital area, or response to oral antibiotic treatment. Orbital computed tomography showed a lesion involving the upper margin of the orbit, and bone destruction at the orbital roof. Biopsy performed revealed the presence of Langerhans cell histiocytosis. The lesion was surgically debulked and corticosteroids were used intraoperatively. The lesion responded to treatment.

Discussion: The orbital involvement of Langerhans cell histiocytosis, despite its low incidence, should be considered in the examination of acute peri-orbital swelling. It usually presents as an osteolytic lesion, and it is confirmed with a histological examination and immunohistochemical techniques for CD1a and S100. An interdisciplinary approach is recommended to rule out multifocal or multisystemic diseases, as well as to develop an appropriate treatment strategy.

Caso clínico de histiocitosis de células de Langerhans presentándose como una celulitis orbitaria

Palabras clave:
Histiocitosis de células de Langerhans
Granuloma eosinofílico
Masa orbitaria
Celulitis orbitaria

RESUMEN

Caso clínico: Paciente de 10 años de edad, con inflamación palpebral superior derecha, sin malestar general ni fiebre, de 3 semanas de evolución y sin respuesta a tratamiento antibiótico oral. La tomografía computarizada orbitaria reveló una lesión de tejidos blandos en el margen superior de la órbita extraconal con destrucción ósea del techo orbitario. El estudio anatomopatológico fue compatible con histiocitosis de células de Langerhans. Tras descartar afectación sistémica se realizó curetaje de la lesión orbitaria e infiltración de corticoides intraoperatoriamente con buena respuesta.

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Discusión: La afectación orbitaria de la histiocitosis de células de Langerhans, a pesar de su baja incidencia, debe tenerse en cuenta en el diagnóstico diferencial de un cuadro inflamatorio orbitario. Suele presentarse como una lesión osteolítica y se confirma con examen histológico y técnicas de inmunohistoquímicas positivas para CD1a y S100. Se recomienda un enfoque interdisciplinario para descartar afectación sistémica y desarrollar una estrategia de tratamiento apropiada.

Langerhans cell histiocytosis (LCH) comprises a clinic range of diseases of unknown cause characterized by anomalous clonal proliferation of Langerhans cells.[1] Orbital compromise by LCH usually presents in isolation in the pediatric population as a soft tissue lesion and bone compromise of osteolytic appearance.[2] In some cases, it expresses as periorbital acute edematization, suggesting periorbital growth giving rise to rapid inflammatory response. For this reason it could be easily confused with an infectious process of the cellulitis type, rupture of a dermoid cyst or orbital inflammatory disease.[3]

Clinical case report

The clinical case of a 10-year-old girl is presented, referred for assessment of upper right palpebral inflammation with 3 months evolution coinciding with a trip to the USA, without history of upper respiratory infection, fever or general discomfort. She had been treated with oral antibiotics for probable orbital cellulitis, initially with amoxicillin and subsequently, due to poor response, with cefuroxime without improvement. Ophthalmological examination produced mechanical blepharoptosis with 2 mm of palpebral opening due to the presence of a soft, inflammatory-like lesion in the central portion of the upper right eyelid, with slight pain upon touch and slight inferior displacement of the right ocular globe. Visual acuity was of one in both eyes and the rest of the examination was normal (Fig. 1).

Orbital computerized tomography (CT) was requested, which showed lesion in the soft parts of the upper margin of the right extraconal orbit that caused bone destruction at the level of the orbital roof as well as downwards ocular globe displacement (Fig. 2). Due to the nonspecific radiological result that did not discard possible inflammatory-infectious origin or neoplasic processes, it was decided to conduct an orbitotomy through transcutaneous approach at the level of the upper palpebral cutaneous fold under general anesthesia. Intrasurgery findings included a brownish granulomatous gel-like content in the most anterior part of the lesion which could be aspired and subsequently the mass was more elastic towards the orbital roof. Samples were taken for pathological anatomy and microbiology. Microbiology result was negative, but the histopathological analysis reported a proliferation of Langerhans cell groups together with predominantly eosinophilic inflammatory infiltrate that also contained histiocytes, some neutrophiles, lymphocytes and occasionally giant cells. The immunohistochemistry study was positive for CD1a and S100 at the level of the Langerhans cells.

With the certain diagnostic of orbital LCH, the patient was referred to the Pediatric Oncology department for extension study. Extraorbital compromise was discarded. As the lesion was localized only at the orbital level, it was decided to respect

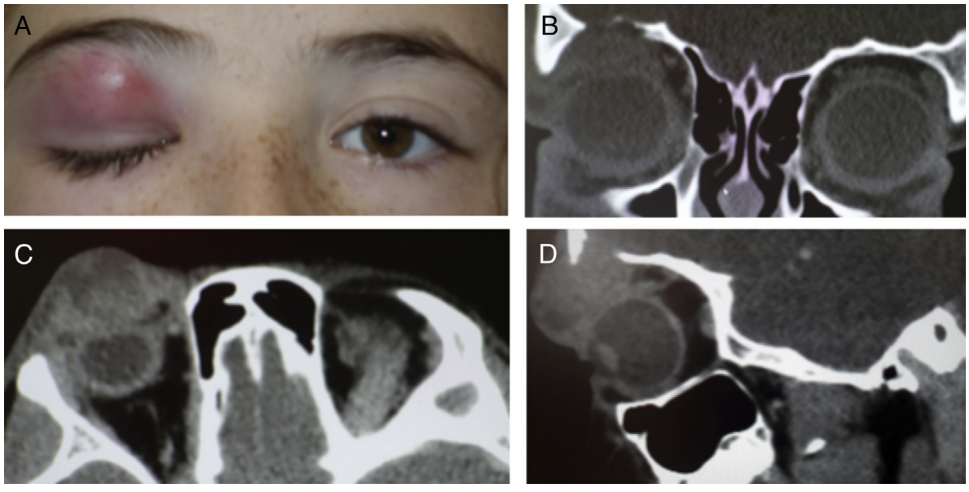

Fig. 1 – Presurgery clinic findings. (A) Clinic photography of the patient with mechanical blepharoptosis caused by soft, inflammatory-like lesion in the upper eyelid of the right eye. (B) Coronal CT image showing the lesion in the right orbital region producing bone destruction in the orbital roof. (C and D) Axial and sagittal image, respectively, of CT showing extraconal heterogeneous lesion.