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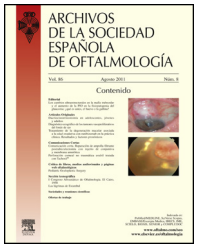

Short communication

Necrobiotic xanthogranuloma. Differential diagnosis, treatment and systemic involvement. Case report☆

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ABSTRACT

Case report: A 48-year-old male was referred to our hospital for further evaluation of eyelid edema with bilateral yellowish ulcerated nodules. Suspecting a xanthogranulomatosis, imaging tests and biopsy were performed with diagnosis of necrobiotic xanthogranuloma. IgG monoclonal gammapathy was diagnosed in a systemic study. Systemic corticosteroids and cyclosporine were initiated unsuccessfully; therefore, intra-lesional injections of triamcinolone were started, which controlled the orbital disease.

Discussion: Necrobiotic xanthogranuloma is a rare condition that usually affects the eyelids and anterior orbit. Its diagnosis is important as it is associated with malignant lymphoproliferative processes. Intra-lesional corticosteroids were effective in our patient.

Xantogranuloma necrobiótico. Diagnóstico diferencial, tratamiento e implicaciones sistémicas. A propósito de un caso

Palabras clave:
Xantogranuloma
Xantogranuloma necrobiótico
Xantogranulomatosis
Corticoides
Gammapatía monoclonal

RESUMEN

Caso clínico: Varón de 48 años remitido por edema palpebral con nódulos ulcerados amarillentos en ambos párpados superiores. Las técnicas de imagen demostraron infiltración en ambas órbitas anteriores que fueron biopsiadas con el resultado de xantogranuloma necrobiótico. En el estudio sistémico se halló una gammapatía monoclonal IgG no conocida. Se inició tratamiento con corticoides y ciclosporina orales sin éxito, por lo que se pautaron inyecciones intralesionales de triamcinolona que controlaron la enfermedad orbitaria.

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Discusión: El xantogranuloma necrobiótico es una rara entidad que suele afectar a párpados y órbita anterior. Su diagnóstico es importante porque se asocia a procesos linfoproliferativos malignos. Los corticoides intralesionales fueron efectivos en su manejo.

Introduction

There are 4 types of orbital xanthogranulomae: necrobiotic xanthogranuloma (NBX), adult onset xanthogranuloma (AOX), asthma-associated adult periocular xanthogranuloma (AAPOX) and the Erdheim–Chester disease (ECD).

AOX affects the eyelids and the anterior orbit without systemic involvement.[1,2] AAPOX exhibits periocular and anterior orbit lesions, typically associating asthma, benign lymphadenopathy and IgG polyclonal paraproteinemia IgG.[2]

ECD is the most severe form, which is lethal between 3 months and 15 years despite different therapies and is characterized by medium and posterior orbital xanthogranulomatous fibrosclerosis with proptosis and progressive visual acuity loss,[1,3] together with fibrosclerosis of long bone metaphysis and of deep organs.[1,3]

NBX is characterized by bilateral palpebral, orbital and periocular lesions, including the conjunctiva, which can infiltrate other bodily areas and deep organs. Said lesions tend to greater ulceration and fibrosis vis-à-vis the previous subtypes.[1,4] Systemic involvement is frequently associated to monoclonal gammopathy and malign lymph proliferative processes.

A necrobiotic xanthogranuloma case is presented with the systemic involvement and good response to intralesional corticoid treatment.

Clinical case

A male, 48, referred to our hospital due to bilateral palpebral tumors of unknown origin with recurring inflammation episodes (Fig. 1). The patient history included smoking habit (20 cigarettes per day) with a diagnosis of idiopathic thrombocytopenic purpura.

Upon exploration the patient exhibited bilateral yellowish injuries in upper eyelids, aponeurotic ptosis and increased intraocular pressure (IOP) (27/26 mmHg) in both eyes, in combined treatment with cusimolol and dorzolamide (Fig. 1); in addition, 2 orange infiltration-like lesions were observed in the left elbow and the right pretibial region.

Orbital MR was carried out due to suspected xanthogranulomatosis (Fig. 2), observing occupation of the anterior half of both orbits by multiple soft tissue lesions.

Palpebral biopsy revealed the presence of Touton-type multinucleated cells together with lymphoplasmocitary infiltrates rich in interstitial cells and collagen necrobiosis; biopsy suggested NBX and matched the result of pretibial and elbow biopsies (Fig. 3).

Systemic study revealed IgG monoclonal gammopathy (1960 mg/dl) and elevation of β2-microglobulin (3.4 mg/dl), together with thrombopenia (169,000 platelets/mm^3), leukopenia (2700 leukocytes/mm^3) with neutropenia (800 cells/mm^3) and VSG 100 mm/h. In addition, the patient presented non-nephrotic proteinuria. Bone marrow biopsy was negative for myeloma and skin biopsy was negative for amyloidosis. Accordingly, monoclonal gammopathy of uncertain significance (MGUS) was diagnosed.

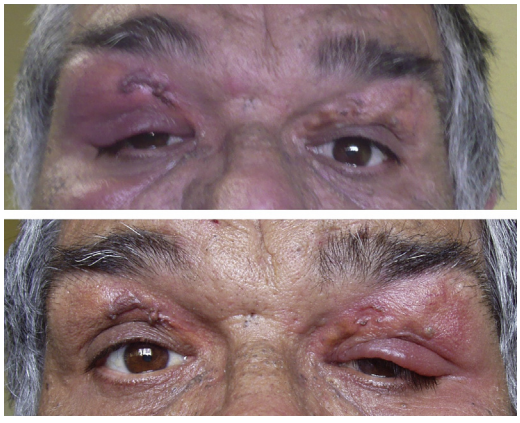

Fig. 1 – Xanthomatous tumors in both upper eyelids, with adjacent soft tissue inflammation and secondary mechanical ptosis. Top image, first visit; bottom image, one month after the first visit.

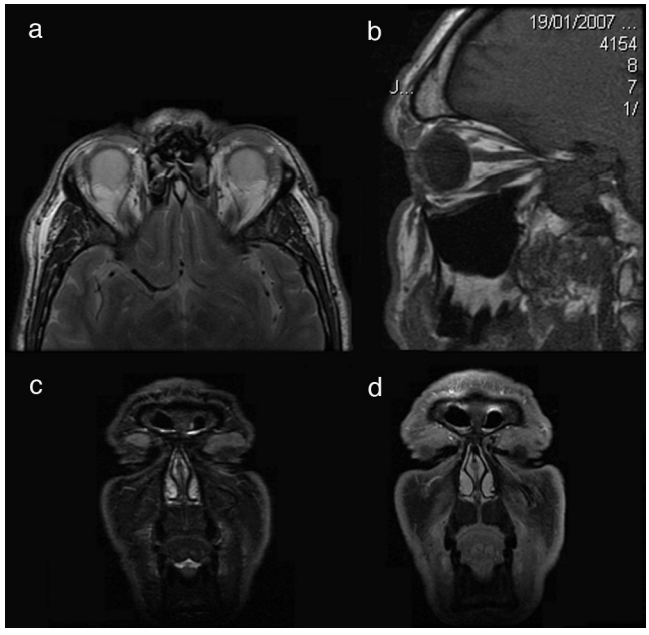

Fig. 2 – Bilateral tumors with soft tissue consistency involved in the anterior portion of the orbit, without surrounding fatty and muscular inflammation. (a) Axial T2; (b) sagittal T1 image; (c) coronal STIR; (d) coronal image with gadolinium.