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Short communication

Granulomatosis with polyangiitis confined to lacrimal gland, a case report☆

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ABSTRACT

Clinical case: A 43 year-old woman consulted due to 2 months of swelling on the superolateral side of the left orbit, with pain and erythema. An excisional biopsy was performed that revealed vasculitis with polyangiitis of the lacrimal gland. A systemic study showed that no other system was compromised.

Discussion: Orbital involvement occurs in up to 60% of patients with granulomatosis with polyangiitis. The involvement of the lacrimal gland is rare and often unilateral. Serological tests are generally negative, both in initial stages, as in localized forms of the disease.

Granulomatosis con poliangeítis localizado en la glándula lagrimal, a propósito de un caso

Palabras clave:
Granulomatosis con poliangeítis
Vasculitis necrotizante
Anticuerpo anti-citoplasma de neutrófilos

RESUMEN

Caso clínico: Mujer de 43 años, consulta por aumento de volumen orbitario superolateral izquierdo, doloroso, eritematoso de 2 meses de evolución. Se realiza biopsia escisional revelando vasculitis con poliangeítis de glándula lagrimal. El estudio sistémico no reveló compromiso de otros órganos.

Discusión: El compromiso orbitario ocurre hasta en el 60% de los pacientes con granulomatosis con poliangeítis. La afección de la glándula lagrimal es rara, y frecuentemente unilateral. Las pruebas serológicas generalmente son negativas, tanto en etapas iniciales, como en las formas localizadas de la enfermedad.

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Introduction

Wegener's granulomatosis, currently known as granulomatosis with polyangiitis (GP), is one of the positive vasculitis for the most common anti-neutrophile cytoplasmic antibodies (ANCA). It compromises the high and low airwaves in 95% and the renal system in 75%. However, during its clinical course it can compromise any organ.[1,2] Occasionally, the lacrimal gland can be affected in isolation and become the initial expression of the disease.[1,3] The purpose of the present article is to describe one case of GP localized in the lacrimal gland.

Clinical case

Female, 43, with a history of arterial hypertension and depressive syndrome, who consulted due to rapidly progressing and painful volume increase in the left periorbitary region, with upper eyelid reddening and 4 months evolution. Uncorrected visual acuity (UVA) was 20/20 in each eye, isochoric pupils with normal reaction. Intraocular pressure was 11 mmHg in each eye. Ocular motility revealed a deficit of −3 for the left eye (LE) elevation and −2 for LE abduction, LE inferior displacement and proptosis of 3 mm. The upper left eyelid exhibited increased volume, erythematous and edematous appearance, indurated and painful to the touch (Fig. 1). No alterations were found in the anterior and posterior segments of both eyes.

Computerized tomography (CT) showed an oval-shaped tumor having a diameter of 24 mm in the superotemporal quadrant with well defined edges and moderate enhancements with contrast and without changes in the adjacent bone (Fig. 2). Excisional biopsy was performed with anterior and lateral orbitotomy, obtaining a mass measuring 26 × 21 × 11 mm with multilobulated surface, poorly defined edges and grayish coffee color (Fig. 3A).

At the histological level, the normal lobular architecture of the lacrimal gland was lost and acinus and ducts were diminished. Some small vessels showed transmural fibrinogen necrosis, in addition to the presence of lymphocytic inflammatory infiltrates, plasma cells, neutrophiles, large areas of patch necrobiosis surrounded by inflammatory infiltrate aggregates comprising lymphocytes, plasma cells, epitheloid histiocytes and multinucleated giant cells (Fig. 3B–D). These findings suggested the diagnostic of vasculitis of the granulomatosis type with polyangiitis in active phase.

The patient evolved positively after surgery. UVA remained at 20/20 in both eyes with discrete upper left lateral eyelid ptosis without compromising the visual axis. LE motility exhibited a deficit of −2 with elevation and −1 with abduction. The histopathological diagnostic prompted questions to the patient who denied other systemic symptoms. Thorax and paranasal cavities tomography and encephalus nuclear magnetic resonance did not reveal alterations. Blood biometry and renal function were within normal limits. cANCA and pANCA were negative.

Due to the localized presence of GP without systemic compromise, it was decided not to initiate immunomodulating treatment for the time being and maintain the patient in follow-up. At present the patient remains stable without systemic symptoms, blood biometry and kidney function within normal limits and negative antibodies 12 months after diagnostic.

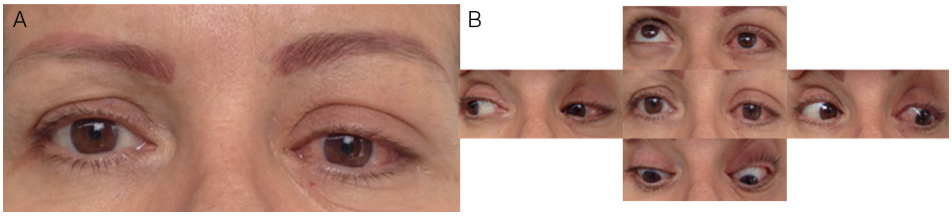

Fig. 1 – Clinic photograph: (A) inferior and medial displacement of the left ocular globe, with erythema and increased superotemporal volume; (B) unaltered RE ocular motility with LE elevation deficit and abduction.

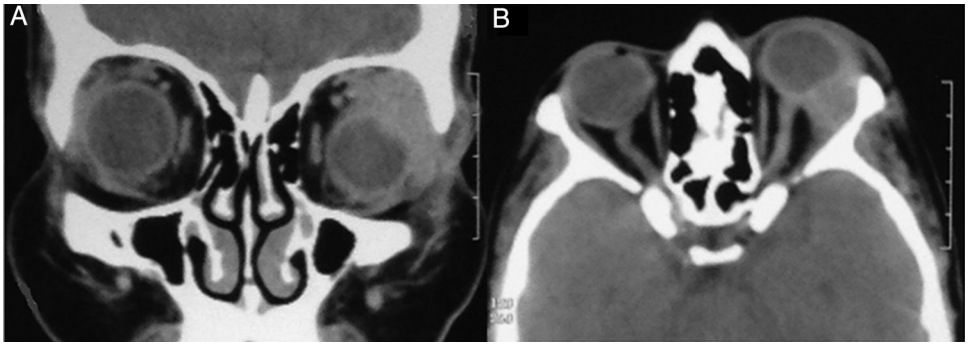

Fig. 2 – Computerized tomography with soft tissue window: (A) coronal section: left superotemporal lesion, with inferomedial ocular globe displacement causing indentation thereof. (B) Axial section, showing left a lesion between ocular globe and lateral rectus, producing proptosis.