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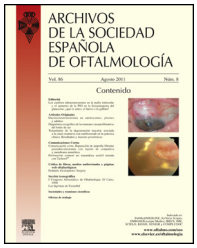

Short communication

Mikulicz's disease. A case report☆

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ABSTRACT

Case report: We report the case of a 48 year-old male with bilateral involvement of the salivary and lacrimal glands, which was diagnosed as Mikulicz's disease by incisional biopsy of the affected lacrimal glands, which was completely resolved after corticosteroid treatment.
Discussion: Sclerosing sialadenitis and/or dacryoadenitis are chronic inflammatory diseases that are currently classified into IgG_4-related diseases. Specifically, Mikulicz's disease is defined by a persistent and symmetrical swelling of the lacrimal and salivary glands together with elevated serum concentration of IgG_4 and tissue infiltration by IgG_4 (+) plasma cells.

Enfermedad de Mikulicz. A propósito de un caso

Palabras clave:
Enfermedad de Mikulicz
Enfermedad asociada a IgG_4
Dacrioadenitis
Enfermedad inflamatoria de la órbita

RESUMEN

Caso clínico: Se presenta el caso de un paciente de 48 años con afectación bilateral de las glándulas lacrimales y salivales que fue diagnosticado de enfermedad de Mikulicz mediante biopsia incisional de las glándulas lacrimales afectadas, presentando resolución completa del cuadro tras tratamiento corticoideo.
Discusión: La sialadenitis y/o dacrioadenitis esclerosante son enfermedades inflamatorias crónicas que, en la actualidad, se clasifican dentro de las enfermedades asociadas a IgG_4. En concreto, la enfermedad de Mikulicz se define por agrandamiento persistente, bilateral y simétrico de las glándulas lacrimales y salivales junto a unas concentraciones elevadas de IgG_4 sérica e infiltración tisular importante por células plasmáticas IgG_4 (+).

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Introduction

The term Mikulicz disease (MD) refers to a bilateral, symmetrical, painless edema of unknown origin that affects the lacrimal and salivary glands;[1] the term Mikulicz syndrome is only used when the cause of these findings (sarcoidosis, tuberculosis or lymphoma) is known.[2]

It was initially considered a subtype of Sjögren's syndrome (SS) due to histopathological similarities. However, this idea was later rejected due to the clear clinical differences between the two conditions.[1–3]

In 2003, Kamisawa et al.[4] proposed that the clinicopathological term for IgG_4-related autoimmune disease encompass a number of conditions with hypergammaglobulinemia and elevated serum concentration of IgG_4; this group of diseases has included Mikulicz's disease since 2004, thanks to the contribution made by the Yamamoto group.[3]

MD diagnostic criteria for Sjögren's syndrome were defined by the Japanese Company in 2008[5] and include: symmetric and persistent edema for over 3 months in at least 2 pairs of lacrimal, parotid and/or submandibular glands, in addition to elevated serum concentration of IgG_4 ($\geq$135 mg/dl) or histopathological findings of lymphocytic infiltration and IgG_4 (+) plasma cells (IgG_4/IgG plasma cells >50%) with tissue fibrosis or sclerosis.

Case report

We report the case of a 48-year-old male patient referred to our center due to increased volume of both upper eyelids with 3-year progression along with non-painful submandibular tumors for 2 months.

The only previous history reported at admission was an episode of abdominal pain of unknown origin in the previous year.

Ophthalmological examination revealed 0.4 visual acuity in the right eye and 0.3 in left eye, without alterations in the anterior or posterior ocular pole. Also, bilateral ptosis of mechanical origin is observed, resulting from two firm and well-defined tumors in the upper outer quadrant (Fig. 1), not causing ocular motility impairment. Exophthalmometry was 18 mm in the right eyeball and 14 mm in the left (Fig. 2). No adenopathies were palpated; however, 2 symmetrical submandibular tumors were, with characteristics similar to palpebral and increased volume of the lower lip (Fig. 3). Orbit CT showed symmetrical lacrimal fossa lesions with density similar to that of soft tissue, not affecting adjacent structures (Fig. 4).

Given clinical suspicion of MD, serum IgG and IgG_4 analyses were requested, which tested positive for IgG (2114.28 mg/dl) and negative for IgG_4 (19 mg/dl). We decided to perform an incisional biopsy of both lesions, which confirmed the diagnosis (Fig. 5).

Treatment was initiated with oral prednisone, 1 mg/kg/day for 2 months, with descending posology 3 months thereafter. Response to treatment was good, with no recurrence of symptoms following discontinuation (Fig. 6a and b). Likewise, no malignant transformation was detected after 15 months of follow-up, although demonstrating this would require a new biopsy of the glandular remnants, which we ruled out for the time being, as there were no signs or symptoms warranting it.

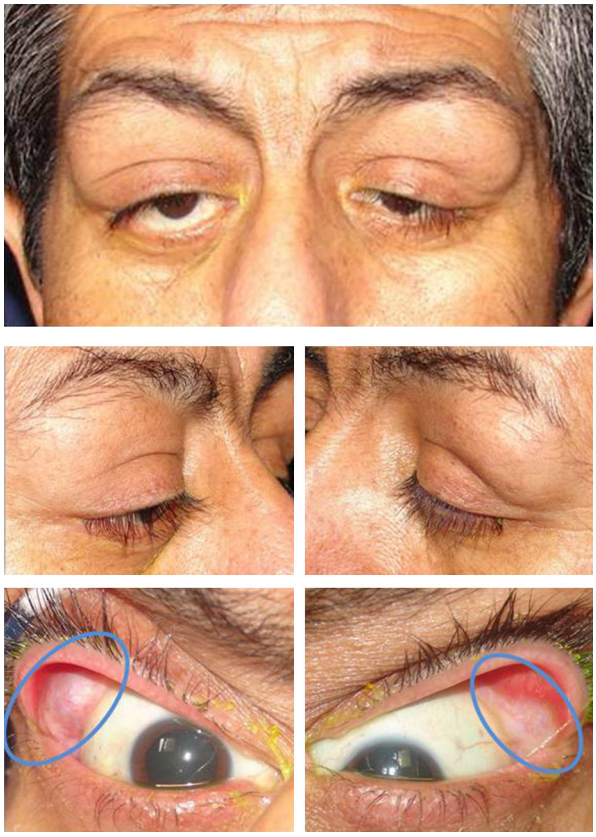

Fig. 1 – Clinical images of patient on admission. Tumors are shown in the outer third of both upper eyelids with secondary mechanical ptosis. Detail of these lesions in lateral vision and with manual elevation of eyelids.

Discussion

IgG_4-related disease is a recently reported clinical condition characterized by elevated serum levels of IgG_4 and tissue infiltration by IgG_4 plasma cells (+) linked to fibrosis.[1,6] They can affect various organs, with frequent lacrimal involvement presented as Mikulicz disease and chronic sclerosing dacryoadenitis related to IgG_4.[7]

Orbital involvement events are more common in women and often have persistent and painless palpebral edema

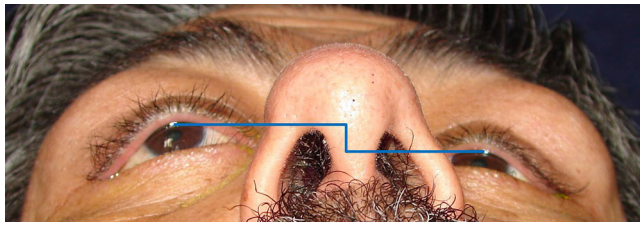

Fig. 2 – Lower perspective of the patient. We observed relative proptosis of right globe compared to the left.