APICAL ORBITAL INFLAMMATION IN GIANT CELL ARTERITIS

INFLAMACIÓN ORBITARIA APICAL EN ARTERITIS DE LA TEMPORAL

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ABSTRACT

Case report: Our purpose is to describe a case of an acute optic neuropathy with apical muscle thickening in a patient already diagnosed with giant cell arteritis. Loss of visual acuity and perimetric concentric constriction responded rapidly to intravenous glucocorticoid therapy. There has been no relapse during continued long-term therapy with cyclophosphamide.

Discussion: Giant cell arteritis is a systemic, idiopathic vasculitis; among its less frequent complications is orbital pseudotumor. Our patient required urgent treatment to avoid visual acuity loss due to compressive neuropathy and perineuritis (Arch Soc Esp Oftalmol 2007; 82: 47-50).

Key words: Orbital pseudotumor, apical orbital inflammation, temporal arteritis, compressive optic neuropathy.

RESUMEN

Caso clínico: Nuestro propósito es discutir la etiología de una neuropatía óptica aguda con engrosamiento muscular apical en una paciente ya diagnosticada de arteritis de células gigantes. La pérdida de agudeza visual y la constricción perimétrica respondieron con rapidez a terapia intravenosa con glucocorticoides. No ha presentado recidivas con ciclofosfamida de mantenimiento.

Discusión: La arteritis de células gigantes es una vasculitis sistémica idiopática, entre cuyas complicaciones menos frecuentes se encuentra el pseudotumor orbitario. Presentamos un caso que requirió tratamiento urgente para evitar la pérdida de agudeza visual por neuropatía compresiva y perineuritis.

Palabras claves: Pseudotumor orbitario, inflamación orbitaria apical, arteritis temporal, neuropatía óptica compresiva.
INTRODUCTION

Non-specific orbitary inflammation is a benign process (1) comprising several categories including posterior, apical and perineuritic forms (2).

This paper presents a neuropathic visual loss and acute inflammatory orbit pathology in a patient who had already been diagnosed through a biopsy of giant cell arthritis without previous ophthalmological complications.

Giant cell arthritis is a granulomatous idiopathic systemic vasculitis involving small and medium arteries of the supra-aortic main arteries. The most typical ophthalmological complication is anterior ischemic optic neuropathy.

CASE REPORT

A 69 year-old woman admitted due to fatigue in the course of an arthritis temporal episode in maintenance treatment with glucocorticoids.

The patient was referred urgently to the ophthalmologist for visual acuity (VA) reduction, pain, tears and difficulty to open the right eye (RE) eyelids, associated to ipsilateral exophthalmos of uncertain beginning.

VA was of 0.3 in the RE and 0.6 in the left eye (LE) with a relative afferent pupillar defect of 3+/4+ in the RE. In addition, the patient exhibited slight conjunctival hyperemia, restricted RE supraduction with vertical diplopia and slight ipsilateral exophthalmos with edematous eyelids upon stress (fig. 1). Intraocular pressure (IOP) was 15 mmHg and 17 mmHg in supraduction for the RE and 14 mmHg in the LE. The macula was normal and the papillar did not exhibit edema or paleness.

Dos months before, VA was of 0.8 in both eyes with incipient cataracts.

In the presence of this condition, an urgent orbitary CAT was requested, which revealed thickening (not clearly cone-shaped) of the inferior and lateral straight muscles of the RE, mainly in the two distal thirds, with discrete increase of orbitary fat as well as occupation of the posterior ethmoidal cells and the ptherigomaxillary fossa (fig. 2).

It was decided to establish treatment with IV methylprednisolone 250 mg at 6-hour intervals for 3 days. The patient was assessed 2 days later, exhibiting significant improvements. In the visual field a concentric reduction was observed, which subsequently returned to normal (fig. 3).

After 8 months, the VA of the RE operated for cataracts is of 1, with normal pupil reflexes and without papillar paleness. CAT and NMR revealed a non-specific occupation of the ptherigopalatine fossa and the orbitary vertex, with slight distal thickening of the inferior and lateral straight muscles, compatible with orbitary pseudo-tumor (fig. 4).

DISCUSSION

The main differential diagnostics we considered were thyroid orbit pathology and orbitary pseudo-tumor or apical orbitary inflammation in the context of systemic vasculitis, because the clinical development, a response to glucocorticoids without relapses and systemic history allowed to discard causes such as orbitary lymphoma and metastasis.

A diagnostic of Graves disease seemed improbable for the following reasons: 1) Only 4-5% of
patients develop severe orbitary pathology; 2) thyroid hormones and anti-receptor antibodies of TSH have been negative in many occasions, which occurs in only 6% of cases (although it is estimated that in the future the probability of detecting thyroid dysfunction will be of 25% in 5 years); 3) the absence of early signs such as palpebral retraction or edema when waking up; 4) exophthalmia is usually bilateral (85-90%), although asymmetric; 5) the typical muscle involvement consists in cone-shaped hypertrophy with up to 8-times size increase with tendinous involvement. The inferior and middle straight muscles are most frequently involved; 6) the typical muscle sign in T2 of NMR is iso- or hyper-tense; 7) complications such as restrictive strabismus, optic neuropathy due to compression and ocular hypertension appear in chronic stages; 8) between 25% and 65% of cases have high IOP; 9) optic neuropathy occurring in Graves disease is of the compressive type due to apex muscle thickening of up to 7 times the normal diameter and/or due to increased IOP (3).

In contrast, an orbitary pseudotumor usually appears unilaterally and abruptly, with inflammatory mechanical phthisis, muscle hyposignal in T2, restrictive strabismus of any muscle and VA loss due to compressive mechanisms and/or direct involvement of the optic nerve in the form of perineuritis (4).

The origin of the orbitary pseudotumor is idiopathic after an exclusion diagnostic process. It is occasionally associated to systemic diseases such as the Churg-Strauss disease in which the origin of the inflammation is a granulomatous vasculitis. In these cases, it has been suggested to call it «orbitary inflammation» (5). There are very few cases published in the context of temporal arthritis.

REFERENCES


Fig. 3: Acute stage of visual field concentric reduction in RE, which returned to normal after 6 months.

Fig. 4: NMR 8 months later showing muscular thickening with hyposignal in T2.