SCLEROTIC INFLAMMATION OF THE ORBIT AND MULTIFOCAL FIBROSCLEROSIS SYNDROME

ESCLEROSIS INFLAMATORIA ORBITARIA Y SÍNDROME DE FIBROSIS MULTIFOCAL

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ABSTRACT

Clinical case: A fifty-two-year-old woman consulted our department because of a bilateral proptosis. The appearance of acute symptoms suggestive of a digestive disorder during her admission to hospital for the study of the proptosis, led to a diagnosis of multifocal fibrosclerosis with simultaneous affliction of orbital tissue, mesenteric fat and the brain stem.

Discussion: Idiopathic Sclerotic Inflammation of the orbit is a rare pathological entity, with similar clinical and radiological characteristics to orbital pseudo-tumour. The differential diagnosis is important because both these conditions differ in treatment and prognosis (Arch Soc Esp Oftalmol 2008; 83: 263-266).

Key words: Proptosis, sclerotic inflammation of the orbit, inflammatory pseudotumour of the orbit, sclerosing mesenteritis, multifocal fibrosclerosis syndrome.

RESUMEN

Caso clínico: Mujer de 52 años que consulta en nuestro Servicio por proptosis bilateral. La aparición de sintomatología aguda digestiva durante el ingreso para el estudio de la proptosis, condujo al diagnóstico de un cuadro de fibrosclerosis multifocal, con afectación simultánea de los tejidos orbitarios, grasa mesentérica y tronco de encéfalo.

Discusión: El pseudotumor esclerosante orbitario es una rara entidad clinicopatológica, cuyas características en la clínica y en las pruebas de imagen pueden ser similares a las del pseudotumor inflamatorio idiopático. El diagnóstico diferencial es importante porque ambos difieren tanto en su tratamiento como en su pronóstico.

Palabras clave: Proptosis, pseudotumor esclerosante, pseudotumor inflamatorio, mesenteritis esclerosante, fibroesclerosis multifocal.
INTRODUCTION

Sclerotic orbital pseudo tumor expresses as a cicatricial infiltration, with mass effect and chronic progressive inflammatory signs determined by fibrosis (1).

In addition to the low prevalence of said tumor, the instant case report is characterized by a rare joint expression involving the brain stem and mesentery.

CASE REPORT

A 52-year old woman who visited our service due to bilateral proptosis and visual acuity reduction (fig. 1).

The exploration revealed a symmetric bilateral exophthalmos with restriction of all extraocular movements; slight conjunctival hyperemia in the anterior segment and a slight blurring in the papillary edge. Her corrected visual acuity (VA) was of 0.2 in the right eye and 0.1 in the left one. Similarly, a faulty perception of red-green was detected.

The ocular echography showed a retro-orbital intracone bilateral tumoration over 3 cm in diameter which encircled the optic nerve. The radiology service reported that the most likely diagnosis was lymphoma and idiopathic inflammatory pseudo-tumor. An orbitary Nuclear Magnetic Resonance (NMR) confirmed the findings of the echography (fig. 2).

The patient exhibited persistent headache, for which reason a cranial NMR was done. It evidenced an extra-axial lesion 21 mm in front of the brain stem (fig. 3), reported as a tumor with a signal intensity similar to the orbitary lesions, suggesting the same disease. In the absence of another neurological focal point, it was decided to maintain a conservative approach.

Tests of thyroid hormones as well as monoclonal markers and antibodies were negative.

The patient was admitted for corticoid treatment and exhibited during her stay a constitutional condition with epigastralgia. An abdominal Computerized Axial Tomography (CAT) abdominal showed a mass surrounding the superior mesenteric vessels (fig. 4). The possible existence of a lymphoma involved a postponement of corticoid treatment.

The pathological anatomy of the mesenteric lesion was reported as idiopathic sclerosing inflammation. During her stay, visual acuity diminished to hand movement in the left eye and remained stable in the right one.

Fig. 1: Bilateral Proptosis.

Fig. 2: Axial NMR of orbits showing bilateral intracranial tumoration surrounding the optic nerve.

Fig. 3: Sagittal cranium NMR observing extra-axial lesion in front of the brain stem.
With the diagnostic of mesenteric fibrosis, treatment was initiated with corticoids at a dosage of mg/kg/day. Considering the initially favorable response, evidenced by lessening exophthalmos and visual acuity improvement (0.3 in both eyes), it was decided to abstain from immune suppressors and to continue with the follow-up in out-patient visits.

During the evolution the patient exhibited a lack of response to corticoid treatment, which is characteristic of the disease, together with an acute and intense involvement of VA and bilateral visual fields (bulk perception and DM: -8dB evidenced in campimetry, respectively), with obvious involvement of the optic nerve. This determined the decision to surgically decompress the orbitary channel, which was performed through the trans-cranial pathway by the Neurosurgery service, which obtained an intra-op biopsy.

The pathological anatomy was reported as idiopathic sclerosing inflammation, confirming the multifocal fibrosis condition. The post-op assessment showed an improvement of VA (0.1 in both eyes).

**DISCUSSION**

As some authors suggest (2), orbitary sclerosing pseudo tumor is different from inflammation pseudo tumor. The following points should be taken into account: firstly, it involves loss of vision in a few months after the onset of fibrosis (1), second, it is characterized by a deficient response to corticoid treatment (1,2), which requires considering other therapies, and thirdly its possible association with other systemic diseases (1) which constitutes an extremely uncommon condition known as multifocal fibrosis. The literature describes only 22 cases of orbital sclerosing inflammation with extra-orbitary extension (3).

Even though the confirmation diagnostic was made on the basis of the pathological anatomy, the clinical assessment and imaging tests could point to either diagnostic (1). The unilateral involvement points to an inflammatory process, while bilateral involvement points to fibro sclerosis; in relation to imaging techniques, in NMR T1 the lesion is usually isointense and in T2 hyper intense in fibrosclerosing processes, whereas inflammatory pseudo tumors in T2 are hypotense (1).

Even a negative response to empirical treatment with corticoids, in the case it occurred to confirm the inflammatory involvement, could point to a sclerosing process (2).

Corticoids and radio therapy is prescribed as treatment for sclerosing disease of the orbit, although its results are poor. Kennerdall recommends combining said treatment with surgery, steroids and radiotherapy at an early stage and in an aggressive manner. The literature also describes the use immunosuppressants such as azathioprine and cyclophosphamide (3).

The patient exhibited involvement of the mesentery together with the orbitary involvement. It is easy to understand the importance of this finding because sclerosing mesenteritis coexists with other
malign processes in up to 69% of cases (lymphoma, melanoma and breast, lung and colon cancer) (4).

This case report emphasizes the importance of differentiating two independent clinical entities, the knowledge of which could allow us to change the natural history of the patient both at the ophthalmological and the systemic level (5).

REFERENCES