Idiopathic orbital myositis

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Abstract

Idiopathic orbital myositis is an inflammatory disease of unknown aetiology, with an increasing knowledge due to the new neuroimaging techniques. It should always be an exclusion diagnosis. The authors describe a case of a 69 years-old male patient, with right periorbital pain and diplopia of sudden onset, and one week later loss of homolateral visual acuity. Neurological examination showed, on the right side, an almost total loss of vision, ophthalmoplegia, proptosis and hypalgesia in the territory of the first division of the Vth cranial nerve. The CT scan findings and the response to corticosteroids therapy are highlighted because of its major importance in the diagnosis of this disease.

Key words: idiopathic orbital myositis, pseudotumour.

Introduction

The expression pseudotumour of the orbit applies to a heterogenous entity, characterized by an inflammatory process of the structures of the orbit, which includes idiopathic orbital myositis (IOM), a condition of unknown etiology located in the extrinsic muscles of the eye (uni- or bilaterally). [1,2] IOM is manifested by a clinical condition of acute or subacute installation, with periorbital pain, proptosis, chemosis, conjunctival injection, ptosis and palpebral edema, presenting a rapid response to corticotherapy (generally evident at the end of 48 h). These aspects are characteristics of this entity, and are present in this clinical case. The association with previous respiratory infections has been highlighted by some authors, and the possibility that it is an autoimmune process is currently under discussion.[3-5] The knowledge of this pathology is significantly altered with the advent of modern imaging technologies, which enable the injured tissues to be identified and non-specific inflammatory diseases to be classified according to the tissues involved.

The clinical case presented here exemplifies the pathological situation in question, and the methodology that led to its diagnosis.

Case report

Male patient, 69 years, white, born and residing in Leiria, a retired farm worker, admitted in February 1995 with a clinical condition of sudden onset, occurred a month previously, and characterized by periorbital pain in the right eye, aggravated by ocular movements, and diplopia, followed one week later by decreased homolateral visual acuity. The patient denies any other symptoms, including general malaise, fever, vomiting, anorexia, weight loss or respiratory infections.

Personal history includes heavy use of alcohol since adolescence, and significant levels of smoking of 20 cigarettes per day.

On objective examination, the patient presented good general state and nutrition, apparent age coinciding with real age, mucosa normal color and hydrated, jaundiced sclerotic, apyrexia, blood pressure of around 130/70 mmHg, pulse of 64 bpm. No periorbital sounds on auscultation, temporal regions painless on palpation, without inflammatory masses or signals. Thyroid palpable and normal size, with preserved consistency, without masses. Cardiopulmonary auscultation showed no alterations. Two palpable supraclavicular ganglions, bilaterally, painless, with elastic consistency, not adhering to the deep muscle layers, of 0.4 cm on the right and 0.5 cm on the left.

In the neurological and neuro-ophthalmological exam, accentuated decrease in visual acuity on the right is highlighted (RE: < 1/10 and LE: 10/10), without alterations in fundoscopy; in the visual fields,

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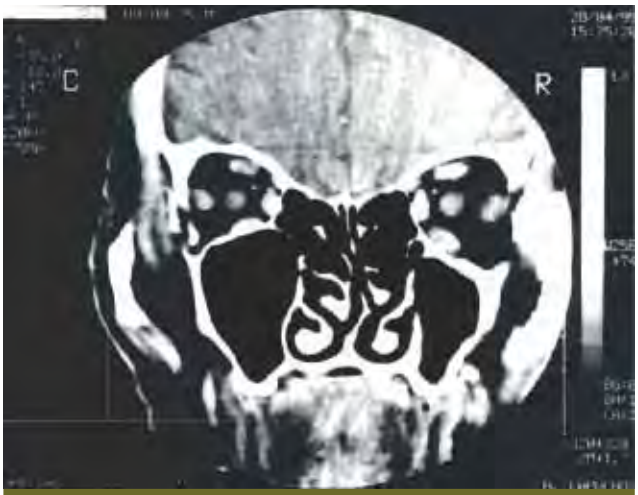
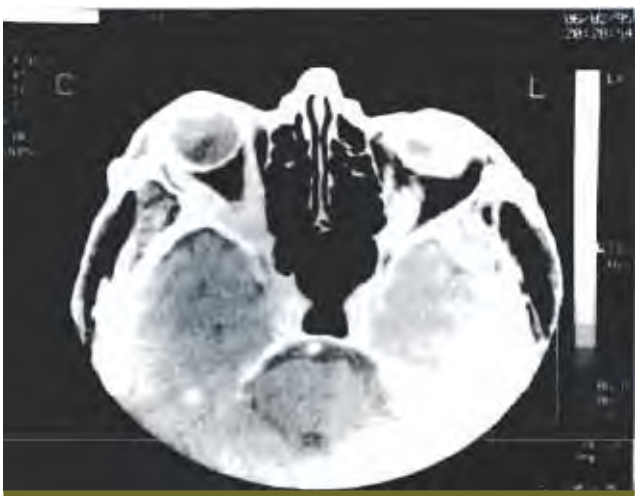


Orbital CT scan prior to the start of therapy.

FIG. 1

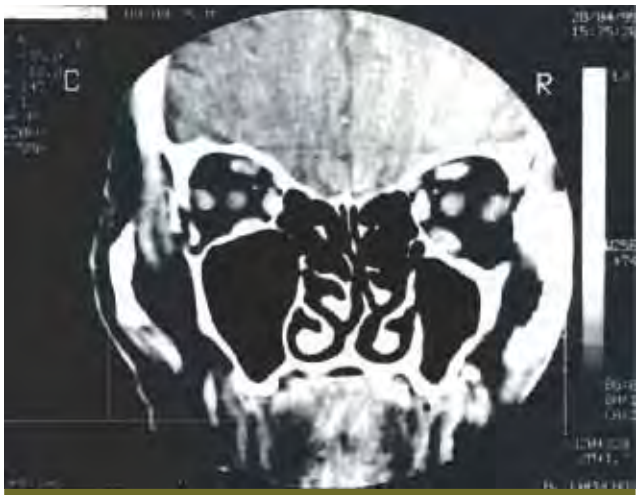


Orbital CT scan prior to the start of therapy

FIG. 2

the patient presented tunnel vision in the RE and increased blind spots in the LE; anisocoric pupils with left diameter slightly smaller than the right, light reflexes and accommodation preserved; proptosis, conjunctival injection, chemosis, palpebral ptosis, edema and inflammatory signals in both eyelids on the right side, maintaining lower vertical movements of the ocular globe only homolaterally.

Of the relevant complementary exams carried out (*Table 1*), an erythrocytic macrocytosis of 102,3 fL is visible, with normal hemoglobin values and red blood



Orbital CT scan after 3 months of therapy

FIG. 3

cell count. Computed tomography (TC) of the orbit carried out at the start of the condition (*Figs. 1 and 2*) revealed diffuse and global thickening of the extrinsic ocular muscles of the RE, and of the lower and medial rectus muscles of the LE, extending to the tendinous insertions and causing extrinsic compression of the right optic nerve at the level of the apex. These aspects were compatible with pseudotumour of the bilateral orbit, with greater expression on the right.

In view of these results, oral therapy was started with corticoids, initially at a dose of methylprednisolone 32 mg/day, which is currently maintained (end of the 3rd month) at a dose of 10 mg/day, with complete recovery of the neuro-ophthalmological signs, which was significant from the 2nd day. The orbit CT scans carried out for control purposes at 15 days, 1 month and 3 months of therapy (*Fig. 3*) show progressive normalization of the imaging alterations described.

Comments

The signs and symptoms presented by the patient (pain, proptosis, edema of the eyelid, conjunctival injection and diplopia), although highly suggestive of orbital inflammatory pseudotumour, may also be present in various other processes, including carotid--cavernous fistula, cavernous sinus thrombosis, arteriovenous fistula, infectious cellulitis, neoplastic infiltrate, granulomatoses (tuberculosis and sarcoidosis), vasculitis and in particular, myositis of the dysthyroid ophthalmology.[4,6]

TABLE I

Hemogram: macrocytosis (Hb: 16.2 g/dL; MGV: 102.3 fL)
Sedimentation rate: 10 mm/1st h
Thyroid function tests (T3, T4, HRT): normal
Ionogram (including CA2+): normal
Total CK: normal
ACE: normal
Serology for vasculitis: negative
X-ray of the chest and paranasal sinuses: no alterations
Mantoux Test: positive

The neuroimaging evaluation seeks to reduce diagnostic uncertainty in relation to the hypotheses of dysthyroid ophthalmopathy and IOM, both of which are inflammatory processes of the extraocular muscles.[3] The appearance of the orbital CT scan can assist in the differential diagnosis between these two pathologies, since the involvement of the tendinous insertion and the greater irregularity of muscle thickening, as occurs in our clinical case, are good indicators of idiopathic myositis (although its absence is not sufficient to rule out this condition).[4,7] The differential diagnosis between the two pathologies is, however, more frequently made by the clinic, myositis of thyroid dysfunction having more insidious onset and absence of rapid response to corticotherapy. This latter factor practically establishes the diagnosis of idiopathic orbital myositis, particularly if significant on the 2nd day of therapy.[3,4,7] as occurred in the present case. In relation to the laboratory diagnosis between the two situations, the overload test with HRT and/or inhibition by T3 should have been carried out (besides the determinations of T3, T4 and TSH) but this was not possible.

The particular characteristic that we found in our patient, of decreased visual acuity and campimetric defects due to compression of the optical nerve in the orbital apex, with increased volume of the muscles, is curious, as few cases have been reported in which these symptoms are observed, and they constitute an element in favor of dysthyroid ophthalmopathy.[4]

The frequency with which this inflammatory situation is associated with respiratory infections (occurring, on average, 2 weeks before the onset of the symptoms), has raised the possibility that the process is mediated by autoimmune mechanisms.[3-5] This epidemiological data was not found in the patient presented. ■

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