

Case Report

A Rare Case of Ocular MyositisCARMEN VALERIA ALBU¹, B. CĂTĂLIN², CORNELIA ZAHARIA¹¹Neurology Clinic, University of Medicine and Pharmacy of Craiova,
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ABSTRACT : We report the case of a 43 year old man who presented recurrent left abducens palsy. His medical history included arterial hypertension, ischemic cardiomyopathy, dyslipidemia, rhinitis, maxillary sinusitis. Physical examination revealed a overweight patient, horizontal gaze diplopia, left nerve VI paresis, mild left retro-orbital pain. The orbital MRI also didn't offer new information: mild external edema on the left eye, with normal tendon aspect, no thickening or enhancement of the muscle belly and also normal aspect of the bony orbit. Recurrent palsy of EOMs can be caused in rare cases by ocular myositis.

KEYWORDS: ocular myositis, unilateral, corticotherapy.

Introduction

First described by Gleason in 1903, orbital myositis (OM) or orbital pseudotumor is a benign idiopathic inflammatory disease that may affect any structure in the orbit [1]. Ocular myositis is now seen as a subgroup within the entity of idiopathic orbital inflammatory syndrome (IOIs) [2] and it just represent 8% of all idiopathic orbital inflammations [3].

Ocular myositis affects mostly females (2:1 females to males ratio) around 32 years old but IOIs has been reported at any age [4]. While orbital myositis is an inflammation of any orbital structure, ocular myositis describes a rare inflammatory disorder of extraocular eye muscles [2,5] but the two have a lot of overlaps and so are hard to distinguish. Most cases of OM have subacute orbital painful diplopia which is caused by handicapped contraction and distraction of affected eye muscles not by neurogenic associated affections [6]. Pain can be exacerbated by eye movement. In most of the patients, beyond orbital discomfort, no additional signs or symptoms are present [2].

Two major forms of ocular myositis have been described, one with few ocular symptoms and with an additional conjunctival injections called limited oligosymptomatic ocular myositis (LOOM) and a second form with a severe exophthalmic ocular involvement with additional ptosis, chemosis, and proptosis, called severe exophthalmic ocular myositis (SEOM). Also signs and symptoms of other subgroups of IOIS such as dacryoadentitis, periscleritis and perineuritis may be evident [2].

Many inflammatory, vascular, neoplastic or infectious disease can affect the extra-ocular muscles and mimic orbital myositis. Thyroid-

related orbitopathy is seen as the most important differential diagnosis [4]. Muscle biopsies of EOMs have reporting mixed infiltrates of plasma cells, lymphocytes, macrophages and polymorphonuclear cells while chronic forms of idiopathic ocular myositis are associated with fibrosis of the affected muscles [2]. Specific myositis either by bacterial or viral infections can be associated to orbital myositis while others can be a local manifestation of a systemic immunomediated disease [7].

While specific forms of ocular myositis will be resolved with specific systemic treatment, idiopathic ocular myositis is treated with corticosteroids or with radiotherapy [4].

Case Report

We present the case of a 43 years old Caucasian male, ex-smoker (15PA), stopped 6 years ago, was admitted in our hospital in September 2012 for a history of recurrent palsy of left abductor nerve with left gaze diplopia. These symptoms began in March 2012 were treated first by his general practitioner with non-steroidal anti-inflammatory drug and B complex vitamins for 10 days with no results. After this the treatment was changed to methylprednisolone 32 mg. Under methylprednisolonum the nerve palsy remitted but reappeared when the dose was lowered. The patient had a history of arterial hypertension, ischemic cardiomyopathy, dyslipidemia, rhinitis, maxilar sinusitis and he was treated with calcium blocker channels, indapamidum and statin. Physical examination revealed a overweight patient (MC=26 kg/m²), horizontal gaze diplopia, left nerve VI paresis, mild left retroorbital pain, no dizziness, no nystagmus, no

motor deficits, no difficulty in swallowing liquids or solids, normal osteotendinous reflexes, bilateral plantar flexion, normal language. Blood pressure was 120/60mmHg, pulse was 74 bpm, respiratory rate was 18 breaths/minute, oxygen saturation was 99% while breathing ambient air; the rest of the examination were in normal limits. At this point several diagnoses were taken into consideration, such as thyroid disease, ocular myasthenia gravis, infection, ocular-faringial syndrome and ocular myositis.

Results of laboratory tests revealed a moderate hypercholesterolemia (237mg/dl) and hypertriglyceridemia (147mg/dl); the rest of the results were in normal limits. The patient was tested for different pathologies in parallel as following: TSH=0,4mIU/ml, FT4=10,6pmol/l, T3=1,01ng/ml, IgM antibodies for Lyme were 2.3AU/ml, IgG for Borrelia were less than 5 AU/ml, anti MUSK antibodies were less than 5 nmol/l, anti-acetylcholine receptor (AChR) antibody less than 0,1 nmol/l, HbA1c=5%, ANCA, anticardiolipin, anti-dsDNA, HIV were

all negative. Electrocardiogram revealed normal sinus rhythm and no abnormalities were found on cardiac ultrasound. The chest X-ray showed no active pulmonary lesions, so it didn't raise the suspicion of sarcoidosis or malignancy. The thyroid ultrasound suggested normal dimensions with a slightly anterior engaged left lobe. The otorhinolaryngology exam revealed signs of maxillary chronic sinusitis, chronic amigdalitis, deviated nasal septum and chronic rhinitis. The ophthalmology consult observed just edema in the left orbit, with no other modifications. Head CT -scan performed revealed no pathological lesions. The orbital MRI also didn't offer new information: mild external edema on the left eye, with normal tendon aspect, no thickening or enhancement of the muscle belly and also normal aspect of the bony orbit (Fig.1). Cerebral angiography: normal aspect at injection in both internal and external carotid artery (Fig.2). Also functional testing through visual evoked potentials using a pattern-reversal stimuli was normal (Fig.3).

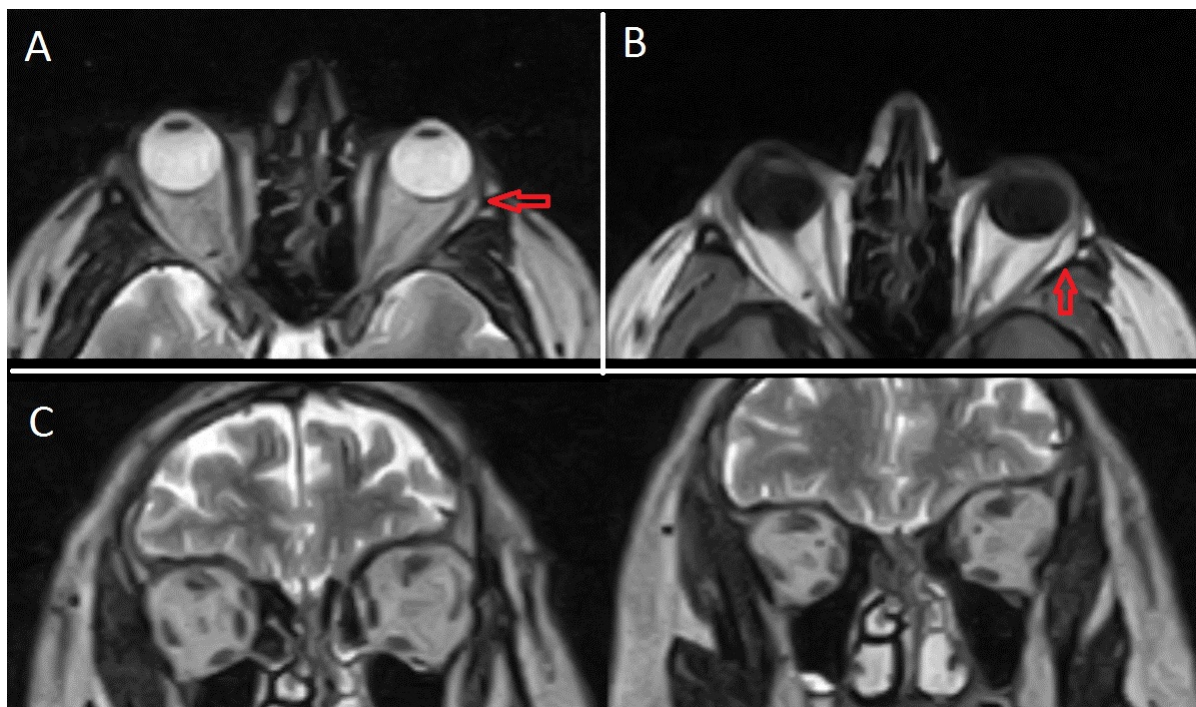


Fig.1. Orbital MRI showed for both T2 (A) and T1 (B) mild external edema on the left eye, with normal tendon aspect, no thickening or enhancement of the muscle belly and also normal aspect of the bony orbit (C).

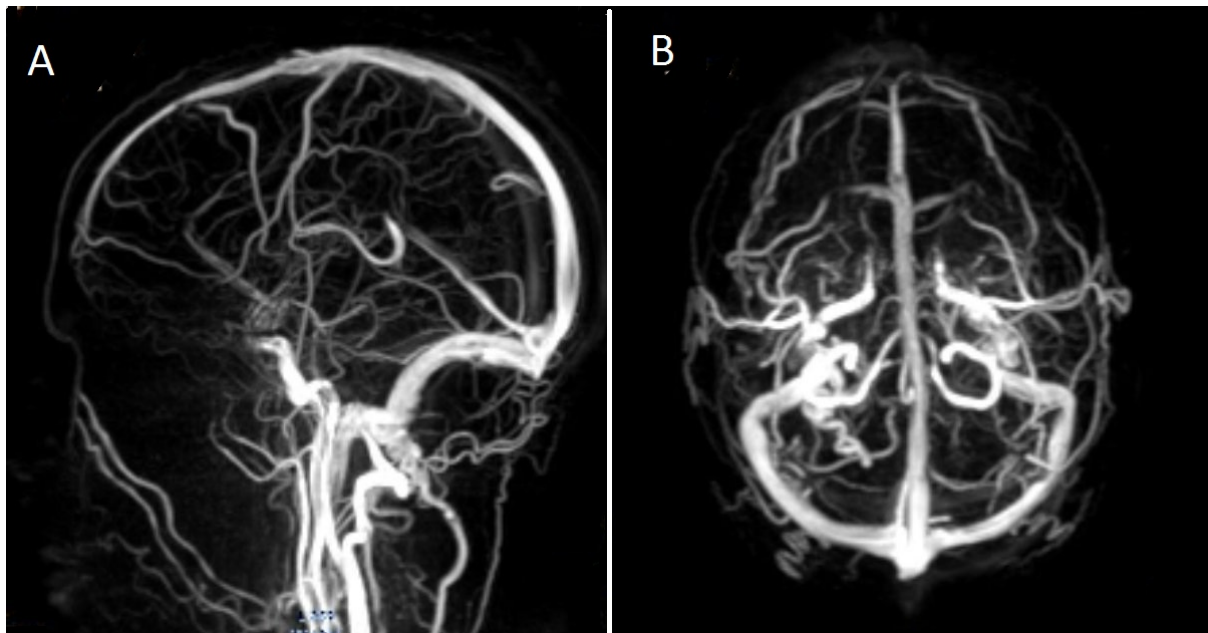


Fig.2. Cerebral angiography with normal aspect in both internal and external carotid artery.

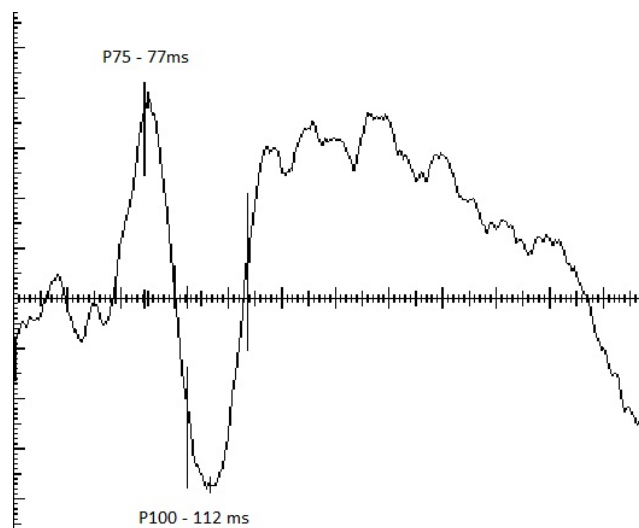


Fig.3. Normal left eye visual evoked potential according with the standard of the lab.

Given these results a diagnose of ocular myositis was made without having the biopsy confirmation. Corticosteroid treatment was reinitiated with high doses as 80mg/day of methylprednisolonum for 1 month with a slow decrease in dosage. All the fallow-up evaluation found the patient struggling between asymptomatic periods and diplopia with left nerve VI paresis periods, according to the corticosteroid dosage.

Discussions

It is not precisely known why extraocular muscles (EOMs) are preferentially affected by inflammation in IOIS [2,4]. EOMs have a

unique myofibrillar protein isoform composition reflecting their structural and functional properties [8]. Compared with skeletal muscle, EOMs have smaller motor unit sizes, higher motor neuron discharge rates, higher blood flow with higher mitochondrial volume fractions which can suggest that the energy requests and susceptibility to mitochondrial dysfunction are higher as compared with skeletal muscle [2,9]. Due to the high blood flow and vascularisation, inflammatory blood cells can access more easily this specialized body compartment [2, 10].

Thyroid orbitopathy is the most common cause of orbital disease with a insidious onset is rather than acute leading to lid lag and lid

retraction in down-gaze which is characteristic for this disorder. Usually patients will have bilateral eye involvement with signs of inflammation like edema leading to secondary fibrosis and in a few cases to optic neuropathy [4].

Because 50%-80% of myasthenia gravis initial manifestation can be an isolated eye muscle weakness with blepharoptosis or ophthalmoparesis this diagnostic has to be taken in consideration in all patients presenting with diplopia. Usually asymmetric ptosis is frequent associated with myasthenia gravis and up to 60% of the patients have acetylcholine receptor antibodies present [11].

Other pathologies can be taken in consideration in this patient, some can be excluded by careful conducted history interview like Tolosa-Hunt syndrome, while others need specific tests to confirm (oculopharyngeal muscular dystrophy, myotonic dystrophy type 1, congenital cranial dysinnervation disorders and congenital fibrosis of extraocular muscles and mitochondrial myopathies).

Although not an often pathology, when dealing with a patient with recurrent palsy of EOMs, ocular myositis must be always be taken in consideration.

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