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Orbital myositis: description of a cohort of seven patients with atypical features

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INTRODUCTION

Orbital myositis (OM) is a very rare disease characterized by a non-infectious, inflammatory involvement of one or more extraocular eye-muscles (EOMs). The clinical picture is characterized by an acute/subacute onset diplopia, orbital pain and paresis of the affected EOMs associated to signs ocular inflammation. Two clinical forms are recognized: the Limited Oligosymptomatic Ocular Myositis (LOOM) with only additional conjunctival injection, and the Severe Exophthalmic Ocular Myositis (SEOM) with ptosis, chemosis, pupillitis and proptosis[1]. MRI is the most valuable diagnostic tool showing the enlargement and contrast enhancement of the affected EOMs and offering important clues for the differential diagnosis with other OM mimicking diseases. Timely treatment is greatly important as OM promptly responds to steroids; nevertheless partial recovery or relapses may occur in up to 80% of cases[2].
We describe, a cohort of OM patients diagnosed at our institute, showing some atypical features as absence of pain or association with another neuromuscular disease, myotonic dystrophy type 2 (DM2) that complicated the clinical picture.

MATERIALS & METHODS

Seven OM patients (3M:4F) were diagnosed at our centres in the past 15 years according to clinical and neuroradiological features.
Their clinical data were retrospectively analysed after a mean follow-up period of 3 years (range 1-10 yrs) (Tab. 1).
They have all received steroids as first line drug (prednisone 1 mg/Kg/die for 2 weeks, then tapered according to the clinical response); alternative immunosuppressants (MTX, AZA, IvIg) were used in non-responsive/relapsing cases (Fig. 3).

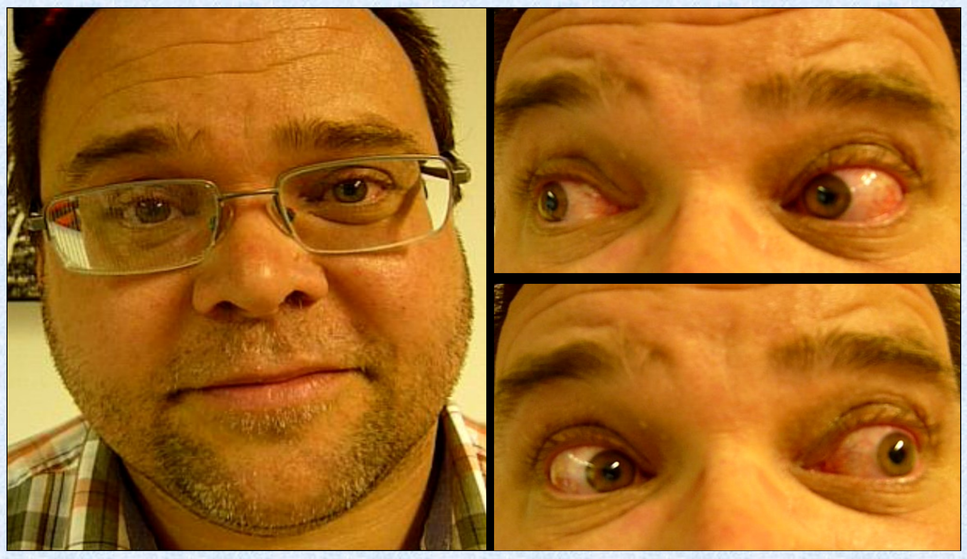

Fig. 1 Clinical features of Patient 1

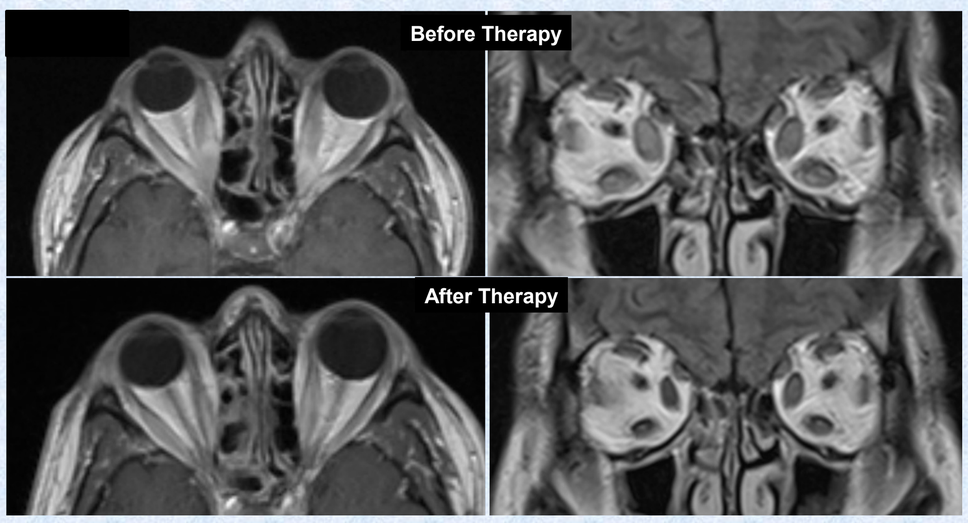

Fig. 2 Typical MRI findings in OM (Pt. 1)

Pt	Sex/ Age	Symptoms	Ocular signs	Associated diseases	EOMs involvement	Diagnostic Delay (months)	Therapy I° line	Therapy II° line	Relapses	Outcome
1	M 43	Double vision + pain	CI (LOOM)	Sinusitis	MR (bil.) IR (bil.)	1	prednisone 1mg/Kg/die	MTX, AZA	2	Partial remission
2	F 48	Double vision	CI (LOOM)	No	IR (left)	10	prednisone 1mg/Kg/die	MTX	1	Partial remission
3	M 39	Double vision	no (LOOM)	No	LR (bil.), MR (bil.)	6	prednisone 1mg/Kg/die	No	1	Complete remission
4	M 44	Double vision + pain	CI +papillitis (SEOM)	Sinusitis	IR (bil.)	8	prednisone 1mg/Kg/die	No	No	Partial remission
5	F 54	Double vision + pain	no (LOOM)	DM2	SR (right)	2	prednisone 1mg/Kg/die	AZA, IvIg	1	Complete remission
6	F 60	Double vision	CI+proptosis (SEOM)	Grave´s disease + surgery	IR (left)	2	prednisone 1mg/Kg/die	No	No	Complete remission
7	F 23	Double vision + pain	CI (LOOM)	No	MR+LR (right)	7	prednisone 1mg/Kg/die	MTX	1	Partial remission

Tab. 1 Clinical features of patients with orbital myositis – CI: conjunctival injection; LOOM: limited olygosymptomatic orbital myositis; SEOM: severe exophthalmic ocular myositis; MR: medial Rectus; IR Inferior rectus; LR: lateral rectus; SR: superior rectus

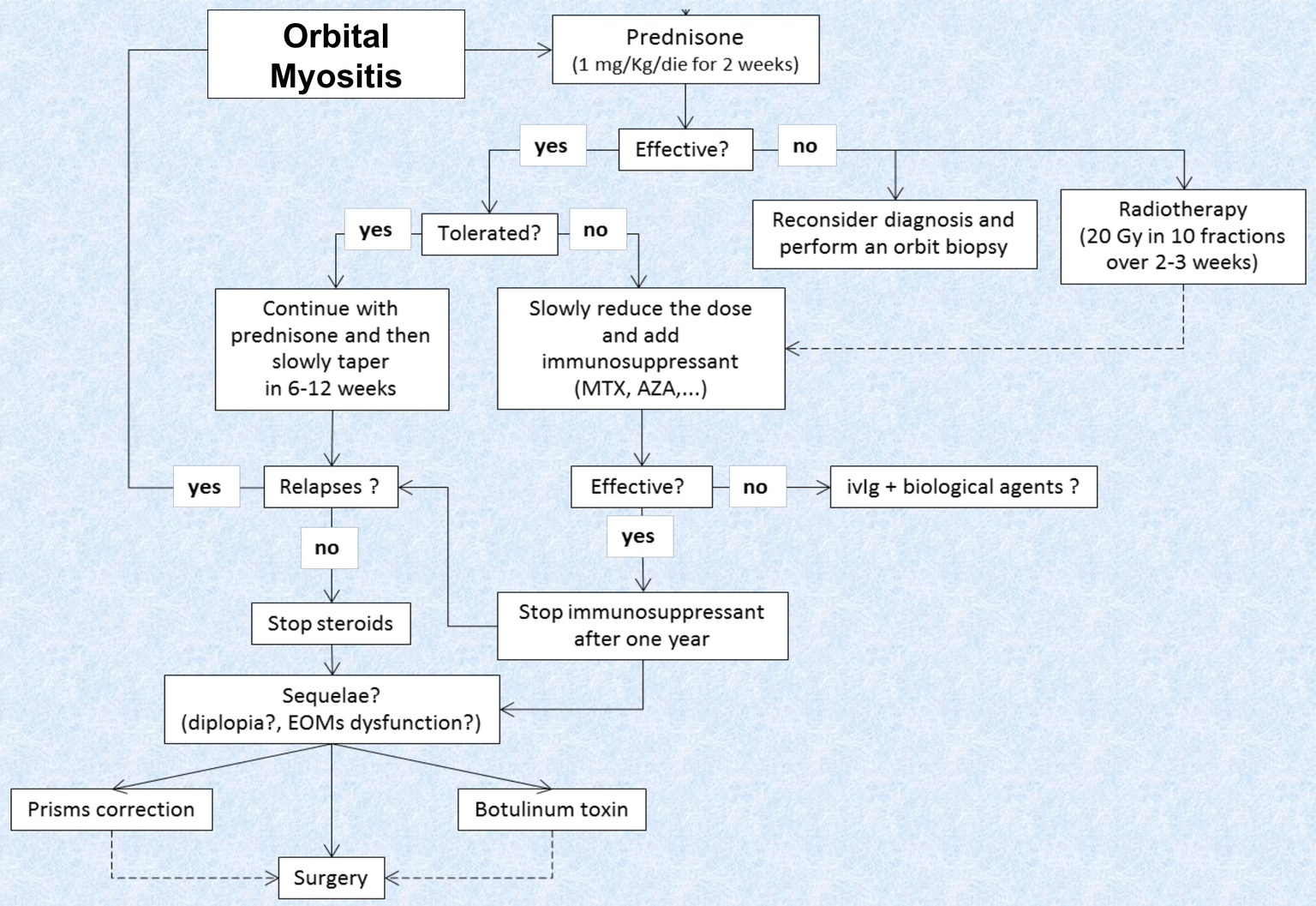

Fig. 3 Suggested therapeutic algorithm in OM

RESULTS

Mean age at onset was 44 years (range 23-60). All patients presented double vision but pain was absent in 3/7 cases (43%). A LOOM form occurred in 5/7 and a SEOM in 2. Associated diseases were: sinusitis (2/7), previous Grave´s disease (1/7), DM2 (1/7). MRI showed a unilateral single EOM involvement in 3/7 patients (43%) and multiple EOMs involvement in the others. The most frequently affected EOMs were the horizontal recti (3/7) and inferior rectus (4/7).
All patients showed a prompt response to steroids but relapses occurred in 5/7 (70%) and an immunosuppressant was added (AZA or MTX). Only in 3/7 (43%) a complete remission of symptoms could be obtained, persisting in the majority of patients a mild diplopia. Patients with single EOMs involvement and early therapy start seem to have a better prognosis.

DISCUSSION

From the presented cohort emerge some relevant atypical aspects that should be considered for the OM diagnosis and management. In a significant proportion of patients, orbital pain may be absent, often delaying the diagnosis; OM may occur in the context of other diseases complicating the clinical picture, as in our DM2 case or in patients with a history of thyroid disease leading to difficulties in the differential diagnosis with thyroid associated orbithopathy (TAO).
In comparison with previously described cohorts[2,3], many of our patients had involvement of the inferior rectus which is quite unusual for OM and more typical for TAO.
The occurrence of relapses (70%) is similar to what reported in other studies[2,3] but partial remissions and sequels were more frequent in our cohort and seem related to delayed diagnosis/ therapy and multiple EOMs involvement.

CONCLUSIONS

Our study helps better characterizing this rare, heterogeneous, under-recognized muscular disease, thus helping the general neurologist in its early diagnosis. This is the first report of OM associated to DM2; given the higher incidence of autoimmune diseases in DM2[4], a causal relationship can be hypothesized. A prompt therapy start is recommended to prevent disease relapses and permanent EOM dysfunction.

References

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