

Bilateral Ocular Myositis as a Late Complication of Dermatomyositis

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ABSTRACT. The association of bilateral ocular myositis with primary inflammatory muscle disease is rare. We describe a 30-year-old man with a 2-month history of eyelid edema, erythema, and orbital pain, occurring in the course of previously undiagnosed and untreated dermatomyositis (DM). Although ocular myositis is a very rare manifestation of DM, it might be overlooked by clinicians who are not aware of this complication. (J Rheumatol 2005;32:379–81)

Key Indexing Terms:

OCULAR MYOSITIS

DERMATOMYOSITIS

IMMUNOGLOBULIN THERAPY

The clinical classification of idiopathic inflammatory myopathies includes polymyositis (PM), dermatomyositis (DM), amyopathic DM, juvenile DM, myositis associated with neoplasia or with collagen vascular diseases, and inclusion body myositis. The forearm, hand, leg, and foot muscles are spared in all but 25% of cases^{1,2}. Ocular muscles are not affected except in the rare patient with both PM and myasthenia gravis³. Although orbital myositis may be associated with several conditions^{4,5}, to our knowledge the association of bilateral ocular myositis with primary inflammatory muscle disease has not been reported.

CASE REPORT

A 30-year-old man presented in December 2001 with a 2-month history of eyelid edema, erythema, and orbital pain, which developed initially at the left eye and subsequently affected both eyes. Notably, his history included a 6-month period of symmetrical weakness of limb-girdle muscles occurring 7 years before, which had resolved with brief oral prednisolone treatment. In addition, he reported 2 self-limited episodes of jaw lymphatic gland swelling and low-grade fever 3 and 7 years previously, as well as non-itching erythema on his back for the last 7 years. Biopsy from the affected gland at that time disclosed some grade of immunoreaction, but no specific diagnosis had been made. His family history was unremarkable for neurological or connective tissue diseases.

Admission examination revealed bilateral proptosis and exophthalmos with no signs of diplopia, in addition to the erythema of his back and the eyelid signs. He had no signs of any other muscle weakness, dysphagia, or fatigue. Laboratory evaluation revealed normal findings except for slightly elevated white blood cell count (11,500/ μ l), a C-reactive protein of 6.7 mg/l, and elevation of β -globulin (0.9 g/dl, upper normal limit 0.8 g/dl) in protein electrophoresis. Creatine phosphokinase and aldolase levels were within normal limits. Serologic tests for antinuclear antibodies, antibodies

to extractable nuclear antigens (RNP, SSA, SSB, Scl-70, and Jo1) and acetylcholine receptors, antineutrophil cytoplasmic antibodies, and rheumatoid factors were negative, while complement concentrations were also within normal limits. The serum did not contain anti-Epstein-Barr virus, anti-cytomegalovirus, anti-herpes simplex virus, or anti-Toxoplasma antibodies. Extensive investigation for sarcoidosis and malignancy was negative.

Orbital magnetic resonance imaging (MRI) disclosed enlargement of the lateral and inferior rectus in both eyes, and the superior rectus in the left eye (Figure 1A). Electromyography of the limb muscles was within normal limits, but the mimic muscles (frontalis and orbicularis oculi) were affected, with short duration multiphase motor units (mean duration 3.2 ms; lower normal limit 5.4 ms), which were compatible with myopathic-like changes⁶. Nerve conduction velocities were normal, while repetitive stimulation test for myasthenia gravis was negative. Muscle biopsy findings (quadriceps femoris) revealed mild myopathic changes; nonspecific inflammatory changes were found on skin biopsy.

On the basis of these findings and his history of symmetrical weakness the diagnosis of DM was established. He was then treated with azathioprine (daily dose 2 mg/kg) and 12 monthly doses of intravenous immunoglobulin (IVIG)⁷⁻¹⁰; however, because of gastric intolerance, azathioprine was subsequently discontinued. Initially, a marked improvement of eyelid edema and erythema signs was noted; a new electromyography test of the mimic muscles performed at 6 months disclosed a slight improvement of duration of the motor units (mean duration increased to 4.94 ms). However, a second orbital MRI performed at 12 months was not encouraging for continuing this therapy, since a deterioration of both lateral rectus and superior rectus in the left eye and enlargement of the superior rectus in the right eye were observed (Figure 1B). Thus, we decided to discontinue IVIG and treat our patient with methotrexate (10 mg weekly) and oral methylprednisolone (8 mg daily). This therapy resulted in stabilization of the disease; no further enlargement of the involved muscles was observed in a third MRI performed at 24 months (Figure 1C).

DISCUSSION

Orbital myositis is a rare, focal inflammatory muscle disease of unknown etiology, usually presenting with eye pain, head pain, diplopia, conjunctival and lid hyperemia, and proptosis¹¹. It may occur in the context of a systemic disease, most frequently systemic lupus erythematosus and sarcoidosis for adults. Although our patient was first seen for the signs of focal bilateral ocular myositis, he had a mild DM that was untreated for years. Enlargement of the inflamed muscles is

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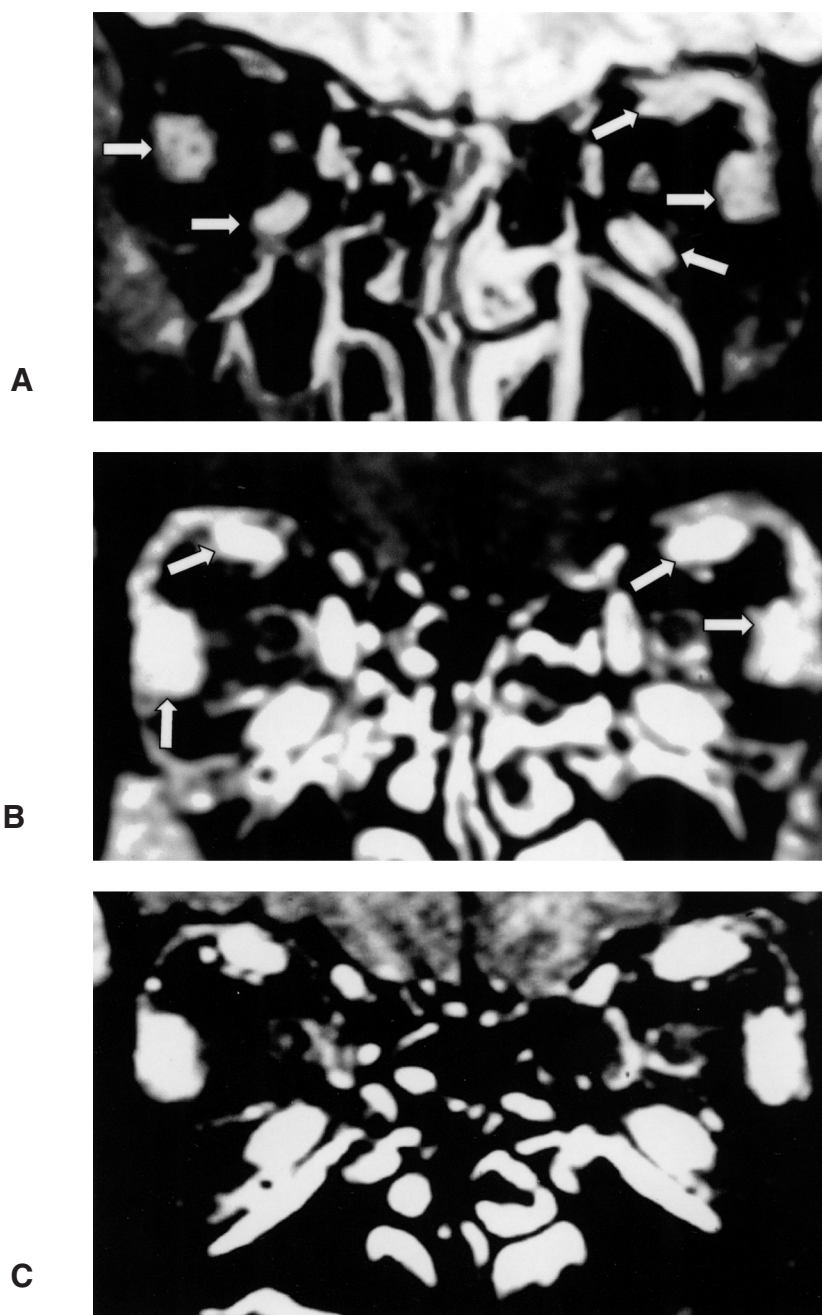


Figure 1. Orbital MRI (T1 sequence) at presentation, showing enlargement of the lateral and inferior rectus in both eyes, and the superior rectus in the left eye (A). Deterioration of both lateral rectus and the superior rectus in the left eye, and enlargement of the superior rectus in the right, are noted after 12 months of monthly IVIG therapy (B). The lack of any significant improvement in muscle size is observed after an additional 12-month period of daily methylprednisolone and weekly methotrexate therapy (C).

a common feature of DM¹², as well as in the rare cases of focal myositis¹³, therefore the orbital MRI findings were not unexpected. The electromyography findings of the limb muscles were not indicative for PM, but the facial muscles showed changes compatible with inflammatory muscle dis-

ease. These findings were also improved by the immunoglobulin therapy⁷⁻¹⁰. Muscle biopsy from a muscle with no clinical or electromyography signs of inflammation disclosed mild abnormalities suggesting that a subclinical form of damage was present in more muscles. Therefore our

patient had a history of symmetrical muscle weakness and rash, as well as muscle biopsy and electromyography evidence of myositis, establishing the diagnosis of definite DM^{1,2}.

We describe the presence of bilateral orbital myositis, occurring in the course of a previously undiagnosed and untreated dermatomyositis. Although ocular myositis is a very rare manifestation of DM^{4,5}, it might be overlooked by clinicians who are not aware of this complication.

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