RECTUS SUPERIOR AND LEVATOR PALPEBRAE SUPERIORIS (RS+LPS) MUSCLE COMPLEX MYOSITIS

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SUMMARY – A patient with acute isolated rectus superior and levator palpebrae superioris (RS+LPS) muscle complex myositis is presented. A 50-year-old woman presented with a one-day history of left mild palpebral edema, pseudoptosis and orbital pain. On clinical examination, the patient exhibited left palpebral edema and pseudoptosis with preserved levator function and eye movements. Echography and computed tomography (CT) scan findings showed isolated enlargement of the left rectus superior RS/LPS muscle complex. Oral steroid and NSAID therapy led to complete resolution of all abnormal clinical and echography findings within two weeks. Isolated RS+LPS muscle complex myositis is a rare disease; however, it is important for differential diagnosis in patients with acquired blepharoptosis even if the muscle function is preserved. This was a case of acute isolated RS+LPS muscle complex myositis with preserved muscle function and mild palpebral edema with pseudoptosis and orbital pain as the only clinical manifestations of the disease. The diagnosis was confirmed by pathognomonic echography and CT scan findings of isolated enlargement of the RS+LPS muscle complex and symptomatic improvement on corticosteroid and NSAID therapy.

Key words: *Myositis – diagnosis; Myositis – etiology; Myositis – drug therapy; Orbital diseases – diagnosis; Orbital diseases – drug therapy*

Introduction

Orbital myositis is an idiopathic, nonspecific inflammation of one or more extraocular muscles[1]. The disease is typically unilateral, usually involves one muscle, mostly superior or lateral rectus muscle. It usually occurs in early adult life, in most cases in female patients[2]. The symptoms include edema of the eyelid, ptosis, weakened function of the muscle, diplopia, mild proptosis and intensifying pain caused by extension of the muscle when trying to look in the opposite direction of the muscle action[1,2]. Echographic scan, computed tomography (CT) and magnetic resonance imaging (MRI) are employed to confirm the diagnosis. Differential diagnosis includes orbital cellulitis, dysthyroid myopathy and Tolosa-Hunt syndrome[1]. The course of the disease can be acute/subacute without relapse, with spontaneous recovery within 6 weeks. Otherwise, the disease takes a chronic course characterized by a persisting episode that lasts over 2 months or by recurrent attacks. Recommended therapy involves systemic steroids, nonsteroidal anti-inflammatory drug (NSAID) therapy, and possibly radiotherapy[1].

A case of a patient with acute rectus superior and levator palpebrae superioris (RS + LPS) muscle complex myositis is presented.

Case Report

A 50-year-old woman presented for examination because of the mild left upper eyelid edema, pseudoptosis and orbital pain that persisted for one day. Clinical examination showed visual acuity 1.0 bilaterally, consecutive retraction of the right upper eyelid, edema and pseudoptosis of the left upper eyelid along with normal active and passive eyelid movement, normal bulbomotoricity and photomotoricity. Slit-lamp examination of

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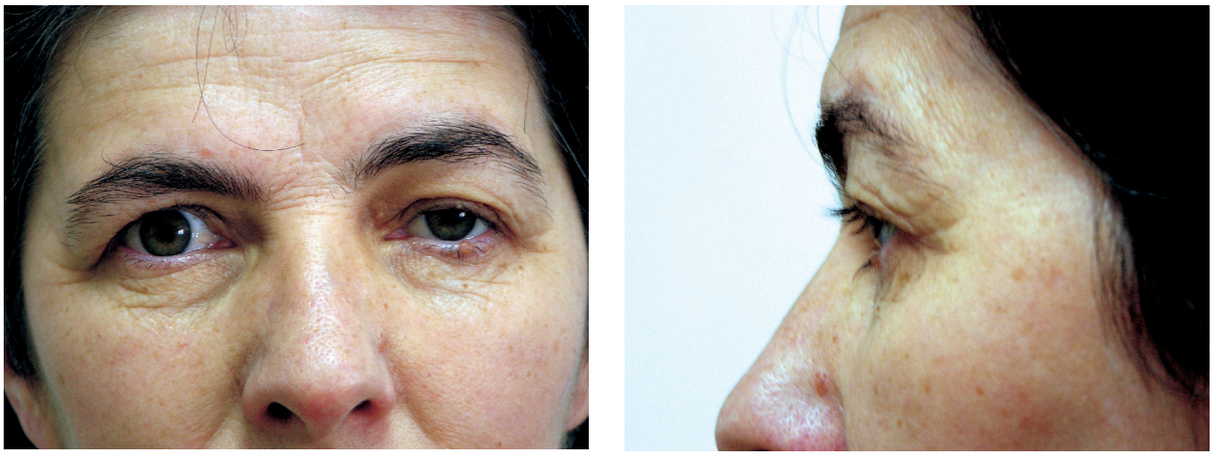

Fig. 1. Pseudoptosis and edema of the left upper eyelid and consecutive retraction of the right upper eyelid.

the anterior segment and fundus showed normal finding with bilateral intraocular pressure value of 18 mm Hg.

Echographic scan showed enlargment of the left rectus superior muscle (RS) and levator palpebrae superioris (LPS) muscle complex along its length, with perimyogenic and perisinew edema with accentuation of the muscle membranes. The right and left eye RS+LPS muscle complex diameter was 4.83 mm and 7.48 mm, respectively. CT scan showed isolated enlargment of the RS+ LPS muscle complex.

The patient was administered a single dose of 4 mg dexamethasone parabulbarly, oral prednisolone 60 mg with gradual dose reduction by 10 mg every 4 days, and 25 mg indomethacin 3 times daily. This therapy produced significant improvement of the patient's clinical condition and regression of symptoms after only 4 days of treatment. Two weeks later, complete resolution of the disease was observed, with normal clinical and echography findings. The follow up values of the right and left RS+LPS muscle complex diameter were 4.84 mm and 5.33 mm, respectively (Figs. 1-4).

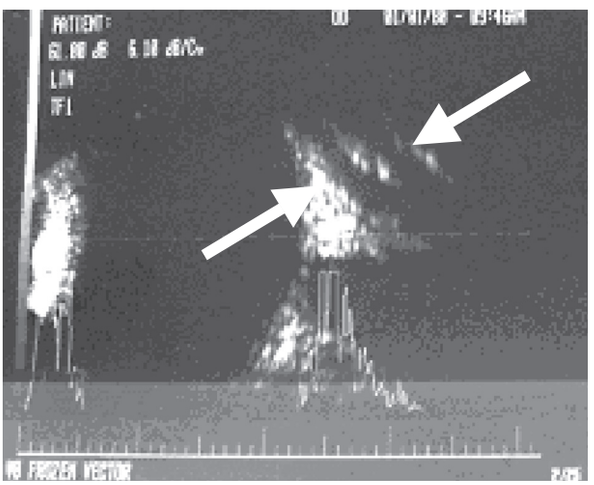

Fig. 2. B scan ultrasound showing enlargement (arrows) of the left rectus superior muscle and levator palpebrae superioris muscle complex.

Discussion

Isolated myositis of the RS+LPS muscle complex is a very rare condition. It is usually manifested by ptosis, edema of the upper eyelid and muscle function damage of a variable degree[3,4]. In our patient, the disease exhibited only mild edema of the eyelid and pseudoptosis with preserved muscle function. The preserved function of the muscle should have most probably been ascribed to the short span of the disease.

In the early stage of the disease, there is minimal enlargement and muscle inflammation which enables preservation of the normal function. During the first week, in untreated myositis further enlargement and muscle inflammation lead to weakening of the function and development of paresis of the muscle. If the disease persists longer or, in other words, changes from acute to chronic phase, it can lead to gradual development of muscle fibrosis and restrictive myopathy[5].

The exact mechanism and etiology of the inflammation are unknown; it has been speculated to be an idiopathic inflammatory condition. Myositis of the extraoc-

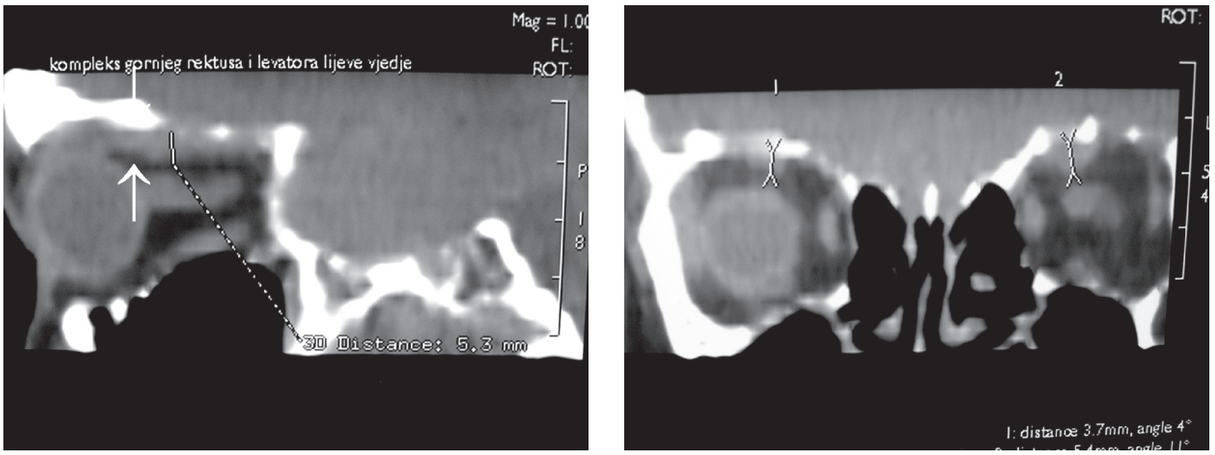

Fig. 3. CT scan showing enlargement (arrow) of the upper rectus muscle and levator palpebrae superioris muscle complex.

ular muscles, however, can follow numerous systemic diseases including rheumatoid arthritis, possibly suggesting an autoimmune mediatory condition[3,6-8]. Cases of orbital myositis development after an acute infection of upper respiratory tract have also been reported[9,10]. In our patient, the disease occurred in otherwise healthy condition, with no previous acute or chronic illness.

The best method in diagnosing orbital myositis is echography[5]. The echographic characteristics of extraocular muscle myositis are low reflexivity and normal inner structure, reflecting inflammatory homogenization of the muscle visible on A-scan; and muscle tube-shaped enlargement due to prominent swelling of the inserting sinew and moderate swelling of the muscle belly visible on B-scan[11].

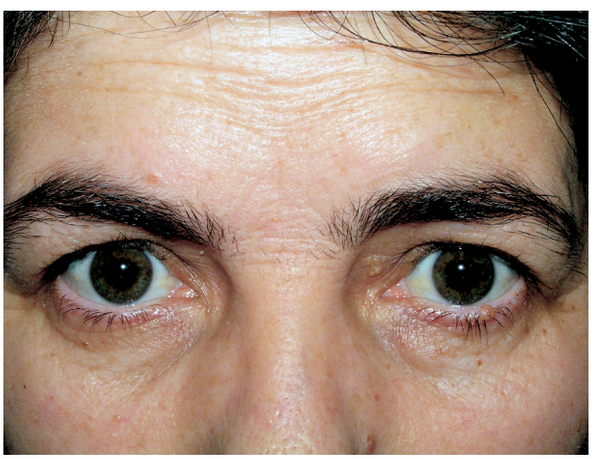

Fig. 4. Normal globe appearance after treatment.

Sometimes episcleritis can be observed near the muscle insertion; the muscle swelling is smaller than the big functional excess in comparison to other muscle disorders[11]. CT and MRI show enlargement of the affected muscle in all its length including the sinew and the muscle belly. Systemic steroid therapy and initial administration of 60-120 mg prednisone with gradual reduction of the dose over several weeks or months are recommended as the essence of orbital myositis therapy.

Prompt improvement is expected within 5 days of therapy introduction[5]. NSAIDs are useful because of their anti-inflammatory effects. Radiotherapy is recommended in patients with severe, refractive inflammation[6,12,13].

Our patient responded with significant alleviation of symptoms after 4-day oral therapy with 60 mg prednisolone and 75 mg indomethacin *per* day.

Conclusion

Even though myositis of the RS+LPS muscle complex is a rare condition, it is important to consider it on differential diagnosis of acquired blepharoptosis, even when the function of the muscle is preserved, which can be seen in the early stages of the disease. The patient presented had acute RS+LPS muscle complex myositis with preserved muscle function and mild palpebral edema with pseudoptosis and orbital pain as the only clinical manifestations. The diagnosis was confirmed by pathognomonic echography and CT findings of isolated enlargement of the RS+LPS muscle com-

plex and prompt improvement upon steroid and NSAID therapy.

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Sažetak

MIOZITIS SPLETA GORNJEG UZDUŽNOG OČNOG MIŠIĆA I PALPEBRALNOG LEVATORA

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Opisuje se bolesnica s akutnim miozitisom spleta gornjeg uzdužnog očnog mišića i palpebralnog levatora (RS+LPS). Žena u dobi od 50 godina primljena je s jednodnevnom anamnezom blažeg otoka vjeđe, pseudoptoze i boli u lijevoj orbiti. Kliničkim pregledom utvrdio se edem vjeđe na lijevom oku, te pseudoptoza s očuvanom funkcijom levatora i očnim pokretima. Nalazi dobiveni ehografijom i kompjutoriziranom tomografijom (CT) pokazali su izolirano povećanje lijevostranog mišićnog spleta RS/LPS. Terapija oralnim kortikosteroidima i NSAID dovela je do potpunog nestanka svih nenormalnih kliničkih i ehografskih nalaza kroz dva tjedna. Izolirani miozitis mišićnog spleta RS+LPS rijetka je bolest, ali je važna za diferencijalnu dijagnozu u bolesnika sa stečenom blefaroptozom, čak i onda kad je mišićna funkcija očuvana. Ovo je bio slučaj akutnog izoliranog miozitisa mišićnog spleta RS+LPS s očuvanom mišićnom funkcijom i blagim otokom vjeđe uz pseudoptozu i bolove u orbiti kao jedinim kliničkim pojavnostima bolesti. Dijagnoza je potvrđena patognomonskim ehografskim i CT nalazom izoliranog povećanja mišićnog spleta RS+LPS i simptomatskim poboljšanjem uz terapiju kortikosteroidima i NSAID.

Ključne riječi: *Miozitis – dijagnostika; Miozitis – etiologija; Miozitis – terapija lijekovima; Bolesti orbite – dijagnostika; Bolesti orbite – terapija lijekovima*