Superior oblique myositis causing acquired Brown's syndrome as the first manifestation of primary Sjögren's syndrome

Sirs,

Orbital myositis (OM) is classified as an orbital inflammatory disease and is defined by inflammation of the extraocular muscles. However, superior oblique (SO) myositis is very rare, to our knowledge, and has never been reported in primary Sjögren's syndrome (pSS). And SO myositis can cause Brown's syndrome characterised by restriction of the SO trochlea-tendon complex which results in limited upgaze of the adducted eye. We describe a case of pSS where SO muscle involvement was the first manifestation and which caused acquired Brown's syndrome.

A 49-year-old woman was referred to the ophthalmic department with a day history of swelling of the left eyelid and periocular pain. She complained of difficulties during elevation of the left eye (LE) in the adducted position. She had a history of hypertension. At presentation, general examination was normal. Ophthalmological examination showed that her visual acuity was 20/20 in both eyes. Her left eyelids were swollen, and the left bulbar conjunctiva was injected. Her LE elevation was restricted to 3- and 4- in adduction (on a scale of 1–4) (Fig. 1A). Computed tomography (CT) of the orbits demonstrated significant enlargement of the left SO muscle and the posterior part of its tendon (Fig. 1B and 1C).

Biological examination showed an increased erythrocyte sedimentation rate at 38 mm/hr and C-reactive protein at 0.18 mg/l(normal 0.47 mg/l). Thyroid function was normal, and thyroid-stimulation hormone-receptor antibody and anti-microsomal antibody were both negative. Antinuclear antibody was strongly positive (3+) and anti SS-A/Ro, SS-B/La antibodies were moderate (63.9 unit) and weakly (29.5 unit) positive, respectively. Complement factor C3 and C4 were normal at 92 mg/dl(normal 76–139 mg/dl) and 16 mg/dl(normal 12-37 mg/dl), respectively. Serum immunoglobulin G (IgG) rate was increased at 2260 mg/dl(normal 870–1700 mg/dl), but IgG4 serum level was low at 910 mg/L(normal 30–2010 mg/dl). Labial minor salivary gland biopsy confirmed focal lymphocytic sialadenitis and salivary gland scintigraphy revealed the relatively impaired function of both parotid glands.

A diagnosis of left SO myositis causing Brown's syndrome secondary to pSS was made. Oral prednisolone 40 mg/day and hydroxychloroquine 200 mg were initiated. Over the ensuing 3 months, her clinical condition improved.

We report a case of SO myositis and Brown's syndrome as the first manifestation of pSS. Recognized causes of OM are autoimmune thyroid disease, systemic lupus erythematosus, anti-neutrophil cytoplasmic antibody-associated vasculitis and sarcoidosis (1-3). However, orbital myositis of the SO has been reported to occur in only 2% of patients with the orbital inflammatory syndrome (4). This could be due to the relative lack of muscle fibers in strap muscles or because of under-reporting due to technical difficulties in imaging (5).

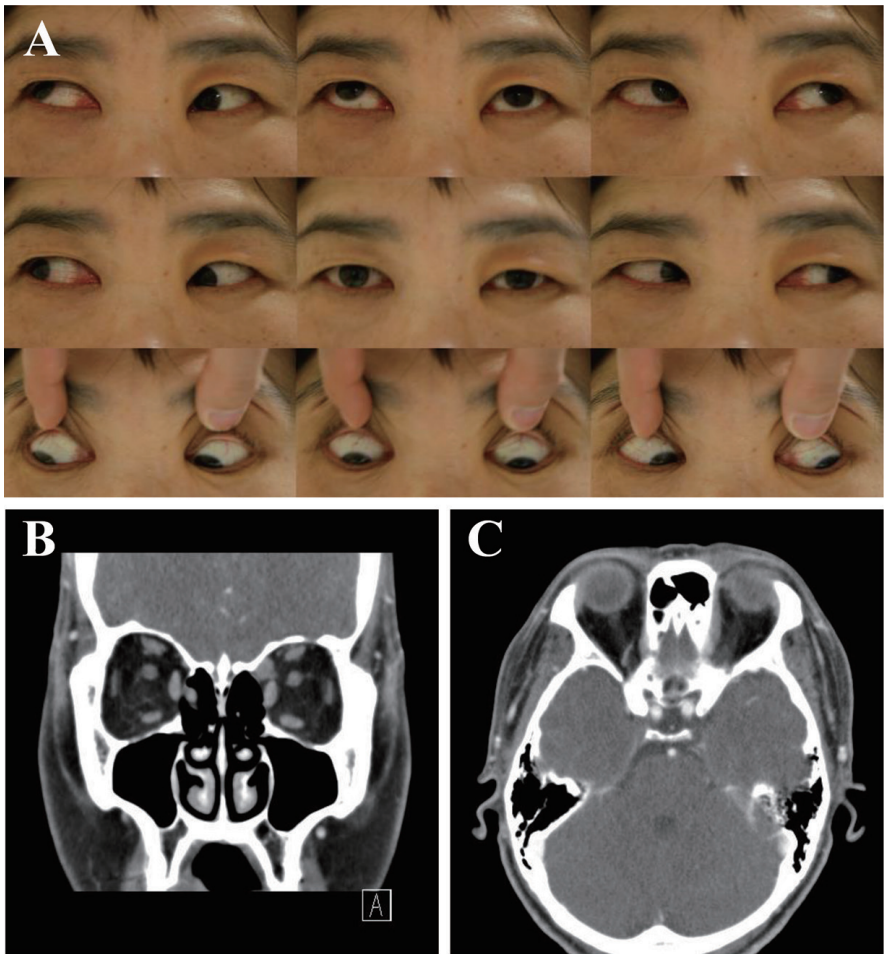

Fig. 1. Clinical manifestations and image study of this case. (**A**) The patient had a restriction in the left eye elevation in the adducted position. (**B**) Coronal and (**C**) axial images of the orbital CT scan show left superior oblique muscle enlargement with involvement of the posterior part of the tendon.

Brown's syndrome is caused by mechanical limitation of the SO tendon. In this case, an orbital CT scan showed thickening of the SO muscle involving the posterior part of the tendon, which caused mechanical limitations. Treatment of both inflammatory OM and Brown's syndrome depend on the cause. Systemic steroids, non-steroidal anti-inflammatory drugs and local steroid injections have been used successfully (6, 7).

Sicca features are the central clinical manifestations of pSS. However, recent studies have confirmed that pSS has a systemic expression, including extraglandular manifestations that contribute to confusion and delay in diagnosis (8). The clinical significance of muscular involvement in pSS has also been reported. Inflammatory myositis or inflammatory perivascular infiltrates have been confirmed by muscle biopsy (9). Although any of the extraocular muscles can be involved in pSS, the literature regarding the incidence of individual extraocular muscle involvement as a consequence of pSS is sparse. Although various ophthalmic features have been reported in pSS, our case suggest that uncommon ocular manifestations and extraocular muscle involvement could be the first signs of pSS.

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