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Letter to the Editor

Levator palpebrae superioris myositis: An uncommon cause of ptosis

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Dear Editor,

We would like to report a case of unilateral ptosis due to levator palpebrae superioris myositis. It illustrates an uncommon cause of a common neurological symptom.

A 48 year old man presented to us in October 2018 with acute left partial ptosis for one day. There were no diplopia, headache or orbital pain, red eye or change in visual acuity. He did not complain of excessive lacrimation. His past health was unremarkable and was not using any long-term medication.

On examination, there was partial ptosis with impaired levator function over the left eye. Pupils were equal and reactive to light. Visual acuity was unremarkable. No ophthalmoplegia could be demonstrated. There was no lid lag on downgaze. There was no anhidrosis nor fatigability. The eyelids had no erythema. Other cranial nerve examinations were all normal. Neurological examination of the limbs was unrevealing.

Serum autoimmune antibody screening showed negative anti-acetylcholine acetylcholine receptor antibody, anti-nuclear antibody, anti-neutrophil cytoplasmic antibody and anti GQ1b IgG. Serum thyroid function was normal. Fasting glucose did not suggest diabetes. Tensilon test was negative. Nerve conduction study was normal. Repetitive nerve stimulation test did not suggest neuromuscular junction disease.

Brain CT and CT cerebral angiogram showed no significant finding. Brain MRI with gadolinium contrast was done and reported to be negative. Orbit MRI was subsequently done. Hyperintense T2W signal with contrast enhancement was seen over left levator palpebrae superioris muscle with mild involvement of the superior rectus muscle involving its distal portion (Fig. 1). Diagnosis of left orbital myositis (OM) with levator palpebrae superioris involvement was made. The ptosis resolved completely after about one week before any treatment was given.

OM is the inflammation of orbital extraocular muscles including the levator palpebrae superioris. It is under the subgroup of idiopathic orbital inflammatory syndrome. It has a female predominance and most commonly affects middle aged adults [1,2]. The pathogenesis is still unknown, but an autoimmune mechanism has been proposed [3]. Associations with systemic diseases such as rheumatoid arthritis, systemic lupus erythematosus, sarcoidosis and ANCA associated vasculitis have been reported. Common presentations include ptosis, diplopia, conjunctival injection and eye pain exacerbated by eye movement. Less common features include visual disturbance and proptosis [4–6]. Ptosis can be due to orbital oedema, as a result of inflammation, or direct involvement of levator palpebrae superioris. Lateral, superior and medial recti are more commonly involved than inferior rectus and oblique muscles [2]. Isolated involvement of these extraocular muscles is relatively common, reported to be 68% in a study of 75 OM patients [7]. However, isolated involvement of levator palpebrae superioris is relatively rare [2,8,9]. A recent review has classified OM into typical and atypical forms. Typical form is usually acute with good responsiveness to steroid. Atypical form includes idiopathic chronic or recurrent OM associated with systemic autoimmune and infective conditions [10]. Our case probably belongs to the typical form as the presentation was an acute single episode and was not associated with systemic autoimmune or infections. For treatment of OM, systemic corticosteroid is the mainstay option [11,12]. However, treatment was not required in our patient, nor was it required in two other patients with isolated levator myositis reported before [5,6]. Although there are proposed management algorithms for OM, there is lack of support by literature data on treatment efficacy, especially in accordance with severity and phases of the disease.

Our case highlights that destinated orbit MRI can be very useful in neuro-ophthalmological diagnosis. Orbit MRI can give an excellent view of the optic nerve and extraocular muscles. Fat suppression sequences of orbit MRI are ideal to look for abnormalities in the orbit which is a fatrich structure. The classical MRI findings of OM include contrast enhanced thickening of extraocular muscle(s), usually involves surrounding fat, tendon and myotendinous junction [13]. Orbit MRI imaging also allows us to distinguish OM from few differential diagnoses that affect extraocular muscles. The first is thyroid orbitopathy, which typically spares the myotendinous junction [2]. The second is orbital cellulitis, which usually shows sinus involvement and subperiosteal abscess in about half of the cases [14]. The third is orbital lymphoma, which may also appear as hyperintense T2W lesion at extraocular muscle, but is usually brighter on DWI and has lower ADC values compared to OM [2]. Orbit MRI should be considered for cases with relevant neuro-ophthalmology presentation especially if brain MRI is

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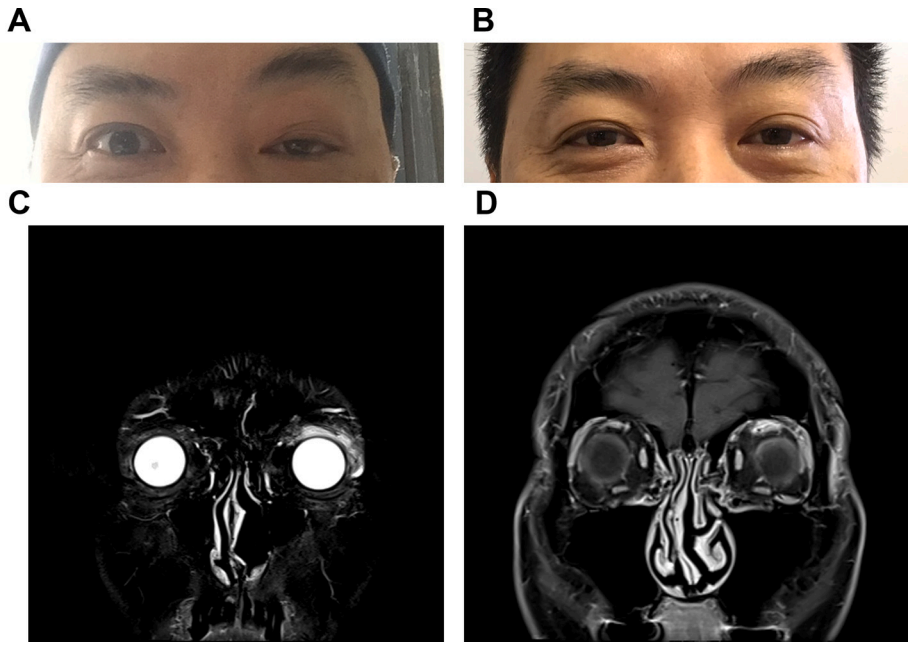

Fig. 1. Caption: (A) and (B) Initial presentation of left eye incomplete ptosis which spontaneously resolved after about one week. (C) Orbit MRI coronal T2-weighted fat-suppressed image showing hyperintense T2 signal in the left levator palprebrae superioris (D) Orbit MRI coronal section showing contrast enhancement of left levator palpebrae superioris in post contrast T1-weighted fat-saturated sequence.

negative.

In short, OM could be an uncommon cause of ptosis. Prognosis is usually favorable. Orbit MRI is the investigation of choice, especially if other neuroimaging results are negative.

Declarations of interest

None.

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