LETTERS TO THE EDITOR

Acquired Brown's syndrome and primary Sjorgren's syndrome

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Brown's syndrome is characterised by limitation of elevation of the adducted eye. This disorder has been described in association with a number of connective tissue disorders including rheumatoid arthritis,¹ juvenile chronic arthritis,² and systemic lupus erythematosus.³ We report here the case of a patient with primary Sjorgren's syndrome who developed unilateral Brown's syndrome. To our knowledge this association has not been described previously.

Case report

A 21-year-old woman presented with a 1 day history of an acute onset of diplopia noted when she looked upwards. She had been diagnosed as having primary Sjorgren's syndrome 1 year before this presentation, based on objective evidence of dry eyes and mouth, bilateral parotid swelling, and antibodies to Ro and La antigens. Current medication was chloroquine 250 mg orally on alternate days which she had been taking for 1 year. For her dry eye symptoms she was using artificial tears (four to six times a day).

At presentation general and neurological examinations were normal. Her visual acuity was 6/5 (aided) in both eyes. Examination of eye movements revealed heterotropia in the primary position without diplopia. There was no sign of an abnormal head posture. On attempted dextro-elevation of the eyes she complained of diplopia.

The left eye failed to elevate in adduction in both versions and ductions. Hess chart (Fig 1) analysis was consistent with the ocular movement examination – that is, V pattern exotropia, no vertical deviation in the primary position, and no overaction of the other ocular muscles. The left trochlea was prominent to palpation compared with the right at the time of presentation but was not tender. Forced duction test (under topical local anaesthesia as described by Mein and Trimble⁴) was positive, the left eye exhibited significant limitation of passive elevation in adduction compared with the right eye.

At the time of her presentation, her erythrocyte sedimentation rate (ESR) was 43 mm/h, C-reactive protein 96 mg/l (normal up to 1.7 mg/l) indicating an acute inflammatory response. Computed tomography of the orbits and cranial contents did not reveal any abnormality.

Acquired Brown's syndrome was diagnosed. She was initially treated with oral prednisolone 60 mg daily and this was gradually tapered off to 10 mg daily over 6 weeks. Chloroquine was discontinued. She was commenced on oral azathioprine 50 mg daily as the dosage of prednisolone was lowered. ESR and C-reactive protein levels returned to her normal levels on commencement of the above treatment. Two months after presentation, the diplopia and ocular motility abnormality has decreased since her initial presentation (Fig 2). The patient had

Figure 1 Hess chart at presentation.

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Figure 2 Hess chart 2 months after presentation.

only minimal diplopia in dextrolevation which was not troublesome.

Comment

Brown⁵ described the superior oblique tendon sheath syndrome which simulates an isolated inferior oblique palsy. Though this patient did not have a clocking sensation when she dextro-elevated the eye (which has been described in Brown's syndrome) other features in her examination and investigations – V pattern exotropia, absence of vertical deviation in the primary position, positive forced duction test, absence of a compensatory head posture, failure of the eye to elevate in ductions and versions, and absence of secondary muscle sequelae are consistent with Brown's syndrome.

An isolated left inferior oblique palsy is a differential diagnosis of Brown's syndrome and its features are an A pattern exotropia, development of muscle sequelae, significant vertical deviation in the primary position, development of muscle sequelae, marked abnormal head posture, and negative forced duction test.

Chloroquine toxicity resulting in a left inferior oblique palsy was considered as a differential diagnosis. Chloroquine is a neuromyotoxin⁶ affecting cardiac muscle, skeletal muscle, and nerve, in addition to its retinal toxicity. In muscle it produces a vacuolar myopathy,⁶ which is reversible after discontinuation of chloroquine. There have been no reports of ocular muscle chloroquine toxicity in the literature.

Various theories have been proposed to explain this syndrome and both Sandford-Smith⁷ and Mein⁸ in their reviews have suggested stenosing tenosynovitis to be the most likely cause. Tenosynovitis is also a common feature of primary Sjogren's syndrome,⁹ and we propose

that inflammation in the tendon sheath of the superior oblique muscle was responsible for this abnormal ocular movement in this patient.

Acquired Brown's syndrome is usually self-limiting, some cases of Brown's syndrome have resolved with systemic and local steroid treatment.¹⁰ The improvement of the ocular motility abnormality in this patient with systemic steroids suggests tenosynovitis of the superior oblique tendon sheath but definite proof is lacking.

Primary Sjogren's syndrome may present with many different ophthalmic features including keratoconjunctivitis sicca, uveitis, optic neuritis, retinal vasculitis. Brown's syndrome should now be added to this list.

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