LETTERS TO THE EDITOR

Brown's syndrome: an important cause of diplopia in systemic lupus erythematosus

Brown's syndrome is caused by a mechanical limitation of the superior oblique tendon. This simulates a palsy of the inferior oblique muscle with limitation of elevation in adduction. The syndrome can be congenital or acquired. Acquired Brown's syndrome has been described in collagen vascular diseases including rheumatoid arthritis and Still's disease.1,2 Two case reports have described Brown's syndrome in systemic lupus erythematosus (SLE) in adults,3,4 but no cases of Brown's syndrome in a juvenile with SLE have been reported to date.

A 14 year old girl was referred to the ophthalmic department with a two day history of diplopia and arthalgia of her hands. She had a three year history of SLE that had been complicated by rashes, arthritis, cerebral vasculitis (two years previously), and was recently under investigation for haematuria. She had been receiving prednisolone in variable doses since the diagnosis was made.

On general examination, the patient had a butterfly rash, cshingoid features, and mild swelling of the small joints of her hands. Ocular examination revealed visual acuities of 6/6 in both eyes. Her eye movements showed restricted elevation in adduction of both eyes. Papillary responses and other ocular examination were normal. There was tenderness over the left trochlear, but no clicking sound or sensation noted on eye movement. A Hess chart revealed limited elevation in adduction, with no superior oblique overaction of the left eye, consistent with a left Brown's syndrome (figure). The patient declined a forced duction test.

Laboratory studies were consistent with active SLE: leucocyte count 2.8 x 10^9/l (neutrophils 2.0 x 10^9/l, lymphocytes 0.7 x 10^9/l); antinuclear factor was increased to 1600 and DNA antibodies to 1650 IU/ml (normal range up to 60 IU/ml). Serum complement component C3 was 0.64 (normal range 0.8–1.9 g/l) and C4 was <0.08 (normal range 0.12–0.4 g/l).

A diagnosis of Brown's syndrome secondary to SLE was made. The patient's steroids were increased from 5 mg of prednisolone on alternate days to 30 mg per day. Her symptoms improved in one week and completely resolved by two weeks, confirmed by normal Hess test.

Six weeks later, renal investigations showed a plasma creatinine of 70 μmol/l, plasma albumin 43 g/l and a normal protein/creatinine index. Renal biopsy revealed mild lupus nephritis (World Health Organisation type 2A). During this hospital stay she had a further episode of vertical diplopia. Computed tomography (CT) and magnetic resonance imaging scans of her head, for presumed intracranial cause, revealed no local or intracranial pathology. With the combination of diplopia and increased markers for SLE activity, intracranial involvement was assumed and a course of cyclophosphamide was subsequently given on an outpatient basis. Although no ophthalmic assessment was made, a recurrence of her Brown's syndrome was a possibility, and this diagnosis may have prevented the need for cyclophosphamide.

The causes of Brown's syndrome can be divided into two main categories—inflammation and trauma—both involving the muscle, tendon, trochlear, or surrounding tissue of the superior oblique muscle.1

Two descriptions of Brown's syndrome associated with SLE have been of a 27 year old woman and a 30 year old man,3,4 the former successfully treated with a combination of indomethacin and prednisolone orally, the latter with intravenous alone. In our patient, inflammatory markers indicated active SLE and her diplopia was resolved by increasing her oral steroid.

Diplopia in patients with SLE has usually been attributed to nervous system disease caused by a microvasculitis. The exact cause for Brown's syndrome in SLE is unknown, but is thought to be a tenosynovitis of the superior oblique tendon.

Clinically, patients with Brown's syndrome present with vertical diplopia that is worse on adducting and elevating the affected eye, often with associated pain and tenderness in the trochlear region. Examination, Hess chart, and forced duction test confirm restricted elevation in adduction. An orbital CT scan may reveal local pathology, but this is not always the case.

Treatment of inflammatory Brown's syndrome is dependent on the cause. In addition to oral steroids and non-steroidal anti-inflammatory drugs,3,4 local steroid injections have been used successfully.5 In those patients who fail to respond, superior oblique tenotomy combined with inferior oblique recession has been advocated.6

This case report illustrates the importance of involving the ophthalmologist in cases of SLE with diplopia. The true incidence of Brown's syndrome in SLE is probably much greater than the few reported cases would suggest. Ophthalmic assessment would help in distinguishing between nervous system disease and this local mechanical problem found in these patients.

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Mechanism of haematuria in lupus nephritis

While the renal manifestations of systemic lupus erythematosus (SLE) vary with the severity of the renal lesion, proteinuria is present in the vast majority of cases. Haematuria, which is less common, is usually microscopic, but may be macroscopic in an occasional patient. Haematuria seems to result from the passage of red blood cells through anatomical gaps in the glomerular basement membrane (GBM). While such gaps have been described in patients with various forms of glomerulonephritis, including those with lupus nephritis associated with haematuria,7 the passage of red blood cells (RBCs) through these gaps has been demonstrated only in patients with focal proliferative glomerulonephritis, mild proliferative glomerulonephritis, 7,8 membranous nephropathy,7 and mesangial proliferative nephropathy.7 Passage of RBCs has also been demonstrated in our model of experimental glomerulonephritis.7 We now report a patient with SLE and lupus nephritis in...