Acquired Brown's syndrome in a patient with systemic lupus erythematosus

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Abstract

A 27 year old woman with systemic lupus erythematosus (SLE) developed vertical diplopia with an apparent bilateral inferior oblique muscle palsy, resulting in a limitation of elevation of the globe in adduction. It resolved with systemic steroid treatment. A transient tenosynovitis affecting the superior oblique tendons was the probable underlying pathological mechanism. This is the first described case of Brown's syndrome associated with SLE.

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Brown's syndrome is characterised by limitation of elevation of the adducted eye. It is due to a mechanical limitation of movement of the superior oblique tendon. Clinically, it simulates a palsy of the inferior oblique muscle with vertical diplopia on upward and inward gaze. This disorder has occasionally been described in association with inflammatory collagen vascular diseases including rheumatoid arthritis, juvenile chronic arthritis, and Still's disease.

We report here the case of a patient with systemic lupus erythematosus (SLE) who developed bilateral Brown's syndrome. To our knowledge this association has not previously been described.

Case report

The patient, a 27 year old woman, presented with a six day history of diplopia. She had a history of SLE which was first diagnosed four years before her present admission. She was then treated with prednisone by mouth and her disease went into remission and remained stable with a low daily dose of prednisone for three years, at the end of which the drug was stopped. One month before admission she had had fever and arthritis in her wrists and left ankle, which resolved on treatment with naproxen.

On admission, the patient was asymptomatic except for diplopia. This was at a maximum on upward and lateral gazes. Ophthalmological examination showed that she was unable to elevate her eyes completely when they were in the adducted position (figs 1 and 2). All other ocular movements were intact. Pupillary responses and fundusoscopic examination were normal. No clicking sound or sensation on movement of the eyes was elicited. The red glass test and a Hess chart were consistent with the ophthalmological examination. An edrophonium chloride (Tensilon) test was negative. Physical examination was otherwise normal.

Laboratory studies showed a haemoglobin concentration of 105 g/l, a leucocyte count of $3.6 \times 10^9/l$ with 29% lymphocytes, a platelet count of $440 \times 10^9/l$, and erythrocyte sedimentation rate of 56 mm/hour (Westergren). Plasma protein electrophoresis showed a slight increase in the α_2 , β , and γ globulins.

Serum complement component C3 was 610 mg/l (normal 550–1200) and C4 was 140 mg/l (normal 200–500). Serological findings included a positive antinuclear antibody titre (1/640) and a positive titre of antibodies to double stranded DNA (1/160). IgG anticardiolipin antibody was positive using a enzyme linked immunosorbent assay (ELISA). Coagulation studies were compatible with the presence of lupus anticoagulant. Results of the following studies were within normal limits: biochemical profile, urine analysis, thyroid function, Venereal Disease Research Laboratory (VDRL) test, rheumatoid factor, antibodies to Ro (SS-A), La (SS-B), Sm, RNP, and cerebrospinal fluid analysis. Computed tomography and magnetic resonance imaging of her orbits and head showed no abnormality.

Figure 1 Limitation of elevation of the right eye in adduction.

Figure 2 Limitation of elevation of the left eye in adduction.

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The patient was diagnosed as having Brown's syndrome. She was initially treated with indomethacin 50 mg every eight hours for three weeks, but there was no benefit. She was then given prednisone, 20 mg daily, with rapid resolution of the diplopia within 48 hours. Over the next few weeks the dose was gradually tapered and one year after starting prednisone she is free of symptoms, except for occasional diplopia on extreme upward gaze, and receives 7.5 mg/day prednisone.

Discussion

In 1950 Brown described the superior oblique tendon sheath syndrome, subsequently known as Brown's syndrome.¹ It is the most common cause of an isolated apparent palsy of the inferior oblique muscle.

The oblique muscles have a complementary action in moving the eyes in the vertical plane. Normally, when the inferior oblique muscle moves the globe upward and inward, the superior oblique muscle relaxes. If the superior oblique tendon cannot lengthen or slide freely, the affected eye cannot be raised completely in full adduction. The patient reports diplopia on upward gaze.

Various theories have been proposed about the nature of the abnormality. Brown initially suggested that the syndrome was a developmental anomaly following congenital paralysis of the inferior oblique muscle, producing a contracture of its check ligament, the sheath of the anterior segment of the superior oblique.¹ Later, as an attempt to explain specifically the intermittent cases, those associated with a clicking sound or sensation and those showing spontaneous resolution, it was suggested that there is a circumferential constriction of the trochlea and sheath preventing a locally enlarged tendon from sliding freely in the sheath.²⁻⁴ The various theories of the aetiology of the syndrome were well reviewed by Sandford-Smith,⁵ who was the first to suggest the marked anatomical and clinical similarities between the tendon sheath syndrome and stenosing tenosynovitis.⁴

Mein reviewed 49 cases of Brown's syndrome and found that swelling of the posterior part of the superior oblique tendon was the most common mechanism, and also considered stenosing tenosynovitis to be the most likely cause of the tendon sheath syndrome.⁶

Despite the common association of tenosynovitis at other sites with rheumatoid arthritis (for example, trigger finger), this particular complication has only been reported in a few patients with rheumatoid arthritis,^{4 7-11} juvenile chronic arthritis,¹⁰⁻¹⁴ or adult Still's disease.¹⁵ The disorder is usually self limited and transitory, and specific treatment is not usually required. Some cases have resolved with systemic steroid treatment, however,^{9-12 15} and an adult patient with rheumatoid arthritis with

bilateral symptoms of Brown's syndrome showed a resolution of symptoms only after local corticosteroid injections into the region of the trochlea.⁸

Brown's syndrome has been also described in association with various non-rheumatic disorders including frontal sinusitis or frontal sinus surgery,¹⁶ trauma,¹⁷ focal metastatic lesions,¹⁸ Sneddon's syndrome,¹⁹ and hypogammaglobulinaemia.²⁰

Although swelling of the superior oblique tendon has occasionally been noted on computed tomography scanning of the orbits,^{14 15 20} in our patient computed tomography and magnetic resonance imaging findings were normal. The resolution of Brown's syndrome associated with the use of steroids, however, points to a tenosynovitis of the superior oblique tendons as the cause, though definitive proof is lacking. A true and selective palsy of the two inferior oblique muscles is exceptional.

Diplopia in patients with SLE has generally been attributed to nervous system disease. In our opinion, it is important to consider Brown's syndrome as a possible cause of diplopia in patients with SLE.

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