Case report

Transient Brown's syndrome in juvenile chronic arthritis

A. S. KEMP, CATHIE SEARLE, AND SUSAN HORNE

From the Departments of Immunology and Orthoptics, Royal Alexandra Hospital for Children, Camperdown, Sydney, NSW 2050, Australia

SUMMARY A 9-year-old boy with systemic juvenile chronic arthritis and Brown's syndrome (limitation of elevation of the adducted eye due to limitation of movement of the superior oblique tendon) is described. The resolution in association with steroid treatment suggested a transient tenosynovitis involving the superior oblique tendon as the cause.

Brown's syndrome is a limitation of elevation of the adducted eye due to limitation of movement of the superior oblique tendon.¹ Most cases in childhood are congenital, though acquired cases have been described.² This syndrome has occasionally been described in adults with rheumatoid arthritis,^{3, 4} and Jacobs mentions a case in childhood.⁵ We report a case of acquired Brown's syndrome in systemic juvenile arthritis which resolved in association with steroid treatment.

Case history

A 9-year-old boy developed systemic juvenile arthritis characterised by a rash, fever, and arthritis involving the wrists, ankles, elbows, and small joints of the hands and fingers. Treatment with the non-steroidal anti-inflammatory drugs, aspirin and Naprosyn (naproxen), failed to control the disease. Six months after the onset of the disease he was admitted to our institution for further treatment. Investigation at that time showed an ESR of 78 mm/h Westergren, C-reactive protein 21.5 mg/dl (<0.8 mg/dl) (mg/dl $\times 10 =$ mg/l), negative antinuclear factor and rheumatoid factor and raised liver enzymes, SGPT 307 U/l (10-50), and SGOT 298 U/l (10-40). While on anti-inflammatory treatment with salicylate, 100 mg/kg per day, and Naprosyn, 125 mg twice daily, he complained of double vision and pain over the inner angle of the left eye on looking upwards and to the right. Examination of his eyes

showed limitation of elevation in adduction of the left eye and a Hess chart (Fig. 1) confirmed a pattern of eye movements typical of Brown's syndrome. No swelling above the medial canthus in the region of the trochlea and no clicking sound or sensation on movement of the eye were elicited. Prednisolone 10 mg on alternate days was added to

Fig. 1 Hess chart, left eye, at time of presentation.

Fig. 2 Hess chart, left eye two weeks later.

his treatment regimen. Two weeks after the patient presented the ocular symptoms had improved and the Hess chart was normal (Fig. 2). On review two months later there was still evidence of active systemic disease, with a rash, arthritis, and fever; however, the ocular symptoms had not recurred.

Discussion

Brown's syndrome may be congenital or acquired. Shortening of the anterior sheath of the superior oblique tendon restricting the elevation of the adducted eye by the inferior oblique muscle has been considered to be the mechanism of congenital Brown's syndrome,¹ though such a defect was not

confirmed at operation in a series of 25 cases in childhood.⁶ Rarely cases have been intermittent in nature, and it has been proposed that tenosynovitis involving the trochlea and tendon sheath, together with an associated tendon swelling, limits movement of the superior oblique tendon through the trochlea.⁷ In some cases a clicking sensation with a variable weakness has been observed, and it is suggested that the click is due to the swollen tendon passing through the trochlea.⁷ This feature was observed in the childhood case associated with systemic juvenile arthritis mentioned by Jacobs.⁵

The present case had symptoms for two weeks, and resolution was associated with the introduction of alternate-day prednisolone. A previous case associated with bilateral symptoms in an adult with seropositive rheumatoid arthritis persisted for one year, but it resolved after local corticosteroid injections into the region of the trochlea.⁴ Tenosynovitis is common in systemic juvenile arthritis, particularly around the wrists, but may involve other tendons.⁵ The transient nature of Brown's syndrome and the resolution associated with the use of steroids point to a transient tenosynovitis of the superior oblique tendon as the cause in the present case.

References

- 1 Brown H W. Congenital structural muscle anomalies. In: Allen J H, ed. *Strabismus: Ophthalmic symposium*, St Louis: Mosby, 1950: 205.
- 2 Scott A B, Knapp P. Surgical treatment of the superior oblique tendon sheath syndrome. *Arch Ophthalmol* 1972; **88**: 282-6.
- 3 Killian P J, McClain B, Lawless O J. Brown's syndrome: an unusual manifestation of rheumatoid arthritis. *Arthritis Rheum* 1977; **20**: 1080-4.
- 4 Beck M, Hickling D. Treatment of bilateral superior oblique tendon sheath syndrome complicating rheumatoid arthritis. *Br J Ophthalmol* 1980; **64**: 358-61.
- 5 Jacobs J C. *Paediatric rheumatology for the practitioner*. New York: Springer, 1983.
- 6 Parks M M, Brown M. Superior oblique tendon sheath syndrome of Brown. *Am J Ophthalmol* 1975; **79**: 82-6.
- 7 Sanford-Smith J H. Superior oblique tendon sheath syndrome and its relationship to stenosing tenosynovitis. *Br J Ophthalmol* 1973; **57**: 859-65.