ACQUIRED BROWN'S SYNDROME: A CASE REPORT*

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Abstract

A case of Brown's syndrome occurring with chronic systemic juvenile rheumatoid arthritis is presented. The patient, a nine year old child, had a sudden onset of diplopia in dextro-elevation occurring simultaneously with a severe inflammatory attack involving several limb joints. He received systemic anti-inflammatory medication which improved his general condition, and coincided with the Brown's syndrome being resolved within three weeks without any specific ocular treatment.

It is suggested that tenosynovitis, which is commonly a feature of juvenile rheumatoid arthritis (JRA), may have been the cause of this patient's Brown's syndrome.

It is recommended that a thorough assessment of ocular motility be included in the ophthalmic examination of young children with JRA.

Key words: *Chronic systemic juvenile rheumatoid arthritis, superior oblique tendon sheath syndrome, pseudoparalysis of the inferior oblique muscle, tenosynovitis, polyarticular arthritis.*

INTRODUCTION

Brown's syndrome may be described as a mechanical limitation of elevation in adduction, of one or both eyes, due to a congenital[1-5] or acquired[3-7] defect of the superior oblique tendon, and/or its sheath. Resolution[8,9] and recurrences[3,6,7] have been reported in both congenital and acquired cases.

This paper presents a case history of acute Brown's syndrome in a child with chronic systemic juvenile rheumatoid arthritis (CSJRA).

Iritis and iridocyclitis are the usual ophthalmic disorders associated with rheumatoid arthritis.[10] Smith,[3] Scott and Knapp,[4] Terrell,[6] and Beck and Hickling,[7] however, have reported acute Brown's syndrome occurring in patients with rheumatoid arthritis.

CASE HISTORY

A nine year old boy was diagnosed as having CSJRA in February 1983. At this time he had a papular rash on his hands, arms, legs and lower abdomen, slight fever and stiffness of the limb joints.

In April 1983, he was admitted to hospital with swollen metatarsals, a painful right ankle and knees which showed bilateral popliteal fullness with tender left popliteal fossa and limitation of flexion and extension. He also had tender temporo-mandibular joints, shoulders, elbows and wrists.

Three days after admission, the patient reported seeing double when he looked up to the right.

An ocular motility examination at this time revealed the following:

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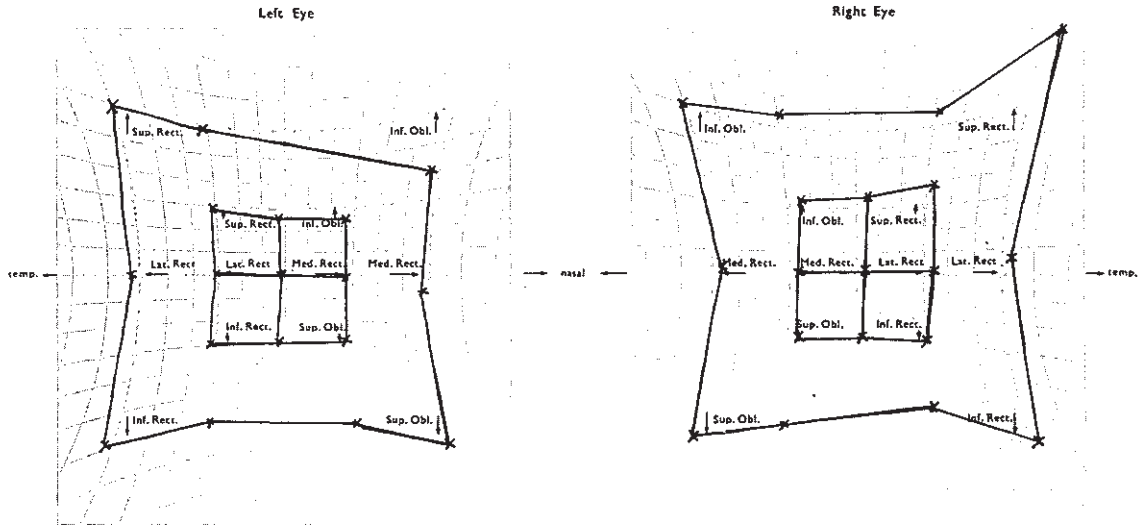

Figure 1: Hess chart showing restriction of the left inferior oblique muscle.

Abnormal head posture: slight elevation of the chin, face turn left, head tilt right.

Cover test 1/3 m: small exophoria with rapid recovery.

Cover test 6 m: orthophoria.

Ocular movements: limitation of elevation in adduction of the left eye, with a corresponding overaction of the right superior rectus.

Visual acuity: right and left = 6/5, N5

One week later a Hess chart was plotted showing a pattern of eye movements typical of Brown's syndrome (Figure 1).

An ophthalmic assessment was also performed. Fundi and media were normal, with no significant refractive error. Palpation of the orbital area above the medial canthus, in the region of the trochlea, revealed no sign of swelling. No tenderness of the area was reported.

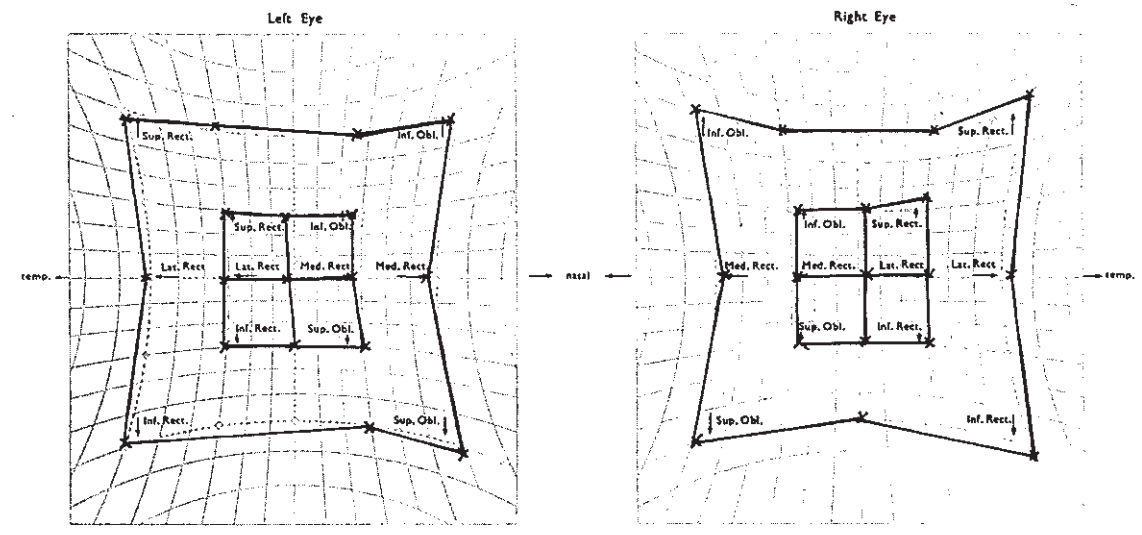

Figure 2: Hess chart showing improvement of left inferior oblique action following systemic treatment for JRA.

A stethoscope was used in an attempt to identify a clicking sound or sensation, which may have suggested the presence of a nodule on the tendon. No such sound was identified nor was a clicking sensation reported by the patient.

Systemic treatment consisted of prednisolone (dosage of 10 mg alternating with 2.5 mg per day); naprosyn (125 mg every 12 hours); and atoxiprin (900 mg every six hours). No specific ocular treatment was given.

Seven days later, the patient's ocular symptoms had significantly diminished: he complained of diplopia only on extremes of dextro-elevation. On examination, there was only a very slight restriction of elevation in adduction of the left eye (Figure 2).

The patient was discharged 16 days after admission. At this time the eye condition seemed to have resolved. There was no abnormal head posture, he had a full range of eye movements and he had no pain or diplopia on dextro-elevation.

DISCUSSION

Juvenile rheumatoid arthritis (JRA), one of fifty or more types of arthritis, is a disease that not only causes swelling and injury to the joints but affects many other parts of the body as well.[1] Our patient was diagnosed as having systemic onset JRA which occurs in about thirty percent of JRA patients and may be characterised by high fever, a rheumatoid rash, polyarticular arthritis, heart, liver, spleen and lymph node involvement and occasionally iridocyclitis.[1] Systemic onset JRA differs from Still's Disease, which is a pauciarticular type of JRA, in that the latter is characterised by having fewer than five joints involved and by having iritis as a common finding.

Tenosynovitis (i.e. frequent inflammation, or involvement of the tendon sheaths of the muscles) may also accompany JRA.[1] Both Mein[5] and Sandford-Smith[2] have postulated stenosing tenosynovitis as a cause of Brown's syndrome. The anti-inflammatory medication administered to our patient relieved the tenosynovitis (as well as the other symptoms and signs of JRA), at the same time as the Brown's syndrome resolved. We suggest, therefore, that tenosynovitis induced Brown's syndrome in our patient.

There are several differences between this patient and those discussed in previous reports of Brown's syndrome occurring in association with arthritis.[2-4] Earlier studies report patients as being adults whose arthritic condition was long-standing. Their ocular motility defects persisted for a much longer period of time, and in some cases, the eye condition was noted to recur—the recurrences coinciding with inflammatory attacks. In most cases, both the general and the ocular condition proved to be difficult to control with medication.

In contrast, our patient was nine years old. After the commencement of antiflammatory treatment, his general as well as his ocular condition improved. The limitation of elevation in adduction of the left eye had almost resolved two weeks after the onset of ocular symptoms. However, because recurrence has been noted in other cases, the possibility of Brown's syndrome recurring has been an important consideration for our patient's long-term management.

CONCLUSION

Iritis and iridocyclitis should not be the only ocular complications to be considered in the investigation of children with JRA. Assessment of the extra-ocular muscle movements should form an important part of the ophthalmic examination.

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