Bilateral Acquired Inflammatory Brown’s Syndrome

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ABSTRACT

An 11-year-old boy presented with fever, skin rash, joint pains and vertical diplopia, and was found to have a right Brown’s syndrome and systemic onset Juvenile Rheumatoid Arthritis (JRA). Later in the course of his illness, he developed a left Brown’s syndrome. Acquired Brown’s syndrome may occur as a complication of JRA during exacerbations of the systemic inflammatory disease.

Introduction

The superior oblique tendon sheath first described by Brown1 in 1950 is characterized by an inability to elevate the affected eye in adduction. In full adduction the eye cannot be elevated above the midline, either actively or passively. There is little, if any, overaction of the ipsilateral superior oblique muscle, although there is often mild down-drift of the affected eye in adduction. The traction test is unequivocally positive and there may be a widening of the palpebral fissure on adduction. There is usually a V-pattern exotropia on straight upgaze. Most patients have good binocular function with or without a compensatory head posture.

Brown has further subdivided the syndrome into true and simulated types.2 The true sheath syndrome, which he considered to be due to a congenitally short anterior tendon sheath, may be typical when there is no underaction of the ipsilateral superior rectus or atypical if this is present. The simulated sheath syndromes include cases which are acquired, intermittent, or show spontaneous recovery. Acquired Brown’s syndrome is uncommon, it is usually unilateral and may present with sudden onset of vertical diplopia. It has been reported in association with orbital trauma,3-6 sinusitis or sinus surgery,7,8 adult rheumatoid arthritis,9-11 Juvenile Rheumatoid Arthritis,12 superior oblique tuck procedures,13 or may be idiopathic.5,14

We would like to report a case of bilateral acquired Brown’s syndrome which developed first in the right eye and later in the left eye during the course of a systemic illness.

Case Report

This 11-year-old boy with a two-month history of fever, joint pains, skin rash, and a two-week history of vertical diplopia and pain around the right eye was admitted to The Hospital for Sick Children for investigation. He had had no previous eye problems. On examination he was pale, febrile (30°C), had a diffuse macular skin rash, lymphadenopathy, and multiple joint effusions.

On eye examination his vision was 20/20 in each eye. Visual fields, pupil reactions, fundoscopy, and slit lamp examination were all normal. The eyes were orthophoric in the primary position and on right gaze, but on left gaze there was a right hypotropia which increased on levo-elevation. There was vertical diplopia on looking up and to the left. Examination of his ocular movements revealed an inability to elevate the right eye in adduction with overaction of the contralateral superior rectus (Figure 1). In full adduction the right eye could not be elevated above the midline. Ductions were full in both eyes. A traction test carried out on the right eye under local anesthetic was positive. Stereo acuity was 40 seconds of arc. There was extreme tenderness over the right trochlea which appeared thickened on palpation. Attempts to elevate the right eye in adduction were accompanied by pain localized to the right trochlea. Examination of old photographs showed no evidence of strabismus or an abnormal head posture.

A provisional diagnosis of systemic onset Juvenile Rheumatoid Arthritis (JRA) complicated by a right Brown’s syn-
FIGURE 1: Ocular versions in nine positions of gaze showing right Brown's syndrome.

FIGURE 2: Ocular versions in nine positions of gaze showing bilateral Brown's syndrome.
drome was made. The erythrocyte sedimentation rate was 95 mm in one hour; rheumatoid factor and antinuclear factor were negative. Other investigations to rule out an infective or neoplastic etiology for his systemic disease were negative. Orbital computed tomography (CT) scan showed a thickened posterior tendon and superior oblique muscle on the right side.

He was treated initially with naproxen, indocin, and aspirin without improvement in his systemic disease or ocular motility. Four weeks after admission he complained of worsening diplopia and pain over his left trochlea. On examination now there was a small esophoria in the primary position and a V-pattern exotropia on straight upgaze. Examination of his ocular movements showed bilateral limitation of elevation in adduction (Figure 2). He was very tender over the left trochlea. There was a further reduction in his field of binocular single vision since his first assessment; there was now diplopia in all positions of upgaze.

The following day he was started on systemic steroids (15 mg prednisolone/dy). This was followed by improvement in his general condition and over the next few weeks his ocular motility improved first in the right eye, and then the left. Six weeks after starting steroids his ocular movements were almost normal (Figure 3) and his field of BSV had expanded so that he had only a small area of diplopia on levo-elevation.

The clinical findings of persistent arthritis, intermittent fever and typical skin rash together with the negative results of extensive investigation to rule out an infectious, neoplastic, or other inflammatory cause for his illness allowed a firm diagnosis of systemic onset JRA to be made. Over the next four months he continued to have active joint disease requiring treatment with systemic steroids, but he had no recurrence of his Brown's syndrome.

**Discussion**

Various theories have been proposed to account for the motility disorder seen in Brown's syndrome. Brown himself proposed that a short anterior tendon sheath was responsible, and noted a thickened tendon sheath at surgery. However, the poor surgical results of procedures confined to the tendon sheath cast doubt on this theory. Parks and Brown suggested that abnormal fibrous bands between the globe and other orbital structures are responsible for true Brown's syndrome but Parks later considered that an abnormally tight or inelastic superior oblique tendon was a more likely cause. Crawford has investigated the effect of various procedures on the superior oblique tendon and sheath and concluded that true Brown's syndrome is caused by a tight superior oblique tendon, and that a complete tenotomy is the most effective procedure in restoring normal ocular motility.

Sevel has identified connective tissue trabeculae between the superior oblique tendon and trochlea which are present in normal embryological development and later regress and has speculated that the persistence of thickened trabeculae after birth may limit the movement of the superior oblique tendon through the trochlea. It has also been suggested that Brown's syndrome is an innervational disorder similar to Duane's syndrome but this is not supported by electromyographic studies. Although the exact mechanism is not clear it appears that true Brown's syndrome is due to a congenital structural anomaly, probably a tight superior oblique tendon, which restricts the ability of the normal...
inferior oblique muscle to elevate the eye in adduction.

Such a structural anomaly cannot account for the motility disturbance seen in acquired Brown's syndrome, nor those cases which show spontaneous recovery. Acquired Brown's syndrome results from localized trauma or inflammation in the area of the trochlea, or follows superior oblique tuck procedures. Traumatic cases may follow orbital injury or sinus surgery. Acquired inflammatory Brown's syndrome may follow sinusitis or be associated with systemic inflammatory disease (Table).

Sandford-Smith reported a case occurring in an adult with rheumatoid arthritis and three further cases have since been reported. He first suggested that acquired Brown's syndrome was due to a stenosing tenosynovitis of the superior oblique tendon and sheath and compared the motility disorder to that seen in "trigger finger," a condition caused by tenosynovitis of the synovial sheaths of the flexor tendons of the fingers. Such a process affecting the superior oblique tendon would result in a constriction of the tendon sheath at the trochlea restricting the passage of a thickened inflamed posterior tendon through the trochlea. Orbital CT scan in our patient demonstrated a thickened posterior tendon and muscle which lends some support to this theory. Helveston et al. have suggested an alternative hypothesis based on their studies of the anatomy of the trochlea. The superior oblique tendon at the trochlea is enclosed in a loose fibrillo-vascular sheath about 0.5 mm thick and between the sheath and the inner aspect of the trochlea is a bursa-like space. Helveston has suggested that local inflammation may result in fluid accumulation in the bursa or vascular engorgement in the sheath which would limit the smooth movement of the tendon through the trochlea.

The clinical features of extreme tenderness and swelling in the region of the trochlea points to an inflammatory etiology for the Brown's syndrome in our case. The ocular motility defect developed at a time when his systemic disease was most active and resolved as his systemic symptoms improved, after the introduction of systemic steroids. Although this improvement may have occurred spontaneously, it is tempting to speculate that the anti-inflammatory effect of the steroids was responsible. Hermann described two cases of acquired Brown's syndrome which resolved after injection of local steroid into the trochlear region. More recently, Beck and Hickling have reported a case of bilateral acquired Brown's syndrome associated with adult rheumatoid arthritis that was successfully treated with injection of local steroid. A trial of local steroid injection would seem to be indicated in those cases of acquired inflammatory Brown's syndrome which do not improve spontaneously and when systemic steroids are not used to treat any associated systemic illness.

The diagnosis of JRA in this case satisfied the criteria laid down by the American Rheumatism Association, that is, persistent inflammation of one or more joints for at least six weeks in a patient in whom other infectious inflammatory or neoplastic causes of arthritis have been excluded. Three different subtypes are recognized depending on whether there is pauciarticular, polyarticular, or systemic onset. Our case falls into the systemic onset group. Ophthalmologists are often asked to see patients with JRA
who may develop chronic uveitis and complicated cataract. These anterior segment complications are most frequently seen in young girls who have pauciarticular onset of arthritis. Optic disc swelling\(^\text{20,21}\) and rheumatoid nodule of the sclera\(^\text{22}\) have also been reported in this condition. Recently, Wang et al.\(^\text{12}\) reported two children with JRA who developed a transient unilateral Brown's syndrome during an exacerbation of their systemic disease. Their two patients were similar to our case in being young boys with systemic onset JRA, who developed their ocular motor abnormality at a time when there was widespread systemic inflammation. In each case there was complete recovery of normal ocular motility which occurred either spontaneously or following treatment with systemic steroids. Brown's syndrome, although rare, should be considered in the differential diagnosis of diplopia occurring in children with systemic inflammatory disease.

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References