Acquired Brown Syndrome Associated With Enteropathic Arthropathy: A Case Report

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INTRODUCTION

Brown1 first described the "superior oblique sheath syndrome" in 1950 as an ocular motility disorder manifesting primarily as a restriction of active and passive elevation in adduction. More recent reports2-4 have disproved Brown's "sheath" hypothesis and have shown the restriction is most likely within the tendon-trocchlea pulley mechanism rather than the tissue surrounding the anterior superior oblique tendon.

Most cases of Brown syndrome are congenital, with acquired Brown syndrome thought to be rare. However, increasing numbers of this form of strabismus are being seen, and the etiologies are quite variable. Adult and juvenile rheumatoid arthritis (JRA),5,6 systemic lupus erythematosus (SLE),7 sinusitis,8 trauma,9 and metastasis to the trocchlea10 have been implicated in causing an acquired Brown syndrome. Inflammation of the tendon-trocchlea complex manifested as a "stenosing tenosynovitis," which normally resolves spontaneously, is thought to be the common pathophysiologic mechanism among these causes.

Enteropathic arthropathy is a symptom complex of peripheral arthritis, sacroilitis, and spondylitis associated with inflammatory bowel disorders such as Crohn disease.11 To our knowledge, this is the first reported case of acquired Brown syndrome associated with an enteropathic arthropathy.

CASE REPORT

A 37-year-old white man with a history of ulcerative proctitis and spondyloarthropathy since 1987 noticed an intermittent vertical diplopia associated with flares of his lower abdominal and back pain since 1988. These symptoms would last approximately 3 days and resolve spontaneously without sequelae. In November 1991, the patient was seen by an ophthalmologist during an acute attack in which the diplopia had not resolved. Examination at that time revealed a corrected visual acuity of 20/20 in both eyes and a 7-prism-diopter right hypertropia in primary gaze which increased in left gaze and decreased in right gaze. He preferred a chin-up posture and a left face turn. The patient was noted to have normal Worth four-dot and Titmus stereo acuity tests of 40 arc-seconds in his head posture of choice. His motility examination was remarkable for significant restriction of elevation of the right eye in adduction with no other deficits. His symptoms improved over several months with ibuprofen, methocarbamol (Robaxin), and mesalamine (Rowasa) enemas.

The patient was referred to the Ophthalmology Clinic, Fitzsimons Army Medical Center, in April 1992 for evaluation. His vertical diplopia was symptomatic only on upgaze and he had had no other ocular complaints. Past surgical history was significant only for hemorrhoidectomy in 1986 and vasectomy in 1988. Family history was negative for ocular disease. Ophthalmologic examination at that time was normal except for restricted elevation of the abducting right eye. Forced duction testing verified restriction of passive movement of the right eye. Laboratory evaluation was significant for a positive HLA B-27. Orbital magnetic resonance imaging (MRI) demonstrated intense enhancement of the right trocchlea (Figure). Rheumatologic evaluation confirmed the diagnosis of sacroilitis, and colonoscopic biopsies were positive for Crohn disease. The patient was treated with sulfasalazine (Azulfidine), and a nonsteroidal medication.

Previously reported causes of acquired Brown syndrome include JRA, SLE, trauma, sinusitis, and trocchlear metastasis.5-10 The common etiologic factor is a stenosing tenosynovitis of the superior oblique resulting in impaired passage through the fibrocartilaginous trocchlea.2 Some patients with rheumatic disease may complain of

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FIGURE: Post-contrast coronal T1-weighted magnetic resonance image with fat suppression shows a ring-like enhancement of the right trochlea. Note the hypointense central portion, corresponding to the nonenhancing tendon of the superior oblique tendon.

worsening of their diplopia symptoms with flares in their rheumatic disease as well as “early morning stiffness of the eyes” and a trigger finger-type “click” on extraocular movement when the inflamed superior oblique tendon is pulled through the trochlea.6

Acute episcleritis, scleritis, uveitis, marginal keratitis, and other ocular inflammatory conditions have been described in 4% to 10% of Crohn patients.12 Patients with colitis are more likely to suffer from ocular inflammation (24%), than patients with small-bowel disease alone (3%). Sacroiliitis has been reported in up to 16% of patients with Crohn disease and the HLA B-27 is positive in up to 75% of those patients with Crohn disease who develop sacroiliitis. In addition, patients with Crohn disease and arthritis have a higher incidence of ocular inflammation (29%), than patients without joint involvement (6%).

Although not previously described, we believe the acquired Brown syndrome case presented herein was caused by trochlear inflammation secondary to an enteroopathic arthropathy (Crohn disease associated with HLA B-27 positive sacroiliitis). The inflammation, as illustrated in the orbital MRI (Figure) is described as a stenosing tenosynovitis of the fibrocartilagenous pulley (trochlea),13 and is similar in appearance to the inflammation of the sacroiliac joints. This correlation, along with the clinical presentation and history of our patient, implicates enteroopathic arthropathy as another etiology of acquired Brown syndrome.

REFERENCES