

# Acquired Brown's Syndrome in a Child with Poststreptococcal Reactive Arthritis

ALIX O. FAUST, JAY M. GILLENWATER, and FRANK T. SAULSBURY

**ABSTRACT.** Acquired Brown's syndrome is a disorder of ocular mobility characterized by the inability to elevate the affected eye in full adduction owing to inflammatory tenosynovitis of the superior oblique tendon. We describe a child who developed Brown's syndrome as a complication of poststreptococcal reactive arthritis. (J Rheumatol 2001;28:2748-9)

*Key Indexing Terms:*

BROWN'S SYNDROME

POSTSTREPTOCOCCAL REACTIVE ARTHRITIS

Brown's syndrome is a congenital or acquired disorder of ocular mobility characterized by the inability to elevate the affected eye in full adduction<sup>1</sup>. In addition to orbital tumors, trauma, and surgery, acquired Brown's syndrome can be caused by inflammatory tenosynovitis of the superior oblique tendon/trochlea complex<sup>2</sup>. Acquired Brown's syndrome has been reported as an uncommon complication in a variety of inflammatory arthropathies, including juvenile rheumatoid arthritis<sup>3-6</sup>, adult Still's disease<sup>7</sup>, rheumatoid arthritis<sup>8,9</sup>, systemic lupus erythematosus<sup>10,11</sup>, HLA-B27 associated arthropathy<sup>12</sup>, and psoriatic arthritis<sup>13</sup>. We describe a child who developed Brown's syndrome as a complication of poststreptococcal reactive arthritis (PSRA). To our knowledge, this is the first report of Brown's syndrome associated with PSRA.

## CASE REPORT

A previously healthy 9-year-old boy developed fever, myalgia, and arthralgia one week after complaining of a sore throat. Examination was normal except for pharyngitis. A throat culture was positive for Group A beta hemolytic streptococcus and he was treated with penicillin. Ten days after the initial visit, he developed swelling of the left elbow and the right wrist. He was treated with ibuprofen, but during the ensuing 3 weeks, he remained febrile, and the swelling of the right wrist and left elbow persisted.

Five weeks after the onset of joint symptoms, he was referred for rheumatologic evaluation. Examination revealed limitation of motion and pain in the right shoulder and the right hip. The left elbow and the right wrist were swollen, warm, and restricted in motion. In addition, he had prominent tenosynovitis of the extensor tendons of the dorsum of the right wrist and hand. The cardiovascular examination was normal. Laboratory studies revealed a white blood cell count of 12,000/ $\mu$ l with 75% neutrophils and 20% lymphocytes. The hemoglobin was 10 g/dl, the platelet count 577,000/ $\mu$ l, and the erythrocyte sedimentation rate 114 mm/h. Tests for antinuclear antibody and rheumatoid factor were negative. The antistreptolysin O titer was elevat-

ed at 659 IU/ml. Serologic studies showed no evidence of infection with *Borrelia burgdorferi*, Epstein-Barr virus, or human parvovirus B19. An electrocardiogram and cardiac echocardiogram yielded normal results.

A diagnosis of poststreptococcal reactive arthritis was made and he was treated with naproxen 18 mg/kg/day and penicillin prophylaxis. Despite therapy he continued to have intermittent low grade fever and polyarthritides.

Four weeks later, he presented with pain in the right supraorbital region and mild swelling of the right upper eye lid. He complained of painful diplopia with upward gaze. Examination revealed inability to elevate the right eye in full adduction. Ophthalmologic consultation confirmed the diagnosis of acquired Brown's syndrome by clinical examination and by the Hess screen. There were no other ophthalmologic abnormalities. Magnetic resonance imaging (MRI) of the orbits showed slight enhancement of the right superior oblique tendon/trochlea complex. In addition to the Brown's syndrome, the patient had active arthritis of the left elbow, right ankle, and left hip.

He was treated with prednisone 1.5 mg/kg/day. Three days later he was much improved, and one week after starting prednisone, the Brown's syndrome had resolved completely. In addition, the arthritis resolved completely after one week of prednisone therapy. The prednisone dose was tapered over the next 2 weeks and then discontinued without recurrence of symptoms. Two months after the onset of Brown's syndrome and 4 months after the onset of arthritis, the naproxen was discontinued. Examination at that time was normal. There has been no recurrence of the Brown's syndrome or arthritis during followup observation of 9 months.

## DISCUSSION

The patient described in this report developed Brown's syndrome as a complication of PSRA. Brown's syndrome is characterized by the inability to elevate the affected eye in full adduction (Figure 1). The orbital oblique muscles have complementary actions in moving the eyes in the vertical plane. When the inferior oblique muscle moves the globe upward and inward, the superior oblique muscle relaxes. If the superior oblique tendon cannot relax and slide freely through the tendon sheath and the trochlea, the affected eye cannot be elevated while in full adduction.

Sanford-Smith<sup>2</sup> described Brown's syndrome in an adult with rheumatoid arthritis and suggested that acquired Brown's syndrome is similar to trigger finger, a condition caused by tenosynovitis of the synovial sheaths of the flexor and extensor tendons of the finger. Acquired Brown's syndrome has

*From the Department of Pediatrics, University of Virginia Health System, Charlottesville, Virginia, USA.*

*A.O. Faust, MD; J.M. Gillenwater, MD; F.T. Saulsbury, MD.*

*Address reprint requests to Dr. F.T. Saulsbury, Department of Pediatrics, University of Virginia Health System, Box 800386,*

*Charlottesville, VA 22908, USA. E-mail: fts@virginia.edu*

*Submitted February 19, 2001; revision accepted June 25, 2001.*

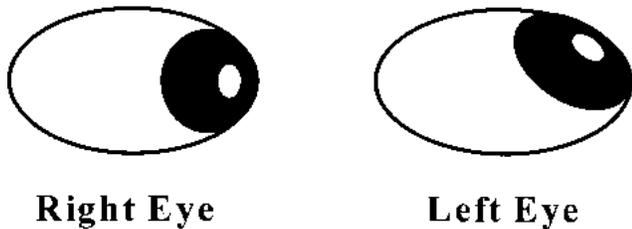


Figure 1. Schematic representation of Brown's syndrome affecting the right eye. The right eye is unable to be elevated while in full adduction.

been reported as an unusual complication in patients with a variety of inflammatory arthropathies<sup>3-13</sup>. In addition to the obvious limitation of ocular mobility, acquired Brown's syndrome owing to tenosynovitis of the superior oblique tendon/trochlea is often associated with diplopia on upward gaze and orbital pain and swelling in the region of the trochlea.

The clinical diagnosis of acquired Brown's syndrome is generally not difficult. Imaging modalities such as computed tomography or MRI may be helpful to confirm the diagnosis of Brown's syndrome. Some, but not all patients show evidence of inflammation of the superior oblique tendon/trochlea complex on imaging studies<sup>5-7,11-13</sup>. MRI of our patient showed mild enhancement of the right superior oblique tendon/trochlea. He had rapid resolution of the Brown's syndrome following institution of prednisone therapy and others have also reported that systemic corticosteroid or locally injected corticosteroid therapy is highly effective in treating acquired Brown's syndrome<sup>4,10,13</sup>.

Our patient developed Brown's syndrome as a complication of PSRA. Goldsmith and Long in 1982<sup>14</sup> first described PSRA in a report of 12 children who developed prolonged arthritis following a streptococcal infection. Since then, there have been a number of reports of PSRA in children and adults<sup>15-19</sup>.

PSRA differs from acute rheumatic fever in several respects. In PSRA, the interval between the streptococcal infection and the onset of arthritis is short, often 7–10 days<sup>17,19</sup>. The arthritis of PSRA involves both large and small joints and it is frequently nonmigratory<sup>15,17,19</sup>. The duration of arthritis is often prolonged, lasting up to several months, and arthralgia may persist for several months after the resolution of objective arthritis. In addition, tenosynovitis is a prominent feature of PSRA<sup>15,17,18</sup>. PSRA is relatively unresponsive to nonsteroidal antiinflammatory agents, whereas the arthritis of acute rheumatic fever responds promptly to therapy<sup>17,19</sup>. Lastly, the incidence of carditis in PSRA is much lower than in patients with acute rheumatic fever, but carditis has been reported in roughly 5% of patients with PSRA<sup>15,19</sup>. Thus, patients with PSRA should receive penicillin prophylaxis<sup>15,19</sup>.

We describe a child who developed Brown's syndrome as a complication of poststreptococcal reactive arthritis and review the clinical features of PSRA and the association of inflammatory arthritis with Brown's syndrome.

## REFERENCES

1. Brown HW. True and simulated superior oblique tendon sheath syndromes. *Doc Ophthalmol* 1973;34:123-36.
2. Sanford-Smith JH. Intermittent superior oblique tendon sheath syndrome. *Br J Ophthalmol* 1969;53:412-7.
3. Wang FM, Wertebaker C, Behrens MM, Jacobs JC. Acquired Brown's syndrome in children with juvenile rheumatoid arthritis. *Ophthalmology* 1984;91:23-6.
4. Kemp AS, Searle C, Horne S. Transient Brown's syndrome in juvenile chronic arthritis. *Ann Rheum Dis* 1984;43:764-5.
5. Moore AT, Morin JD. Bilateral acquired inflammatory Brown's syndrome. *J Pediatr Ophthalmol Strabismus* 1985;22:26-30.
6. Roifman CM, Lavi S, Moore AT, Morin DJ, Stein LD, Gelfand EW. Tenosynovitis of the superior oblique muscle (Brown syndrome) associated with juvenile rheumatoid arthritis. *J Pediatr* 1985;106:617-9.
7. Kaufman LD, Sibony PA, Anand AK, Gruber BL. Superior oblique tenosynovitis (Brown's syndrome) as a manifestation of adult Still's disease. *J Rheumatol* 1987;14:625-7.
8. Killian PJ, McClain B, Lawless OJ. Brown's syndrome. An unusual manifestation of rheumatoid arthritis. *Arthritis Rheum* 1977;20:1080-4.
9. Cooper C, Kirwan JR, McGill NW, Dieppe PA. Brown's syndrome: an unusual ocular complication of rheumatoid arthritis. *Ann Rheum Dis* 1990;49:188-9.
10. Alonso-Valdivielso JL, Alvarez Lario B, Alegre Lopez J, Sedano Tous MJ, Builtrago Gomez A. Acquired Brown's syndrome in a patient with systemic lupus erythematosus. *Ann Rheum Dis* 1993;52:63-4.
11. Whitefield L, Isenberg DA, Brazier DJ, Forbes J. Acquired Brown's syndrome in systemic lupus erythematosus. *Br J Rheumatol* 1995;34:1092-4.
12. Bradshaw DJ, Bray VJ, Enzenauer RW, Enzenauer RJ, Truweit CL, Damiano TR. Acquired Brown syndrome associated with enteropathic arthropathy: a case report. *J Pediatr Ophthalmol Strabismus* 1994;31:118-9.
13. Thorne JE, Volpe NJ, Liu GT. Magnetic resonance imaging of acquired Brown syndrome in a patient with psoriasis. *Am J Ophthalmol* 1999;127:233-5.
14. Goldsmith DP, Long SS. Streptococcal disease of childhood — a changing syndrome [abstract]. *Arthritis Rheum* 1986;29 Suppl 4:S18.
15. DeCunto CL, Giannini EH, Fink CW, Brewer EJ, Person DA. Prognosis of children with poststreptococcal reactive arthritis. *Pediatr Infect Dis J* 1988;7:683-6.
16. Arnold MH, Tyndall A. Poststreptococcal reactive arthritis. *Ann Rheum Dis* 1989;48:686-8.
17. Fink CW. The role of the streptococcus in poststreptococcal reactive arthritis and childhood polyarteritis nodosa. *J Rheumatol* 1991;18 Suppl 29:14-20.
18. Gutierrez-Urena S, Molina J, Molina JF, Garcia CO, Cuellar ML, Espinoza LR. Poststreptococcal reactive arthritis, clinical course, and outcome in 6 adult patients. *J Rheumatol* 1995;22:1710-3.
19. Ahmed S, Ayoub EM, Scornik JC, Wang CY, She JX. Poststreptococcal reactive arthritis. *Arthritis Rheum* 1998; 41:1096-102.