Frosted Branch Angiitis: The Role of Systemic Corticosteroids

Latif M. Hamed, MD, Ervin N. Fang, MD, Maher M. Fanous, MD, Robert Mames, MD, and Scott Friedman, MD

INTRODUCTION

Frosted branch angiitis is a rare disorder, with only 12 previously reported cases. It is characterized by acute bilateral visual loss with dramatic fundus abnormalities. These include marked sheathing of the retinal veins resembling frosted tree branches, retinal edema, late perivascular leakage on fluorescein angiography, and relative sparing of the optic nerve and retinal arteries. Moderate to severe iridocyclitis and some ocular hyperemia are often present. The etiology is unknown, but is suspected to be viral. All previously reported patients except one have been treated promptly with systemic corticosteroids.

We report a case of bilateral frosted branch angiitis with visual improvement from counting fingers to 20/30 visual acuity with only topical corticosteroid administration.

CASE REPORT

A 5-year-old previously healthy boy developed an acute onset of bilateral decreased vision, ocular injection, and photophobia. Examination 2 days after onset revealed visual acuities of count fingers (CF) at 5 feet in each eye. There was mild external ocular injection. The pupils measured 6 mm each, were sluggishly reactive to light OU, and showed no afferent defect. There were + cell and flare in both anterior chambers; 1 + vitreal cells were also present. The fundi showed diffuse perivenous sheathing; the arterioles were generally spared (Fig 1). Bilateral exudative macular detachments with scattered blot hemorrhages were present. The optic discs were slightly hyperemic.

Laboratory tests revealed no serological evidence of syphilis, Lyme disease, toxoplasmosis, rheumatoid arthritis, or collagen vascular disease. The complete blood cell count and white blood cell differential were normal. Westergren sedimentation rate was 35 mm/hr. A PPD skin test was negative. Levels of angiotensin converting enzyme and serum lysozyme were normal. Cytomegalovirus (CMV) titers as well as herpes simplex virus types 1 and 2 IgG antibody titers suggested prior exposure only; CMV serum cultures were negative. HLA-B27 antigen was negative. Results of magnetic resonance imaging of the head and a chest X-ray were normal.

Topical corticosteroid drops every hour and cycloplicges were administered, and the dosages were tapered to no drops at all during a 10-week period. A trial of Septra and Daramprin for toxoplasmosis was administered for 2 days and then discontinued after serologic results became available. Nine days after presentation, vision improved to CF at 7 feet OD and 20/400 OS. The anterior chamber showed only trace cells. The perivenous sheathing was markedly decreased in both eyes. A small, shallow exudative detachment of the right macula persisted; both maculae showed central pigmentary disturbance. Five weeks after presentation, the visual acuity had improved to 20/300 OU. The anterior segments showed no inflammation. The fundi showed well-defined RPE mottling in the central maculae with mild peripheral RPE granularity (Fig 2). The perivenous sheathing had resolved.

Seven weeks after presentation, the vision had improved to 20/30 OU. The central maculae showed circumscribed RPE mottling. The photopic electroretinography (ERG) was decreased to 50% of normal OD and 30% of normal OS, with increased latency. The scotopic ERG was 25% of normal OU. Electroc-oculography revealed L/D ratios of 1.67 OD and 1.41 OS, both below normal for age.

DISCUSSION

Twelve previous cases of frosted branch angiitis have been previously reported—seven in the English literature. The age range of affected patients is 5 to 33 years, our patient being the youngest.

Previous authors have advocated prompt treatment with systemic corticosteroids, citing eventual improvement of visual acuity and fundus abnormalities after its administration. Eleven of the 12 previously reported cases had been treated with systemic corticosteroids.
FIGURE 1: Dramatic sheathing of retinal venules was present at initial presentation. (A) Right eye. (B) Left eye.

FIGURE 2: Residual, circumscribed central macular pigmentary changes were present in both eyes, 7 weeks after onset and subsequently. (A) Right eye. (B) Left eye.

Unfortunately, scrutiny of previously reported cases does not allow precise assessment of the time course of the disease after corticosteroid therapy. While fundus abnormalities in one case reportedly showed some improvement even 1 day after commencing treatment, visual recovery typically occurred several weeks to months later. Our patient, like the patient of Vander and Masciulli, received only topical corticosteroids, which we believe did not significantly affect the posterior segment findings. Nonetheless, both patients showed considerable improvement, comparable to that seen in systematically treated patients. Whether the residual macular pigmentary changes in our patient might have been less remarkable if systemic corticosteroids were administered is unclear.

While highly recommended by previous authors, we believe that the role of systemic corticosteroids in the treatment of frosted branch angiitis requires further clarification.

REFERENCES