Idiopathic Frosted Branch Angiitis in a 2-Year-Old Boy

To the Editors:

A 2-year-old boy presented to our clinic with a history of not being able to identify and follow objects for the past 4 days. There was no history of prematurity or any congenital disorder. There was no definite history of any febrile episode, upper respiratory tract infection, or diarrhea.

On examination, the child was not able to fixate on a target. There was no deviation of eyes. The anterior chamber showed 3+ cells. On fundus evaluation, the media was hazy due to vitritis in both eyes. All visible retinal veins of both eyes showed significant perivascular exudation (Figure 1A). A diagnosis of bilateral frosted branch angiitis was made.

Hematology investigations revealed normal hemoglobin (10.8 gm) and a normal total leukocyte count of 10,000 per microliter. Differential leukocyte count showed elevated lymphocytes (72%). The peripheral smear showed normal blood film morphology. Blood and urine culture were sterile. A chest x-ray was also normal.

We administered intravenous acyclovir 15 mg/kg/dose four times a day for 1 week followed by oral acyclovir at the same dose for the next 8 weeks. Oral corticosteroids were administered at a dose of 1 mg/kg 2 days after starting antiviral therapy. A full dose of steroids was given for the first 2 weeks and then slowly tapered over the next 2 months. There was clearing of the media with resolution of the severe vasculitis over the next 4 weeks (Figure 1B). The child is now able to recognize and follow objects normally.

We found only one report of frosted branch angiitis in a 2-year-old child.1 Also, there is only one such case of frosted branch angiitis reported from India.2 It is difficult to image the fundus of children at this age even with the RetCam (Clarity Medical Systems, Inc., Pleasanton, CA), a digital imaging system for imaging mainly fundus of neonates. Despite hazy media, the image showed the frosted branch appearance of the vasculitis (Figure 1A). We photodocumented the occurrence of this rare disease in such a small child.

Before making a diagnosis of primary idiopathic frosted branch angiitis, infiltrative disorders such as leukemia and lymphoma, infective causes such as cytomegalovirus retinitis, and diseases such as Behcet’s and sarcoidosis, which may cause widespread vasculitis, should be ruled out. Immune recovery uveitis in cytomegalovirus retinitis has also been reported to present as frosted branch angiitis.3 The normal blood investigations, immunocompetent status, and age of our patient made us rule out most of the secondary causes.

There is no definite treatment protocol for this disease. Previous studies have not given any medica-

Figure 1. (A) Marked perivenular exudation along all visible veins is seen through hazy media. (B) At 4 weeks of follow-up, the media cleared with no evidence of vasculitis or its sequela along the veins.
tion and some have only used steroids, whereas some have used a combination of steroids and acyclovir. Walker et al.\(^4\) tabulated various treatment strategies in a study on frosted branch angiitis. We received a good response with combination of acyclovir and oral steroids. Following 4 weeks of therapy, the anterior uveitis, vitritis, and vasculitis completely regressed, leaving no morphological sequelae (Figure 1B).

REFERENCES

Rohan Chawla, MD, FRCS(Glasg)
Ravi Bypareddy, MD, DNB
Pradeep Venkatesh, MD
Ankit Singh Tomar, MBBS
New Delhi, India

The authors have no financial or proprietary interest in the materials presented herein.

doi: 10.3928/01913913-20150623-02