

the scarring and the accumulation of mucopolysaccharide material proximal to a possible local enzyme block.

The treatment of acromesomelic dysplasia is directed toward the specific symptom and physical characteristics seen in each patient. Treatment may require the coordinated efforts of a team of specialists such as pediatricians, orthopedists, physical therapists, and ophthalmologists.<sup>1,3</sup> Ophthalmic treatment includes corrective lenses for errors of refraction and polarized lenses for symptoms of glare. The patient still has good visual acuity and keratoplasty is, therefore, not indicated yet. Follow-up is necessary to assess the progression of the disease and to decide on appropriate management of symptoms.

#### References

1. Clarke WN, Munro S, Brownstein S, et al. Ocular findings in acromesomelic dysplasia. *Am J Ophthalmol* 1994; 118: 797-804.
2. Ianakiev P, Kilpatrick MW, Daly, MJ, et al. Localization of an acromesomelic dysplasia on chromosome 9 by gene mapping. *Clinical Genetics* 2000; 57: 278-283.
3. Jones K. *Smith's Recognizable Patterns of Human Malformation*, 4th ed. Philadelphia: WB Saunders Co., 1988; 308-309.
4. Sharrard W. *Pediatric Orthopedics and Fractures*, 3rd ed. London: Blackwell Scientific Publications, 1993; 155-156.

## Frosted-branch angiitis

Anthony F. Felipe, MD  
Farlah Angela M. Salvosa, MD  
Harvey S. Uy, MD  
Juancho Francisco C. Remulla, MD

*Department of Ophthalmology and Visual Sciences  
University of the Philippines-Philippine General Hospital  
Manila, Philippines*

### ABSTRACT

#### Objective

*To report a case of frosted-branch angiitis.*

#### Method

*This is a case report of frosted-branch angiitis seen at the University of the Philippines-Philippine General Hospital.*

#### Results

*A 42 year-old male presented with progressive blurring of vision of the left eye. Indirect funduscopy showed dilated retinal veins with perivascular sheathing, giving the appearance of frosted-branches of a tree.*

#### Conclusion

*Frosted-branch angiitis is a rare form of retinal vasculitis with various etiologies. Despite the severe retinal appearance, the prognosis is usually good, with rapid recovery of visual acuity after prompt steroid treatment.*

FROSTED-BRANCH angiitis is a rare form of retinal vasculitis characterized by white perivascular sheathing of retinal blood vessels. The first case reported in 1976 involved a 6-year-old boy who had severe white sheathing of all retinal vessels presenting an appearance similar to the frosted branches of a tree.<sup>1</sup> Affecting more males (52%) than females (48%), frosted-branch angiitis is mostly seen in children and young adults. It usually affects individuals 6 to 16 years old in Japan and 23 to 29 in other countries. It is typically bilateral although unilateral cases have been reported.

This case involved a 42-year-old male who consulted at the University of the Philippines-Philippine General Hospital (UP-PGH) because of a 4-month history of progressive blurring of vision in the left eye. Visual acuity was 20/20 for the right eye and 20/40, improved to 20/25 on pinhole, for the left eye. Intraocular pressures were within normal limits for both eyes (OU). The anterior segment was normal.

Indirect ophthalmoscopy for the right eye was normal. The left eye, seen through a hazy medium, showed dilated and tortuous retinal veins with perivascular sheathing peripherally. There were some intraretinal foci of inflammation with scattered hemorrhages mostly in the inferior nasal periphery, and numerous vitreous opacities.

Fluorescein angiography (FA) of the left eye showed dilated veins with leakage of dye from the retinal vessels on late phase and multifocal areas of perivenular staining. There were areas of capillary nonperfusion on the infero-nasal arcade with foci of hyperfluorescence. Systemic work-up for possible etiology and polymerase chain reaction of the aqueous humor yielded negative results.

It is still unclear whether frosted-branch angiitis is a unique disease entity by itself or a clinical presentation resulting from several causes as reported by Kleiner.<sup>2</sup> Its characteristic features are:<sup>3</sup>

- Severe sheathing of retinal vessels appearing like frosted branches of a tree in one or both eyes;
- Acute visual disturbance associated with signs of anterior-chamber and vitreous inflammation;
- FA demonstrates no occlusion or stasis of sheathed vessels, but late staining and/or leakage along vessels;
- Otherwise healthy patient;
- Prompt response to corticosteroid;
- Typically no recurrence.

In 1998, Kleiner et al.<sup>4</sup> classified the disease into 3 subgroups: idiopathic, those associated with hematologic malignancies like leukemia and lymphoma, and those caused by viral infection or autoimmune disease.

Most cases of frosted-branch angiitis are idiopathic, as in the case of our patient. An immune-mediated mechanism is believed to be the main cause as evidenced by localized ocular vasculitis sparing other organs. This

suggests that the immune response is directed to an inciting agent in the eye. The presence of positive titers for herpes simplex, varicella, rubella, cytomegalovirus, Epstein-Barr virus, and antistreptolysin O in some patients affected by frosted-branch angiitis suggested that viral or bacterial infection can be the triggering antigen. The prompt response to systemic steroids also indicates a probable immune-mediated mechanism.

In frosted-branch angiitis associated with viral disease like cytomegalovirus and human immunodeficiency virus, it has been theorized that viral antigens form immune complexes and deposit in retinal vessels causing vasculitis. A direct viral invasion may also be responsible for the pathogenesis. Immune complexes are also responsible for retinal vasculitis secondary to autoimmune disease. Direct infiltration of retinal vessels by malignant cells is believed to be the cause of frosted-branch angiitis among patients with leukemia and lymphoma.

Diagnosis is mainly by ophthalmoscopy and fluorescein angiography. Ophthalmoscopy shows the typical sheathing of retinal vessels mostly the veins giving the appearance of frosted-branches of a tree. Fluorescein angiogram shows normal blood flow but with late staining, leakage of dye from vessels, and optic-disc hyperfluorescence. Laboratory examinations usually do not show abnormalities and are mostly useful to rule out the possibility of associated systemic diseases.

Despite the severe retinal appearance, the prognosis is usually good, with rapid recovery of visual acuity after prompt steroid treatment to suppress intraocular inflammation and prevent visual loss and long-term complications such as capillary nonperfusion, retinal neovascularization, neovascular glaucoma, macular scarring, and retinal detachment. Recovery usually starts 2 to 3 weeks after treatment is initiated. Steroid-sparing agents may be used when there is significant steroid toxicity or persistent relapse at high dose of steroids. The ones commonly used are cyclosporine and azathioprine.<sup>5</sup>

Our patient was started on high-dose oral prednisone at an initial dose of 1 mg/kg/day for 5 weeks and responded well with improvement in visual acuity and fundus findings after 2 weeks. Funduscopy showed decreased vitreous haziness and opacities, less dilatation and tortuosity of the veins, and decreased perivascular sheathing. Prognosis is relatively good.

#### References

1. Ito Y, Nakona M, Kyu N, Takeuchi M. Frosted-branch angiitis in a child. *Jpn J Clin Ophthalmol* 1976; 30: 797-803.
2. Kleiner RC. Frosted-branch angiitis: clinical syndrome or clinical sign? *Retina* 1997; 16: 370-371.
3. Liu IT, Chung YM, Liu JH, Hsu WM. Frosted-branch angiitis in two Chinese young girls. *J Chin Med Assoc* 2003; 66: 501-504.
4. Kleiner RC, Kaplan HJ, Shakin JL, et al. Acute frosted retinal periphlebitis. *Am J Ophthalmol* 1988; 106: 27-34.
5. Walton RC, Ashmore ED. Retinal vasculitis. *Curr Opin Ophthalmol* 2003; 14: 413-419.

## A mysterious case of bilateral stromal keratitis

Michelle D. Lingao, MD  
Ruben Lim Bon Siong, MD  
Mario J. Valenton, MD

Department of Ophthalmology and Visual Sciences  
University of the Philippines-Philippine General Hospital  
Manila, Philippines

### ABSTRACT

#### Objectives

To describe a rare case of bilateral stromal keratitis and demonstrate the effectiveness of penetrating keratoplasty in the management of toxocara keratitis.

#### Method

This is a case report.

#### Results

A 53-year-old male farmer had a 10-month history of bilateral corneal opacity, photophobia, redness, foreign body sensation, and eye pain. The diagnosis was central microbial keratitis with the following etiologies considered: Epstein-Barr virus, herpes simplex, fungal, syphilis, tuberculosis (TB), mycobacteria other than TB, and acanthamoeba. Despite treatment with topical steroids and antibiotics, both eyes worsened. Penetrating keratoplasty markedly improved the patient's visual acuity. Histopathology of the left corneal button revealed toxocara keratitis.

#### Conclusion

Good history taking, complete systemic and ocular examinations, and a histopathology of the corneal tissues are vital to the diagnosis of toxocara keratitis. Penetrating keratoplasty was shown to be effective in its management. Emphasis is given on prevention to decrease the incidence of the disease.

TOXOCARA keratitis is one of the many presentations of ocular toxocariasis. It results from invasion of the eye by the roundworm toxocara canis, a parasite that completes its life cycle in dogs and other canids, via the hematogenous route.

Only 2 cases of toxocara keratitis have been reported. Baldone and colleagues reported the presence of a nematode larva with the morphological appearance of toxocara in the corneal stroma.<sup>1</sup> However, no histopathologic examination was done because the larva was moving too swiftly to be surgically removed. The second case was the only one in a study by J. Altchek et