Frosted branch angiitis is a rare retinal vasculitis mostly seen in children or young adults. The first case was reported by Ito et al.\(^1\) in 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. The mean age was 9 years in the initial reports from the Japanese group of patients,\(^1-5\) whereas Kleiner and associates have reported this disease in an older age group (range, 23-29 years).\(^6\) The characteristic features of frosted branch angiitis can be summarized as follows: (1) severe sheathing of the retinal vessels appearing like frosted branches of a tree in one or both eyes;\(^7-9\) (2) acute visual disturbance associated with signs of anterior chamber and vitreous inflammation; (3) fundus fluorescein angiography that demonstrates no occlusion or stasis of the sheathed vessels, but late staining and/or leak age along the vessels; (4) otherwise healthy patients; (5) prompt response to corticosteroids; (6) typical no recurrence. The only report in Taiwan was described by Chan et al.\(^10\) Here, we studied two additional cases of frosted branch angiitis.

CASE REPORTS

Case 1

A previously healthy 9-year-old girl suffered from acute red eyes with blurred vision and painful sensation. She also complained of headache sometimes. She was referred to us four days after the onset of symptoms. The ocular examinations showed the best corrected visual acuity of right eye: 6/20 and left eye: 6/60. Marked inflammatory reactions with cells and flare were noted in the anterior chambers. One patient even had hypopyon in both eyes. Initial visual acuity ranged from 6/6.7 to 6/60. Both patients showed rapid improvement with systemic and/or topical corticosteroid therapy, and all of the affected eyes regained a visual acuity of 6/6. The clinical appearances and courses of these two patients matched the condition previously described in Japan and labeled frosted branch angiitis. Before this article, there was only one case reported in Taiwan.
M were all within normal limits. Mild elevation of erythrocyte sedimentation rate (30 mm/hr, normal < 20 mm/hr) was detected, but no fever was noted throughout the course. The serum tests for toxoplasma, mumps, rubella, cytomegalovirus, and varicella zoster virus were all negative, while immunoglobulin G for mumps and rubella were positive, which is usually present in normally vaccinated children.

A regimen started with topical corticosteroids (1% prednisolone acetate OU Q2H, 0.1% betamethasone ointment OU QN) and 15 mg of oral prednisolone given twice daily for two weeks. Cycloplegic eyedrop (1% atropine) was also given 3 times a day. Two weeks after treatment, there was no more hypopyon, and the perivascular exudates resolved. The best corrected visual acuity of both eyes was improved to 6/7.5. Since the response was rapid and remarkable, corticosteroids were tapered gradually over the following two weeks. One month after the initial examination, all drugs were discontinued, and the best corrected visual acuity was improved to 6/6 on both eyes. The patient had been followed up for two years and no recurrence or complication was ever noted.

Case 2

A 9-year-old girl in good health complained of decreased vision, redness, pain and floaters of both eyes, as well as photophobia of right eye for 2 days. On examination the best corrected visual acuity was R.E.: 6/7.5 and L.E.: 6/6.7. Mild-to-moderate cells and flare were present in the anterior chambers, and fine keratic precipitates in both eyes were noted. Ophthalmoscopy showed edematous retina with hyperemic discs and extensive vascular sheathing from the posterior pole to the periphery in both eyes (Fig. 2). The results of laboratory studies included a normal complete blood cell count and differential, urine routine, chest X-ray, C-reactive protein, erythrocyte sedimentation rate, rheumatoid factor and immunoglobulin A. Antinuclear antibody was negative. Serum tests for syphilis, and immunoglobulin M for infection of TORCH (toxoplasma, measles, mumps, rubella, cytomegalovirus, herpes simplex virus, and varicella zoster virus) were all negative.

The patient was treated with a topical corticosteroid (1% prednisolone acetate OU QID). Ten days later, the visual acuity was 6/6 on both eyes. Only trace cells were noted in both anterior chambers, and both fundi appeared normal. Three weeks after treatment, the regimen was
changed to a weaker corticosteroid eyedrop (0.02% fluometholone OU TID), which was discontinued in the following week. She had been followed up for one and a half years and found no sign of recurrence.

**DISCUSSION**

Frosted branch angiitis in otherwise healthy young patients was thought to be a single clinical syndrome. It was reported to occur in patients aged from 3 to 33 years. The initial visual acuity ranged from 6/6 to only light perception, and most cases were involved bilaterally. There is a distinct possibility that the vasculitis represents an immune response to an underlying stimulus such as a viral infection (herpes group, rubella, etc). Other patients with similar retinal appearances but with an underlying disease were classified by Kleiner into subgroups. The first group includes patients with lymphoma or leukemia arising from infiltration with malignant cells. In his opinion, these patients should be described as having a “frosted branch-like appearance” but not true frosted branch angiitis. The second group is the patients who have associated viral infection (human immunodeficiency virus, cytomegalovirus, etc) or autoimmune disease (systemic lupus erythematosus, Crohn’s disease, etc). In these patients frosted branch angiitis is thought to be a clinical sign, possibly of immune complex deposition, and the fundus could be described as having a “frosted branch response” to the underlying disease or “secondary frosted branch angiitis.”

Our 2 patients, both 9-year-old girls in good health, were similar to the previously reported cases of primary frosted branch angiitis. They both had rapid onset of blurred vision, severe sheathing of the retinal vessels in both eyes, prompt response to corticosteroids, and no recurrence. The fluorescein angiograms (in the first case) showed mild and late staining of the retinal vessels with no evidence of stasis or occlusion. Additionally, both of our patients developed anterior chamber inflammation in varying degrees; the first one had hypopyon, which has rarely been described in this disease, and the second one had fine keratic precipitates.

Most of authors have advocated prompt treatment with systemic corticosteroids, citing eventual improvement of visual acuity and fundus abnormality after its administration. However, there were 3 cases that had not received corticosteroids systematically. Two patients were given corticosteroids topically, whereas the other one received only a single subconjunctival injection of triamcinolone. All of them showed considerable improvement, compared with that seen in systemically treated ones. Our patients dem onstrate that the rule of systemic corticosteroids in the treatment of frosted branch angiitis requires further clarification.

In conclusion, we encountered 2 cases of acute idiopathic (primary) frosted branch angiitis, which is impressive due to severe sheathing of the affected vessels resembling the frosted branches of a tree. Anterior chamber inflammation was always seen, but our patient is the second one that presented with hypopyon as reported in literature. The response of primary frosted branch angiitis to systemic and/or topical corticosteroids was dramatic in our patients. There seems to be a broader spectrum of severity than previously reported in this disease.

**REFERENCES**

7. Sugin SL, Henderly DE, Frieden SM, Jampol LM, Doyle...


