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Recurrent Unilateral Frosted Branch Angiitis

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Abstract: In the left eye of a 27-year-old man we found perivascular creamy sheathing of retinal veins with retinal hemorrhages and, on fluorescein angiography, delayed filling of veins with late leakage. Dramatic recovery of visual acuity and healing of retinal lesions followed intravenous corticosteroid therapy. However, the condition recurred several times within a few months. Fluorescein angiography showed delayed filling of arteries and veins and arteriovenous anastomoses with a widespread capillary nonperfusion area. Eventually, neovascular glaucoma resulted. It is suggested that frosted branch angiitis is related to vascular occlusion. Systemic corticosteroid therapy seems to affect the course of this disease. **Jpn J Ophthalmol 1998;42:56–59** © 1998 Japanese Ophthalmological Society

Key Words: Corticosteroid, delayed filling, frosted branch angiitis, recurrence.

Introduction

In 1976, Ito et al[1] described a bilateral, unusual thick sheathing of the retinal vessels and designated this condition *frosted branch angiitis*. Since then, several similar reports[2–9] have been made, including the unilateral cases of Sugin et al reported in 1991.[7] There was no recurrence in any of these patients. We report an unusual case of this disease with several recurrences.

Case Report

On February 5, 1996, a 27-year-old man visited our hospital because of sudden visual loss that had occurred in his left eye 3 days previously. Systemic and ocular histories were unremarkable. Ocular examinations showed the best corrected visual acuity to be 25/20 in the right eye and 8/200 in the left. Intraocular pressure, the anterior chamber, and the vitreous cavity were normal in both eyes. On biomicroscopic fundus examination, extensive white sheathing of the veins and scattered retinal hemorrhages were visible in the inferior retina of the left eye, with retinal exudate and macular edema (Figure 1). The right eye was normal. Fluorescein angiography of the left eye demonstrated delayed filling of veins (Figure 2) and late leakage (Figure 3). Through systemic examinations, we investigated blood pressure, fasting blood sugar, complete blood cell count, C-reactive protein, erythrocyte sedimentation rate, urine analysis, and serum electrolytes.

Other examinations included enzyme-linked immunosorbent assay for herpes and cytomegalovirus, chest x-ray, and computed tomography of the brain. Results of all these tests were within normal limits. Pathergy test results were negative, while Montoux test results were positive. Human leukocyte antigen typing showed the presence of A24, A34, B7, B57, Cw3, Cw7, DR1, and DR13. The patient showed no signs of Behçet's disease, such as aphthous oral ulcers, genital ulcers, or erythema nodosum of the skin.

Methylprednisolone (1 g/d) was prescribed for 3 days, divided into four doses per day; intravenous injection was followed by oral prednisone (1 mg/kg per day for 11 days). No topical eyedrops were prescribed. Initially, dramatic recovery of visual acuity and healing of retinal lesions were noted. Seven days after therapy the corrected visual acuity was 20/20 in the left eye. One month later, visual acuity in the left eye had decreased gradually to 6/20. New retinal hemorrhages and macular edema without vascular sheathing were seen (Figure 4); 40 mg/d of oral pred-

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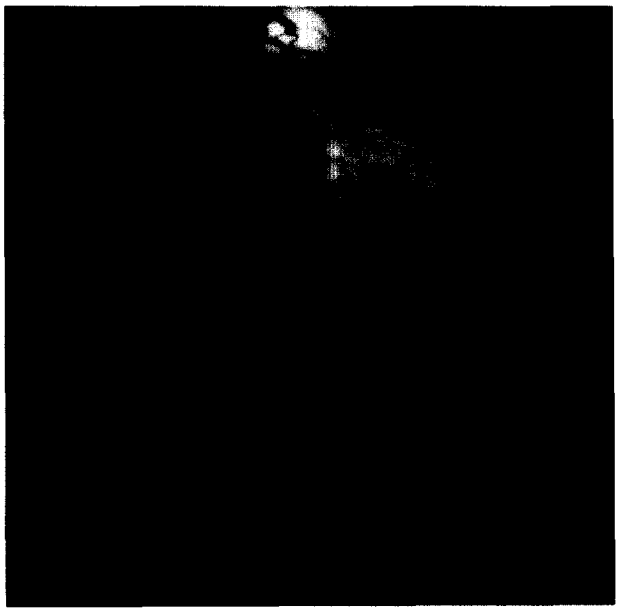

Figure 1. Creamy white venous sheathing and scattered retinal hemorrhages along inferior division of central retinal vein with retinal exudates in left eye of 27-year-old patient.

nisone was prescribed. Four weeks later, visual acuity had improved to 14/20 in the left eye. Follow-up was interrupted when, on June 26, the patient returned to our hospital because of sudden recurrence of vision loss in the left eye 3 days previously. On examination, there were several cells in the anterior chamber and the visual acuity was 4/200. Mild vascular sheathing of retinal arteries and veins and extensive retinal hemorrhages had recurred (Figure 5).

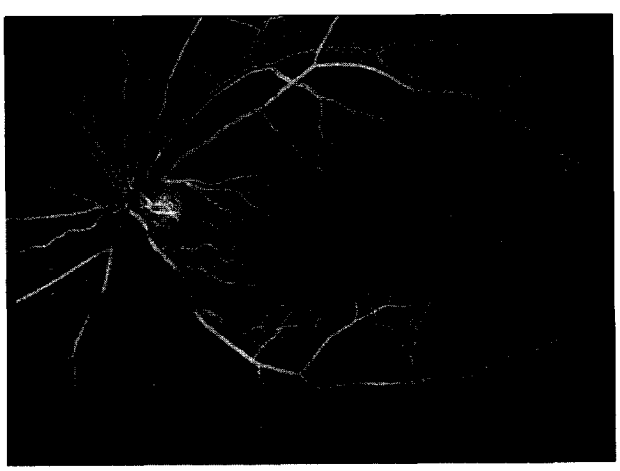

Figure 2. Fluorescein angiography shows delayed filling of involved retinal veins in early phase.

Figure 3. Later view shows leakage of dye from retinal veins and foveal involvement resulting in visual acuity of 8/200.

Optic disc swelling was visible with dilatation and beading of the superior division of the central retinal vein in the left eye. The right eye showed no abnormality.

The patient was lost to follow-up again, but eventually returned to our hospital on August 23, 1996. Remaining retinal hemorrhages were seen with more prominent vascular sheathing (Figure 6). Fluorescein angiography demonstrated delayed filling of retinal vessels (Figure 7) and two arteriovenous anas-

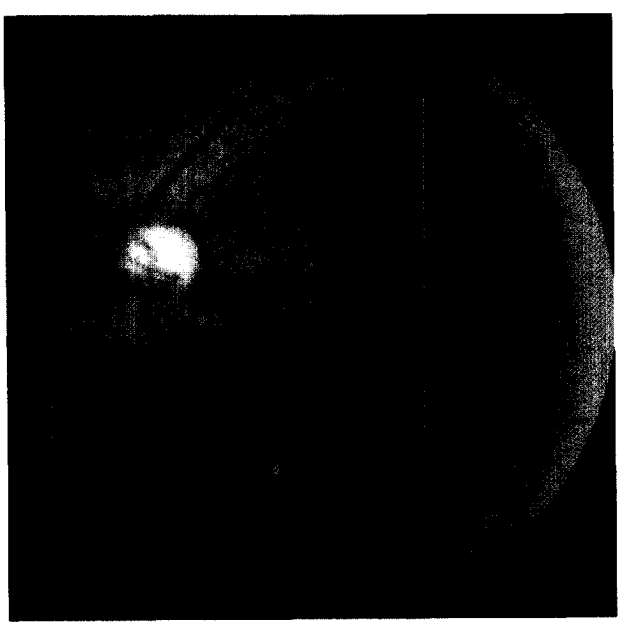

Figure 4. Retinal hemorrhages and macular edema recur without any vascular sheathing.

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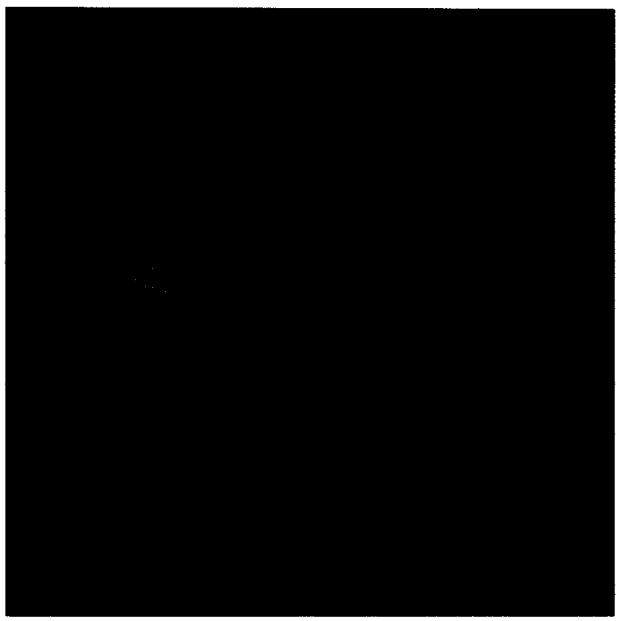

Figure 5. Extensive retinal hemorrhages and exudates recur; optic disc swelling with dilatation and beading of superior division of central retinal vein.

tomoses with widespread capillary nonperfusion areas (Figure 8). Eight weeks later, the patient complained of pain in the left eyeball and headache. Neovascular glaucoma was diagnosed in the left eye; intraocular pressure was 42 mm Hg and visual acuity

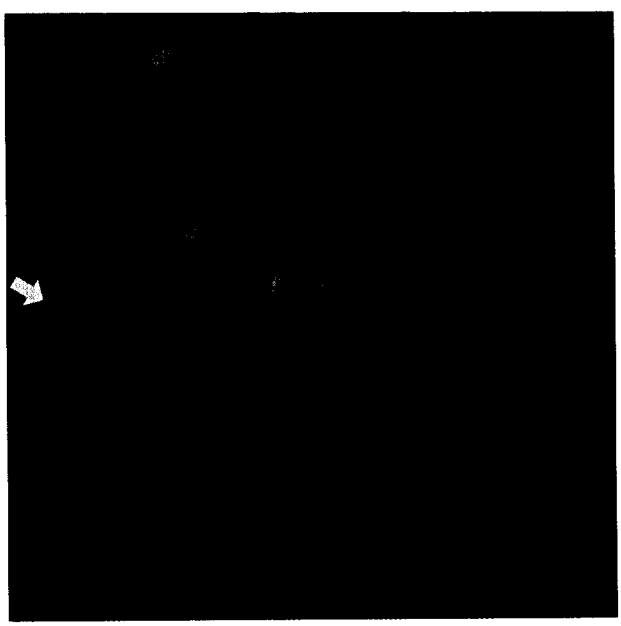

Figure 6. More prominent sheathing of retinal arteries and veins and remaining retinal hemorrhages with recovered veins (arrow).

Figure 7. Fluorescein angiography shows delayed filling of involved retinal arteries (arrow) in early phase.

was 4/200. Successful retinal photocoagulation was carried out using an argon laser. Four weeks later, cyclophotocoagulation with a CW-YAG laser was necessary to control the intractable neovascular glaucoma. Six weeks later, vitreous hemorrhage occurred, resulting in a visual acuity of 25/20 in the right eye and only hand motion detectable by the left. Intraocular pressure was 16 mm Hg in the right eye and 38 mm Hg in the left. The fundus of the right eye was normal.

Figure 8. Widespread capillary nonperfusion area below two arteriovenous anastomoses.

Discussion

The frosted branch angiitis and similar conditions[1–9] reported in 13 patients manifested with acute visual loss in healthy young persons which was usually bilateral. The fundus finding was characterized by severe white sheathing of the retinal vessels, mainly of veins. Fluorescein angiography demonstrated late vascular leakage without any evidence of stasis or occlusion. The response to systemic corticosteroids was favorable and there was no recurrence in any of these patients.

In the present case, frosted branch angiitis was unilateral and involved the retinal arteries and veins. Fluorescein angiography showed delayed filling of involved vessels, arteriovenous anastomoses, and a widespread capillary nonperfusion area below the vascular anastomoses. Despite dramatic response to intravenous corticosteroids, the condition recurred. The effect of systemic corticosteroid therapy on frosted branch angiitis in controversial. It has been reported to be useful in accelerating visual recovery and resolution of macular edema. There have been many reports[1–9] of visual recovery and no recurrence with and without systemic corticosteroid therapy. Earlier and better recovery of vision after intravenous corticosteroid therapy[10] occurred in our patient than in these other reports; nevertheless, there was recurrence.

It is suggested that frosted branch angiitis is related to vascular occlusion and systemic corticosteroid therapy seems to have an effect on the course of this disease. It is, therefore, recommended that patients with frosted branch angiitis be treated with moderate doses of systemic corticosteroids to achieve early recovery. With systemic corticosteroid therapy, however, there is always the potential for side effects, so patients should be followed up carefully. Because this disease occurs in otherwise healthy persons, corticosteroids are usually well tolerated.

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