

A RARE CAUSE OF RETINAL VASCULITIS: ACUTE FROSTED RETINAL PERIFLEBITIS

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SUMMARY

Frosted branch angiitis is a rare disorder. It is characterized by acute bilateral or unilateral visual loss with dramatic fundus abnormalities. These include marked sheathing of the retinal veins resembling frosted tree branches, retinal edema, late perivenous leakage on fluorescein angiography, and relative sparing of the optic nerve and retinal arterioles. In this report, we present a case of unilateral acute frosted retinal periflebitis in a 22 - year - old woman. The patient responded to topical and oral corticosteroid therapy 10 days after presentation.

Key Words: Frosted Branch Angiitis, Retinal Periphlebitis, Vasculitis, Acute Uveitis.

INTRODUCTION

In 1976, Ito et al reported a case of bilateral acute uveitis with severe and extensive sheathing of the retinal vessels in a 6 - year - old boy, which they termed frosted branch angiitis (1). Similar findings were reported in Japanese children in subsequent years, all characterized by acute bilateral visual disturbance that responded to corticosteroid therapy (2).

Kleiner et al were the first to report this condition in non-Japanese patients (3). They observed that the sheathing involved mainly veins, and proposed the term "acute retinal periphlebitis". Other subsequent cases from Western countries differed from the first cases in Japanese children in that patients were older and the condition was sometimes unilateral. To date, approximately 30 cases of frosted branch angiitis have been reported.

In this report, we present a case of unilateral acute frosted retinal periflebitis in a 22 - year - old woman.

This is the second case report of acute frosted retinal periflebitis in Turkey.

CASE REPORT

A 22 - year - old previously healthy woman developed an acute onset of unilateral blurred vision, pain and photophobia in her left eye. Examination 3 days after onset revealed visual acuity of 20/20 in the right and 20/30 in the left eye. There was mild ocular injection in the left eye. The pupils were measured 3 mm. each and were reactive to light and showed no afferent defect. There were 2+ cells and flare in the left anterior chamber; 2+ vitreal cells were also present. The fundus examination of the left eye showed diffuse perivenous sheathing; the arterioles were generally spared. There were also scattered intraretinal hemorrhages (Fig.1).

Laboratory tests revealed no serological evidence of syphilis, toxoplasmosis, rheumatoid arthritis, or collagen vascular disease. The complete blood cell count and white blood cell differential were normal. Sedimentation rate was 27 mm/hr. PPD skin test showed 10 mm. induration. Levels of angiotensin converting enzyme and serum lysozyme were normal. HLA - B27 antigen was negative. Chest X-ray was normal.

Topical corticosteroid drops 6 times a day and cycloplegics were administered. Oral 60 mg. methylprednisolon was also started. 10 days after presentation, left visual acuity improved to 20/20 and the anterior chamber showed only trace cells. The perivenous sheathing was markedly decreased in the left eye. 6 weeks after presentation, the anterior segment showed no inflammation, the perivenous sheathing and intraretinal hemorrhages had resolved. Topical and oral corticosteroids were tapered and stopped at all during a 10-week period. The patient did not experience a recurrence over a 18 months follow-up period.

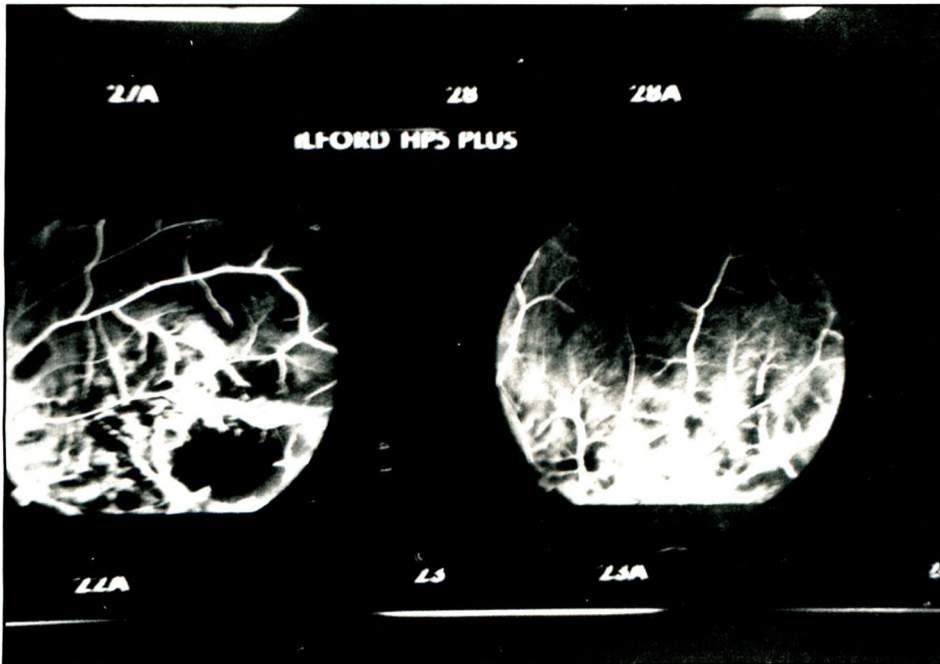


Fig. 1:
Scattered
intraretinal
hemorrhages
and diffuse
perivenous
sheathing
in the left eye.

DISCUSSION

Frosted branch angiitis is a rare disorder, with only approximately 30 previously reported cases (2). It is characterized by acute bilateral or unilateral visual loss with dramatic fundus abnormalities. These include marked sheathing of the retinal veins resembling frosted tree branches, retinal edema, late perivenous leakage on fluorescein angiography, and relative sparing of the optic nerve retinal arterioles. Moderate to severe iridocyclitis and some ocular hyperemia are often present. The cause of frosted branch angiitis remains unknown, but is suspected to be viral. It has been associated with fever and upper respiratory infection and cytomegalovirus retinitis in patients with AIDS (2, 4, 5).

Previous authors have advocated prompt treatment with systemic corticosteroids, citing eventual improvement of visual acuity and fundus abnormalities after its administration (4-6). In our unilateral case, after treatment with systemic methylprednisolone the periphlebitis began resolving and the visual acuity returned to 20/20 within 10 days. 6 weeks after presentation, the anterior segment showed no inflammation, the perivenous sheathing and intraretinal hemorrhages had resolved completely. Systemic work-up initiated at the time of the patient's first examination, and results of all of the studies were negative. As far as we know, this is the second reported case in Turkey.

There is a strong temptation to initiate some form of treatment in cases of sudden, severe vision loss, especially when it occurs bilaterally. Although a short

course of systemic corticosteroids is usually well tolerated, the potential for serious side effects does exist. This is especially true when the treatment extends over several weeks, as in this and other reports. In this inflammatory condition of uncertain cause, corticosteroid therapy is a rational approach that may be very helpful in the recovery of vision.

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