

Acute Idiopathic Frosted Branch Angiitis*

Akut İdiyopatik Donmuş Ağaç Dalı Anjiitisi

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Case Report

Olgu Sunumu

ABSTRACT

Five years old boy presented with bilateral visual loss for 3 days. His visual acuity was hand motion in both eyes. Ophthalmic examination revealed bilateral anterior uveitis, vitritis, periphlebitis resembling "frosted branch" and serous retinal detachment in the left eye. Systemic evaluation did not show any pathological findings. Topical and systemic steroids were started following periocular steroid injection. As the fundus findings did not improve, intravenous acyclovir was added to the treatment protocole on the 3rd day of steroid treatment. Fundus findings improved at the first week of acyclovir and steroid treatment. 4 months later, the visual acuity was 0.4 in his both eyes and macula showed pigmentary alterations. Frosted branch angiitis is a rare entity. Systemic acyclovir treatment may be helpful along with steroids in cases not responding to steroid treatment.

Key Words: Acyclovir, bilateral, frosted branch angiitis, serous retinal detachment, steroid.

ÖZ

Beş yaşında erkek hasta, her iki gözde üç gün önce başlayan görme kaybı şikayeti ile başvurdu. Görme keskinliği her iki gözde el hareketi düzeyinde olan hastanın muayenesinde her iki gözde ön üveit, vitritis, "donmuş ağaç dalı" görünümünde periflebit ve sol gözde seröz retina dekolmanı saptandı. Sistemik değerlendirmede herhangi bir patoloji saptanmayan hastaya, perioküler steroid enjeksiyonunu takiben topikal ve sistemik steroid başlandı. Fundus bulgularında gerileme olmaması üzerine, izlemin 3. gününde tedaviye intravenöz asiklovir eklendi. Asiklovir ve steroid tedavisinin 1. haftasında fundus bulgularında gerileme izlendi. Hastanın 4. aydaki muayenesinde görme keskinliğinin her iki gözde 0.4 olduğu, makulada pigmenter değişikliklerin ortaya çıktığı görüldü. İdiyopatik donmuş ağaç dalı anjiitisi nadir görülen bir durumdur. Steroid tedavisine yanıt vermeyen olgularda, steroid tedavisine sistemik asiklovir eklenmesi fayda sağlayabilmektedir.

Anahtar Kelimeler: Asiklovir, bilateral, donmuş ağaç dalı anjiitisi, seröz retina dekolmanı, steroid.

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INTRODUCTION

Frosted branch angiitis (FBA) is a rare entity which is characterized by retinal vasculitis with severe sheathing, producing the appearance of frosted branches of a tree. Up to now, nearly 60 cases have been reported, mainly from Japanese literature.¹ Our case is an acute idiopathic frosted branch angiitis responding to steroid and anti-viral therapy.

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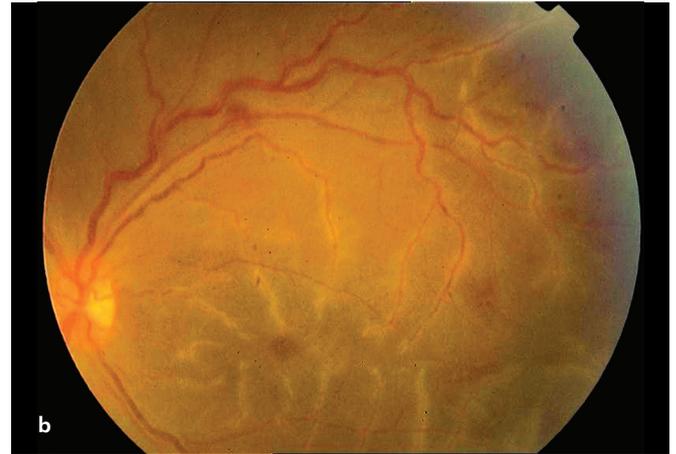
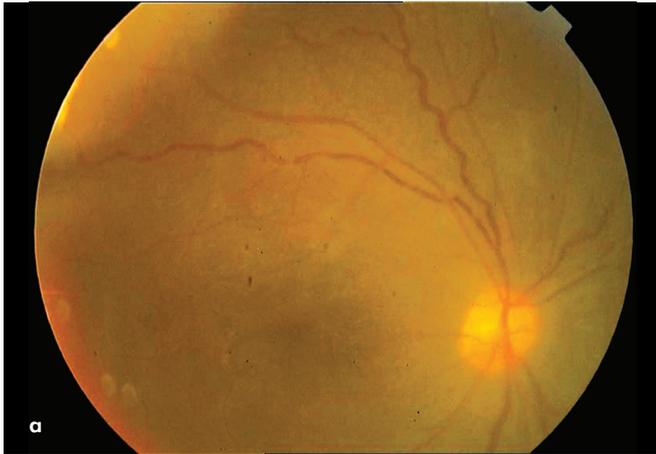


Figure 1a: Fundus photographs showing perivascular sheathing resembling frosted branch angiitis and retinal edema in the right eye **b:** and the left eye.

CASE REPORT

A five years old boy presented with bilateral visual loss for 3 days. His medical history did not reveal any systemic diseases including preceding viral infections. His visual acuity was hand motions in both eyes. Ophthalmic examination revealed bilateral anterior uveitis (4+), vitritis, periphlebitis resembling “frosted branch” and serous retinal detachment in the left eye (Figure 1a, b).

Fundus fluorescein angiography showed late leakage from retinal vessels (Figure 1c). Systemic evaluation regarding sarcoidosis, systemic lupus erythematosus, Wegener’s granulomatosis, leukemia and lymphoma did not show any pathological findings.

Serology for rubella, cytomegalovirus, herpes simplex, syphilis, tuberculosis and brucellosis were negative. Topical dexamethasone gtt 24x1, cyclopentolate 4x1 and intravenous methyl prednisolone (30 mg/kg/day) were started following periocular steroid injections.

As the fundus findings did not improve, intravenous acyclovir (30 mg/kg/day) was added on the third day. Anterior chamber reaction, periphlebitis and serous retinal detachment regressed by the first week. Following intravenous treatment, oral prednisolone (1 mg/kg/

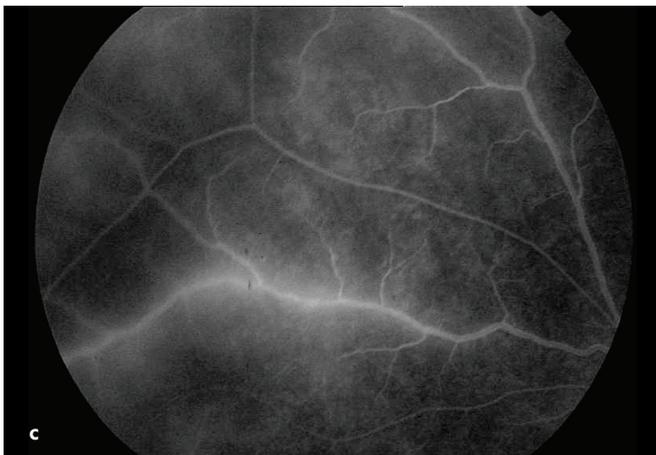


Figure 1c: Fluorescein angiography showing late perivascular leakage in the left eye.

day) and oral acyclovir (70 mg/kg/day) were started at the second week and tapered and discontinued over 2 months. At his 4 month follow-up visit, visual acuity was 20/50 in his both eyes, macula showed pigmentary alterations (Figure 2).

DISCUSSION

Acute idiopathic FBA is a rare entity predominantly affecting the young and healthy patients with one peak in childhood and a second in the third decade.¹ The most common symptoms are visual loss, floaters and photopsiae. Fundus appearance is characteristic with widespread retinal vasculitis and retinal edema. Intra-retinal hemorrhages, vitritis, iritis and papillitis may also be seen. Fundus fluorescein angiography shows vascular leakage in the late phase.¹ Although most of the cases are bilateral, unilateral and atypical presentations have also been reported.²⁻⁴

The etiology of acute idiopathic FBA is unknown. The history of preceding prodromal illness in many cases along with good response to steroids suggest the possibility of an immune mechanism.^{1,5,6} It is not clear whether this disorder is a distinct clinical syndrome or a clinical sign accompanying a number of inflammatory or neoplastic diseases.

Some authors suggest to classify FBA as primary (idiopathic) form and secondary form which is associated with a concurrent inflammation (CMV retinitis, toxoplasmosis, syphilis, sarcoidosis, systemic lupus erythematosus and others) and masquerades (leukemia and lymphoma).^{1,5} Systemic steroid treatment has been effective in many FBA patients with good visual recovery.¹ There are few cases having steroid and acyclovir treatment in the literature but the therapeutic effect is not clear.^{1,7}

In our case, on the third day of steroid treatment, the lack of improvement in visual acuity and fundus findings led us to discuss about starting anti-viral treatment despite the negative serology for viral infections.

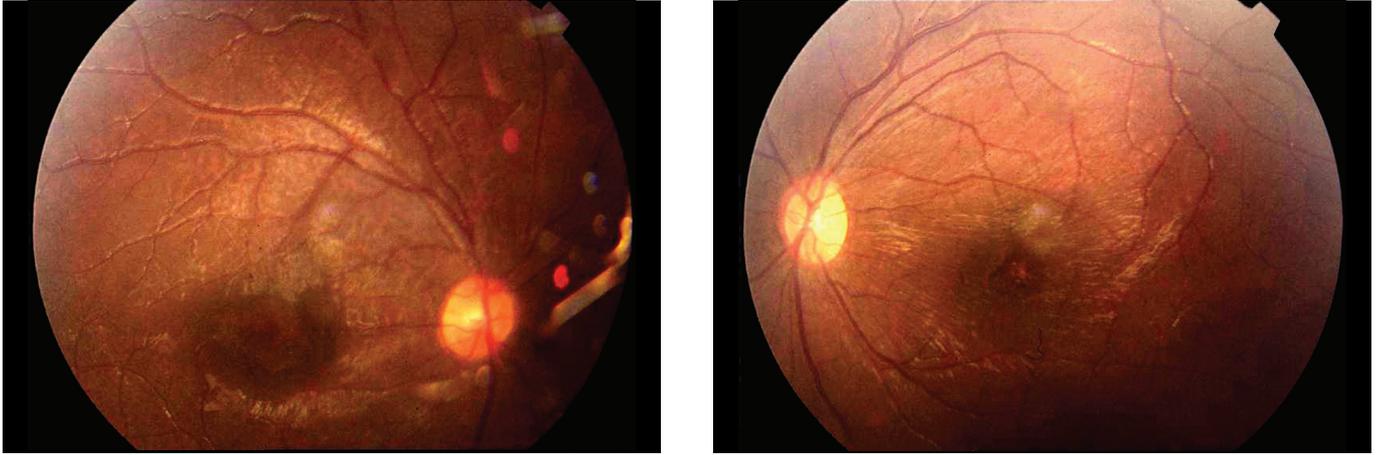


Figure 2a: Fundus photographs showing macular pigmentary alterations at 4-month follow-up in the right eye, **b:** and the left eye.

Fundus findings improved at the first week with steroid and acyclovir treatment and remained stable for 4 months. The improvement of clinical findings could be due to the late response to systemic steroids as well as antiviral treatment. We believe that the preferred approach should be the analysis of intraocular fluid if available before giving anti-viral treatment in case of negative blood serology.

In conclusion, acute idiopathic FBA is a rare disease and it is important to rule out concurrent inflammation or masquerades in FBA patients. Since the natural course of the disease is not well-known, prompt treatment with steroids is recommended to protect the eye against macular scarring.⁸ Acyclovir treatment along with steroids may also be beneficial in certain cases.

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