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LETTER TO THE EDITOR

Bilateral Central Retinal Artery Occlusion in Behçet Disease

Ebru Esen, MD, Selcuk Sizmaz, MD, Ayna Sariyeva, MD, and Nihal Demircan, MD

Department of Ophthalmology, Çukurova University, School of Medicine, Adana, Turkey

ABSTRACT

Purpose: To report a case of bilateral central retinal artery occlusion as the primary ocular manifestation of Behçet disease.

Methods: A 38-year-old patient, with no history of previous ocular complaints, admitted with bilateral sudden vision loss. His ophthalmologic examination revealed a visual acuity of no light perception in both eyes and the diagnosis of bilateral central retinal artery occlusion. A detailed systemic investigation was conducted.

Results: With the history of recurrent oral and genital ulcers, and the presence of ocular and skin findings, the diagnosis of Behçet disease was made. Despite a potent immunosuppressive therapy the patient became absolutely blind.

Conclusions: Even though retinal arteries are rarely affected in Behçet disease, in the cases of retinal artery occlusion, it should be considered among the differential diagnosis. To our knowledge this is the first reported case of bilateral central retinal artery occlusion as the primary ocular manifestation of Behçet disease.

Keywords: Behçet disease, bilateral, central retinal artery occlusion, retinal vasculitis, vision loss

Behçet disease (BD) is a chronic idiopathic inflammatory disorder characterized by obliterative and necrotizing vasculitis that affects both arteries and veins of many organ systems.[1] The eye is the most commonly affected organ and ocular involvement is closely related to the patients' quality of life. The typical ocular manifestation of BD is a nongranulomatous uveitis and obliterative retinal vasculitis.

Retinal artery occlusion is closely related to systemic conditions, and in most of the patients there is an underlying disorder leading to thrombosis, embolic impaction, or other causes of vaso-occlusion. Occlusive vasculitis is the hallmark of BD, and thrombotic events are described to be in association with the disease.[2] So during the course of BD vaso-occlusive pathologies may occur in various organs, including the eyes. But veins are predominantly affected and occlusion of retinal vein is not an uncommon situation, whereas retinal artery occlusion due to BD has been reported rarely in the literature. There are only few cases with central and branch retinal artery occlusion in BD.[3–6] We reported a patient who developed bilateral central retinal artery occlusion (CRAO) as the primary manifestation of BD before any other ocular symptoms, resulting in bilateral permanent vision loss.

A 38-year-old male patient presented with sudden onset painless visual loss in both eyes for 2 days' duration. The patient had had no previous ocular complaints until he lost his vision. He had been hospitalized for diarrhea in another clinic 10 days prior to his vision loss, but the workup had been unable to ascertain the cause. The medical history also revealed recurrent oral and genital ulcerations, skin lesions, arthralgia, and swelling of the knees. On his ophthalmic examination visual acuity was no light perception in both eyes. The pupils were mydriatic and unresponsive to light. The intraocular pressures were normal and anterior segment slit-lamp examination revealed 1 + cells in the anterior chambers in both eyes. Fundus examination of the right and left eyes showed marked retinal edema, optic disc swelling,

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Correspondence: Ebru Esen, MD, Çukurova Üniversitesi Tıp Fakültesi, Göz Hastalıkları Anabilim Dalı, 01330 Adana, Turkey. E-mail: ebrublg@yahoo.com

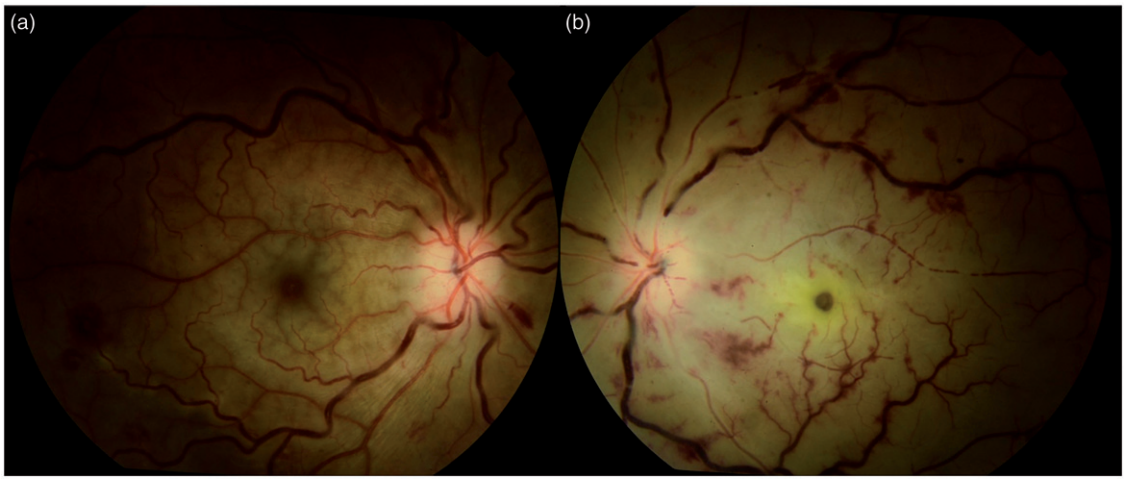


FIGURE 1. Fundus photographs of the right (a) and left (b) eyes at presentation, showing marked retinal edema, optic disc swelling, dot-blot hemorrhages, arteriolar narrowing, tortuous veins, and cherry red spot on the fovea.

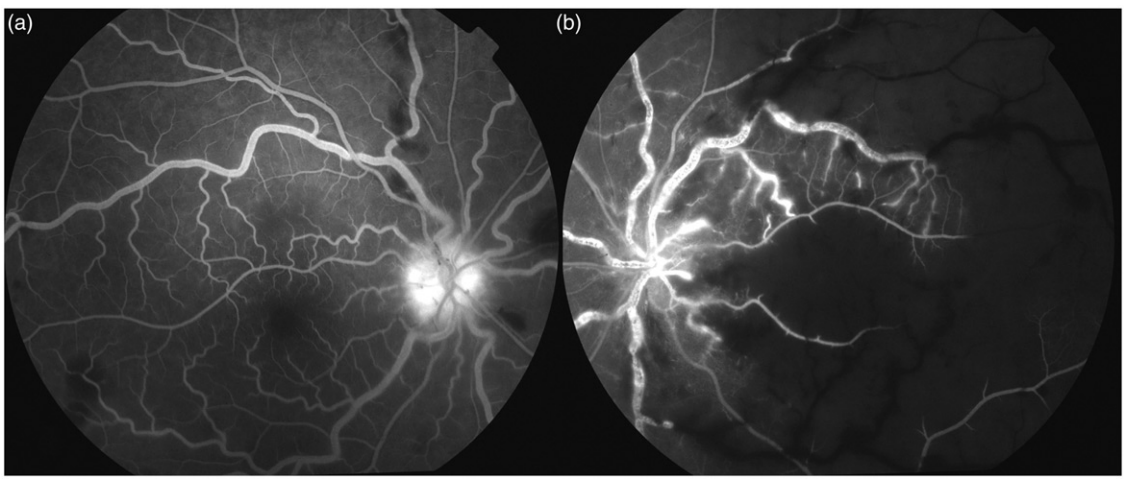


FIGURE 2. Fundus fluorescein angiograms of the right (a) and left (b) eyes at presentation, showing bilateral delay in the filling of retinal vasculatures and dilated and tortuous retinal veins. Occlusion of retinal arteries and capillary nonperfusion, staining of vessels, and vascular leakage are remarkable in the left eye.

dot-blot hemorrhages, arteriolar narrowing, tortuous veins, and cherry red spot on the fovea (Figure 1). Fundus fluorescein angiography (FFA) revealed bilateral delay in the filling of retinal vasculatures. Occlusion of retinal arteries and capillary nonperfusion compatible with central retinal artery occlusion (CRAO) and staining of vessels and vascular leakage consistent with occlusive vasculitis were remarkable in the left eye. Dilated and tortuous retinal veins were thought to occur as a result of severe optic disc edema (Figure 2).

The patient was hospitalized and administered topical and systemic medications for lowering intraocular pressure. Meanwhile, a thorough workup was conducted based on the clinical diagnosis of bilateral CRAO. Laboratory data were significant for white blood cell count (13,700/mL), erythrocyte sedimentation rate (ESR = 120 mm/h), and C-reactive protein (CRP = 22.3 mg/L); peripheral blood smear and urine analysis were normal. Blood serology results for infectious diseases were unremarkable. Antinuclear antibody, anti-double-stranded DNA, antiphosphoplipid antibody, anti-neutrophil cytoplasmic antibody, cryoglobulins, and cryofibrinogen were all negative and complement proteins (C3 and C4) were at normal levels. It was remarkable that plasma homocystein level was elevated (16.2 μmol/L). His electrocardiogram and echocardiogram were normal. Carotid artery ultrasound did not show any atherosclerotic plaque or thrombosis. Sinuses (Waters' view), chest x-ray, and brain and orbital magnetic resonance imaging revealed normal findings. The patient was referred to the dermatology and rheumatology departments. The pathergy test was negative but his physical examination revealed acne-like papulopustular lesions on the skin. With the presence of ocular and skin findings and the history of recurrent oral and genital ulcers, the diagnosis of BD was made according to the International Study Group for Behçet's Disease criteria.[7]

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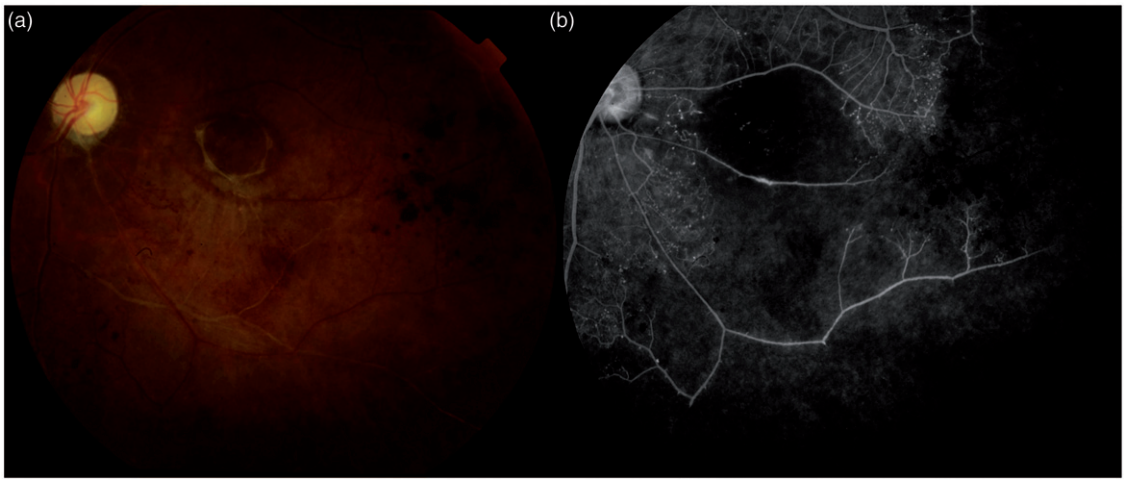


FIGURE 3. (a) Fundus photograph of the left eye at the third month after admission, showing optic atrophy, vascular sheathing, ghost vessels, gliosis of the retina and macular scarring. (b) Fundus fluorescein angiogram of the same eye revealing a wide retinal ischemic area.

The patient was treated with intravenous pulse methylprednisolone 1 g/day for 3 consecutive days, followed by oral prednisone 1 mg/kg/day with a tapering dose regimen. Cyclophosphamide was administered on the seventh day of steroid treatment. The patient was also prescribed colchicine 0.5 mg and acetylsalicylic acid 100 mg per day. The treatment included folic acid 5 mg/day for lowering blood homocysteine level. Despite potent immunosuppressive treatment, the visual acuity didn't improve and unfortunately the patient became absolutely blind. In the following days the patient received 2 additional courses of cyclophosphamide infusion, which was later replaced by interferon-α therapy. Within several weeks optic atrophy developed in both eyes. Reperfusion of the retina was maintained in the right eye, but fundus examination of the left eye revealed vascular sheathing, ghost vessels, gliosis of the retina, and macular scarring with a wide ischemic area detected on FFA (Figure 3). Panretinal photocoagulation is planned for the left eye of the patient. Erythrocyte sedimentation rate, CRP, and plasma homocysteine level decreased to 21 mm/h, 0.29 mg/L, and 11.4 μmol/L, respectively. Oral and genital ulcers gave good response to the immunosuppressive treatment.

Our patient presented with a devastating and an unfortunate situation, acute bilateral CRAO, which is an extremely rare cause of sudden bilateral vision loss. Previously, such cases were described in patients with systemic lupus erythematosus, giant cell arteritis, Churg-Strauss vasculitis, and Wegener granulomatosis.[8–11] The common characteristic of these diseases is a severe vaso-occlusive disorder like BD. In the systemic workup all potential risk factors for CRAO were investigated, but because the angiographic features were consistent with an occlusive vasculitis, the differential diagnosis consisted especially of infectious and noninfectious diseases, immune-mediated disorders that can be the reason of retinal vasculitis. Laboratory data did not point out any specific disease; the diagnosis of BD was decided according to the clinical findings and medical history. Abnormal laboratory data included elevated ESR and CRP levels that were reported to be in correlation with disease activity by some authors.[12] However, there is not a consensus about an original laboratory marker for both diagnosis and disease activity in BD.

Two mechanisms are postulated for arterial vaso-occlusion in BD. It is thought to occur either by focal arteritis and the infiltration of the vessels by inflammatory cells leading to an arterial thrombosis, or the compression of the artery by the focus of retinitis.[4–5] In our patient high plasma homocystein level was remarkable. Hyperhomocysteinemia itself is known as a risk factor for both venous and arterial thrombosis, and it was thought to contribute to the thrombotic events in BD, and induce proinflammatory cytokines during the course of the disease.[13,14] Thus, folic acid was prescribed to our patient to decrease blood homocysteine level and reduce the risk of thrombosis. Patients with BD who have other risk factors proved to facilitate thrombotic events may be more likely to experience such complications.

Although rare, BD should be among differential diagnoses of patients with acute CRAO. This condition may develop in an already diagnosed patient as well as a primary manifestation of BD. To the best of our knowledge this is the first case of BD with bilateral CRAO as the primary ocular involvement.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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