CASE REPORT

Cilioretinal Artery Occlusion Combined with Impending Central Retinal Vein Occlusion in Incomplete Behçet's Disease

İnkomplet Behçet Hastalığında Olası Santral Retinal Ven Tıkanıklığı ile Kombine Silyoretinal Arter Tıkanıklığı

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SUMMARY

A 28-year old woman presented with painless sudden blurring vision in her left eye for 3 hours. With a history of recurrent oral aphtae more than 3 times in a month and her sibling has been being under the diagnosis of Behçet's disease. Fundus findings in her left eye, demonstrated occlusive resembling cilioretinal artery occlusion combined with impending central retinal vein occlusion and fluorescein angiography showing delayed filling of the central retinal vein and prolonged arteriovenous fillig time. Labratory examination for HLA-B51 was positive. The patient was diagnosed incomplete Behçet's Disease (BD) and administrated with oral prednisone. After oral prednisone treatement, visual acuity in the left eye recovered from 20/63 to 20/20 as well as resolution of the retinal findings.

Key Words: Cilioretinal artery occlusion, impending central retinal vein occlusion, Behçet's disease.

ÖZ

Yirmi sekiz yaşında kadın hasta sol gözünde yaklaşık 3 saattir devam eden ağrısız, ani görme bulanıklığı şikayeti ile başvurdu. Bir ay içinde 3'ten fazla tekrarlayan oral aft öyküsü ile beraber kardeşinin de behçet hastalığı tanısı ile tedavi altında olduğu öğrenildi. Fundus muyanesinde sol gözde olası santral retinal ven tıkanıklığı ile beraber siliyoretinal arter tıkanıklığına benzeyen görünüm mevcut olup fundus anjiografisinde santral retinal vende dolum defekti ile beraber arteriovenöz dolum zamanının uzamış olduğu saptandı. HLA-B51 testti. Olguya inkomplet Behçet hastalığı tanısı konup oral steroid tedavisi başlandı. Oral prednison tedavisinden sonra, sol gözde görme seviyesinin 20/63'ten 20/20'ye yükselmesiyle beraber retina bulgularında düzelme olduğu gözlendi.

Anahtar Kelimeler: Silyoretinal arter tıkanıklığı, olası santral retinal ven tıkanıklığı, Behçet hastalığı.

INTRODUCTION

The essential pathology in Behçet's disease (BD) is an obliterative and necrotizing vasculitis that affects both arteries and veins in all system, moreover ocular involvement in BD occurs in around 70% of patients and is associated with a high risk of visual loss.[1]

Posterior segment involvement is a major prognostic factor in ocular BD which is seen in up to 93% of patients with ocular disease and recurrent attacks of posterior segment can cause severe retinal damage and loss of vision.[2] Both retinal arteries and veins could be involved in BD, much as venous involvement is more common.[3]

In this article, we reported an unusual case of unilateral retinal vascular events resembling cilioretinal artery occlusion combined with impending central retinal vein as the primary ocular manifestation of Behçet disease.

CASE REPORT

A 28-year old woman presented with sudden painles blurring of vision in her left eye for 3 hours. On ocular examination revealed that the best corrected visaul acuity (BCVA) was 20/20 in the right eye and BCVA was 20/63 in the left eye. Intraocular pressure (IOP) was 15 mmHg in the right eye and IOP was 14 mmHg in the left eye. Both eyes had normal subjects in slit-lamp biomicroscobic evaluation. Fundus examination of the left eye showed increased tortuosity and venous dilatation on the retinal vessels and blurred optic disc margin particularly temporal side. In addition, well demarcated retinal whitening of the superior quadrant, sparing the fovea along the course of a cilioretinal artery was revealed without any signs of vitritis (Figure 1a). Fluorescein fundus angiography (FA) of the left eye demonstrated with delayed filling of the central retinal vein and prolonged arteriovenous fillig time and slightly optic disc staining and dye leakage particularly at the temporal side of the disc area (Figure 1bc). The fundus examination and fluorescein fundus angiography appearence of the right eye was unremarkable (Figure 2a,b). Systemic blood pressure was 130/70 mmHg. The patient had a history of recurrent oral aphtae more than 3 times in a month and her sibling has been being under the diagnosis of Behçet's disease with ocular involvement for 2 years and has been entering prolonged remission following treatment with systemic cyclosporin. Haemothologic studies for inflammatory markers were increased (erythrocyte sedimentation rate, 32 mm/1 h; C-reactive protein, 18 mg/L). The laboratory tests for C3 and C4, angiotensin-converting enzyme, antiphospholipid antibody, rheumatoid factor, antineutrophilic cytoplasmic antibodies were negative. The tests results which indicate impairing in systemic coagulation that including activated partial thromboplastine time, prothrombin time, protein S and C, antithrombin iii, homocysteine were normal. Human leukocyte antigen

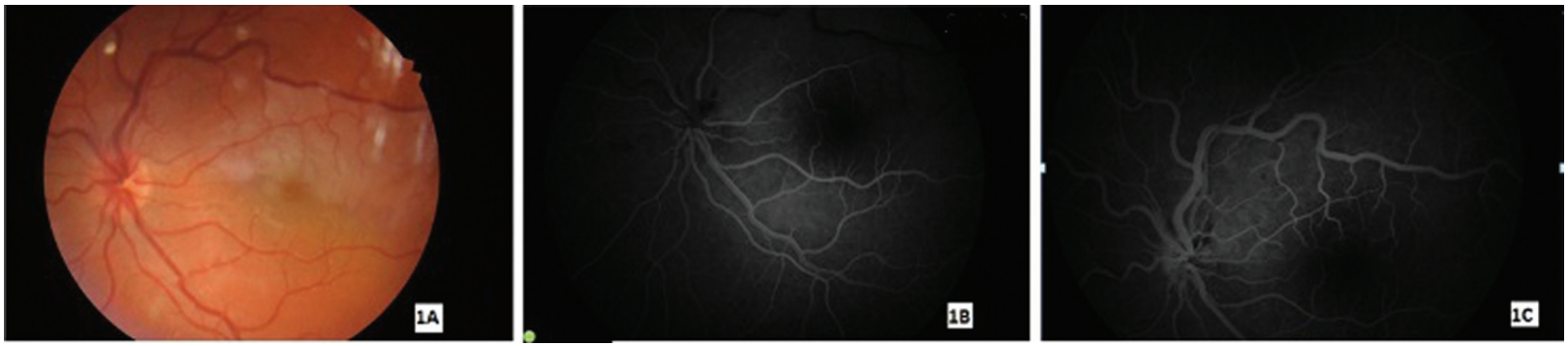

Figure 1: *1A: Increased tortuosity and veneous dilation of retinal vessels and blurred disc magrin particularly temporal side. Retinal whitening of the superior quadrant along the course of a cilioretinal artery (a), Delayed filling of the central retinal vein and prolonged arterivenous fillig time (b), Dye leakage particularly the temporal side of the disc area (c).*

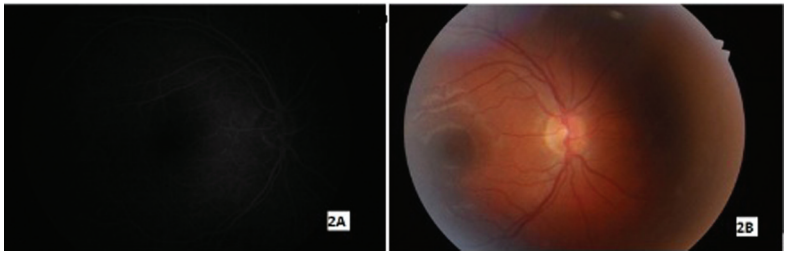

Figure 2a,b: *Fluorescein angiography which has normal dye pattern belongs to the right eye (a), Fundus imagine which has normal appearence belongs to the right eye (b).*

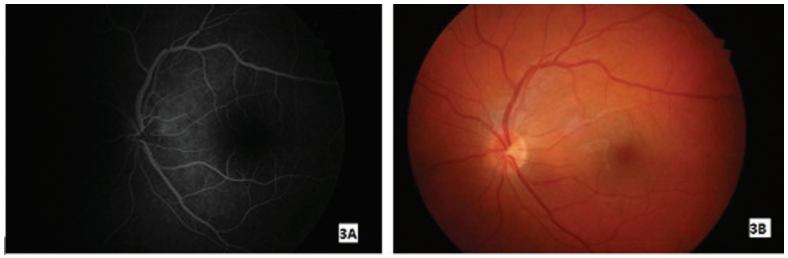

Figure 3a,b: *After the 3 months, normal dye transit time as well as no abnormal fluorescence (a), After the 3 months, resolution of the disc edema, tortuosity and dilation of the retinal veins an macula (b).*

typing was positive for B51. The result of pathergy test, was performed with using an intradermal 21-gauge needle puncture on the skin, was suspicious positive. Neurological evaulation was normal.

The patient was diagnosed with incomplete Behçet disease (BD) and treated with oral prednisone (1 mg/kg/day). Treatment was tapered gradually which was given until 10 mg/daily approximately during the 3 months as long as new attack didn't occur. Two weeks later, visual acuity was improved from 20/63 to 20/32 and 3 months later visual acuity was recovered to 20/20, including resolution of the disc edema, tortuosity and dilation of the retinal veins (Figure 3a). Fluorescein angiography revealed normal dye transit time as well as no abnormal fluorescence (Figure 3b). She has been taking oral steroid at the maintenance dose without any new attacks for the last 4 months.

CONCLUSIONS

The diagnosis of Behçet's disease (BD) can be divided into complete disease when 4 major criteria are present and imcomplete disease when there are 3 major features, 2 minor, or typical recurrent ocular symptom plus 1 major or 2 minor features.[4] Our patient was presented with resembling a cilioretinal artery occlusion and impending CRVO, had recurrent oral ulcers and suspicious positive patergy test result which shows hyperreactivitiy of the skin. She is best classified as incomlpete BD.

HLA-B51 is the most strongly associated known genetic factor to BD. A genetic contribution to BD is also supported by the high siblings recurrence risk ratio, estimated to be between 11.4 and 52.5 in the Turkish population.[5] Both a careful history and a positive HLA-B51 can help make a diagnosis of underlying Behçet's disease.

Behçet's disease (BD) is a multi-systemic inflammatory disorder which can affect all types and sizes of blood vessels. Fei Y at al.,[6] reported that in BD disease, vascular lesions correlated with a high frequency of cardiac involvement and a low incidence of ocular lesions, genital ulcers, and arthritis, thus we didn't need to do intensive cardiac and vascular investigation.

Ocular involvement in BD always progresses to both eyes and it may be presenting initial manifastation of the disease in 10-20% of cases, Although the ocular disease may initially starts unilateral. Posterior segment involvement in BD is a significant cause of severe visual loss unless diagnosed early and treated properly. In the posterior segment, vitreal inflammatory cells always present during the acute attacks. Vasculitis is the essential procces involving veins and arteries with tendency to thrombosis and repeated vasculitic attacks result in permanent vascular occlusion.[6] Tian et al.,[7] reported a case which had bilateral central retinal artery occlusion and recurrent papillitis due to obliterative retinal vasculitis in incomplete BD. In our patient, unilateral cilioretinal artery occlusion (CLRAO) and impending central retinal vein occlusion (CRVO) was the first ocular findings that led to clinical evaluation and diagnosis of BD and had no vitreous inflammatory cells. We think as the clinical signs of our patient has not been established during the early period of the disease.

Occlusion of retinal vein in BD may occur at any location from the central retinal vein to the peripheral small branches.[8] Branch retinal vein occlusion and ischemic retinal vasculitis have been reported as the first presentation of ocular BD in 28% and 21%, respectively, while central vein (4%) and artery (1%) occlusions are less common.[9] Abu-Ameerh MA at al.,[10] have reported a low prevalance for retinal vascular occlusion in BD (16.7%) as CRVO and BRVO.

Ozdal PC at al,.[11] declerating in a serie including 21 eyes with occlusion of the retinal veins in BD , It has been distributed as 15 with BRVO, 2 with hemisphere vein occlusion, 4 with small macular vein occlusion. Although retinal artery occlussion rarely seen in BD, few case presentations have been reported in literature.[12-13] To the best of our knowledge, this is the first report case of showing a cilioretinal artery occlusion associated with impending CRVO as the primary ocular manifestation in incomplet BD.

A cilioretinal artery occlusion associated with branch retinal vein occlusion was only published in one patient with Behçet's uveitis, besides this article, the author advocated the pathomechanisms of CLRAO that an increasement of the intraluminal pressure in the retinal capillaries due to CRVO which is exceed the pressure in the cilioretinal artery could lead to its occlusion.[14] As differentiation , the CLRAO occured in our patient was functional as a result of destructive impact of active ocular inflammation.

The recognized FA features of BD include capillary dropout, diffuse vasculitis, staining and hyperfluorescence of the optic disc, macular edema and ischemia, and irreguler delayed filling of choriocapillaries.[15] Bottos M et al.,[16] showed in FA disclosed delayed filling of the central retinal vein and prolonged arteriovenous filling time without capillary non perfusion in a patient which is diagnosed impending CRVO associated with CLRAO. In our patient we observed in FA, not only slight dye leakage at the optic disc but also prolonged arteriovenous transit time without capillary nonperfusion.

Systemic corticosteroids have proven effective in the treatment of acute ocular attacks in patients with BD and it should be used only in patients with active inflammation with an threatening to vision. Tapering corticosteroids slowly is important because rebound attacks may be more severe than the natural course of the disease.[16]

In our patient, we administrated oral prednisolone in 1 mg/kg/day during the first month. In the follow-up period, the patient's clinical symptoms and ocular findings were improved and medication for 10 mg/daily for 3 months to reduce up to continued this dosage. Following systemic corticosteroid therapy, begining signs of resolution of the ocular inflammation and reperfusion of the occluded artery were observed in our patient.

Early and appropriate treatment with anti-inflammatory medication may help resolution of the active ocular inflammation, reperfusion of the occluded artery and improve the visual acuity.

In summary, This case report brings into literature a new retinal vascular findings as the primary ocular manifestation in BD. Impending CRVO combined with CLRAO may occur in association with a part of BD, which could initially be unilateral and result in decreased visual acuity. It has to be considered in the differential dagnosis of retinal vascular occlussions especially in countries where the disease is prevalent.

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