Vitrectomy for Optic Disc Traction Syndrome in Central Retinal Vein Occlusion Due to Factor V Leiden Mutation

Faktör V Leiden Mutasyonu Varlığında Santral Retinal Ven Tıkanıklığına Bağlı Optik Disk Traksiyon Sendromu Gelişen Olguda Vitrektomi

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Case Report *Olgu Sunumu*

ABSTRACT

A 45-year-old woman came to retina clinic with loss of vision in the right eye. Ischemic central retinal vein occlusion (CRVO) was diagnosed. The laboratory tests for the etiology revealed Factor V Leiden mutation (FVL, R506Q mutation). B-scan ultrasonography revealed vitreopapillary traction associated with disc edema and peripapillary retinal detachment. She underwent vitrectomy, posterior hyaloid stripping and peripapillary subretinal tissue-plasminogen activator (t-PA) injection to relieve the traction, facilitate blood removal and increase the retinal perfusion. Optic nerve head elevation and retinal detachment resolved within the first postoperative week. Retinal hemorrhages cleared within the first 2 weeks and her vision improved from light perception to 20/400. Optic disc traction may complicate the course of ischemic CRVO and may benefit from vitrectomy combined with focal delivery of thrombolytic agents.

Key Words: Optic disc traction syndrome, central retinal vein occlusion, factor V mutation, tissue-plasminogen activator.

ÖZ

Sağ gözde görme kaybı ile başvuran 45 yaşında bayan hastaya iskemik santral retinal ven tıkanıklığı (SRVT) tanısı kondu. Etiyolojiye yönelik araştırmalar sonucu Faktör V Leiden (FVL, R506Q mutasyonu) mutasyonu saptandı. B-scan ultrasonografik incelemede vitreopapiller traksiyona eşlik eden disk ödemi ve peripapiller retina dekolmanı izlendi. Retina içi kanamaların çekilmesini hızlandırmak, kan akımını artırmak ve traksiyonu rahatlamak için olguya vitrektomi, arka hyaloid soyulması ve peripapiller retina altı doku plazminojen aktivatörü (t-PA) uygulandı. Operasyonu takip eden birinci haftada, optik sinir başı kabarıklığının ve retina dekolmanının yatıştığı izlendi. Operasyonu takiben ikinci haftada, retina içi kanamalar çekildi ve görme el hareketleri seviyesinden 0.05'e yükseldi. Optik disk traksiyonu, iskemik SRVT'nın bir komplikasyonu olarak karşımıza çıkarak hastalığın seyrini etkileyebilir ve vitrektomi ile birlikte bölgesel trombolitik ajanların verilmesinden fayda sağlanabilir.

Anahtar Kelimeler: Optik disk traksiyon sendromu, santral retinal ven oklüzyonu, faktör V Leiden mutasyonu, doku plazminojen aktivatörü.

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INTRODUCTION

Central retinal vein occlusion (CRVO) is more common in elderly especially with a history of smoking, diabetes, hypertension, atherosclerosis and glaucoma. Primary hypercoagulable states, such as antithrombin III, protein S or protein C deficiencies and hyperhomocysteinemia, are more often seen among young (<50) patients with CRVO.[1]

90% of patients with hereditary activated protein C (APC) resistance have a point mutation in factor V gene (Factor V Leiden mutation, FVL) which renders factor V resistant to inactivation by APC complex.[2]

FVL increases the risk for thromboembolic events 5-10 fold in heterozygotes and up to 50-100 folds in homozygotes.[3] Vitreopapillary traction may occur during the evolution of the posterior vitreous detachment and also as a part of proliferative diabetic retinopathy.[4,5] It has recently been recognized as a feature that may complicate the course of CRVO by disrupting the axoplasmic flow and blood supply to optic nerve head.[6] In this report, the use of vitrectomy to relieve the vitreopapillary traction is described in a patient with ischemic CRVO due to FVL. Surgical encounter restored the retinal perfusion, alleviated the disc edema and resulted in an ambulatory vision.

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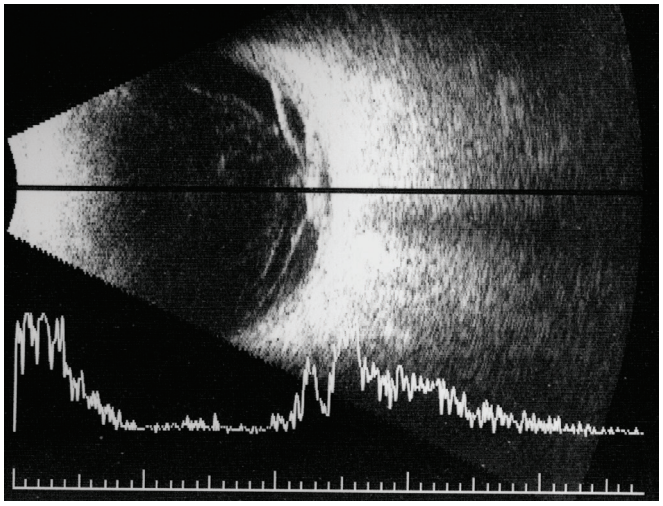

Figure 1: B-Scan ultrasonography shows incomplete posterior vitreous detachment and significant vitreopapillary traction. Optic nerve head is elevated approximately 2.5 mm and surrounded by localized peripapillary tractional retinal detachment.

CASE REPORT

A 45-year-old white female presented with an 8-week history of visual loss in the right eye. Her past medical history was remarkable for two stillbirths. Several members of her family had experienced cerebrovascular or cardiovascular thromboembolic events at early ages. Our subsequent laboratory analysis revealed APC resistance due to a point mutation (R506Q) in one of the factor V alleles.

At presentation, her visual acuity was light perception and she had a marked right afferent pupillary defect. Dense vitreous hemorrhage prevented the visualization of the fundus. However, a B-scan ultrasonography revealed optic nerve head elevation and peripapillary retinal detachment due to significant anteroposterior traction (Figure 1). The patient underwent a standard three-port pars plana vitrectomy.

During the surgery posterior hyaloid was mechanically stripped from the optic nerve head along with residual epiretinal membranes (Figure 2a, b). Optic nerve head vasculature perfused immediately after reliving the traction on the optic nerve head (Figure 2c). Surgery was coupled with peripapillary subretinal t-PA (25 μg in 100 μl) injection to lyse residual clots and facilitate the subretinal blood removal. She also received intravitreal triamcinolone acetonide (4 mg in 100 μl) to alleviate the macular edema.

Optic nerve head elevation and retinal detachment resolved within the first postoperative week. Subretinal and intraretinal hemorrhages disappeared rapidly and her vision improved to 20/400 at one month (Figure 2d). She was put on life-long anticoagulation prophylaxis by her hematologist. Two of her distant family members with recent thromboembolic events were also found to be positive for the same mutation and treated accordingly.

DISCUSSION

Vitreopapillary traction may occur during the evolution of the posterior vitreous detachment in the absence of diabetes mellitus or other forms retinovascular diseases. Optical coherence tomography (OCT) is a valuable tool for illustrating vitreous traction on the optic nerve. Hedges et al[7] reported two patiens with idiopathic vitreopapillary traction demostrated by OCT. The rare association Optic Disc Traction Syndrome with CRVO has recently been recognized and OCT findings in patients with optic disc traction associated with central retinal vein occlusion were described.[6] Similar to our case, these were severely ischemic CRVO cases characterized by development of optic nerve head elevation and localized peripapillary retinal detachments due to the anteroposterior traction of the posterior hyaloid and/or vitreopapillary fibrous membranes. Although it is not clear why some patients with ischemic CRVO develop Optic Disc Traction Syndrome, it may be due to the synergistic work of two factors:[1] Severe ischemia that may sequestrate inflammatory cytokines into the vitreous and result in cellular infiltration and contraction of the vitreous[8] and[2] Abnormal vitreoretinal attachment sites that may exert significant traction on the disc and retina.

Vitreopapillary traction further complicates the course of the CRVO by disrupting the axoplasmic flow and/or blood supply to optic nerve head. Early recognition of this condition before optic nerve head ischemia and atrophy develops is vital in maximizing the patients' potential for visual recovery. Surgical intervention with vitrectomy and vitreopapillary membrane dissection may yield visual recovery before irreversible optic nerve changes occur. Recent studies have focused on resolving venous occlusions with focal delivery of thrombolytics. Intravitreal tPA administration and direct injection of tPA into the retinal veins have been tested in patients with CRVO previously.[6,9,10] Direct injection of tPA into the retinal veins has been reported to result in modest improvement in vision.[9] In this case presentation, intervention with vitrectomy and vitreopapillary membrane dissection combined with intravitreal triamcinolone and subretinal tPA injection also showed a prompt anatomical and functional improvement in a patient with Optic Disc Traction Syndrome secondary to ischemic CRVO. However, it is not clear whether the lysis of epipapillary adhesions, hyaloid separation, intravitreal triamcinolone or tPA administration in an eye with increased permeability contributed most to the success of this procedure. In conclusion, in eyes with optic disc traction syndrome due to CRVO, early vitrectomy combined with focal delivery of thrombolytic agents such as t-PA should be considered to increase the resorption of retinal and subretinal hemorrhages. The combination of these treatment maneuvers seemed to accelerate the resolution of obstructive changes. Further studies are warranted to demonstrate the efficacy of this approach in the treatment of patients with Optic Disc Traction Syndrome due to ischemic CRVO.

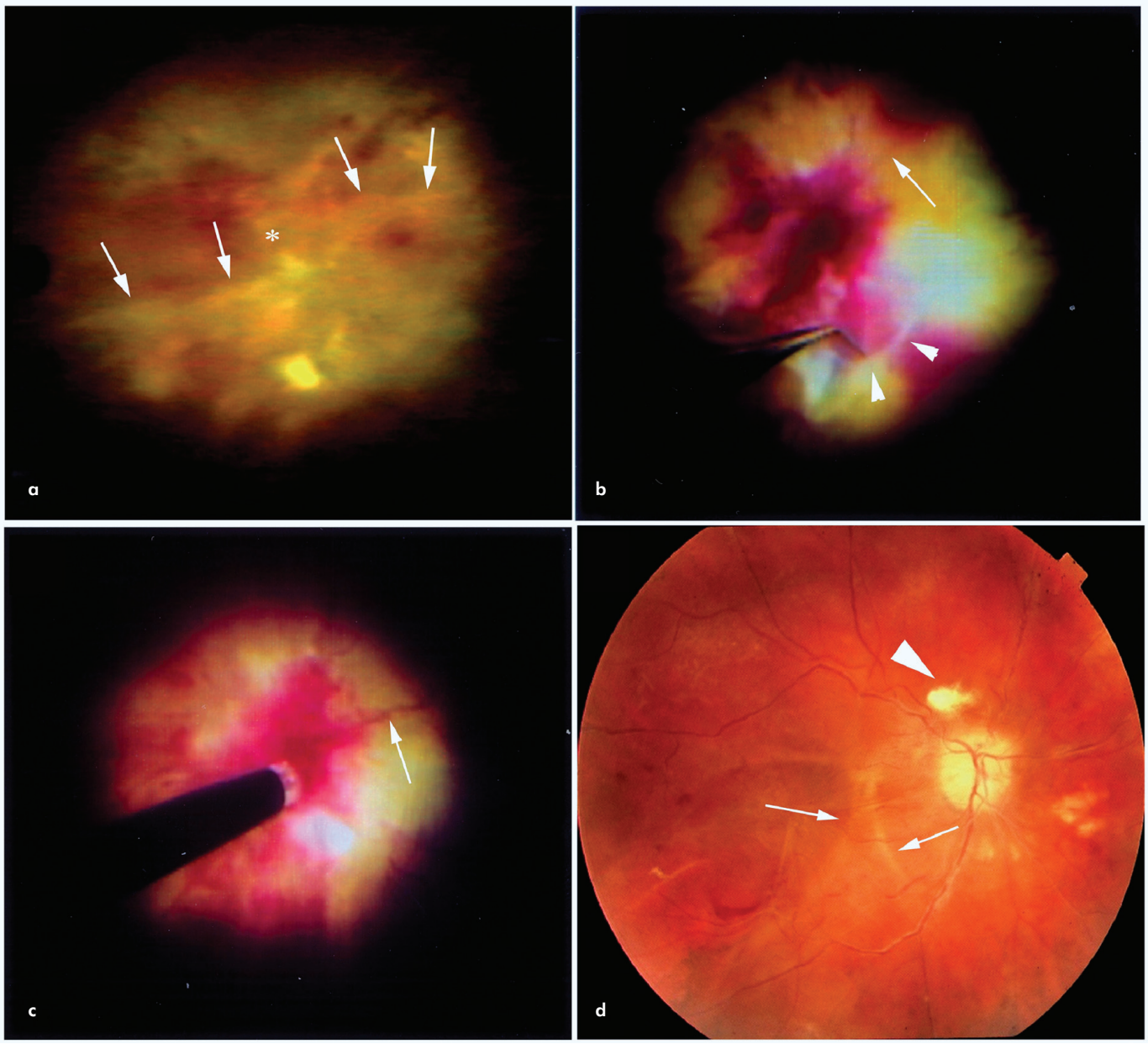

Figure 2: a) Color photograph shows thickened posterior hyaloid (arrows) that attached strongly on the disc (asterix) and peripapillary retina. **b)** Note the attenuated retinal vessels (arrow) at the optic nerve head prior to removal of the membrane (arrowheads). **c)** Removal of the posterior hyaloid relieved the traction and restored the blood flow in retinal vessels (arrow). **d)** Fundus photograph of the patient after surgery shows rapid disappearance of retinal hemorrhages. Mild subretinal fibrotic strands (arrows) persisted without interfering with the overlying retina.

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