Central retinal vein occlusion resulting from anomalous retinal vascular anatomy in a 24-year-old man

Shaheen C Kavoussi  
James E Kempton  
John J Huang

Yale University Department of Ophthalmology and Visual Science, New Haven, CT, USA

Abstract: An otherwise healthy 24-year-old male non-smoker with no prior medical or ocular history presented with 2 days of painless loss of vision in the left eye. Best-corrected visual acuity was 20/20 in the right eye and 20/80 in the left eye. Motility, pupils, intraocular pressure, and anterior segment exam were unremarkable in both eyes. The right fundus was normal. Left fundus exam revealed retinal hemorrhages in all four quadrants with venous dilation and tortuosity and a small vitreous hemorrhage (Figure 1). There was no macular edema or thickening on optical coherence tomography. Fluorescein angiography revealed delayed venous filling without leakage, vasculitis, or choroidal ischemia; suggestive of non-ischemic central retinal vein occlusion (CRVO, Figure 2). Blood pressure monitoring and body mass index were within normal limits. Compressive, autoimmune, and hematologic work-up including complete blood count, a full lipid profile, antinuclear antibody, anti-neutrophil cytoplasmic antibody, ACE, homocysteine, syphilis studies, PROC and PROS levels, anti-cardiolipin and anti-phospholipid antibodies, Factor V Leiden, ATIII, and magnetic resonance imaging (MRI) of the brain and orbits were negative.

Case

A 24-year-old male non-smoker with no prior medical or ocular history presented with 2 days of painless loss of vision in the left eye. Best-corrected visual acuity (BCVA) was 20/20 in the right eye and 20/80 in the left eye. Motility, pupils, intraocular pressure, and anterior segment exam were unremarkable in both eyes. The right fundus was normal. Left fundus exam revealed retinal hemorrhages in all four quadrants with venous dilation and tortuosity and a small vitreous hemorrhage (Figure 1). There was no macular edema or thickening on optical coherence tomography. Fluorescein angiography revealed delayed venous filling without leakage, vasculitis, or choroidal ischemia; suggestive of non-ischemic central retinal vein occlusion (CRVO, Figure 2). Blood pressure monitoring and body mass index were within normal limits. Compressive, autoimmune, and hematologic work-up including complete blood count, a full lipid profile, antinuclear antibody, anti-neutrophil cytoplasmic antibody, ACE, homocysteine, syphilis studies, PROC and PROS levels, anti-cardiolipin and anti-phospholipid antibodies, Factor V Leiden, ATIII, and magnetic resonance imaging (MRI) of the brain and orbits were negative.
Aspirin 81 mg was started as prophylaxis, and the case did not require intravitreal anti-vascular endothelial growth factor therapy. At 1 week and 1 month follow-up, BCVA was 20/40 in the left eye. By 2 months, BCVA returned to 20/20-2. Residual venous tortuosity was present. Upon closer review of the nerve anatomy, the arteries were found to lie directly over the veins with the vessels tightly intertwined in the left eye (Figure 3). More specifically, an undilated nasal retinal vein was firmly wrapped around the arterial bifurcation. Since the veins that emanated from directly underneath the site of the intertwined vessels were dilated and tortuous, this anatomical relationship was postulated to be the cause of his otherwise idiopathic CRVO. No such vascular intertwining was present in the unaffected right eye (Figure 4).

CRVO typically occurs in patients older than 50 years with risk factors including hypertension, diabetes, and open angle glaucoma. CRVO in a young patient merits work-up for underlying compressive, hematologic, or autoimmune etiology. Primary pulmonary hypertension has also been reported as the hemodynamic basis of CRVO in three patients in whom significant morbidity and mortality followed the vein occlusion. CRVO has been reported to occur in two young patients with anomalous retinal anatomy at the level of the optic nerve, with proposed mechanism for vein occlusion to include a combination of turbulent flow, increased intravascular volume, and pressure from the arteriolar side of the circulation causing a mechanical mass effect on the

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**Figure 1** Color fundus photograph of the left eye demonstrates dilated, tortuous retinal veins with intraretinal hemorrhages in all four quadrants.

**Figure 2** Fluorescein angiogram, left eye.

**Notes:**

(A) Arterial phase of fluorescein angiography of the left eye demonstrates normal arterial filling with blockage of choroidal fluorescence by retinal hemorrhages.

(B) Delayed venous filling and venous tortuosity and dilatation are evident in the arteriovenous phase.

(C) Normal choroidal perfusion persists through the venous phase without evidence of leakage or vasculitis.
The tight anatomical relationship is accentuated by an undilated nasal retinal vein firmly wrapped around the arterial bifurcation (arrowhead). The veins that emanate from underneath the artery are dilated and tortuous.

These changes, over time, are thought to damage the vessel wall of the central retinal vein and lead to thrombosis and occlusion. These cases highlight the importance of careful inspection of the retinal vascular anatomy to aid in assessing the risk of retinal vein occlusion in any age group. Thorough work-up and co-management with the patient’s primary care physician is recommended.

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References