

# Impending central retinal vein occlusion associated with cilioretinal artery obstruction

Juliana Mantovani Bottós  
Fabio Bom Aggio  
Eduardo Dib  
Michel Eid Farah

Vision Institute, Department of  
Ophthalmology, Federal University of  
São Paulo, Brazil

**Abstract:** We report a case of a patient with an impending central retinal vein occlusion (CRVO) with cilioretinal artery obstruction, which may help to understand the nature of the retinal hemodynamic changes associated with CRVO which shows direct evidence of arterial vasospasm, suggesting an increased contractility of retinal arteries. The clinical course, with initial retinal whitening along a cilioretinal artery followed by signs of venous stasis, seems to confirm the pathogenesis hypothesis concerning a primary arterial affection due to arterial vasospasm.

**Keywords:** retinal vein occlusion, cilioretinal artery obstruction, arterial spasm, fluorescein angiography

Cilioretinal artery obstruction (CLRAO) has been reported to occur either isolated or combined with central retinal vein obstruction (CRVO) or ischemic optic neuropathy (Brown et al 1983).

The pathomechanisms of CLRAO combined with CRVO are not well established. Two hypotheses have been proposed: (1) CLRAO occurs secondary to the raised capillary pressure caused by CRVO (McLeod and Ring 1976; Zylbermann et al 1981; Glacet-Bernard et al 1987; Schatz et al 1991; Brazitikos et al 1993; Keyser et al 1994) or (2) a primary reduction in perfusion pressure of the cilioretinal and retinal arteries (McLeod and Ring 1976; Glacet-Bernard et al 1987; Brazitikos et al 1993; Keyser et al 1994), leads to decreased retinal circulation and subsequent venous stasis and thrombosis (Hayreh et al 1971). Systemic blood pressure decrease (Hayreh et al 1983) and inflammatory or atherosclerotic retinal arterial disease (Keyser et al 1994) have been suggested as possible causes of reduced arterial perfusion pressure.

Herein, we report a case of a patient with impending CRVO with CLRAO. This case may help to understand the nature of the retinal hemodynamic changes due to CRVO.

## Case report

A 29-year-old man noticed a sudden onset of pericentral microscotomas in the visual field of his right eye after awakening that morning. His medical history was unremarkable. He had rhegmatogenous retinal detachment in the left eye 15 years before presentation, and was treated with a scleral buckling procedure. The best-corrected visual acuity was 20/40 in the right eye and 20/50 in the left. Slit-lamp biomicroscopy of the anterior segment was unremarkable in both eyes. Intraocular pressure was 12 mmHg in the right eye and 14 mmHg in the left eye. Funduscopy examination of the right eye revealed one small intraretinal hemorrhage in the papillomacular bundle, as well as diffuse increased tortuosity and venous dilation. Retinal whitening was seen in the papillomacular bundle, along the course of a cilioretinal artery. Fluorescein angiography (Figure 1) disclosed delayed filling of the central retinal vein

Correspondence: Juliana Mantovani  
Bottós  
Rua Mirassol 236 ap 22, Vila Clementino,  
São Paulo – SP 04044-010, Brazil,  
Tel +55 11 5908 0494  
Fax +55 11 8186 6804  
Email jubottos@gmail.com

and prolonged arteriovenous filling time, without capillary nonperfusion. Blood pressure was 120/70 mmHg. A systemic workup did not reveal any underlying disease. No specific treatment was performed. One week later, best-corrected visual acuity in the right eye was 20/20. Funduscopy of the right eye was normal. Three months after the first evaluation, visual acuity and funduscopy examination were unchanged. Fluorescein angiography revealed normal dye transit time as well as no abnormal fluorescence (Figure 2).

## Discussion

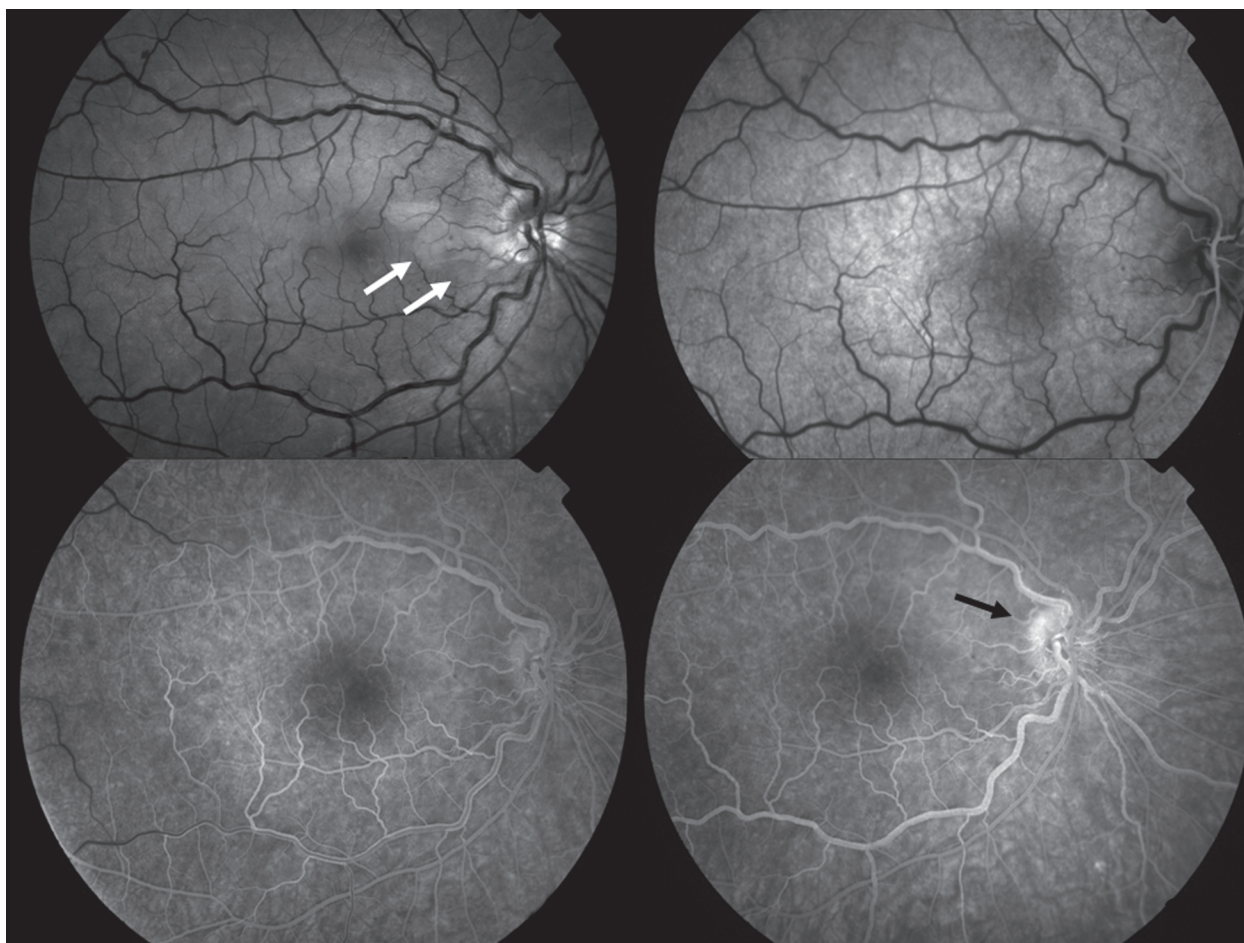
Early in the course of CRVO, arterial and venous flow alterations are present, irrespective of vein dilation or fundus hemorrhage. The mechanisms underlying these hemodynamic abnormalities remain speculative.

In 1904, Coats (1904) noted that angiosclerosis was associated with CRVO since common systemic vascular disorders such as hypertension, arteriosclerosis, and diabetes have been

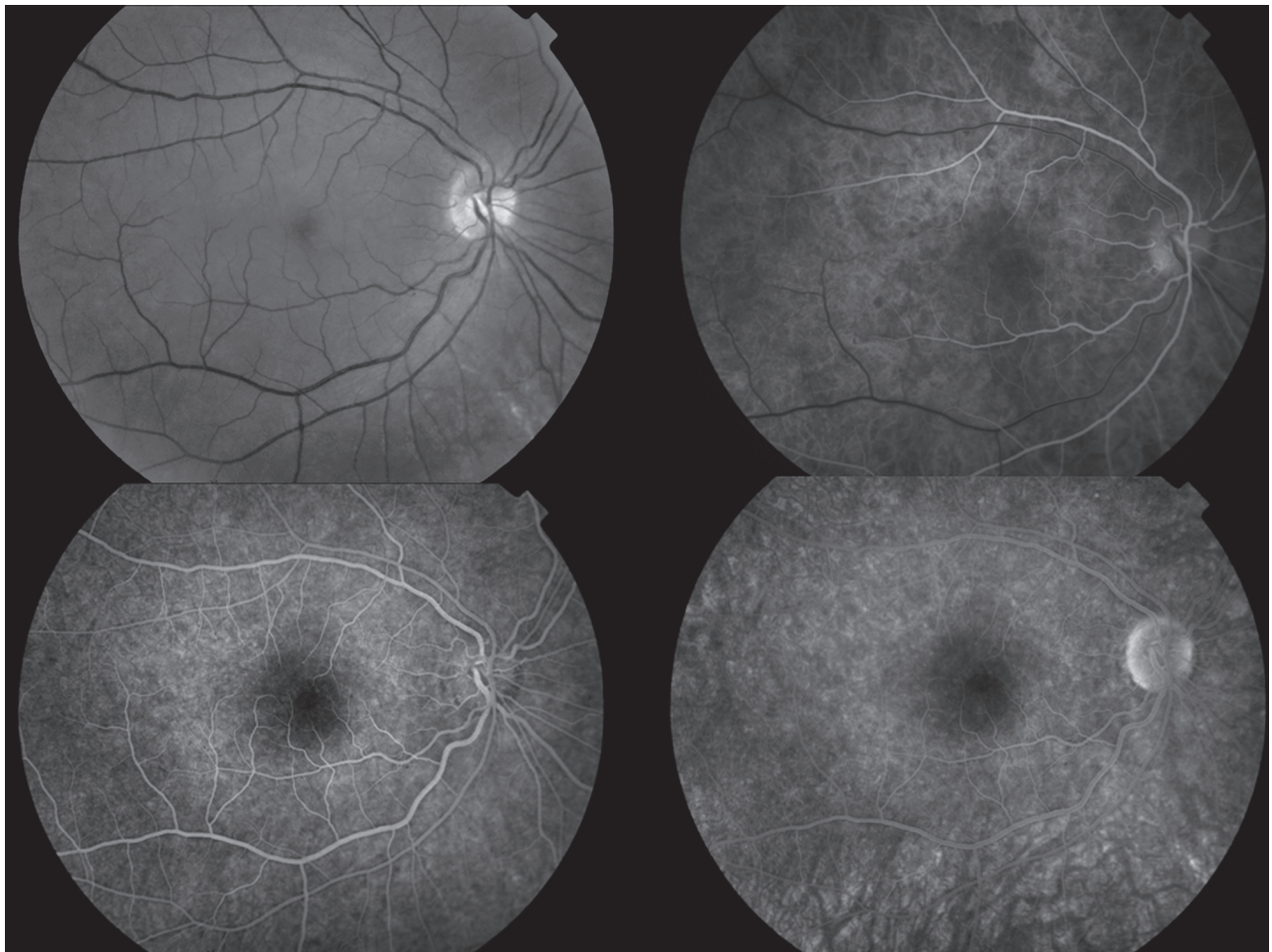
associated with the condition. The thickening of the arterial wall from sclerosis could reduce the caliber of the vein, possibly causing a disturbance in the pulsatile blood flow relation that exists between the central retinal artery and vein.

Hayreh and colleagues (1978) postulated that central retinal artery occlusion is an essential component in the pathogenesis of CRVO and that the ischemic form of CRVO is due to a combination of arterial and venous occlusion. Evidence for that was provided by experimental studies on monkeys. Occlusion of the central retinal vein and temporary occlusion of the central retinal artery at their emergence from the optic nerve produced a clinical appearance in the retina with some similarities to CRVO, which did not occur following occlusion of the central retinal vein alone at this site.

Arterial flow impairment, which manifests itself by pulsatile filling of retinal arteries, best evaluated by dynamic angiography, may be caused either by the retrograde transmission of elevated venous pressure through the capillaries



**Figure 1** Fluorescein angiography at the first visit. Red-free fundus photography (top left) showed retinal whitening along the course of a cilioretinal artery (arrows) as well as a small intraretinal hemorrhage in the papillomacular bundle. The retinal veins appeared slightly dilated. Fluorescein angiogram (top right and bottom) demonstrated delayed filling of the central retinal vein and prolonged arteriovenous filling time, without capillary nonperfusion. The late-phase fluorescein photograph (bottom right) showed leakage in the temporal aspect of the optic nerve (arrow).



**Figure 2** Fluorescein angiography at the third month follow-up visit: normal appearance of the retinal vessels and normal ocular transit time were noted.

(Hayreh 2005) or an increase in arteriolar resistance by vasospasm. The presence of retrograde systolic venous flow could suggest the presence of an intermittent compression of the central retinal vein (Paques et al 2001; Catier et al 2003).

Previous studies suggest that arterial vasospasm may have a role in the pathogenesis of CRVO. Catier and colleagues (2003) used dynamic videoangiography to show angiographic evidence of retinal arterial vasospasm at the initial phases of CRVO. This case also suggests that arterial vasospasm may occur at the early phase of CRVO, since occlusion of the cilioretinal artery occurred with only one intraretinal hemorrhage and mild increase in retinal vascular tortuosity. Besides, these findings improved promptly in the few days following initial evaluation, a presentation referred to in the literature as impending CRVO (Gass 1997) or venous stasis retinopathy (Hayreh 1976).

The case presented herein shows direct evidence of arterial vasospasm, suggesting an increased contractility

of retinal arteries. The clinical picture, with initial retinal whitening along the course of a cilioretinal artery followed by signs of venous stasis, seems to confirm the hypothesis of a primary arterial affection in the pathogenesis of CRVO. These features are in accordance with experimental studies that have reported the presence of arterial constriction within minutes of acute venous obstruction, which was attributed to a decrease in the local concentration of nitric oxide (Donati et al 1997). Besides, increased arterial contractility may contribute to acute retinal ischemia observed in specific presentations of CRVO, such as a cilioretinal artery obstruction associated with CRVO (Schatz et al 1991) or a combined central artery and vein occlusion.

It has been difficult to precisely determine the pathogenesis of CRVO, but there is evidence for a role of hyperviscosity, raised intraocular pressure, arteriosclerosis, and perhaps thrombosis of the vein (Williamson 1997). Although the chronic nature of the 'occlusive' process makes the condition amenable to therapeutic intervention, further innovation, or

reassessment of existing strategies is required. An increase in arteriolar resistance by vasospasm may have therapeutic implications, since vasospasm may be relieved by vasodilators (Jehn et al 2002).

Additional studies are necessary to elucidate the correct mechanisms of CRVO as well as the possible therapeutic targets.

## References

- Brazitikos PD, Pournaras CJ, Othenin-Girard P, et al. 1993. Pathogenetic mechanisms in combined cilioretinal artery and retinal vein occlusion: a reappraisal. *Int Ophthalmol*, 17:235–42.
- Brown GC, Moffat K, Cruess A, et al. 1983. Cilioretinal artery obstruction. *Retina*, 3:182–7.
- Catier A, Paques M, Gaudric A. 2003. Retinal vasospasm in a case of impending central retinal vein occlusion. *Retina*, 23:415–17.
- Coats G. 1904. Thrombosis of the central vein of the retina. *Roy London Ophthal Hosp Rep*, 16:62–122.
- Donati G, Pournaras CJ, Pizzolato GP, et al. 1997. Decreased nitric oxide production accounts for secondary arteriolar constriction after retinal branch vein occlusion. *Invest Ophthalmol Vis Sci*, 38:1450–7.
- Gass JDM. 1997. Stereoscopic atlas of macular diseases. In: *Diagnosis and Treatment*, 4th ed. St. Louis: CV Mosby, pp. 546–55.
- Glacet-Bernard A, Gaudric A, Touboul C, et al. 1987. Occlusion de la veine centrale de la rétine avec occlusion d'une artère cilio-rétinienne: A propos de 7 cas. *J Fr Ophthalmol*, 10:269–77.
- Hayreh SS. 1971. Pathogenesis of occlusion of the central retinal vessels. *Am J Ophthalmol*, 72:998–1011.
- Hayreh SS. 1976. So-called 'central retinal vein occlusion.' II. Venous stasis retinopathy. *Ophthalmologica*, 172:14–37.
- Hayreh SS, van Heuven WA, Hayreh MS. 1978. Experimental retinal vascular occlusion. I Pathogenesis of central retinal vein occlusion. *Arch Ophthalmol*, 96:311–23.
- Hayreh SS. 1983. Classification of central retinal vein occlusion. *Ophthalmol*, 90:458–74.
- Hayreh SS. 2005. Prevalent misconceptions about acute retinal vascular occlusive disorders. *Prog Retina Eye Res*, 24:493–519.
- Jehn A, Dettwiler BF, Fleischhauer J, et al. 2002. Exercise-induced vasospastic amaurosis fugax. *Arch Ophthalmol*, 120:220–22.
- Keyser BJ, Duker JS, Brown GC, et al. 1994. Combined central retinal vein occlusion and cilioretinal artery occlusion associated with prolonged retinal arterial filling. *Am J Ophthalmol*, 117:308–13.
- McLeod D, Ring CP. 1976. Cilio-retinal infarction after retinal vein occlusion. *Br J Ophthalmol*, 60:419–27.
- Paques M, Garmyn V, Catier A, et al. 2001. Analysis of retinal and choroidal circulation during central retinal vein occlusion using indocyanine green videoangiography. *Arch Ophthalmol*, 119:1781–7.
- Schatz H, Fong AC, McDonald R, et al. 1991. Cilioretinal artery occlusion in young adults with central retinal vein occlusion. *Ophthalmology*, 98:594–601.
- Williamson TH. 1997. Central retinal vein occlusion: what's the story? *Br J Ophthalmol*, 81:698–704.
- Zylbermann R, Rozenman Y, Ronen S. 1981. Functional occlusion of a cilioretinal artery. *Am Ophthalmol*, 13:1269–72.