

# Concurrent branch retinal artery occlusion in central retinal vein occlusion: 3 cases reports and literature review

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## Abstract:

Concurrent branch retinal artery occlusion (BRAO) and central retinal vein occlusion (CRVO) are the infrequently encountered clinical entities, with only a handful of cases elaborated in the literature. This case series describes three patients who presented with a dramatic fundus appearance that led to the diagnosis of unilateral BRAO and CRVO. Hypertension could be identified as a predisposing factor in only one patient. All three patients had relatively good visual acuity at presentation. The role of spectral-domain optical coherence tomography in confirming the simultaneous presence of BRAO is also illustrated.

## Keywords:

Branch retinal artery occlusion, central retinal vein occlusion, spectral-domain optical coherence tomography

## INTRODUCTION

Central retinal vein occlusion (CRVO) may be accompanied by a central retinal artery occlusion (CRAO) or a cilioretinal artery occlusion, both of which are well-described entities. However, simultaneous occurrence of a branch retinal artery occlusion (BRAO) in CRVO is extremely rare, despite the fact that both these conditions have common underlying associations such as hypertension, coagulopathies, and rheumatologic diseases.<sup>[1]</sup> Only a handful of cases have been reported in the last three decades in spite of CRVO being one of the most commonly encountered retinal vascular disorders.<sup>[1-8]</sup> This case series depicts three cases of concurrent BRAO in CRVO.

## CASE REPORTS

### Case 1

A 52-year-old female presented with decreased vision in her right eye for the past 2 weeks. Best-corrected visual acuity (BCVA) was 20/60 in the right eye. Anterior-segment examination

was unremarkable. Fundus examination showed venous dilatation with retinal hemorrhages. In addition, an area of retinal whitening was noted superotemporally [Figure 1a]. No cherry red spot or intra-arterial emboli were seen. The left eye had a BCVA of 20/20 with a normal fundus.

Fundus fluorescein angiography (FFA) revealed delayed filling of the superior branch of the central retinal artery (CRA) with a global delay in the arteriovenous transit time [Figure 1b]. Late staining of the superotemporal vein with adjacent capillary nonperfusion was noted [Figure 1c]. Spectral-domain optical coherence tomography (SD-OCT) demonstrated increased inner retinal hyperreflectivity in the superior part of the macula with minimal macular edema [Figure 1d].

After 1-month follow-up, BCVA of the right eye improved to 20/40, and fundus showed resolved retinal hemorrhages and a sclerosed superior branch of the CRA [Figure 1e]. SD-OCT depicted macular thinning, especially superiorly [Figure 1f].

### Case 2

A 48-year-old female presented with decreased vision in her right eye for the past 10 days. BCVA

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was 20/40 in this eye. The right fundus showed a hyperemic disc, prominent peripapillary hemorrhages, dilated and tortuous retinal veins, and few scattered retinal hemorrhages. There was intense superficial retinal whitening along the superotemporal retinal artery [Figure 2a]. FFA demonstrated delayed venous filling pointing toward a CRVO [Figure 2b]. Further, staining at the optic disc and of the branches of the superotemporal retinal artery was observed [Figure 2c]. There was no capillary nonperfusion. The demonstration of inner retinal hyperreflectivity in the superior macula on SD-OCT confirmed a superior BRAO [Figure 2d]. There was no macular edema.

### Case 3

A 45-year-old female with decreased vision in her right eye for the past 1 week was found to have BCVA 20/60 in this eye. Fundus examination showed a hyperemic disc, peripapillary and scattered intraretinal hemorrhages, venous dilatation, and cotton wool spots inferiorly. A clearly demarcated area of retinal whitening was present in the distribution of the superotemporal branch of the CRA [Figure 3a]. FFA showed delayed venous filling [Figure 3b] and peripheral retinal capillary nonperfusion. In addition, delayed filling of the superotemporal retinal artery was picked up with nonperfusion in its distribution [Figure 3c]. SD-OCT revealed superior inner retinal hyperreflectivity in the absence of macular edema [Figure 3d].

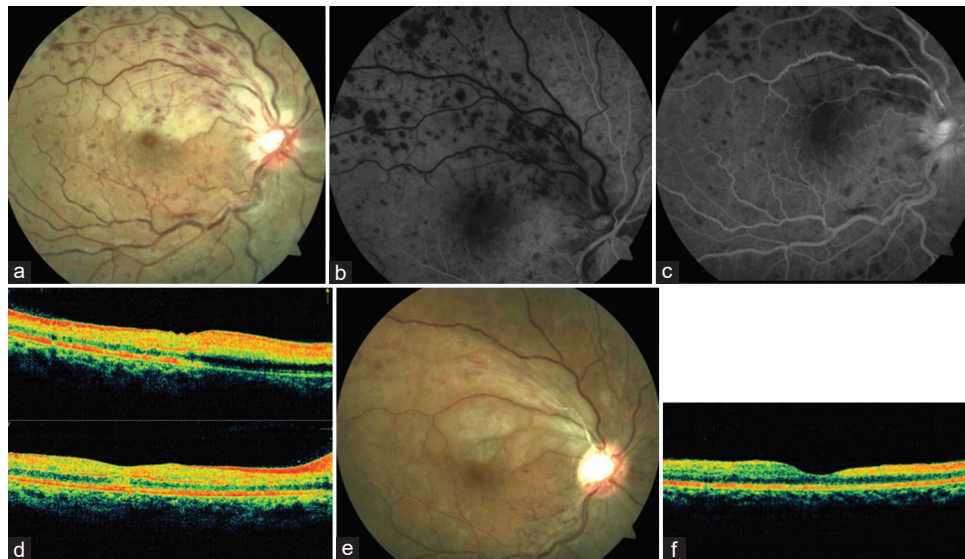
All three patients underwent extensive cardiovascular, hematological, and medical assessment to search for an underlying cause. There was no medical history of hypertension, diabetes, or autoimmune disease. They were nonsmokers and not on oral contraceptive pills. Complete blood counts, erythrocyte sedimentation rate, C-reactive protein,

prothrombin time, partial thromboplastin time, fibrinogen, complement factors C3 and C4, antithrombin III, protein C activity, protein S antigen, activated protein C resistance, factor V Leiden mutation, serum protein electrophoresis, lipid profile, and plasma and urine homocysteine quantitative tests were within normal limits. Antiphospholipid antibodies and c-ANCA were negative. Chest X-ray, carotid Doppler ultrasound, electrocardiography, and echocardiography were normal.

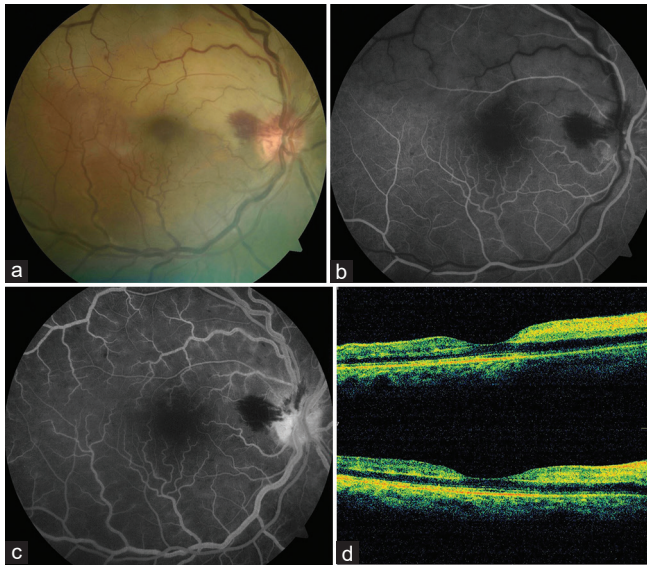
## DISCUSSION

The above three cases displayed a similar yet unique fundus appearance of retinal whitening in the territory of the affected artery along with a dilated and tortuous venous system. In Case 1, there was a predilection of the retinal hemorrhages for the quadrant that contained the artery obstruction [Figure 1a]. While the precise mechanism of these combined obstructions cannot be elucidated, it has been hypothesized that either the arterial blockage could be the initiating event followed by a low-flow state in the central retinal vein or the venous obstruction could be the inciting event followed by external compression of an adjacent branch retinal artery by the ensuing optic disc edema.<sup>[1]</sup> In fact, a primary optic nerve inflammation resulting in secondary impairment of the retinal vasculature has also been suggested, especially in young patients with concomitant CRVO and BRAO, with no underlying etiologic factor.<sup>[1]</sup>

Not more than 16 cases of simultaneous BRAO and CRVO have been reported in the literature.<sup>[1-8]</sup> In addition to hypertension,<sup>[1]</sup> hyperhomocysteinemia,<sup>[2,3]</sup> inherited plasminogen deficiency and high lipoprotein(a) levels,<sup>[4]</sup> and pegylated interferon plus ribavirin therapy for chronic hepatitis C<sup>[6]</sup> have been found as



**Figure 1:** Case 1. (a) Fundus photograph showing dilated, tortuous veins and diffuse retinal hemorrhages, more numerous in the area of retinal whitening superotemporally. (b) Fluorescein angiography showed delayed filling of the superotemporally retinal artery. (c) Late staining of the superotemporally vein was noted with adjacent hypoperfusion. (d) Vertical and horizontal spectral-domain optical coherence tomography scans through the macula revealed inner retinal hyperreflectivity superiorly. (e) A month later, retinal hemorrhages resolved and the superotemporal artery was seen as a sclerosed vessel. (f) A vertical spectral-domain optical coherence tomography scan showing superior macular thinning



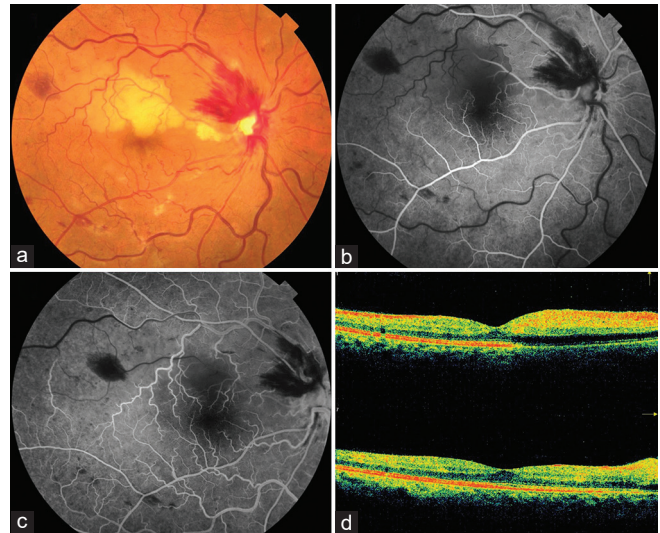
**Figure 2:** Case 2. (a) Fundus photograph showing dilated, tortuous veins with retinal whitening superotemporally. The disc was hyperemic with prominent peripapillary hemorrhages. (b) Fluorescein angiography showing delayed filling of the superotemporal retinal artery and delayed venous filling. (c) Late staining of the capillaries was noted superotemporally suggestive of ischemia. (d) Vertical and horizontal spectral-domain optical coherence tomography scans through the macula demonstrated inner retinal hyperreflectivity in the superior macula confirming a superior BRAO

predisposing factors. A good visual outcome has been seen in most cases, except if neovascularization complicates the clinical course.<sup>[1]</sup> While no systemic abnormalities could be found in our cases, presenting visual acuity was good in all. The nonischemic nature of the CRVO could be an important factor in these patients yielding a good prognosis.<sup>[2]</sup>

CRVO, especially nonischemic, can be associated with areas of similar macular whitening in certain cases. These areas of retinal whitening show hyperreflective bands straddling the inner nuclear layer on SD-OCT and have been described as paracentral acute middle maculopathy (PAMM) recently.<sup>[9]</sup> SD-OCT features of retinal artery occlusion, on the other hand, include hyperreflectivity and increased thickness of the inner retinal layers with decreased reflectivity of the outer retinal layers, owing to an optical shadowing effect.<sup>[10]</sup> SD-OCT is thus useful to distinguish concomitant PAMM and arterial occlusions in the setting of nonischemic CRVO.

While a detailed systemic workup is warranted in any case of retinal vascular occlusion, the battery of tests varies between arterial and venous occlusions. In the setting of arterial occlusions, it is mandatory to rule out thromboembolic phenomena and systemic source of emboli. The systemic prognosis also varies between arterial and venous occlusion. It is therefore important to identify arterial occlusions in the setting of CRVO and investigate accordingly.

To conclude, BRAO occurring in the presence of CRVO is a rare entity. The visual outcome in these cases is relatively good



**Figure 3:** Case 3. (a) Fundus photograph showing dilated, tortuous veins with retinal hemorrhages superior to the optic disc. A well-demarcated area of pale retina was present superotemporally. The inferior macula showed cotton wool spots and a white centered hemorrhage. (b) Fluorescein angiography showing absence of filling of the superotemporal retinal artery 2 disc diameters temporal to the disc margin with adjacent nonperfusion. (c) Delayed venous filling was present. (d) Vertical and horizontal spectral-domain optical coherence tomography scans through the macula demonstrated inner retinal hyperreflectivity in the superior macula

and may not require prompt intervention. SD-OCT can help identify affected pathological areas and establish a complete diagnosis. A close follow-up is warranted to check for the development of macular edema.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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