

Case Report

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# Clinical Features and Treatment Outcome of a Concurrent Central Retinal Vein Occlusion and Cilioretinal Artery Occlusion

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## Keywords

Central retinal vein occlusion · Cilioretinal artery occlusion

## Abstract

We describe the clinical features and treatment outcome of a patient with combined central retinal vein occlusion and cilioretinal artery occlusion. A 52-year-old female presented to our clinic with decreased vision in the right eye for 4 days. Visual acuity and intraocular pressure were count fingers at 2&1/2M and 14 mm Hg in the right and 20/20 and 16 mm Hg in the left eye, respectively. Funduscopic exam and optical coherence tomography (OCT) of the right eye confirmed the diagnosis of concurrent cilioretinal artery occlusion and central retinal vein occlusion with segmental macular pallor in the territory of the cilioretinal artery, corresponding marked inner retina thickening on OCT and signs of vein occlusion. The patient received an intravitreal injection of bevacizumab and at 1-month follow-up, vision improved to 20/30 with corresponding anatomical improvement. It is very important to recognize combined central retinal vein occlusion and cilioretinal artery occlusion as they could be treated with intravitreal injections of anti-vascular endothelial growth factors with favorable treatment outcomes.

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## Introduction

Central retinal vein occlusion (CRVO) associated with cilioretinal artery occlusion (CLRAO) is a variant of retinal vascular disease, which was first reported by Oosterhuis in 1968 [1]. Since then, there have been many case reports and case series describing these concurrent retinal vascular events [2–8]. While the central retinal artery arises directly from the ophthalmic artery and has efficient enough autoregulation to maintain retinal circulation in the advent of fall in perfusion pressure, the cilioretinal artery (CLRA) stems from the posterior ciliary artery (PCA) or the choroidal vascular system with no autoregulation. The CLRA may supply part of the macula, one quadrant, half of the retina, or rarely the entire retina [3]. In a recent systematic review and a prospective cross-sectional study, the prevalence of CLRAs ranged from 6.9% to 49.5% depending on the identification method, with fluorescein angiography having the highest values followed by fundus photography and ophthalmoscopy [9].

CLRAO comprises 5% all retinal arterial occlusions [10]. Three different types of CLRAO have been described: an isolated form and two combined forms which present in association with either CRVO or with anterior ischemic optic neuropathy [4]. The purpose of reporting our case is to describe the clinical features of a concurrent CRVO and CLRAO and treatment outcome of these dual vascular conditions with intravitreal bevacizumab (Avastin).

## Case Report/Case Presentation

A 52-year-old female was referred to our clinic with the diagnosis of CRVO after she noticed decreased vision in the right eye for 4 days duration. She had a complete ophthalmology examination including best-corrected visual acuity at 3M, intraocular pressure with applanation tonometry, slit-lamp biomicroscopy, fundus photography, optical coherence tomography (OCT), and OCT-angiography (OCT-A) (Topcon 3D, OCT-1 Maestro II, Tokyo, Japan), in Roha Specialized Eye Clinic in June 2021.

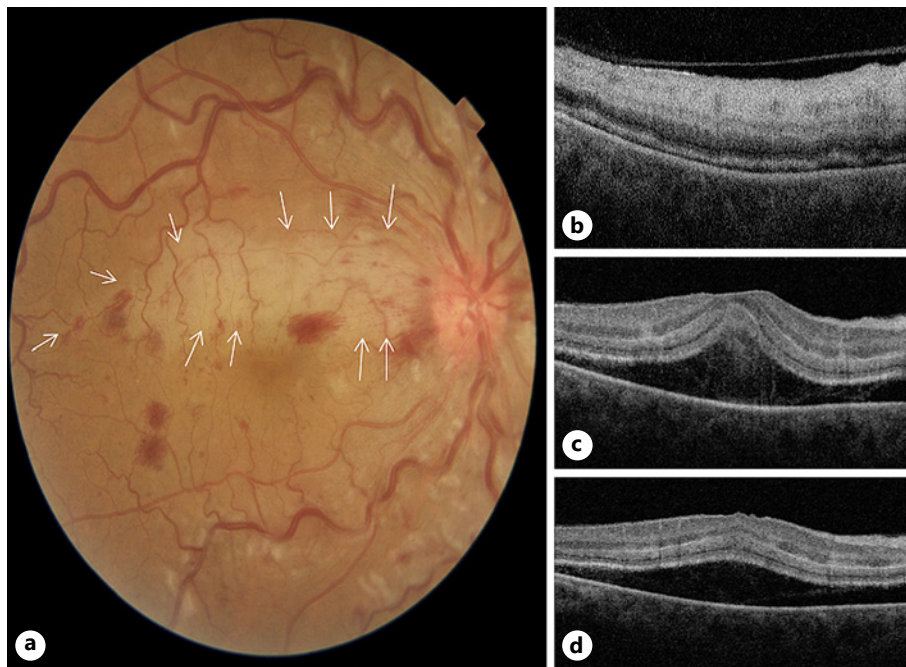
Visual acuity and intraocular pressure were CF 2&1/2M and 14 mm Hg in the right, and 20/20 and 16 mm Hg in the left eye, respectively. Pupils were regular and reactive and there was no relative afferent pupillary defect. Anterior segment slit-lamp examination was normal.

Dilated fundus exam of right eye showed optic disc edema, intraretinal dot blot and flame hemorrhages, dilated and tortuous vessels, and cotton wool spots. In addition, a horizontal stripe of whitish opacified edema was present in the superior macula.

OCT showed segmental densely hyperreflective thickening corresponding to the area of whitish edema and subretinal fluid through the fovea (shown in Fig. 1), OCT-A taken 3 mm × 3 mm centered on fovea revealed paucity of the parafoveal and part of the perifoveal vascular networks and decreased flow signs on all layers of the macula superior to the fovea (shown in Fig. 2). Funduscopy, OCT, and OCT-A of the left eye were normal.

We consulted an internist for systemic disease evaluation. The cardiovascular assessment including carotid Doppler studies was reportedly normal. She was advised to have lifestyle modification as blood pressure was 150/85 mm Hg.

With the working diagnosis of a combined CRVO and CLRAO, the patient received intravitreal injection of bevacizumab 1.25 mg/0.05 mL in the right eye. At 1-month follow-up, visual acuity had improved to 20/30. Follow-up fundus photography showed resolution of the horizontal stripe of opacified edema from the CLRAO and improvement in the CRVO with resolving retinal hemorrhages, disc edema, and cotton wool spots (shown in Fig. 3). OCT showed resolution of subfoveal subretinal fluid, and OCT-A showed improvement of the blood flow and vascular network in the superficial and deeper retinal layers (shown in Fig. 3, 4).

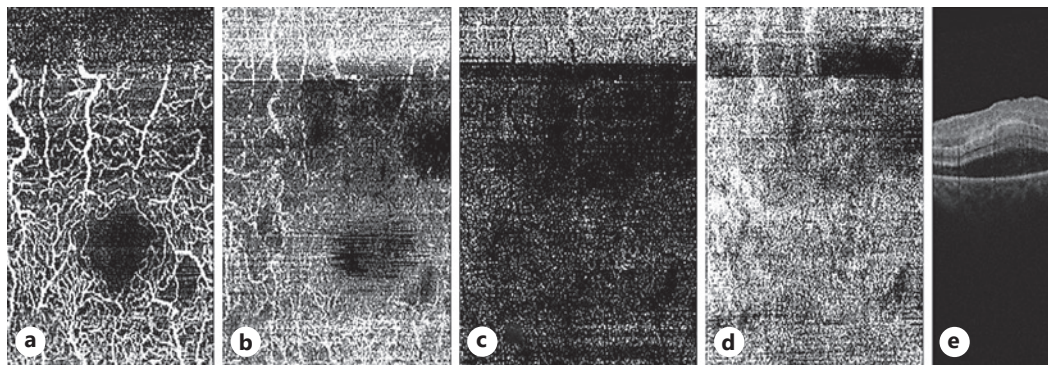


**Fig. 1.** Color fundus photograph (a) showing a segmental pallor of superior macula extending from the superior temporal disc margin to the temporal retina (white arrows) associated with dilated and tortuous vessels, intraretinal hemorrhage, cotton wool spots, and disc edema suggestive of combined CRVO and CLRAO. SD-OCT taken through the opacified retina displays a prominent inner retinal thickening with some subretinal fluid (b); OCT through fovea showed both inner retinal thickening and large subretinal fluid (c, d).

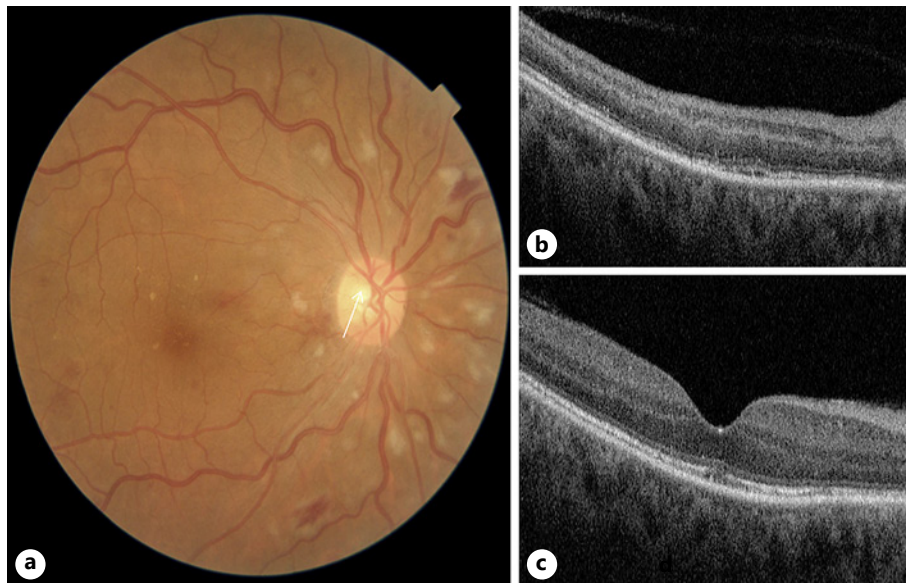
At 8-month follow-up, OCT-A showed a smooth foveal avascular zone and good blood flow in both the superficial and deep capillary plexuses (shown in Fig. 5).

## Discussion

Generally, retinal vein occlusion is the second most common retinal vascular disease, following diabetic retinopathy [11]. Despite the presence of a CLRA in more than 30% of all eyes [9], concurrent CRVO and CLRAO are not as common as CRVO or CLRAO alone, although Schatz et al. [12] in their study on CLRA occlusion with CRVO in young adults postulated that this clinical condition could be more common than appreciated. From our literature review, the hypotheses outlined below may explain how some eyes with CLRAs do not develop CLRAO with CRVO. The slow development of CRVO allows time for collaterals to develop in the optic nerve so that the intraluminal pressure in the retinal capillaries never gets high enough to cause a hemodynamic block in the CLRA [3]. The CLRA may be a branch of one of the short PCAs, which may arise from the trunk of the ophthalmic artery and therefore could have blood flow autoregulation as the central retinal artery does. The PCA is the main source of blood supply to the optic nerve head and the choroid up to the equator, the outer retina, and, when a CLRA is present, the entire thickness of the retina in that region [13]. Identifying CLRAO in the severe form of CRVO with significant retinal hemorrhages and retinal infarction is challenging [12]. A small CLRAO on the peripapillary region may be overlooked and could even be mistaken for a cotton wool spot. Furthermore, the timing of the examination is also crucial. The hemodynamic dysfunction is transient, lasting from a few hours to several days,



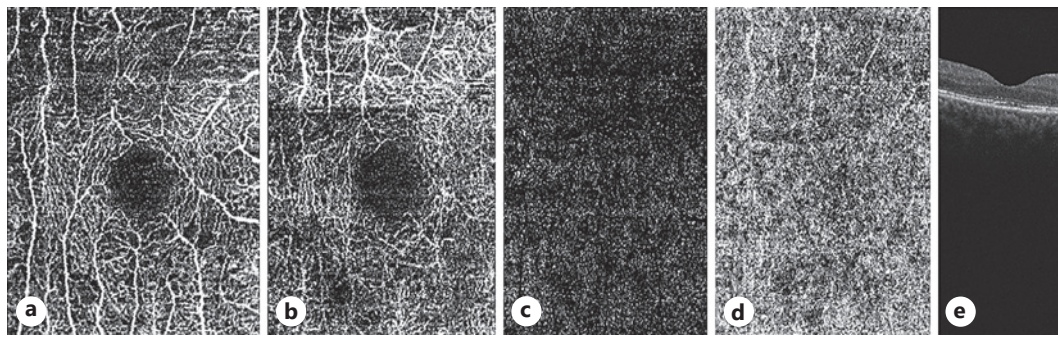
**Fig. 2.** OCT-A depicting paucity of the parafovea, part of perifoveal vascular networks, and reduced flow signs on superficial (a) and deep capillary plexuses superior to fovea (b). Indistinct vascular images in the outer retina (c) and choriocapillaris (d), and motion artifact likely due to thickened inner retina shown in the corresponding B-scan (e) and poor fixation from profound loss of vision.



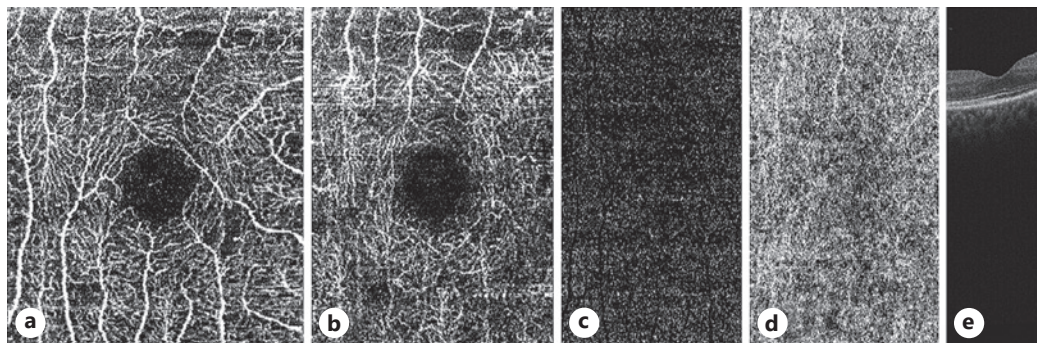
**Fig. 3.** Color fundus photography (a) 1 month after treatment with Avastin: resolution of intraretinal hemorrhage, macular opacity, vascular dilation, and tortuosity. Note the hook-shaped arteriole emerging from superior temporal optic disc most likely representing CLRA (upward arrow). OCT B-scan through the opacified retina shows segmental atrophy of superior perifovea (b). B-scan through the center of macula shows preserved fovea (c).

depending upon how severe the retinal venous stasis is and how rapidly the collateral circulation is established. Therefore, medical attention many days after the onset of symptoms may not help identify CLRAO as the vessel regains some function.

Several hypotheses have been put forward to explain the pathogenesis of CLRAO in conjunction with CRVO. A hemodynamic block is the most popular hypothesis proposed by Hayreh et al. [3]. Sudden occlusion of the central retinal vein results in a marked rise of intraluminal pressure in the entire retinal capillary bed; when that intraluminal pressure rises above that in the CLRA, the result is a hemodynamic block in the CLRA. The perfusion pressure in the choroidal vascular bed is normally lower than that in the central retinal artery



**Fig. 4.** OCT-A 1 month after treatment with Avastin: improvement of the blood flow and vascular network in the superficial (a) and deeper retinal layers (b). The avascular retina showed better image quality (c). Normal flow signs on choriocapillaris (d). Projection artifact on choriocapillaris could be related to segmental atrophy. The corresponding B-scan (e) shows normal foveal contour.



**Fig. 5.** OCT-A 8 months after treatment with Avastin: showing a nearly round foveal avascular zone and good blood flow in both the superficial and deep capillary plexuses (a, b). Corresponding B-scan depicts normal foveal contour (e). The avascular retinal layer is better outlined (c). The horizontally arranged hyporeflective areas of the superior perifoveal area noted on the superficial and deep OCT-A (a, b) images, and the projection artifact on deep capillary plexus (b) and choriocapillaris could be related to segmental atrophy (d).

by approximately 7 mm Hg [3, 14]. During the early stages of combined CRVO and CLRAO, the CLRA fills for a variable distance from the optic disc during systole, but the filling retracts to the optic disc during diastole, extending back and forth from the optic disc to the retina [2]. This hemodynamic theory was further supported by fluorescein angiography that revealed a pulsating filling in the CLRA [5–8]. McLeod has also proposed branch flow exclusion and choroidal steal as factors in combined CLRAO and CRVO [15]. These hypotheses imply that following CRVO, the blood flow from the cilioretinal branch of PCA initially decreases and then may completely stop due to lack of an arteriovenous perfusion gradient across the CLRA (branch flow exclusion), and the flow may be diverted to a system with lesser resistance (choroidal arterial steal) [15].

In our patient, OCT scan of the macula through the distribution of an occluded CLRA demonstrated a marked segmental thickening of the inner retina extending from the superior temporal optic disc margin to the temporal limit of the macula (shown in Fig. 1). This OCT finding, if captured early, is an essential anatomical biomarker to differentiate combined CLRAO with CRVO from an isolated CRVO. As we were not able to perform FA in our clinic, OCT-A was performed to assess blood flow. An initial examination at 4 days after onset of

symptoms showed paucity of parafoveal and part of the perifoveal vascular networks and decrease in flow signs in all layers of the retina superior to the fovea (shown in Fig. 2).

Retinal vein occlusion leads to an increased expression of vascular endothelial growth factors (VEGFs) in the retina and retinal pigment epithelium, causing vascular hyperpermeability with subsequent breakdown of the blood-retina barrier and eventual neovascularization in some cases [14, 16]. It has been shown that anti-VEGF medications decrease the hyperpermeability status of the retinal veins, which can decrease the edema surrounding the occluded vein and thus improve the circulation of the corresponding nearby artery.

Anatomical and functional improvement was observed in our case 1 month after a single intravitreal injection of bevacizumab (Avastin). After treatment with Avastin, there might have been a significant reduction of the retrograde transmission of intravascular pressure to the CLRA. Consequently, the functional block at the CLRA could have been relieved, leading to improved circulation. Only four other cases of combined CRVO and CLRAO treated with anti-VEGF have been reported. Two cases of combined CRVO and CLRAO had anatomical improvement after multiple monthly intravitreal injections of Avastin [17]. Another 2 cases with combined CRVO and CLRAO had functional and anatomical improvement after a single injection of Lucentis [18].

In our current case, there was dramatic improvement in vision from CF to 20/30 after a single intravitreal injection of anti-VEGF. Anatomical improvement with resolution of macular edema by OCT and improved vascular flow signs by OCT-A were observed both in the superficial and deep retinal layers (shown in Fig. 4) 1 month after treatment. The area of retinal opacity and thickening on initial examination subsequently progressed to a segmental stripe of atrophy in the inner retina in the territory of CLRA (shown in Fig. 1, 3) as typical for retinal artery occlusions in which the duration of ischemia exceeds limit of retinal tolerance.

## Conclusion

The segmental thickening of the inner retina seen by OCT may help differentiate combined CRVO and CLRAO from isolated CRVO. Treatment of combined CRVO and CLRAO with anti-VEGF appears to be a viable option.

## Statement of Ethics

This study protocol was reviewed and approved by the Ethics Review Committee of Roha Specialized eye clinic, approval number Roha/IRB/07/2021. Written Informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review upon request.

## Conflict of Interest Statement

The authors have no conflict of interest to declare.

## Funding Sources

The authors have no funding source to declare.

## Author Contributions

Alemu Kerie Tesfaw was involved in drafting of the manuscript, review, and editing. Nikhil N. Batra and Cong T. Phan were involved in the review and editing.

## Data Availability Statement

All data analyzed during this case report are included in the manuscript. Further inquiries can be directed to the corresponding author.

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