THE **RETINA** PARTNERS

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Case of the Month – June 2019

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A 73 year-old female, with a history of hypertension, and high cholesterol, was referred for sudden painless vision loss in the left eye occurring about 2 weeks prior to presentation. At the onset of her symptoms, she presented to an emergency department where vision was noted to be decreased in the left eye to 20/200. Her ocular examination was otherwise reported as normal. MRI/MRA of the brain, EKG, ESR/CRP and basic blood work were all within normal limits. No cause was found. She was discharged home and instructed to follow up with an outpatient ophthalmologist. She presented to our clinic 2 weeks later. Vision was 20/25 in the right eye, and 20/25 with eccentric fixation in the left eye. There was no afferent pupillary defect. Intraocular pressures and slit lamp examination of the anterior segments were unremarkable. Fundoscopic examination of the right eye was normal. Imaging of the left eye is shown below.

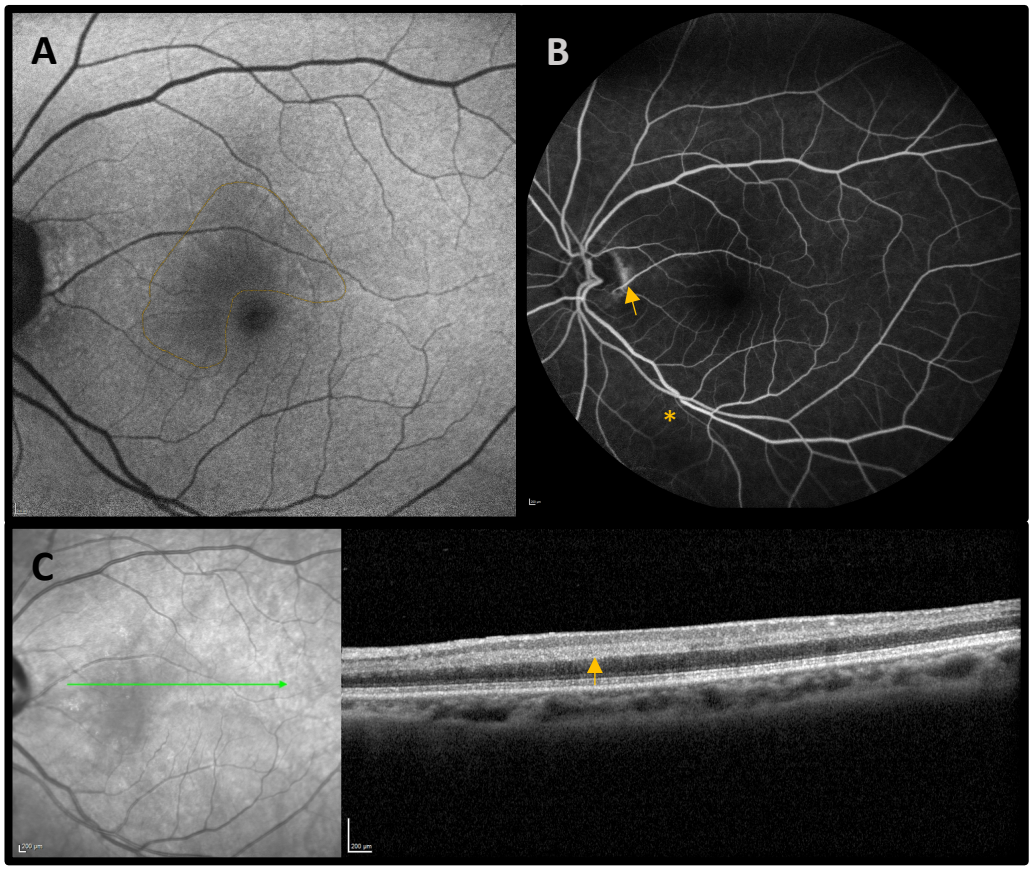
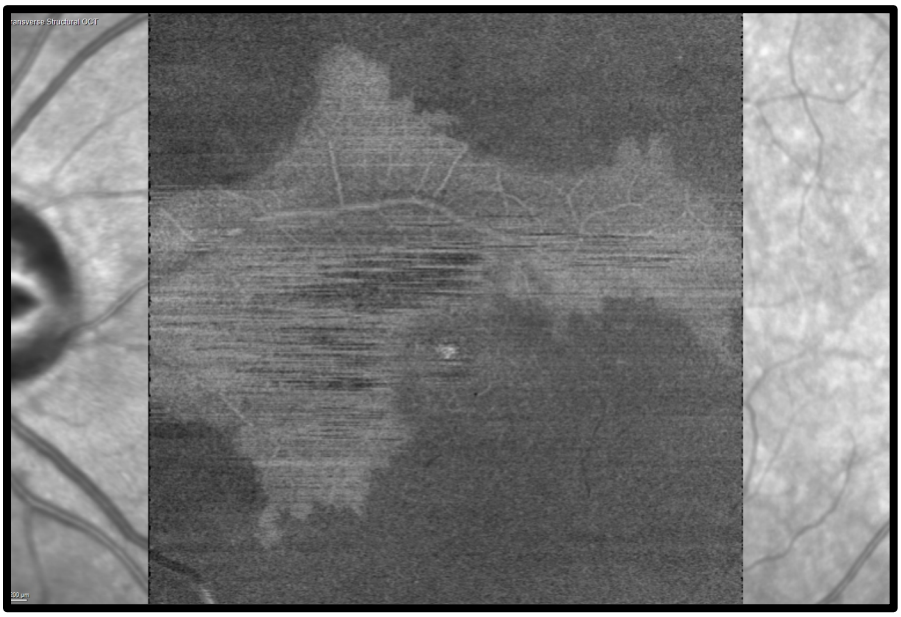


Figure 1: A. Autofluorescence imaging of the left eye shows a hypo-autofluorescent lesion in the nasal macula (outlined in yellow). **B.** Fluorescein angiography shows the presence of a cilioretinal artery (arrow). There is mild AV nicking (asterisk), but no perfusion defects or other abnormalities. **C.** SD-OCT of the left eye shows middle retinal hyper-reflectivity within the region outlined in figure 1A (arrow). This is consistent with paracentral acute middle maculopathy.

Differential Diagnosis: Central retinal artery occlusion, acute macular neuroretinopathy, impending retinal vein occlusion, branch retinal artery occlusion, isolated cilioretinal artery occlusion.

Clinical Course:

Additional imaging with structural *en face* OCT segmented at the level of the deep capillary plexus colocalized the region of ischemia to the distribution supplied by the cilioretinal artery (Figure 2). The patient was diagnosed with an isolated cilioretinal artery occlusion.



F**igure 2:** *En face* OCT segmented at the level of the deep capillary plexus precisely localizes the ischemic injury to the distribution of the cilioretinal artery.

Discussion:

Cilioretinal artery occlusion (CLRAO) is an uncommon variant of retinal arterial occlusion. It may occur in isolation, as in this case, in conjunction with central retinal vein occlusion or in conjunction with anterior ischemic optic neuropathies. This entity is important to recognize as its pathophysiology differs from other retinal artery occlusions.

The retina is supplied by both the retinal vascular system, arising from the central retinal artery, and the choroid. Unlike the retinal vascular system, the choroidal vasculature lacks the ability to autoregulate (constrict and dilate) in response to changing blood flow and oxygen levels. The cilioretinal artery is a branch of the choroidal circulation, arising from either the short posterior ciliary artery or directly from the peripapillary choroid. It is present in about 32% of eyes, and in these eyes, it supplies blood to portions of the nasal macula and fovea. As part of the choroidal vasculature, the cilioretinal artery also lacks the ability to autoregulate. Therefore, if perfusion pressures drop, or if venous outflow resistance increases (as in the case of central retinal vein occlusion), the cilioretinal artery may not perfuse adequately. This results in what has been termed "cilioretinal artery occlusion". Notably, this differs from the more common branch or central retinal artery occlusion which are most often caused by emboli. For this reason, some authors have advocated for a change in terminology to "cilioretinal artery hypo-perfusion" to more accurately reflect its pathophysiology.

Because CLRAO most often occurs as a result of decreased perfusion rather than complete obstruction, blood flow is significantly decreased, but does not cease completely. The superficial retina is often able to extract enough oxygen, even in this hypo-perfused state, to avoid infarction. However, the deeper layers of the retina are not. This variant of "misery perfusion" results in isolated infarction of the middle retinal layers. The subtle, deep whitening that results may be difficult to detect on funduscopic examination, but imaging can be very helpful. Infarction of the middle retinal layers produces a characteristic appearance on OCT imaging which

is known as paracentral acute middle maculopathy (PAMM). PAMM is easily recognized on SD-OCT imaging as a hyper-reflective band isolated to the inner nuclear layer (figure 1C, arrow). In these cases, *en face* OCT can be very helpful in colocalizing the region of infarction in relation to retinal vessels (Figure 2), especially when fluorescein angiography appears normal, as in our case (Figure 1B).

The clinical presentation of CLRAO varies dependent upon whether it occurs in isolation, or in association with other disease states. All patients will experience central or paracentral visual loss, in addition to the symptoms typically associated with their concurrent disease. Likewise, workup should be adjusted dependent upon the clinical picture. For instance, CLRAO associated with CRVO should concentrate primarily on vasculopathic risk factors such as hypertension. In patients with isolated CLRAO, or associated with anterior ischemic optic neuropathy, giant cell arteritis must be ruled out and vascular risk factors should be identified and minimized. Thorough review of medications should be performed as isolated CLRAO was recently associated with phosphodiesterase inhibitor use and nocturnal hypotension as might occur in patients taking blood pressure medications before bed. Although embolic CLRAO is rare, it has been reported, and therefore embolic workup should be completed in cases not associated with CRVO.

Our patient underwent a thorough workup which did not reveal any undiagnosed or untreated inflammatory, vasculopathic or embolic disease. Interestingly, she was on multiple blood pressure medications at the time of presentation. She was advised to avoid taking these medications at bedtime. Fortunately, she has regained much of her central visual acuity.

Take Home Points

- Cilioretinal artery occlusion can occur in isolation or in association with other disease states like central retinal vein occlusion and anterior ischemic optic neuropathy.
- The pathophysiology of CLRAO differs from branch and central retinal artery occlusions. Systemic workup should be tailored to concurrent disease states but should include a search for vasculopathic risk factors, giant cell arteritis, and offending medications. Embolic causes are rare, but have been reported, therefore embolic workup should be considered.
- Optical coherence tomography can be helpful in diagnosis of middle retina ischemia, especially when clinical findings are subtle and fluorescein angiography is normal.

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