Central retinal artery occlusion with cilioretinal artery sparing of fovea

Logan Russ Vincent, Mehdi Siddiqui

CASE REPORT

A 64-year-old white male with a past medical history of hypertension and prostate cancer, treated with radiation in remission for the past 10 years, presented with a 1-day history of “tunnel vision” in the right eye. He described the vision as if “looking through a view finder with no peripheral vision”. He denied eye pain, flashes, floaters, diplopia, or recent trauma at the time of presentation. He denied any prior history of similar symptoms or significant ocular history. Social history was significant for a 24 pack-year history of tobacco use. On review of systems, the patient complained of abdominal pain, diarrhea, and nausea for the past few days, but he denied fatigue, jaw claudication, scalp tenderness, headache, or fever. Outpatient medications included aspirin and metoprolol. The patient had an abnormal stress test with negative heart catheter results within the previous three months. Upon presentation to the emergency department, vital signs showed elevated blood pressure of 189/93 mmHg. Lab results were significant for creatinine 1.8 mg/dL, white blood count 15.23 K/mcl, CRP 66.2 mg/L, and ESR 48 mm/hr.

On ophthalmic examination, both eyes showed no abnormalities externally, visual acuity was 20/25 OD and 20/20 OS, and intraocular pressure (IOP) was 16 OD and 18 OS. The right pupil had a trace afferent pupillary defect (APD), extraocular movements were intact bilaterally, and screening confrontation visual field testing showed peripheral restriction in the right eye while the left eye was full. Slit lamp examination showed no acute anterior segment pathology in both eyes. Dilated fundus examination in the right eye showed 0.3 cup-to-disc ratio, 1+ disc edema, superior and inferior macular ischemia with preservation of the fovea, clear vitreous, and diffuse retinal ischemic changes in the periphery (Figure 1). No abnormalities were found in the left eye on dilated fundus exam. On color plate examination, patient identified 9/14 images with right eye and 14/14 images with left eye. The patient was diagnosed with central retinal artery occlusion with preservation of central vision due to cilioretinal artery sparing of the fovea. The diagnosis was made 25 hours from reported vision loss, which placed the patient outside the timeframe for hyperbaric oxygen therapy established at our institution.

Due to elevated ESR and CRP with acute vision loss, admission for thromboembolism workup was recommended and the patient was started on a 3-day course of methylprednisolone due to concern for giant cell arteritis (GCA). Temporal artery biopsy was performed, but results were negative for GCA. MRI of the brain demonstrated multiple aneurysms of the bilateral cavernous internal carotid arteries and anterior communicating artery with mild to moderate stenosis of

Figure 1: Color fundus photo of the right eye exhibiting 1+ disc edema, superior and inferior macular ischemia with preservation of the fovea due to cilioretinal artery perfusion.

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the proximal basilar artery with remote infarct of the right basal ganglia. No acute neurosurgical intervention was indicated. Carotid ultrasound was negative for stenosis and transthoracic echocardiogram showed no significant findings. The patient was discharged on a taper of oral prednisone and was scheduled for follow up with neuro-ophthalmology. Humphrey visual field 30–2 testing was performed as an outpatient and showed global restriction with preservation of a central island in the right eye and was full in the left eye.

**DISCUSSION**

Central retinal artery occlusion (CRAO) is caused by a sudden blockage of the central retinal artery, most commonly due to emboli, with resulting retinal ischemia. Common embolic sources include atherosclerotic plaque from the carotid arteries and cardiac valves. Brown and Shields presented a series of 187 eyes with retinal artery occlusion with 107 eyes (57.2%) representing CRAO. Of these 107 CRAO patients, 26% exhibited macular sparing. [1]. The incidence of CRAO has been cited as 0.85 to 1 in 100,000 persons per year and risk factors including hypertension, diabetes mellitus, carotid artery disease, coronary artery disease, smoking, optic nerve disc drusen, proatherogenic states (e.g., Factor V Leiden, protein C and S deficiency), and sickle cell disease [2, 3]. The common presenting symptoms of CRAO include painless, monocular acute vision loss of count fingers vision or worse in 74% of patients. On funduscopic exam, early clinical findings include cherry-red spot (90%), retinal opacity in the posterior pole (58%), optic nerve pallor (39%), retinal arterial attenuation (32%), and optic disc edema (22%) [3]. In some instances, cilioretinal artery sparing of the macula can prevent the profound central vision loss of CRAO. The cilioretinal artery, supplied by the posterior ciliary arteries, is a terminal artery that supplies the papillomacular bundle, which contains most of the photoreceptors involved in central vision. A study by Justice et al. examined 2000 eyes under stereo color fundus photography and fluorescein angiography and found that 32.1% had one or more cilioretinal arteries. They attributed the increased incidence of cilioretinal arteries to the use of advanced imaging that may not have picked them up typically [4].

Although a cilioretinal artery has been reported in as high as 49.5% of patients, arteries with significant macular collateral circulation are found only approximately 15% of the population and are associated with better long-term outcomes and milder presentations in the setting of CRAO [3, 5]. A study by Hayreh et al. of 260 eyes with CRAO found that 35 eyes had cilioretinal sparing and had a 67% rate improvement of visual acuity within 7 days compared to improvement of 22% non-arteritic CRAO without cilioretinal artery [6]. Acute management of CRAO has been focused on attempting to restore ocular perfusion and the extent of visual improvement depends heavily on type of CRAO and the duration of non-perfusion. Treatment options include digital ocular massage to dislodge thrombus occlusion, anterior chamber paracentesis, intravenous mannitol, sublingual isosorbide dinitrate, and hyperbaric oxygen. Patients also required evaluation for coronary artery disease, peripheral vascular disease, and cerebrovascular disease at the time of diagnosis [3].

**CONCLUSION**

In general, CRAO typically results in devastating vision loss with few options for treatment that results in regaining visual acuity. Because of this patient’s anatomy, the cilioiretinal artery supplying the macula resulted in preservation of a central island of vision.

**REFERENCES**


**Keywords:** Central retinal artery occlusion, Cilioiretinal artery, Retina

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