Bilateral Ocular Ischemic Syndrome in the Setting of Chronic Angle Closure Glaucoma

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Abstract: We report novel case of a 57-year-old woman who developed bilateral ocular ischemic syndrome in the setting of chronic angle closure glaucoma without associated angle neovascularization. Detailed is a course in which markedly prolonged, elevated intraocular pressure led to significantly reduced arterial perfusion at the level of the central retinal artery, leading to the clinical picture of ocular ischemic syndrome.

Key Words: bilateral ocular ischemic syndrome, OIS, chronic angle closure glaucoma, CPAC

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Ocular ischemic syndrome (OIS) occurs in the setting of arterial insufficiency with symptoms such as gradual vision loss, eye pain, amaurosis fugax, and prolonged light recovery.1 Patients with OIS typically present with carotid stenosis of at least 90% occlusion of the ipsilateral carotid artery, though mild to no stenosis has also been reported.1,2 Other sources of disrupted flow may lead to this syndrome, such as significant flow limitation of the ophthalmic artery.4 At presentation, iris neovascularization (NVA) as a result of limited tissue perfusion is present in less than half of eyes as decreased arterial perfusion is believed to lead to ciliary body hypofunction.1,3,4 Anterior chamber in- ward flow limitation of temporal veins (Supplemental Digital Content 1, FA 1 OS late, http://links.lww.com/IJG/A515; FA1 OD late, http://links.lww.com/IJG/S16; FA 2 OS late, http://links.lww.com/IJG/A517; FA2 OD late, http://links.lww.com/IJG/A518). Macular optical coherence tomography (OCT) was unremarkable. Retinal nerve fiber layer OCT showed severe thinning with floor effect in both eyes.

She was started on acetazolamide 500 mg twice a day, timolol, brinzolamide/brimonidine tartrate, and travoprost in both eyes. CT angiogram of the neck and brain showed no abnormalities, specifically no evidence of vascular stenosis.

Two days later, her IOP had decreased significantly to 9 mm Hg in the right and 7 mm Hg in the left eye. Her visual acuity improved to 20/80 right eye and 20/250 in the left eye. She continued to endorse daily waxing and waning vision changes. She underwent emergent cataract extraction and intraocular lens placement, gonioscopy, and Ahmed (New World Medical, Rancho Cucamonga, CA) tube placement in the right eye followed by micropulse cyclophotocoagulation laser of the left eye. She subsequently underwent Ahmed tube placement in the left eye 1 week later.

Two weeks following her presentation, she experienced a pressure spike of >50 mm Hg in both eyes. Her visual acuity declined to hand motion in the right eye and light perception in the left eye. She continued to endorse daily waxing and waning vision changes. She underwent emergent cataract extraction and intraocular lens placement, gonioscopy, and Ahmed (New World Medical, Rancho Cucamonga, CA) tube placement in the right eye followed by micropulse cyclophotocoagulation laser of the left eye. She subsequently underwent Ahmed tube placement in the left eye 1 week later.

Two months after her initial surgery, her fundoscopic findings including midperipheral hemorrhages had resolved. Macular OCT showed 1 to 2+ cystoid macular edema, trace subretinal fluid, and a trace epiretinal membrane in both eyes. Repeat FA showed normal filling, mild cystoid macular edema, mild peripheral leakage, possible trace ischemia, peripheral microaneurysms, and optic nerve head staining in both eyes. She was started on topical prednisolone drops 8 times a day for suspected Irvine-Gass syndrome.
Her most recent best-corrected visual acuity 5 months following her presentation was 20/70 in the right eye and hand motion in the left eye. Her IOPs were 15 and 14.

DISCUSSION

We present an unusual case of bilateral vision loss and fundus findings consistent with bilateral OIS in the setting of bilateral CACG without associated angle NVA. The midperipheral hemorrhages argue strongly enough for OIS that neuroimaging was performed. It is believed that OIS occurs as a result of chronic insufficient arterial perfusion, most commonly from > 90% carotid stenosis. Bilateral OIS is usually associated with bilateral carotid stenosis or systemic inflammatory disorders, such as giant cell arteritis, moyamoya syndrome, Takayasu arteritis, and increased homocysteine/CRP levels.6–9 One report described bilateral OIS without evidence of carotid stenosis, but failed to include details of IOP on initial presentation or final visual acuity. While OIS can cause CACG from NVA, our case uniquely describes CACG without NVA causing OIS.10 There was no evidence of vascular stenosis or systemic inflammatory disorder and both eyes responded to IOP lowering medications.

To our knowledge, CACG itself has not been reported as a cause of OIS. Rao et al describe a patient with bilateral NLP vision, unilateral macular hemorrhages, bilateral angle closure, and IOP of 6 and 10 mm Hg. The clinical description is not consistent with OIS and the authors do not posit how OIS could occur in this patient with relative hypotony.11 Our case showed classic features of OIS and high-IOPs. It is plausible that central retinal arterial vascular compromise from high-IOPs secondary to poor aqueous outflow contributed to the appearance of OIS in this patient. This is consistent with the hypothesis proposed by Song and colleagues who demonstrate that the sudden increase in IOP in acute angle closure glaucoma results in decreased choroidal thickness which may be evidence of compression of the choroidal blood vessels. The compressed choroidal blood vessels may in turn result in ischemic retinal injury.12 The presence of spontaneous central retinal arterial pulsations at extremely high-IOPs, combined with her unusual delay before seeking treatment (3 months), argues that chronic arterial hypoperfusion was present in both eyes. OIS may initially present with intraocular hyotension secondary to decreased ciliary body blood supply and subsequent hypofunction, however, in the setting of bilateral angle closure with nonfunctioning outflow, high pressures will prevail as they did initially in this patient. The restoration of central retinal arterial perfusion following the lowering of IOP led to the resolution of the clinical findings of OIS.

We strongly considered central retinal venous occlusion and diabetic retinopathy in this case. While the patient showed mild venous tortuosity, she did not have other features consistent with central retinal venous occlusion including hemorrhages in the posterior pole, swollen optic nerves or late venous staining on FA.

In conclusion, we report a patient who developed bilateral OIS in the setting of bilateral CACG with markedly prolonged, elevated IOP. We postulate that this occurred as a result of chronically reduced arterial outflow at the level of the central retinal artery. To our knowledge, this association has not been described previously in literature.

**FIGURE 1.** Spontaneous arterial pulsations (3 reference frames from infrared videography) depicted in the left eye. The superior artery blanches over the three frames (arrows).

**FIGURE 2.** Color fundus photos (right and left eyes) showing midperipheral hemorrhages.
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