Central Retinal Artery Occlusion Secondary to Ocular Ischemic Syndrome

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Background

68 year old white male
Ocular History:
Unremarkable, wears reading glasses, last eye exam 7/22/16
Medical history:
- Per patient report –right carotid endarterectomy about 10 years ago
- Type II Diabetic
- Deep Vein Thrombosis
- Hypertension
- Every day smoker

Case Details

09/14/2016
Chief Complaint:
- Sudden, painless vision loss yesterday morning, right eye.
- Denies fatigue, unintentional weight loss, malaise, anesthesia or paresthesia of extremities, or headache

Visual Acuity:
- Right: Hand Motion
- Left: 20/25

Pupils:
- Equal, Round, 4+ Afferent Pupillary Defect Right Eye

Anterior segment:
- Deep and quiet, (-) NVI, IOP 18 OU

Fundus:
Right: white, edematous retina and nerve, severe boxcarring of vessels, multiple visible plaques, 2 nevi inferior arcade
Left: 1 plaque, 1 cotton wool spot, otherwise unremarkable
Plan: labs, carotid ultrasound, and echocardiogram ordered, start brimonidine BID OU

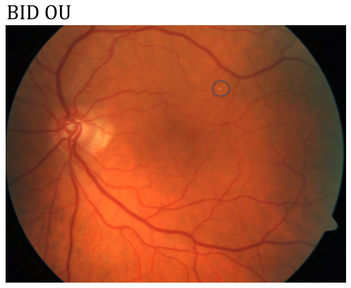

Figure 1: Fundus photos 09/14/2016 left eye

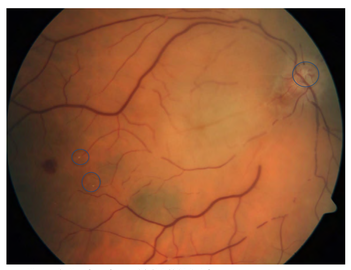

Figure 2: Fundus photos 09/14/2016 right eye

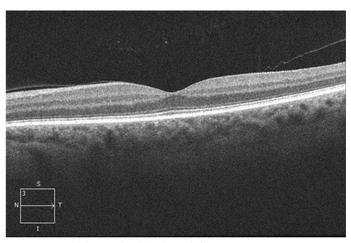

Figure 3: OCT macula 09/14/2016 left eye

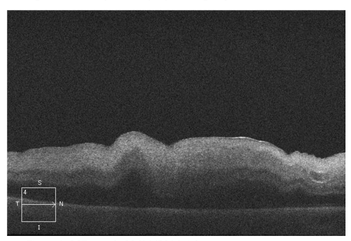

Figure 4: OCT macula 09/14/2016 right eye

Additional Testing

Blood work and cardiac testing 9/2016	
Sedimentary Rate	40 Slightly elevated
C-Reactive Protein	1.345 Slightly elevated
Carotid ultrasound	100% occlusion of the right Almost 100% occlusion of the left
Echocardiogram	unremarkable

Follow up Exam

10/13/16
Chief Complaint: No change in vision, report of pain and photophobia for about 1 week in the right eye
Background: successful endarterectomy of the left carotid artery, no new reports of amaurosis fugax
Visual Acuity:
- Right: Hand Motion
- Left: 20/25

Pupils:
- Equal, Round, 4+ Afferent Pupillary Defect Right Eye

Anterior segment
- Right: 2+ bulbar injection 3-4+ anterior chamber reaction, posterior synechia with neovascularization of the iris
- Left: deep and quiet

Gonioscopy
- Open to Ciliary Body both eyes, no neovascularization of the angle noted

Intraocular Pressure
- Right 12
- Left 13

Plan: Start Prednisolone Acetate and Atropine. Consult with local retinal specialist for further treatment and evaluation.

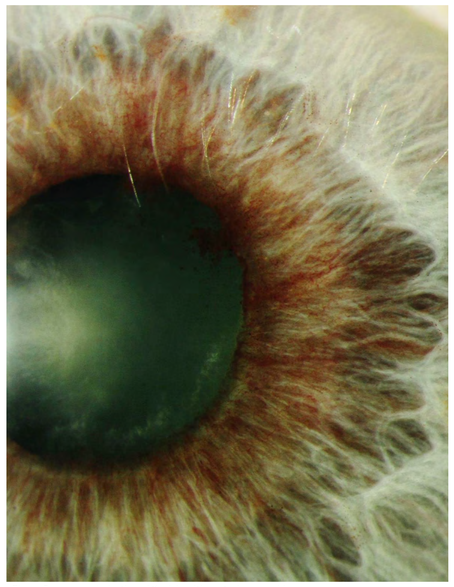

Figure 5: neovascularization of the iris with posterior synechia

Treatment

This patient had a severe central retinal artery occlusion and then went on to develop ocular ischemic syndrome in the right eye. The patient underwent successful left carotid endarterectomy on 9/28/16. Due to complete occlusion of the right carotid artery, endarterectomy was not a viable treatment option.
Pain and intraocular pressure management were initiated:
- Atropine BID: a cycloplegic to stabilize the blood-aqueous barrier and decrease the likelihood of a spontaneous hyphema
- Prednisolone Acetate QID: a steroid to decrease inflammation
- Brimonidine and Cosopt BID: ocular hypotensives to reduce aqueous production and increase aqueous outflow

Prostaglandins are contraindicated considering their inflammatory properties

Currently this patient is also being treated with Antivascular Endothelial Growth Factor injections, to control anterior segment neovascularization. He is scheduled to undergo cataract extraction and synechiolysis, after which they plan to proceed with panretinal photocoagulation as further treatment for neovascularization.

Discussion

Central retinal artery occlusion (CRAO) is a sudden painless loss of vision due to retinal ischemia. It occurs in 1 to 2 per 100,000 patients, who commonly present with count finger vision or worse. A small percentage of patients may present with better visual acuity, due to the presence of a patent cilioretinal artery, which preserves blood flow to the macula. Patients who suffer from CRAO also have a higher incidence of death due to stroke or coronary event for 5 years following the initial incident. Carotid ultrasound and echocardiogram should be completed for all patients with CRAO. Practitioners may also consider a diffusion weighted MRI to rule out concurrent cerebral ischemia. [2]
There is no accepted treatment of CRAO. However, in a study of non-human primates, if treatment was initiated within 100 minutes of onset, no evidence of retinal damage was noted. Treatment between 100 and 240 minutes show variable damage, and after 240 minutes eyes showed irreversible severe damage. Treatment options include:

Dislodge the embolus
- Ocular massage: compression of the globe with digital pressure for 10-20 minutes to increase retinal artery perfusion
- Laser embolectomy: use of an Nd:YAG laser when embolus is visible. This option is controversial and not standard of care

Increase Retinal Artery Perfusion Pressure
- Hypotensive medications: both the use of topical and oral medications
- Anterior chamber paracentesis

Vasodilation
- Hyperventilation: increasing carbon dioxide in the blood prevents vasoconstriction, increasing retinal perfusion

Thrombolysis
- tPA: convert plasminogen to plasmin which dissolves fibrin-based clots [2]

Ocular ischemic syndrome (OIS) is most common in men over the age of 65. It occurs due to systemic atherosclerosis, most often carotid artery stenosis. Patients with OIS can develop anterior uveitis, neovascularization of the iris and/or retina, and mid peripheral retinal hemorrhages. The prevalence of OIS is 7.5 cases per million per year, which is thought to be an underestimation. As with CRAO, patients with OIS have a significantly higher incidence of death following the initial presentation. These patients should also undergo carotid ultrasound.
If significant stenosis is found on carotid ultrasound, these patients may require carotid endarterectomy. However, if the degree of stenosis is too great, carotid endarterectomy is not recommended due to risk of stroke. Some new treatment options include: Extracranial-Intracranial Bypass surgery and carotid artery stenting, which are in trials with the aim to increase blood supply to the ischemic areas. Neither have been shown to restore vision. [1]

Conclusion

Immediate evaluation and treatment following painless loss of vision is vital to potentially restoring vision. In many cases, it is unlikely patients will regain usable vision. Therefore, the management of CRAO and OIS is twofold. First, it is important to address the patient's systemic risk of stroke and cardiac event by ordering immediate bloodwork, carotid ultrasound, echocardiogram and for those able, an MRI. Considering increased mortality rate in those suffering a CRAO or IOS, systemic health problems must be addressed immediately. Then, treatments are aimed at improving quality of life and protecting the vision that remains. Our patient successfully underwent an endarterectomy and is currently undergoing topical therapy and anti-VEGF injections to reduce pain and control neovascularization. In other cases, patients may undergo more invasive treatments to avoid a blind, painful eye, including cycloablation, retrobulbar injection of alcohol or chlorpromazine, and in extreme cases enucleation or evisceration. [1]

References

1. Mendrinos, E., MD, Machinis, T. G., MD, & Pournaras, C. J., MD. (2010). Ocular Ischemic Syndrome.. *Survey of Ophthalmology, 55* (1).
2. Dattilo, M. MD, PhD., Biousse, V., MD, & Newman, N. J., MD. (2017). Update on the Management of Central Retinal Artery Occlusion. *Neurol Clinic, 35.*