Ocular Ischemic Syndrome
Nicole Pogue, OD
Optometric resident-The Dimock Center, Roxbury, MA

Abstract
Ocular ischemic syndrome is an uncommon diagnosis but can tell the clinician many things about a patient’s systemic health, which is usually poor. It can present in many forms and may be misdiagnosed.

I. Case History

CW, a 71 year old black female presented to the clinic in early July 2012 with the chief complaint of blur at distance and near without glasses that she had lost x 5 years. She had no other ocular complaints.

Her ocular history is as follows:
   In 2004 she had trauma to her right eye. The right side of her head, including a portion of her right eye hit the side of a doorframe. OD was not affected however the left eye was “cockeyed” for a month including double vision, eye pain, flashes/floaters, and light sensitivity. No significant findings were noted OS after once the symptoms resolved.

Social history: CW smokes about 1 pack of cigarettes every 3 days and has been a smoker since the age of 18.

Medical history:
HTN and diabetes x 4 mo as well as blood clots in both legs. She was recently hospitalized in March 2012 for elevated blood sugar (>500) and elevated blood pressure (>200).

Family Medical history: unremarkable

Current medications include Metformin, Lisinopril, and Warfarin

II. Pertinent findings

Best corrected visual acuity at distance:
OD: 20/25+1
OS: 20/20-2

All entrance testing was unremarkable

IOP: 16,16 mmHg @2:00pm

Anterior segment examination was unremarkable with the exception of arcus OU and +2 nuclear sclerosis OU.
Fundus examination revealed:
C/D: 0.4, OU
Mild scattered dot/blot hemorrhages, OU
Mild venous dilatation, OU
Peripheral drusen, OU
(-)CSME, OU (OCT performed)
Cotton wool spots sup/temp quadrant, OS
Nasal, mid-peripheral hemorrhages, OS

No physical abnormalities noted and no laboratory studies were performed. The patient was referred back to her primary care physician for a carotid doppler based on the presence of mid-peripheral hemorrhages OS.

III. Differential diagnoses

Differential diagnoses include ocular ischemic syndrome, nonischemic central retinal vein occlusion (CRVO), venous stasis retinopathy, diabetic retinopathy, or hypertensive retinopathy.

IV. Diagnosis and discussion

The patient had multiple diagnoses including ocular ischemic syndrome OS with both eyes manifesting nonproliferative diabetic retinopathy and hypertensive retinopathy. It is believed the retinopathy is resolving from CW’s uncontrolled HTN and diabetes in March.

Ocular ischemic syndrome (OIS) is an uncommon condition resulting from chronic ocular hypoperfusion secondary to ipsilateral atherosclerotic carotid stenosis. The carotid artery usually has 90% or more occlusion. OIS usually occurs in patients 50-70 years old with a male predilection. The condition is rarely bilateral (only 20% of cases) and there are strong systemic associations with hypertension and diabetes. The mortality rate is 40% at 5 years. The leading cause of death is cardiovascular disease, usually myocardial infarction with cerebral infarction being the second leading cause.

Signs and symptoms of OIS

Patients may present with:
- Gradual/decreased visual acuity ranging from 20/20 to NLP
- Transient vision loss
- Ocular pain. This may be present in about 40% of patients but of those with pain, approximately 94% have iris neovascularization
- Photophobia and a prolonged photostress recovery

Anterior segment findings may include:
- Unilateral conjunctival and episcleral injection
- Corneal edema (rare)
- Mild anterior chamber reaction with flare being greater than cells. Keratic precipitates are usually absent.
- Iris atrophy
- Rubeosis
- Cataracts may be seen in advanced cases but this is rare.
- IOP may be elevated

Posterior segment findings:
- Venous dilation without tortuosity
  - CRVO vessels dilated and tortuous
- Arterial narrowing
- Retinal hemorrhages seen in ~80% of affected eyes usually mid periphery and usually dot/blot
- Disc edema is rare
- Disc and retinal neovascularization
- Spontaneous arterial pulsation
- Macular edema
- Cotton wool spots

OIS when compared with a CRVO or diabetic retinopathy has some unique features. The retinal veins will be dilated but not tortuous. In a CRVO the veins are both dilated and tortuous while diabetic retinopathy presents dilated and beaded veins\(^3\). The hemorrhages seen in OIS most commonly present in the mid-periphery\(^3\). There is typically spontaneous arterial pulsation. This is caused by a low diastolic pressure. If not seen it can be induced if pressure is exerted on the globe\(^3\).

Anterior segment ischemia may be observed. Dilated conjunctival and episcleral vessels may be seen. The presence of episcleral injection may be a sign of collateral blood flow from the external carotid artery due to an internal carotid artery occlusion\(^3\). If iris atrophy and neovascularization are seen, this may be indicative of longstanding anterior segment ischemia\(^3\). A mild uveitis may be seen. There will be more flare than cells and rarely keratic precipitates\(^3\).

V. Treatment and management

Once the diagnosis of OIS has been made, the patient needs to be properly referred as there is an increased chance of myocardial and cerebral infarction\(^3\). A carotid doppler should be performed to determine the amount of blockage in the carotid arteries. Magnetic resonance angiography and computed tomography angiography may be helpful as a second line of testing to further confirm the amount and location of the blockage\(^3\). Fluorescein angiography can help establish a diagnosis of OIS\(^3\). There will be slow or patchy choroidal filling (seen in about 60% of eyes) as well as a delayed arteriovenous transit time\(^3\).

Anterior segment manifestations should be treated with the appropriate medications, usually steroids and cycloplegics. If there is proliferative retinopathy, PRP may be
required but it may not always be effective\(^3\).

A carotid endarterectomy (CEA) is usually helpful as it can reverse or prevent progression of chronic ocular ischemia and may increase ocular blood flow\(^3\). As a result, visual acuity may increase or remain stable. However, some studies found it did not help improve the ocular ischemia or vision, but most patients in these studies had poor initial visual acuity and/or the presence of iris neovascularization\(^3\). A CEA may help to prevent cerebral infarctions as well\(^3\). It is recommended in 50-99% of symptomatic patients if perioperative risk of stroke or death is less than 6%\(^3\). It is recommended in 60-99% of asymptomatic patients if perioperative risk of stroke or death is less than 3%\(^3\). For those who are considered high risk for complications after CEA, carotid artery stenting can be used but there is insufficient evidence as to the effectiveness of stents over CEA\(^5\).

**VI. Conclusion**

Ocular ischemic syndrome is uncommon and may be misdiagnosed. It is helpful to understand the differences between OIS and a CRVO and diabetic retinopathy. Once a diagnosis of OIS has been made, it is imperative that the patient receives the proper testing and treatment as their life may depend on it.

**Clinical Pearls:**

- When seeing a relatively mild anterior uveitis in an elderly patient, suspect OIS.
- Look for asymmetry. Suspect OIS when patients present with asymmetric retinopathy, asymmetric cataracts, or a unilateral red eye.
- Always consider OIS in elderly patients with neovascularization of the anterior segment.
- Consider OIS when encountering ocular hypotony, especially if there is concurrent anterior chamber reaction and/or neovascularization.
- The retinal veins in OIS will be dilated, but not tortuous. If tortuous, consider a retinal vein occlusion.
- Refer the patient right away. The carotid arteries need to be imaged as soon as possible to determine blockage.

**References:**
