Ocular Ischemic Syndrome: A Brief Review

Hyder J. Almosawy, OD; Joseph V. Mega, OD; Paul B. Greenberg, MD, MPH; Claire Messina, OD; Amanda M. Hunter, OD

Abstract
We review the etiology, characteristics and management of ocular ischemic syndrome (OIS). Ocular ischemic syndrome is a visual-threatening condition closely linked to advanced cardiovascular disease. Patients with OIS are at risk for ocular neovascularization and carotid artery stenosis. Diagnostic testing to confirm the diagnosis of OIS must be done in order to properly determine treatment and management options for these patients as the risk for permanent visual loss is greater with higher levels of ischemia and advanced cardiovascular disease. Coordination of care with the patient’s primary care provider is necessary to optimize the patient’s cardiovascular risk factors and determine the most appropriate medical and/or surgical treatment.

Introduction
Ocular ischemic syndrome (OIS) is an uncommon but sight-threatening condition caused by the atherosclerotic occlusion of the common or internal carotid arteries or less commonly the ophthalmic artery. Ocular ischemic syndrome typically occurs in the elderly—mean age is 65—and is an important marker of cardio- and cerebrovascular disease, given that up to 3% of this age group have carotid artery disease. Herein, we review the key characteristics and management of patients with OIS.

Characteristics
Ocular ischemic syndrome presents unilaterally in 80% of cases. Vision loss can be gradual over several months to minutes, with patients presenting with a visual acuity ranged from 20/50 to counting fingers. Patients may have a sluggish pupillary response to light on the affected side corresponding to the stenosis. Notable anterior segment findings include neovascularization of the iris (NVI) (Fig. 1) [present in up to 90% of cases], neovascularization of the angle (NVA), iris atrophy, low grade uveitis, and hypotony.

The most common posterior segment finding in OIS is mid-peripheral retinal hemorrhages (Fig. 2), which are present in 80% of cases. Other posterior segment findings include attenuation of retinal arteries, dilated retinal veins, microaneurysms, and posterior segment neovascularization. Cystoid macular edema (CME) is present in 17% of OIS cases.

Diagnostic Testing
When a patient is suspected of having OIS, several ophthalmic diagnostic tests are helpful. Formal visual field testing can uncover deficits due to retinal ischemia. The visual fields can range from normal to severe restriction and can serve as a baseline for future change. Fluorescein angiography (FA) can point to the presence of carotid stenosis and assess the degree of retinal ischemia. In a normal patient, the arm-to-choroid time in FA is normally five seconds; this can increase to one minute in OIS patients due to carotid artery occlusion. In addition to demonstrating retinal hypoperfusion in OIS, FA can show leakage from neovascularization, microaneurysms or CME.

Figure 1. Neovascularization of the iris (NVI) corresponding to advanced retinal ischemia. (Courtesy, North American Neuro-Ophthalmology Society.)
With the assistance of the patient’s primary care provider (PCP), suspected carotid artery stenosis can be verified with carotid ultrasound, also known as the carotid duplex.\textsuperscript{1,2} Patients with OIS typically have 75\% stenosis of the carotid artery.\textsuperscript{1} Carotid duplex, with a sensitivity of up to 89\% and a specificity of up to 84\%,\textsuperscript{2,7,8} is a reliable diagnostic tool in patients with 70 to 90\% carotid stenosis. A carotid duplex may not detect stenosis if it occurs higher up on the carotid artery.\textsuperscript{7} Indeed, for those suspected OIS patients without significant stenosis on carotid ultrasound testing, a magnetic resonance angiogram (MRA) or computed tomography angiography (CTA) may be helpful.\textsuperscript{1,2,8} For detecting 70-99\% carotid artery stenosis, the MRA and CTA have a sensitivities and specificities of up to 95\% and 90\% and up to 91.6\% and 97.4\%,\textsuperscript{7,8} respectively. Unlike CTA, MRA does not expose patients to radiation but it would generally preclude patients with claustrophobia, pacemakers or other metallic implantable devices in the body.\textsuperscript{7}

Management

Ocular disease

The ocular treatment of patients with OIS depends on the presenting signs. Neovascularization of the anterior or posterior segment is typically treated with laser pan-retinal photocoagulation or intravitreal injection of anti-vascular endothelial growth factor (VEGF) agents.\textsuperscript{1} In patients with CME, the main course of treatment is intravitreal anti-VEGF therapy.\textsuperscript{1,2} For patients without ocular NV or CME, continued observation with undilated gonioscopy and dilated fundus exams is necessary to monitor for these complications. The ocular follow-up plan for OIS patients will be based on their exam findings: patients with ocular neovascularization who require treatment will need closer follow-up than patients without ocular neovascularization.

Systemic disease

The systemic treatment of OIS can involve both medical and surgical therapy. Ocular ischemic syndrome is an important marker of cardiovascular disease.\textsuperscript{1,2} To this end, it is important to work with the patient’s PCP to manage any risk factors for cardiovascular disease. This includes treatment of hypertension, hypercholesterolemia or diabetes mellitus, smoking cessation and weight loss through diet and exercise.\textsuperscript{1,2} Additional subspecialty consultation with cardiology or neurology may also be needed.\textsuperscript{2} Anti-platelet agents and statins are effective in treating patients with less than 70\% carotid artery stenosis but can also be used in a concerted effort with surgical intervention.\textsuperscript{9}

The most effective surgical intervention is the carotid artery endarterectomy (CEA), which lowers the risks of stroke in patients with 70-99\% of carotid stenosis.\textsuperscript{1,2,8,10-12} A CEA is often effective in preventing progression and reversing signs of OIS by increasing blood flow to ocular tissues.\textsuperscript{10,12} For these reasons, patients with significant carotid artery stenosis should be referred to vascular surgeons to consider CEA.

Not all OIS patients with this degree of stenosis may benefit from CEA: a CEA is considered a high-risk procedure for perioperative stroke in patients over the age of 80 or who suffer congestive heart failure, chronic obstructive pulmonary disorder, recent coronary artery bypass graft or with complete occlusion of the contralateral carotid artery.\textsuperscript{9,11} Carotid artery stenting (CAS), which involves inserting a stent into the stenotic lumen of an atherosclerotic carotid artery, is an alternative treatment for higher risk patients.\textsuperscript{9,10}

Prognosis

The majority of OIS patients (58\%) will have counting fingers vision or worse visual acuity (VA) one year following diagnosis, irrespective of their presenting vision.\textsuperscript{1,2,6} If NVI is present at the time of diagnosis, 97\% of patients will have a VA of counting fingers or worse after 1 year.\textsuperscript{6} Systemically, patients presenting with OIS have a 40\% mortality rate at five years.\textsuperscript{1,2,4} The most common causes of death in OIS patients are myocardial infarction (67\%) and cerebrovascular accident (19\%).\textsuperscript{1,2,4} Educating patients on OIS and its ocular and systemic manifestations is not only critical in terms of preserving vision but also in reducing the risk for cardiovascular complications.

Conclusion

In sum, OIS is closely linked to a patient’s ocular and systemic health. It is important that eye providers work closely with the PCP to optimize the health outcomes of this high-risk patient cohort. Due to the high mortality rate (40\%) of patients with a diagnosis of OIS, it is imperative that eyecare providers perform a thorough dilated eye examination on every patient annually.

Acknowledgements

We thank Dr. Joseph Sowka, OD for providing the fundus photograph for this manuscript.
References