Ocular ischemic syndrome: A classical presentation of an uncommon condition

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We report a 47-year-old male who presented with acute monocular vision loss, and had classical signs of global ocular ischemia in the right eye. Fundus fluorescein angiography demonstrated delayed choroidal filling and no perfusion of retinal vasculature. Carotid Doppler and computed tomogram (CT) angiography studies revealed extensive bilateral atherosclerotic disease involving the carotid circulation. Ophthalmologists must be aware of the possibility of this potentially fatal condition, which is extremely rare. An astute clinical diagnosis, targeted workup for systemic associations and a prompt referral may turn out to be life-saving.

Key words: Carotid occlusive disease, mono-ocular vision loss, ocular ischemic syndrome

Ocular ischemic syndrome is a vision-threatening condition which may herald a potentially devastating cerebral infarction.[1] It is characterized by both anterior and posterior segment ischemic signs in the eye.[2] The reported incidence is 7.5 cases per million persons every year.[3] A detailed literature search did not reveal any report of such a case from India.

We report an adult male who presented with unilateral loss of vision and was detected to have classical features of ocular ischemia, with bilateral carotid occlusive disease.

Case Report

A 47-year-old male presented with loss of vision, swelling, redness and pain of 15 days’ duration in the right eye. There was no history of ocular trauma, surgery or systemic disease, including hypertension or diabetes mellitus.

The patient had no light perception in the right eye, and the intraocular pressure was 14 mm Hg. A detailed examination revealed the presence of conjunctival chemosis and congestion, simulating the appearance of a bulging eye; whereas exophthalmometry readings and extra-ocular movements were within normal limits. There was presence of anterior segment flare and uveal ectropion, with 360 degrees of iris neovascularization [Fig. 1] and presence of new vessels in the inferior quadrant of the anterior chamber angle on gonioscopy. Both direct and consensual light reflexes were absent. Fundus examination showed a hyperemic disc with blurred margins, blot hemorrhages in all quadrants extending up to the mid-periphery, a single cotton wool spot and opacification of the retina [Fig. 2]. The visual acuity in the left eye was 20/25 and intraocular pressure was 12 mm Hg. Anterior and posterior segment examination of the left eye was unremarkable.

Fundus fluorescein angiography (FFA) revealed a delay in choroidal filling in the right eye, with first signs of appearance of dye after 50 sec [Fig. 3]. There was no filling of retinal vessels, even after 14 min [Fig. 4]. A carotid Doppler study showed patterns suggestive of atherosclerotic changes involving bilateral common carotid arteries and carotid bulb, as well as bilateral internal carotid artery occlusion. There was no flow detected in the right internal carotid artery and a 35-40% stenosis on the left side. A subsequent computed tomogram...
(CT) angiography corroborated these findings, with bilateral proximal internal carotid artery occlusion [Fig. 5] and partial reformation of supraclinoid internal carotid arteries via circle of Willis.

The blood pressure was 110/70 mm Hg, carotid pulses were feeble on palpation on both sides, with no irregularities detected on cardiac auscultation. Further investigations revealed a random blood sugar level of 120 mg/dl, serum triglyceride level of 207 mg/dl and serum cholesterol level of 144 mg/dl, with a normal 2D echocardiography report.

**Discussion**

Signs of anterior segment ischemia (conjunctival chemosis and congestion, a mild inflammatory reaction in the anterior chamber, presence of neovascularization, fixed semi-dilated pupil) coupled with posterior segment ischemic signs (retinal opacification with no cherry red spot, retinal hemorrhages extending up to the mid-periphery) immediately point to the diagnosis of ocular ischemic syndrome. One of the most striking features is the normal intraocular pressure (14 mm Hg) despite anterior segment neovascularization and uveal ectropion, thought to be due to ciliary body ischemia and reduced aqueous humor production.[4] The characteristic findings of delayed choroidal perfusion and non-filling of retinal vessels on FFA confirm the diagnosis.

A detailed workup is warranted in all patients with ocular ischemia to look for systemic associations such as carotid or coronary occlusive disease, atherosclerosis, hypertension and diabetes mellitus. In the absence of any of these, one may look for other, rarer associations such as giant cell arteritis, aortic arch syndrome and Takayasu arteritis.

An interesting feature in this case is the relatively young age of the patient. The mean age at presentation of ocular ischemic syndrome is 65 years, and the condition is rare before the age of 50.[5] The absence of known systemic disease is also notable; systemic arterial hypertension and diabetes mellitus have been found to be present in over half the patients with this condition.[5,7] Moreover, there is no history of episodes of transient visual loss. Though considered to be the hallmark of carotid insufficiency, such a history is present in 10-15% of patients with ocular ischemic syndrome.[5,7] On FFA, the choroid normally fills completely within five seconds of the appearance of the dye in the choroidal arteries. Prolonged choroidal filling time, evident in this patient, is considered...
to be the most specific angiographic sign of ocular ischemic syndrome.[5]

Ophthalmologists need to be especially alert regarding this condition, as it is uncommon, and may present for the first time without any preceding ischemic events elsewhere in the body, as in this case. Ocular ischemic syndrome as the initial manifestation of bilateral occlusion of carotid arteries is rare.[8] It has a dismal visual prognosis.[7,9] The greatest concern is that the five-year mortality rate is 40%.[6] A careful clinical examination combined with targeted ophthalmic investigations like FFA may clinch the diagnosis. Prompt referral to the concerned specialists for appropriate minimally invasive or surgical intervention may be life saving.

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