# Ocular ischemic syndrome as initial manifestation of bilateral internal carotid artery occlusion

## Sindrom očesne ishemije kot začetni znak obojestranske zapore notranjih karotidnih arterij

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## Abstract

**Background:** Ocular ischemic syndrome (OIS) is characterized by ocular symptoms and signs secondary to severe carotid artery stenosis or occlusion.

**Methods and results:** The authors describe a case of a 66-year-old man, a smoker with hyperlipidemia, who presented with a three-day history of pain over his left eye associated with redness. Visual acuity (VA) of the left eye was 6/18, with marked anterior chamber flare, unreactive dilated pupil, rubeosis of the angle and intraocular pressure of 30 mmHg. Fundus examination showed marked venous stasis with cotton-wool spots. VA of the right eye was 6/6, with mild venous stasis. No bruits were heard on carotid auscultation. Carotid Doppler ultrasonography revealed complete occlusion of both internal carotid arteries (ICA's) which was confirmed by CT-angiography. Endarterectomy of ICA's could not be performed. Three months later panretinal argon laser photocoagulation in the left eye was performed. One year later the left eye VA was perception of light with progression of neovascular glaucoma and development of cataract. No deterioration was noted and VA remained stable 6/6 in the right eye.

**Conclusions:** OIS has a poor visual prognosis, and treatment is primarily aimed at management of the underlying carotid occlusive disease. Clinicians should be aware of the signs and symptoms of carotid artery disease in order to facilitate prompt diagnosis and appropriate referral.

## Izvleček

**Izhodišča:** Za sindrom očesne ishemije so značilni očesni simptomi in znaki, ki so posledica hude zožitve ali zapore karotidnih arterij.

**Metode in rezultati:** Predstavljamo primer 66-letnega moškega, kadilca s hiperlipidemijo, ki je bil napoten na pregled zaradi tri dni trajajoče bolečine v levem očesu s pordelostjo. Vidna ostrina na levem očesu je bila 0,3. Prisotna je bila huda uveitična reakcija s široko in nereaktivno zenico, neovaskularizacijo šarenice in zvišanim intraokularnim tlakom do 30 mmHg. Na očesnem ozadju levega očesa so bili znaki hudega venskega zastoja s posamičnimi mehkimi eksudati, na desnem pa znaki blagega venskega zastoja. Barvna doplerska preiskava karotidnih arterij je pokazala popolno zaporo obeh notranjih karotidnih arterij, kar smo potrdili z računalniško-tomografsko angiografijo. Bolnik ni bil kandidat za endarterektomijo notranjih karotidnih arterij. Šest mesecev kasneje je bila narejena laserska fotokoagulacija levega očesa. Po letu dni je na levem očesu prišlo do napredovanja neovaskularnega glavkoma in nastanka katarakte, oko je zaznavalo le svetlobo. Na desnem očesu nismo ugotovili poslabšanja, vidna ostrina očesa je ostala stabilna 1,0.

**Zaključki:** Sindrom očesne ishemije ima slabo prognozo, zdravljenje pa je usmerjeno v oskrbo zapore karotidnih arterij. Prepoznavanje znakov in simptomov bolezni karotidnih arterij omogoča pravočasno diagnozo in ustrezno obravnavo.

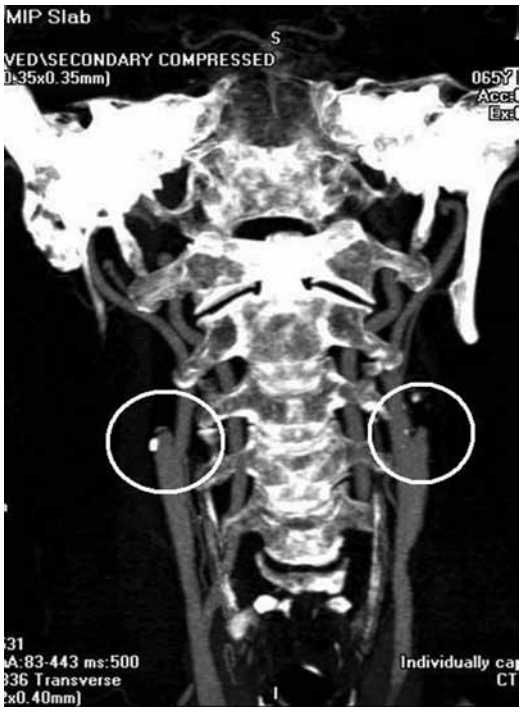


**Figure 1:** CT angiography showing absence of filling of both internal carotid arteries (marked with circles).

## Background

Ocular ischemic syndrome (OIS) is a rare and severe form of ischemia of both anterior and posterior segments of the eye in patients with severe carotid artery stenosis or occlusion. The early and often asymptomatic stage of chronic ocular ischemia is referred to as hypotensive retinopathy, previously known as venous stasis retinopathy.[1]

## Case report

A 66-year-old male patient, a smoker with hyperlipidemia, presented with a three-day history of pain over the left eye associated with redness. Visual acuity (VA) of the left eye was 6/18, with dilated episcleral vessels, marked anterior chamber flare, unreactive dilated pupil, iris neovascularization with angle neovascularization and intraocular pressure of 30 mmHg. The fundus showed marked venous stasis with mid-peripheral microaneurysms, small dot and blot intraretinal hemorrhages and cotton-wool spots. VA of the right eye was 6/6, with signs of mild venous stasis. No bruits were heard on carotid auscultation.

Carotid Doppler ultrasonography revealed complete occlusion of both internal carotid arteries (ICA's). CT-angiography confirmed bilateral occlusion of ICA's immediately after bifurcation (Figure 1 and Figure 2) with ischemic changes of brain cortex in occipital region. Aortocervical digital subtraction angiography demonstrated cerebral blood supply via collaterals from external carotid artery system, while vertebral arteriography showed well maintained cerebral blood supply from vertebrobasilar system via posterior communicating arteries, mainly from the right site. Cardiac echography excluded emboli of cardiac origin. Blood tests were normal except for raised total cholesterol (6.1 mmol/L) and LDL-cholesterol (4.1 mmol/L). Endarterectomy of ICA's could not be performed. Medical treatment included Aspirin, atorvastatin, and advice to stop smoking. Six months later a full retinal laser photocoagulation was performed in the left eye. One year later the left eye VA was perception of light with progression of neovascular glaucoma and development of dense cataract. No deterioration was noted and VA remained stable 6/6 in the right eye.

## Discussion

OIS is a rare form of ischemia of the eye secondary to severe carotid artery stenosis, with reported frequencies between 5 % and 21 % in series of patients with occlusive carotid artery disease.[2] At least 90 % ipsilateral carotid artery stenosis is necessary to reduce the perfusion pressure within the corresponding central retinal artery by about 50 % and induce ocular ischemia.[3] It is uncertain why only some patients develop chronic ocular ischemia. In ICA obstruction, collateral blood flow via branches of external carotid artery may result in reversal of flow in ophthalmic artery to sustain cerebral blood flow. Decreased blood flow to the eye thus results both from the blocked ICA and secondarily induced ophthalmic artery steal effect.[3] Kljin et al. have demonstrated that OIS is associated with compromised cerebral blood flow.[2]

Patients with carotid occlusive disease may develop a variety of symptoms and signs secondary to chronic ocular ischemia.[4] As these usually represent the earliest signs of life threatening carotid artery disease, a

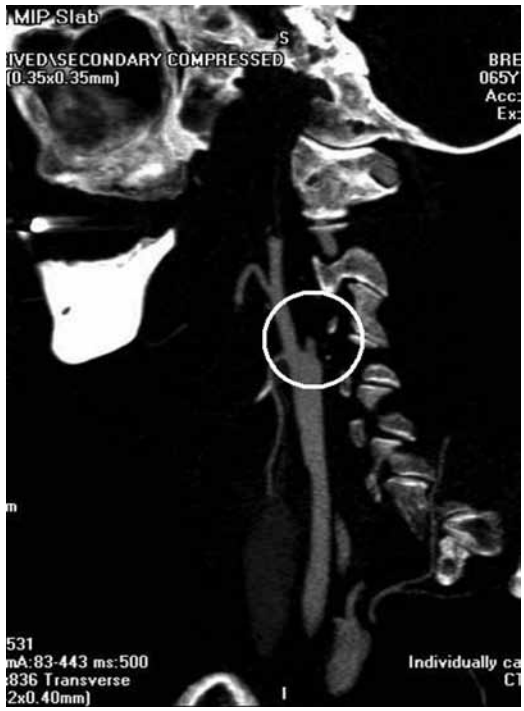


**Figure 2:** CT angiography showing absence of filling of left internal carotid artery (marked with circle).

high index if suspicion is necessary. Clinical presentation may include sudden, gradual, or transient vision loss.[1] At presentation, approximately 43 % of eyes have 6/6 to 6/15 vision, whereas 37 % have counting fingers or less. Pain occurs in 40 % of patients with OIS and is usually described as a dull ache localized either to the eye or the periorbital region that may improve in the supine position. The probable causes of ocular angina are ischemic damage to the branches of the ophthalmic division of trigeminal nerve or increased intraocular pressure due to neovascular glaucoma.[3] Our patient presented with pain over the left eye, while he was unaware of gradual vision deterioration in the same eye (6/18).

Anterior segment signs include dilated episcleral vessels, corneal edema, anterior chamber cells, and pronounced flare. Neovascularization of the iris may be present in approximately two-thirds of eyes with OIS at the time of the initial visit, and is considered an indicator of poor visual prognosis.[1] Intraocular pressure is usually raised, but may be normal or even reduced, presumably due to ciliary body ischemia leading to reduced aqueous production. Posterior segment signs include venous dilatation with or without tortuosity, mid-peripheral retinal hemorrhages and microaneurysms. Ischemic changes include retinal arteriolar narrowing, retinal capillary non-perfusion, macular edema, optic disc neovascularization, and less commonly, retinal neovascularization. In 20 % of cases, the clinical signs of OIS are an incidental, asymptomatic finding.[5]

When carotid artery disease is suspected, a prompt noninvasive vascular workup is mandatory to confirm the carotid disease, and assess its severity. These include carotid duplex scanning, CT angiography, arteriography, digital subtraction angiography, and magnetic resonance angiography.[3] Ocular investigations comprise fluorescein fundus angiography and visual field. Fluorescein fundus angiography signs include delayed and patchy choroidal filling, increased retinal arteriovenous circulation times, areas of retinal capillary non-perfusion, late leakage from retinal vessels, and macular edema. Visual fields vary from normal to central scotomas, centrocaecal scotomas, and nasal defects. Moreover, a timely referral for full medical and neurological evaluation in order to identify systemic risk factors and associated cardiovascular disease is required.[1]

Ocular treatment of OIS remains difficult and controversial. It is aimed at management of anterior segment inflammation, reduction of retinal ischemia, and treatment of raised intraocular pressure and neovascular glaucoma.[1] Panretinal photocoagulation alone may not induce regression of neovascularization or neovascular glaucoma.[4] Recently, successful treatment of neovascular glaucoma secondary to ocular ischemic syndrome with intravitreal injection of monoclonal antibody directed against the vascular endothelial growth factor (bevacizumab) has been reported.[6,7] Additionally, cyclodestructive procedures are performed if medical therapy fails to provide symptomatic relief.[8]

Medical treatment of carotid disease includes antiplatelet therapy, treatment of concomitant diseases such as hypertension or diabetes, and advice to stop smoking.[1] The decision concerning treatment of carotid artery disease involves both the neurologist and vascular surgeon. Carotid endarterectomy has been shown to benefit symptomatic

patients with greater than 70 % internal carotid artery stenosis.[9] However, carotid surgery may not alter long term visual outcome in the affected eye, and the results are often discouraging.[5] Patients with total carotid artery obstruction do not benefit from surgery. In such instances, the thrombus often propagates to the next major vessel.[3] Similarly, extracranial to intracranial by-pass surgery showed no advantage in preventing ischemic strokes.[10]

## Conclusions

OIS has a poor visual prognosis, and treatment is primarily aimed at management of the underlying carotid occlusive disease. Ophthalmologists should be aware of the signs and symptoms of OIS in order to facilitate prompt diagnosis and appropriate referral.

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