

## Case Report

### Ocular ischemic syndrome; A case report

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#### Abstract

**Background:** Ocular ischemic syndrome is not a common condition so most of these cases are often misdiagnosed or treated as a different entity. Therefore, it is very important for the ophthalmologists to have this condition in mind as a differential so that the patients can be diagnosed and treated as early as possible.

A 42 years female presented with painless, progressive diminution of vision in right eye over the period of 1 month. She doesn't give any history of redness of eyes, fever or trauma. There is no history of diabetes mellitus or hypertension as well.

On examination, vision in right and left eyes was 1.78 and 0.30 Log Mar Units respectively. On anterior segment examination, neovascularization of iris (10 4 'o' clock hrs) in right eye was noted. On dilated funduscopy, neovascularization of disc(1/3<sup>rd</sup>) was present in right eye. Cotton wool spots, blot hemorrhages and micro aneurysms were also noted in right eye. Likewise, attenuation of arteries were noted on both sides.

Fundus fluorescent angiography revealed delayed arteriovenous and venous phase. Carotid Doppler was done which showed complete occlusion of right common carotid and bilateral internal carotid artery. These findings lead to the diagnosis as ocular ischemic syndrome so she was then referred to the cardiologist who further confirmed that no active intervention was required at present. The patient was planned for right eye panretinal photocoagulation (PRP) and was completed in 2 sittings and was asked to follow up regularly.

**Conclusion:** Early diagnosis and management is very important for uncommon conditions such as ocular ischemic syndrome to prevent further complications.

**Key words:** ocular ischemic syndrome, cotton wool spots, micro aneurysms, panretinal photocoagulation.

#### Introduction

Ocular ischemic syndrome is an uncommon entity with vision threatening potentials. Severe

carotid artery occlusive diseases leading to arterial narrowing is the main cause of this condition. Atherosclerosis, either due to aging or dyslipidemia is the commonest factor for hardening and narrowing of carotid arteries resulting in ocular hypoperfusion. These patient commonly presents with symptoms of gradual or transient loss of vision and ischemic ocular pain (Terelak-Borys et al., 2012b). This condition is common in the age group of 60

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to 70 years and is less common before 50 years(Mendrinis et al., 2010). In about 20% of the cases, the involvement is usually bilateral. Visual loss is usually gradual and occurs over a period of few weeks or months in about majority of the cases. However, few cases may present with sudden loss of vision over a period of minutes or seconds(Mendrinis et al., 2010).

On examination, anterior segment findings include neovascularization of the iris which is present in about 66% of the patients(Mizener et al., 1997). On dilated fundus ophthalmoscopy examination, the retinal arteries are commonly found to be attenuated as a result of atherosclerosis. However, both retinal arteries and veins may be attenuated in few exceptional cases. Retinal hemorrhages are the commonest findings on fundus examination and are present in majority of the cases. (Terelak-Borys et al., 2012a).

The most important diagnostic tool of ocular ischemic syndrome is Imaging studies of the carotid arteries. Doppler ultrasound(Ho et al., 1992) and ocular plethysmography are commonly used non invasive techniques while invasive techniques such as carotid arteriography are also available. Fundus fluorescence angiography can also aid in diagnosis which shows delay in the arteriovenous and venous phase due to narrowing of the arteries(Malhotra and Gregory-Evans, 2000a).

Early diagnosis is very much important to prevent vision and life threatening complications resulting from carotid arteries obstruction.

### Case presentation

A 45 year old female presented with gradual diminution of vision in right eye for a duration of 1 month. It was not associated with pain, redness and fever. She doesn't give any history of trauma, diabetes mellitus and hypertension as well. She has not been under medication for any disease too.

At the time of presentation, the vision in right eye was 1.78 log mar units and left eye was 0.30 respectively with no improvement in refraction. On examination, the pupil in right eye was mid dilated and fixed. Iris neovascularization was present in the right eye from 1 to 4 'O' clock on slit lamp examination.

On fundus examination in right eye, neovascularization in disc( about 1/4<sup>th</sup>) was present. The arteries were attenuated, cotton wool spots and micro aneurysms were also present (Fig 1). The fundus of left eye was normal except for some amount of arterial attenuation.

Intraocular pressure was found to be 16 and 14 mm of Hg respectively on Applanation tonometry.

On fundus fluorescence angiography(FFA), there was delay in venous phase indicating slowing of arterial and arterio venous phase, hyper fluorescence over disc indicating neovascularization and leakage areas corresponding to the area of microaneurysms in arterial and capillary(AV) phase(Fig 2).

Carotid artery Doppler was done to support the diagnosis which showed the following findings.

- i. Complete occlusion of right common carotid and bilateral internal carotid artery.
- ii. Thickened intima of left common carotid artery
- iii. Stenosis of left common carotid artery.

On blood examination, hyperlipidemia was present with lipid function tests being outside normal limits. Total serum cholesterol was found to be 19 mmol/L(Normal:3.5-5.1 mmol/L).

She was diagnosed as ocular ischemic syndrome on the basis of above findings. The patient was at first referred to a cardiologist and hypo-lipidemic drugs were initiated. The cardiologist further revealed that no active surgical intervention was required at present.



**Figure 1:** Photograph of right fundus showing disc neovascularization, cotton wool spots and microaneurysm.



**Figure 2:** Fundus fluorescence angiography of the right fundus showing disc hyperfluorescence and leakage through microaneurysms (arterial and arteriovenous phase of FFA)

The patient was then planned for right eye pan retinal photocoagulation (PRP) and right eye PRP was done in 2 sittings. The patient was then advised to follow up in every 4 weeks. On subsequent follow up, regression of neovascularization on disc and iris had occurred however the vision remained the same.

## Discussion

Ocular ischemic syndrome is a rare but serious ocular condition resulting from chronic and severe carotid obstruction leading to retinal insufficiency. Because of its variable modes of presentation, most cases are often misdiagnosed or diagnosed late. The most common causative factor is atherosclerosis which is hardening and narrowing of the common carotid arteries either due to aging or dyslipidemia (Mendrinis et al., 2010). However, other possible but uncommon causes may include giant cell arteritis, Bechet's disease, etc (Terelak-Borys et al., 2012b).

The case of ocular ischemic syndrome was first studied and presented by Hedges (Hedges, 1963) in 1963. He studied a case who was found to have peripheral dot and blot hemorrhages and dilated retinal veins on fundus examination the cause of which was found to be retinal hypoxia as a result of carotid arteries narrowing.

Similarly, Kearns (Kearns and Hollenhorst, 1963) and Hollenhorst, also reported the presence of retinal veins dilation along with mid-peripheral dot-blot and flame-shaped hemorrhages, and microaneurysms in some of their patients with carotid artery diseases and referred the condition as venous stasis retinopathy. He also observed the signs of ischemia such as neovascularization in some of those cases in both anterior segment (iris) and retina, and these all findings were eventually referred to as ocular ischemic syndrome.

Studies show that it is bilateral in about 20% of the cases with the male female ratio being about 2:1 (Kearns et al., 1978), the other contributing risk factors being hypertension, hyperlipidemia, uncontrolled diabetes mellitus, ageing and vasculitis as well. A detailed systemic evaluation should be done to rule out these risk factors.

Anterior segment neovascularization may be the only presentation of ocular ischemic syndrome in some cases (Mendrinis et al., 2010). This

may be due to decrease in vascular perfusion resulting in hypoxia leading to ischemia and neovascularization (Kahn et al., 1986).

However, findings in retina such as hemorrhages and neovascularization are more common than findings in anterior segment. Narrowing of arteries and superficial retinal layer hemorrhages are characteristics of this condition. Microaneurysm may also be present which are centered around the macula or mid periphery (Chen and Miller, 2007). Cotton wool spots (nerve fiber ischemia), oedema of nerve fiber layer of optic disc and choroidal atrophy may occur due to choroidal vascular compromise (Mendrinós et al., 2010)

Fundus fluorescent angiography is an important and simple test aiding in diagnosis which reveals prolonged choroidal filling time as well as prolonged arteriovenous phase which is present in about 95 % cases but is not specific (Brown and Magargal, 1988). However, carotid Doppler is an important diagnostic method to study the structure of carotid arteries and its branches (Ho et al.; 1992).

So ophthalmologist should be aware regarding the presentation of this syndrome as it is not so common which can help in preventing ocular morbidities as well as life threatening complications.

Treatment includes panretinal photocoagulation (Malhotra and Gregory-Evans, 2000b) or cryotherapy to reduce the ongoing retinal ischemia but do not show any promising effect in the visual outcome. However, treatment of carotid stenosis by carotid endarterectomy (Dzierwa et al., 2011) or bypass surgery is the main modality of treatment to restore arterial perfusion and prevent visual as well as other life threatening conditions.

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