# Left Eye Central Retinal Artery Occlusion Secondary to Total Obliteration of the Left Internal Carotid Artery- A Rare Cause of a Devastating Ocular Emergency

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**Abstract:** A 48-year-old retired army personnel presented to us with the complaint of sudden onset painless loss of vision in left eye for three days. He also gave a history of severe migraine attacks mainly affecting left temporal area. He had no other co-morbidities, was a non-smoker and denied any history of head and neck trauma. Clinical examination revealed vision in the left eye was no perception to light with Marcus Gunn pupil. IOP was 18mmHg. Fundoscopy revealed a pale optic disc and retina, attenuation of retinal arteries and the typically described cherry red spot sign. Right eye examination was unremarkable, with 6/6 vision. Systemic examination revealed BP of 112/67mmHg, no cardiac murmurs and absent carotid bruit. Blood and imaging studies were performed to determine the underlying cause of CRAO. These include screening for diabetes, dyslipidemia, valvular or cardiac wall abnormalities, vasculitis, coagulopathies and carotid artery stenosis. The blood investigation results revealed that the patient was dyslipidemic. The carotid Doppler demonstrated a total occlusion of left internal carotid artery possibly due to undetected dyslipidemia. An urgent vascular referral was done, and CT carotid angiography confirmed a total obliteration of the left internal carotid artery at the bifurcation of the common carotid artery. Endarterectomy was not required and he was treated conservatively with aspirin and statins.
**Keywords:** central retinal artery occlusion, dyslipidemia, total obliteration of the left internal carotid artery.

## INTRODUCTION

Central retinal artery occlusion (CRAO) is an ophthalmic emergency and analogous to an acute stroke of the eye. It is a rare event with potentially devastating consequences to the patient in terms of systemic and ocular comorbidities. The incidence is estimated to be about 1 in 100,000 per year. The mean age of onset is about 60 years, with a range from the first to the ninth decade of life. Bilateral obstruction occurs in 1-2% of cases and were reported in the setting of Wegener's granulomatosis, temporal arteritis, homocystinuria, sickle cell disease, Henoch-Schonlein purpura, mitral valve prolapse, artherosclerosis and migraine [1].

The retinal artery can be occluded due to embolism, vascular obliteration (atherosclerotic plaques, giant cell arteritis and other types of vasculitis) and compression (a retrobulbar mass i. e hematoma), angiospasm, hemodynamic or hydrostatic arterial occlusion [2]. By far the most common cause of non-arteritic retinal artery occlusion is due to embolism.

## CASE REPORT

A 48-year-old retired army personnel presented to us with the complaint of sudden onset painless loss of vision in left eye for three days. He also gave a history of severe migraine attacks mainly affecting left temporal area. He had no other co-morbidities, was a non-smoker and denied any history of head and neck trauma.

Clinical examination revealed vision in the left eye was no perception to light with Marcus Gunn pupil. IOP was 18mmHg. Fundoscopy revealed a pale optic disc and retina, attenuation of retinal arteries and the typically described cherry red spot sign. Right eye examination was unremarkable, with 6/6 vision. Systemic examination revealed BP of 112/67mmHg, no cardiac murmurs and absent carotid bruit. A diagnosis of left central retinal artery occlusion (CRAO) was made and he was admitted to ward. While waiting for the admission, patient was asked to lie flat in bed, instructed to perform ocular massage and given IV Diamox 500mg STAT.

Blood and imaging studies were performed to determine the underlying cause of CRAO. These include screening for diabetes, dyslipidemia, valvular or cardiac wall abnormalities, vasculitis, coagulopathies and carotid artery stenosis. The blood investigation results revealed that the patient had deranged fasting serum lipids. The carotid Doppler ultrasonography demonstrated a total occlusion of left internal carotid artery (ICA) possibly due to undetected dyslipidemia. An urgent vascular referral was done, and CT carotid angiography confirmed a total obliteration of the left ICA at the bifurcation of the common carotid artery

[Figure 1, Figure 2 and Figure 3]. Endarterectomy was not required and he was treated conservatively with tablet Aspirin 75mg OD. Patient was also referred to physician for his underlying dyslipidemia and started on tablet Atorvastatin 20mg ON. There was no improvement in his left eye vision. Patient's fundus photo one year after the acute attack shows pale optic disc with attenuated vessel [Figure 4, Figure 5]
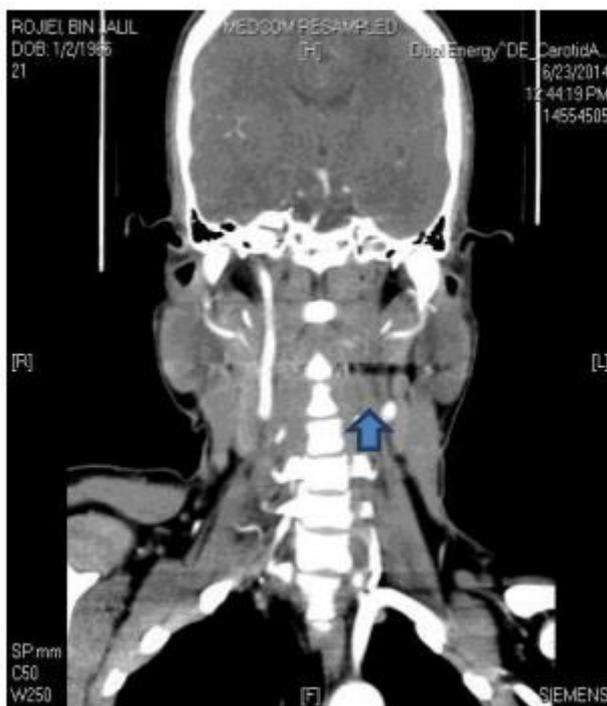


**Fig-1: coronal view: total obliteration of left internal carotid artery**



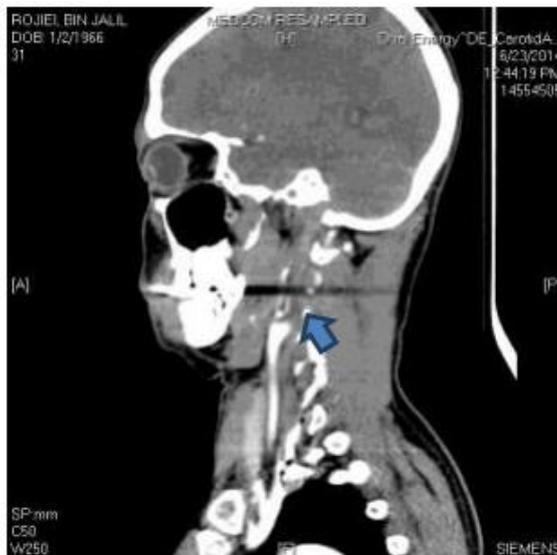**Fig-2: axial view : absence of left internal carotid artery**

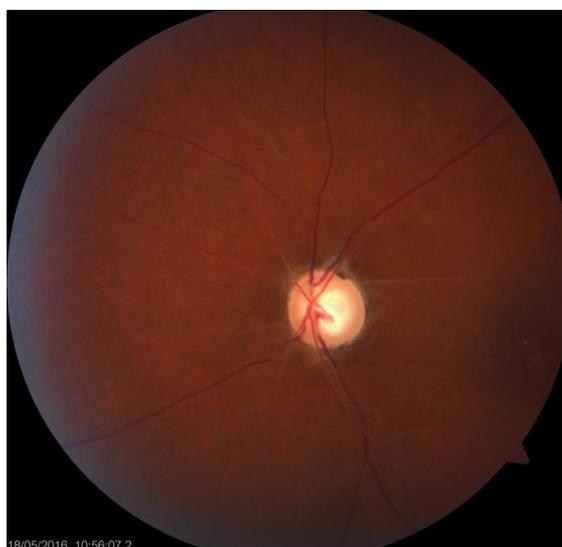**Fig-3: sagittal view: total obliteration of left internal carotid artery**



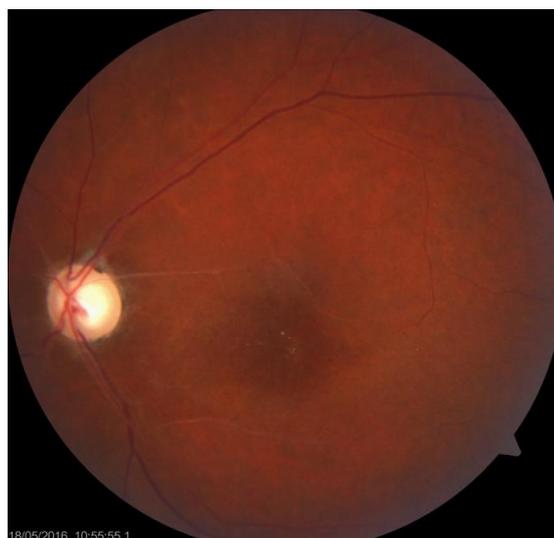**Fig-4: left eye fundus photo : pale optic disc with attenuated vessel**



**Fig-5: left eye fundus photo : pale optic disc with attenuated vessel**

## DISCUSSION

CRAO is the occlusion of central retinal artery (CRA) with resultant infarction of retina and vision loss. It was first described as an embolic occlusion of CRA in a patient with endocarditis by von Graefe in 1859 [3].

Patients with CRAO typically present with an acute painless loss of vision, and 80% of affected patients have a final visual acuity of counting fingers or worse [4, 5]. Visual loss occurs as a result of loss of blood supply to inner retinal layers. Approximately 15-30% of the population has a cilioretinal artery which is a branch of short posterior ciliary artery [6]. It supplies a part or the whole of the fovea, and in those eyes where there is a CRAO, the cilioretinal artery is spared and the visual acuity may be preserved at 20/50 or better, with loss of peripheral vision only.

Embolism is the most common cause of CRAO. The major source of embolism in the carotid artery is an atherosclerotic plaque (66%), whereas a significant (>50%) carotid artery stenosis accounts for only 30% of cases [7]. Statistically, Caucasians, when compared to African Americans have significantly different incidence of ICA stenosis, which is 41% and 3.4% for each group respectively [8]. To date, there was no data produced regarding incidence of CRAO in Asian population. A significant stenosis of the extracranial ICA is the most common identified condition associated with retinal and ocular ischemia. It represents the hemodynamic cause, and especially if associated with nocturnal arterial hypotension , can lead to transient CRAO [9].

In all, 74% of these emboli are shown to be made of cholesterol, 10.5% were calcific material, and 15.5% were fibrin [10] .Once the CRA is occluded, the ability of the retina to recover depends on whether the offending embolus or thrombus is dislodged, and also on the retinal tolerance time [11, 12].

In our case, the carotid Doppler revealed a total occlusion of left internal carotid artery and upon further investigation, CT carotid angiography confirmed a total obliteration of the left internal carotid artery at the bifurcation of the common carotid artery. There are various triggering factors for atherosclerosis and in our case, the patient was found to have dyslipidemia.

In patients diagnosed with CRAO, there are a few reported management options but none of which are of proven benefit. For example, the patient should lie flat, given a stat dose of acetazolamide (500mg) intravenously , and instructed to perform ocular massage. In the same setting, the ophthalmologist may perform anterior chamber paracentesis. In select few centres with appropriate neurological and ophthalmological support, it is possible to consider selective intra-arterial fibrinolytic therapy.

The aim of giving IV acetazolamide is to reduce the intraocular pressure. Ocular massage is performed to dilate the ophthalmic and retinal arteries to help in increasing the retinal perfusion pressure, hence limiting the damage to the retina. It is believed that ocular massage can increase the retinal artery flow by 180% as a result of the vasodilatation that occurs on release of digital pressure [13] and also may facilitate the disintegration of a thrombus, or dislodge an impacted embolus into a more peripheral part of retinal circulation. A small rise in ophthalmic artery pressure, which will influence retinal perfusion pressure in a favourable way, is achieved by lying the patient flat.

Selective intra arterial fibrinolytic therapy i.e. urokinase or tissue plasminogen activator (TPA) administration into the ophthalmic artery has shown considerable success. In a recent meta-analysis, one study has reported a final visual acuity of 6/12 or better in 27% of subjects after the injection compared with 18% to 21% for conventional approaches [14- 16].

Systemic management in cases of CRAO is aimed at reducing morbidity and mortality associated with predisposing factors. The most common cause of deaths in subjects who have suffered acute occlusive events of the retinal arteries is cardiovascular disease [17, 18]. Measures recommended in all cases include cessation of smoking, appropriate dietary advice, managing blood pressure and oral aspirin therapy. Associated disorders should be treated accordingly and specialist referral is indicated in the presence of systemic vasculitis, significant carotid artery stenosis, hypercoagulable states, valvular heart disease etc.

Ophthalmic follow up is essential because of possible retinal and iris neovascularization. The pathogenesis remains poorly understood but there is general consensus that chronic retinal ischemia plays an important aetiological factor [19]. The incidence of ocular neovascularization after CRAO lies between 16.6% and 18.75% , and the majority of these go on to develop rubeotic glaucoma (67.5% to 83.3 %) [20-22]. The vast majority occurs within three months of arterial obstruction [19]. Panretinal photocoagulation has been shown to reduce the risk of rubeotic glaucoma and close ophthalmic follow up is therefore recommended for a minimum period of three months after the occlusive events [20].

## CONCLUSION

Urgent identification of the underlying cause is important as the acute management of CRAO as it can potentially be life- and sight-threatening. Thus, in all cases, ophthalmological referral is indicated for acute management and follow up.

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