Case Report

Central Retinal Artery Occlusion Secondary To High Altitude Exposure

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Abstract

CRAO can be considered as an ocular analogue of stroke or an ocular equivalent of acute myocardial infarction. We present a case report of a 32-years-old serving soldier posted at high altitude area with no premorbidities who presented with symptoms of sudden painless loss of vision in the left eye (LE). Best-corrected visual acuity in the LE was PL+ with PR inaccurate . A relative afferent pupillary defect grade IV was observed in the LE. Ocular fundus examination of LE was suggestive of central retinal artery occlusion. Systemic evaluation revealed Steno occlusive disease of bilateral carotids L>R. Haematological investigations revealed increased haemoglobin. Raised haemoglobin due to long stay in high altitude area is tantamount to a sustained inflammatory state that results in endothelial dysfunction by causing hypercoagulable state culminating in small calibre vessel blockage.

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Introduction

Central retinal artery occlusion (CRAO) was first described by von Graefes in 1859.[1] It is analogous to an acute stroke of the eye and is an ophthalmic emergency. The incidence is estimated to be 1 in 100 000 people and accounts for 1 in 10 000 ophthalmological outpatient visits.[2] A prospective study of 260 eyes with CRAO showed that people suffer profound monocular visual loss, with 80% of patients having a visual acuity (VA) of 20/400 or worse.[3] This reduction in vision increases the fall risk and thus results in increased dependency, and in worst-case scenarios leads to institutional care.[4] CRAO signifies end-organ ischaemia and often the underlying atherosclerotic disease. It is the same underlying atherosclerotic risk factors that in turn place an individual at risk of future cerebral stroke and ischaemic heart disease.

Although analogous to a cerebral stroke, there is currently no guideline-endorsed evidence for treatment. Current options for therapy include the so-called 'standard' therapies, such as sublingual isosorbide dinitrate, systemic pentoxifylline or inhalation of a carbogen, hyperbaric oxygen, ocular massage, globe compression, intravenous acetazolamide and mannitol, anterior chamber paracentesis, and methylprednisolone. None of these therapies have been shown to be better than placebo.[5] There has been recent interest in the use of tissue plasminogen activator (tPA) with two recent randomized controlled trials on the treatment of acute CRAO.[6,7]

Causes of CRAO

1. Classical Non Arteritic CRAO (NA-CRAO) is most common due to permanent occlusion of the CRA, caused by an impacted embolus at the narrowest part of the CRA, where it enters the sheath of the optic nerve8 (not at the lamina cribrosa, as is often erroneously described). The emboli originate from plaques in the carotid artery or the heart rarely, CRAO is due to vasculitis, chronic systemic autoimmune diseases, or thrombophilia.[9]
2. Transient NA-CRAO is most often caused by a migrating embolus, and sometime by a transient marked fall of perfusion pressure in CRAO[10] or high rise of intraocular pressure.
3. Arteritic CRAO is due to thrombosis of the common trunk of the posterior ciliary artery and CRA arising from the ophthalmic artery[11] caused by giant cell arteritis, not of CRA per se.

Carotid artery disease can produce CRAO by the following three mechanisms

1. Embolism, which is by far the most common cause of CRAO[12]
2. A significant stenosis (about 70% or more) or complete occlusion of the internal carotid artery, by markedly reducing the ocular blood flow, can result in development of CRAO.[12] In a study >80% stenosis of the internal carotid artery was seen in 18% of CRAO cases.[13]
3. A study on atherosclerotic monkeys showed that serotonin, a powerful vasoconstrictor, released by platelet aggregation on atherosclerotic plaques in the carotid artery, produces a transient spasm, which can cause transient, complete occlusion, or impaired blood flow in the CRA[14]

Case Report

A 32 years-old-male serving soldier, posted at 18000 feet initially reported with sudden, acute, painless loss of vision in right eye since last 24 hours. There was no headache, vomiting, convulsion or any neurological deficit. He had no precommorbidities.

On general examination, he was conscious, oriented, afebrile with pulse rate of 110/ min, blood pressure of 130/70 mmHg. His peripheral pulsations were well felt, no carotid bruit was heard. He had all deep tendon reflexes brisk on examination. Rest of the general examination was unremarkable. He had no addiction. Other personal history was insignificant. He had no history of any medications or drugs. His family history was insignificant.

On ophthalmological examination

The best-corrected visual acuity (BCVA) was 20/20 in the right eye and light perception, projection of rays was inaccurate in the left eye. Right eye ophthalmologic examination was unremarkable, while the left eye showed a relative afferent pupillary defect gd IV. Dilated fundus ophthalmoscopy revealed the presence of severe arterial narrowing, retinal whitening in the macular region with loss of the physiological macular reflex. Cherry red spot, peripheral areas of retinal pigment epithelium hyperpigmentation was seen (Figure 1). Optical coherence tomography (OCT) showed normal vitreo macular interface ,increased and distorted foveal contour with increased CMT, normal IS-OS junction and normal RPE choroid interface, (Figures 3). CRAO was suspected. Fluorescein angiography (FA) performed, confirmed the diagnosis of CRAO revealing severe delay in the filling of the retinal arteries and a delayed arteriovenous transit time, areas of peripheral capillary nonperfusion, arteriovenous anastomoses, and cherry red spot (Figure 2)

Systemic Examination

Lab tests: CBC showed raised HB of 19g/dl(13-18), MCV 102fl(76-96), MCH 34.8pg(27-32), RDW-CV 15.6%(11.5-14.5). LFT, RFT, Lipid profile, RBS were within normal limits. ANA, ds DNA, and Anti phospholipid antibodies workup was negative & other autoimmune markers were also negative with an ESR of 20 mmhg at 1 hour. Also his ANCA panel was negative, including both myeloperoxidase and perinuclear antibodies. Coagulation profile including Protein C, Protein S and antithrombin III were normal.

ECHO showed EF of 65%, no vegetative growth, clot, PAH. Normal LV function.

CT Cerebral angiography (Figure 4,5) showed calcified plaques bilateral ICA with 30% stenosis on the right and 50% stenosis on the left.

Carotid doppler study showed steno-occlusive disease bilateral carotids(L>R). In right carotid a hypoechoic plaque

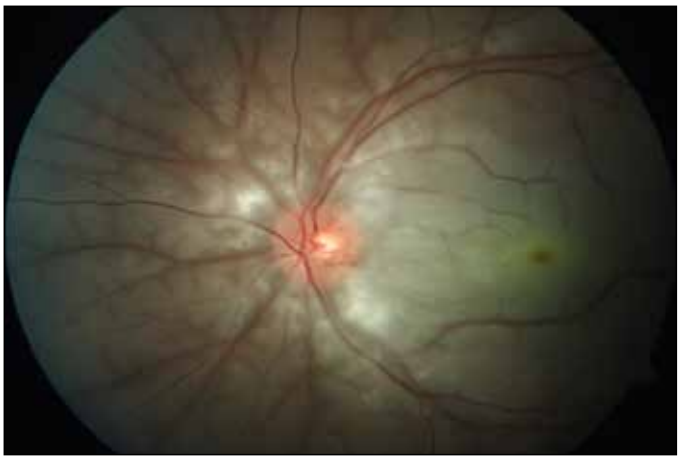

Figure 1: *Fundus photography of the left eye showing the presence of a pale optic disc, diffuse arterial narrowing, a mild 'cherry-red spot' macula and peripheral areas of retinal pigmented epithelium hyperpigmentation.*

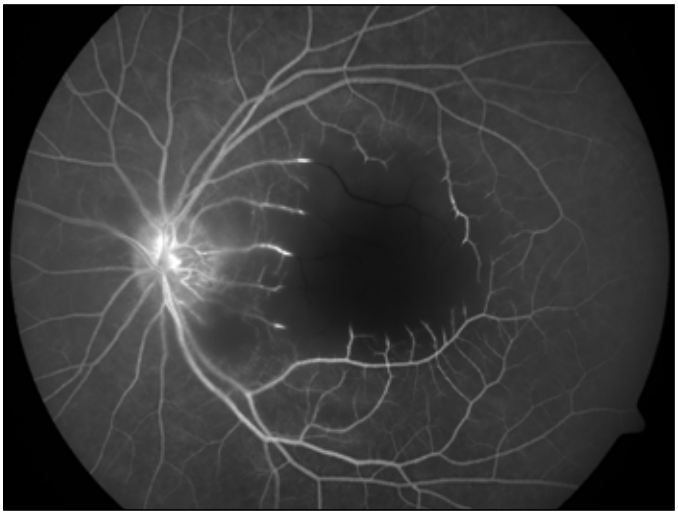

Figure 2: *Fluorescein angiography of the left eye, showing a delayed arrival of the dye in the eye 43 s after the injection , peripheral areas of capillary nonperfusion, arterial narrowing, CRAO, central retinal artery occlusion.*

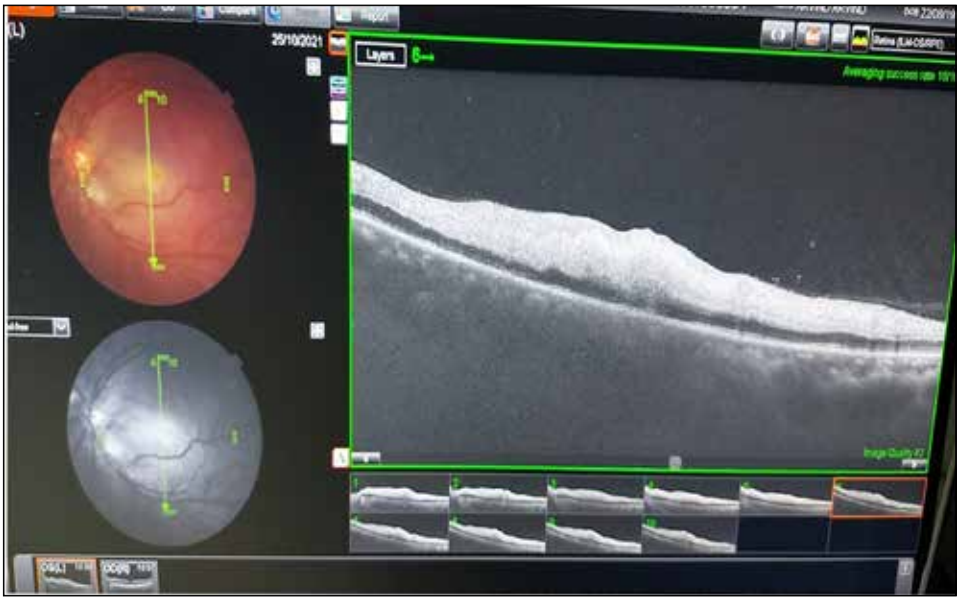

Figure 3: *OCT scan of left eye shows thickening of retinal layers and distortion of foveal contour.*

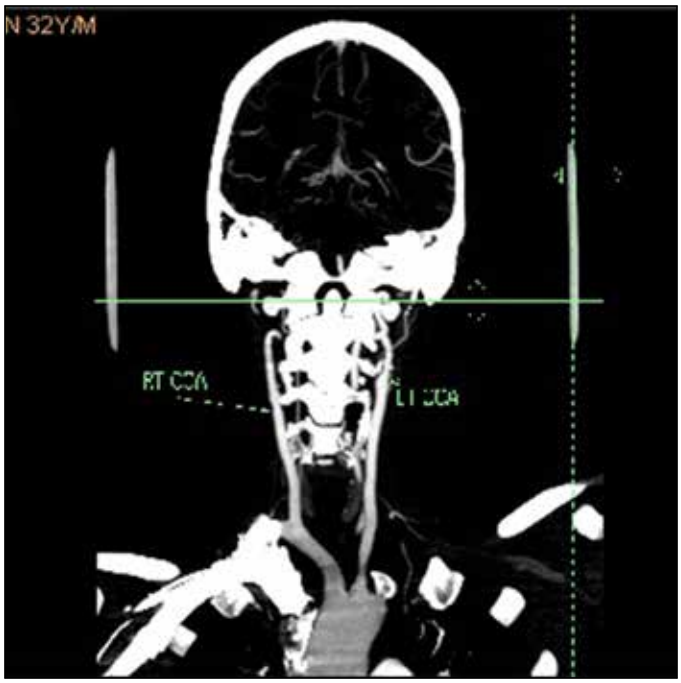

Figure 4: *Cerebral CT Angiography shows a hypoechoic plaque causing 50% occlusion of R CCA*

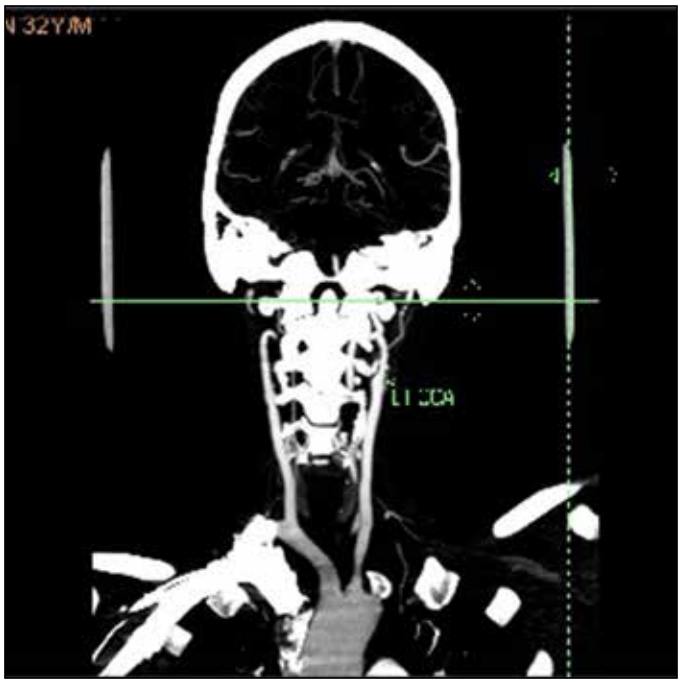

Figure 5: *Cerebral CT Angiography shows a hypoechoic plaque causing 50% occlusion of L CCA*

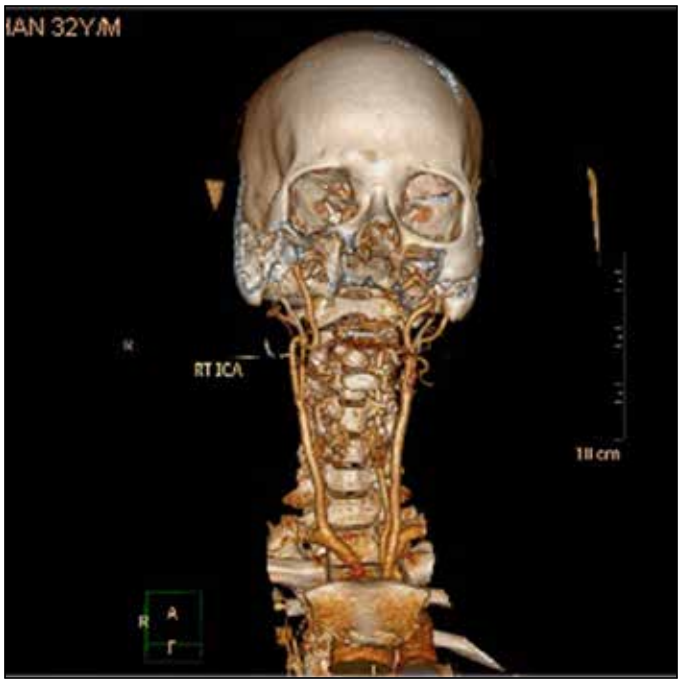

Figure 6: *3D Recon of Cerebral CT Angio showing stenosis of R ICA.*

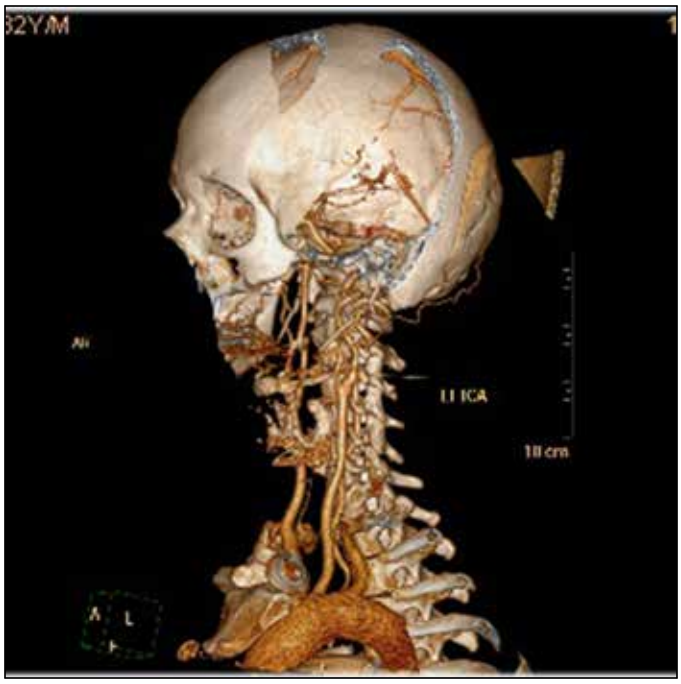

Figure 7: *3D Recon of Cerebral CT Angio showing stenosis of L ICA.*

causing 50% occlusion of lumen of CCA and extending till ICA. In left carotid there was a hypoechoic plaque causing 50-69% occlusion of lumen in left CCA, bulb and extending into left ICA. On colour doppler imaging and spectral waveform evaluation there was increase in velocity in post stenotic segments. The spectral waveform was normal.

Management

Medical review was done. Patient was started on anti-coagulants, anti platelets, statins.

Discussion

Currently, there are only a few reports on retinal artery occlusion secondary to high-altitude exposure. A case of central retinal artery occlusion secondary to bilateral buried optic nerve drusen at high altitude was reported in 1995,[15] while another case of central retinal artery occlusion caused by the expansion of intraocular gas during mountain travel at high altitude was reported in 2002.[16] A recent report has shown that cilioretinal artery occlusion and related central retinal vein occlusion occurred as a complication following high-altitude exposure.[17] It has been speculated that the

reason for a few reports of retinal artery occlusion secondary to high-altitude exposure might be that some clinicians do not suspect retinal vascular occlusion, thus do not inquire the patient's travel history or consider high altitude as a risk factor. Possible reasons for retinal artery occlusion secondary to high-altitude exposure may be related to hematocrit, hemoglobin concentration, and blood viscosity that were all increased in the hematologic examination of high-altitude climbers, which indicated the higher coagulative activity, as the present case shown.[18] The patient was in a state of hypercoagulability. The atmospheric pressure decreases along with the increase of the altitude, after which the retinal arteries and veins tend to dilate. Retinal vascular occlusion in patients with circulatory impairment has been shown to be triggered by reactive vasoconstriction, which may occur during the descent.[19] Furthermore, hypoxia has an important role in the development of retinal artery occlusion. Hypobaric hypoxia caused thrombosis, which further decreased the oxygen transport capacity.

There has been a great interest and controversy in its management ever since CRAO has been known. As with most instances of ischemia, the colloquialism "time is vision" may apply here as well. Best outcomes likely result when applied within the first eight hours from the onset of visual impairment.[21] Usually, one or a combination of the following conventional modes of treatments have been advocated in acute CRAO and have claimed success.

These include

(i) ocular massage, in an effort to dislodge the embolus in the CRA;
(ii) a reduction of IOP by paracentesis, massage of the eyeball, administration of acetazolamide, and so on to improve blood flow;
(iii) vasodilation of the CRA;
(iv) inhalations of 95% oxygen and 5% carbon dioxide;
(v) rebreathing of expired CO2 in a bag; and
(vi) retrobulbar vasodilators.

Except for ocular massage, which occasionally dislodges the embolus, there is no evidence that the rest show any significant benefit.[22]

Conclusion

Prompt recognition of CRAO symptoms should be followed by a detailed neurologic and vascular evaluation for concurrent stroke and carotid artery stenosis or occlusion in patients who are working, or travelled to high altitude. These patients are also at risk for ischemic events after treatment as prolonged stay in high altitude areas is tantamount to a sustained inflammatory state that results in endothelial dysfunction by causing hypercoagulable state. This case highlights the high risk to these patients, which requires intensive workup and admission from the emergency room and promt treatment, both ocular and systemic.

Abbrevations

CRAO	: Central Retinal Artery Occlusion
PL	: Perception of Light
PR	: Projection of light
CRA	: Central Retinal Artery
CMT	: Central macular thickness
IS-OS	: Inner space-outer space junction

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