Introduction
Giant cell arteritis (GCA) is a systemic vasculitis with a myriad of ocular manifestations such as Anterior Ischaemic Optic Neuropathy (AION), Central Retinal Artery Occlusion (CRAO), choroidal infarction, occipital lobe infarction and ocular motor nerve palsies. The predominant systemic features include fever, polymyalgia rheumatica, jaw claudication, weight loss and fatigue. Occult GCA accounts for a minority of cases presenting without systemic symptoms. It is a potential cause of bilateral blindness unless a high index of suspicion is contemplated. A 60 year old female presented with CRAO with no systemic symptoms of GCA. Cardiovascular examination, electrocardiography, echocardiography and carotid doppler were unremarkable. Erythrocyte Sedimentation Rate (ESR), C Reactive Protein (CRP) and platelets were normal. Bilateral prominent temporal arteries were noted but were non tender with subtle pulsations, leading to a suspicion of GCA. Temporal artery biopsy disclosed transmural granulomatous inflammation and fragmentation of internal elastic lamina, ascertaining the diagnosis. Systemic steroids were administered promptly. Although an unusual etiology of CRAO, GCA must be ruled out with utmost priority in patients older than 50 years. The diagnosis can be elusive in occult GCA and CRAO may be the inceptive manifestation, further compounding the diagnostic enigma. Urgent temporal artery biopsy is warranted in occult cases of GCA and substantiates the diagnosis in dubious cases. Occult GCA constitutes an ophthalmic emergency and the imperative use of urgent systemic steroid therapy to prevent visual loss in the other eye cannot be overemphasized.

Case report
A 60 year old female presented with sudden painless loss of vision in left eye one week back. She had consulted elsewhere and was diagnosed as Central Retinal Artery Occlusion (CRAO). There was no history of amaurosis preceding the visual loss. There was no significant history of cardiac illness or autoimmune disease. There was no headache, scalp tenderness, jaw claudication or systemic symptoms of GCA. On presentation, visual acuity in left eye was 1/60. Relative afferent pupillary defect was noted. Fundus examination of left eye revealed mild optic disc edema, attenuated arterioles, cattle trucking in retinal veins and cherry red spot at the macula. An isolated cotton wool spot was present at the bifurcation of retinal vein at supero-nasal quadrant. There was no visible emboli (Figure 1). Examination of right eye was unremarkable. Carotid pulsation was equal on both sides. Cardiovascular examination, electrocardiography, echocardiography and carotid doppler were unremarkable. Erythrocyte Sedimentation Rate (ESR), C Reactive Protein (CRP) and platelets were normal. Bilateral prominent temporal arteries were noted but were non tender with subtle pulsations, leading to a suspicion of GCA (Figure 2). Biopsy of the left temporal artery was performed to confirm the diagnosis, so as to justify the long term steroid use in the elderly patient. High dose systemic steroids with an initial dose of intravenous Methyl prednisolone 1 g per day for three days,

Figure 1: Fundus picture of left eye showing Central Retinal Artery Occlusion (CRAO)
followed by oral prednisolone in a dose of 1.5 mg per Kg was administered promptly. Temporal artery biopsy (3 cm specimen) disclosed segmental inflammation of vessel wall and a fibrointimal plug with collection of lymphocytes. There was disruption of internal elastic lamina and transmural inflammation. Prominent calcification in the media was observed with occasional giant cells, leading to the diagnosis of giant cell arteritis (Figure 3-5). Systemic steroid therapy was continued with gradual weekly tapering. The visual acuity remained unchanged on follow up examination after four weeks, and fundus showed optic disc pallor.

**Discussion**

The diagnostic criteria of GCA requires age more than 50 years, recent onset of temporal headache, tenderness and reduced pulsation in temporal artery, jaw claudication, ESR exceeding 50 mm/hr and typical histologic findings (granulomatous inflammation) on temporal artery biopsy. Elevated CRP and systemic symptoms like fever, weight loss and fatigue may be associated in many cases. Ophthalmic complications have been reported in approximately 40-50% of patients with GCA. The visual loss secondary to GCA is almost universally irreversible. It is widely believed that patients with GCA present with systemic features including fever, headache, jaw claudication and that the absence of such manifestation rules out GCA. This unfortunate impression can be a harbinger of a misdiagnosis and consequent blindness. 20% cases of GCA are occult and are characterised by absence of systemic features. The characteristic clinical sign of a tense, tender, inflamed, nodular and non pulsatile temporal artery may be absent in a small proportion of cases and a vigilant approach must be exercised to circumvent a missed diagnosis. The aetiology of Central Retinal Artery Occlusion (CRAO) may be multifactorial. Although an unusual etiology of CRAO, GCA must be ruled out with utmost priority in patients older than 50 years. GCA can be a devastating disease when not addressed immediately. It is a life threatening condition which can lead to irreversible blindness in the affected eye, or subsequently in both eyes. Laboratory markers like ESR and CRP may offer corroborative evidence in the diagnosis of GCA but are generally nonspecific. Though high levels of ESR are generally obtained in GCA, 20% biopsy-proven GCA patients have been reported to present with normal ESR. CRP generally parallels ESR and may be of value when the ESR is equivocal. Hence, normal ESR and CRP does not exclude GCA in this patient.

Biopsy provides a definitive diagnosis but occasional false negative results are obtained due to ‘skip lesions’. The specificity is considered to be 100% when a giant cell is identified. Two histopathological patterns are considered diagnostic for GCA which include active arteritis and healed arteritis. In active arteritis, there is transmural thickening of the vessel wall with a preponderance of lymphocytes, macrophages and giant cells. Healed arteritis shows fibrosis of vessel wall with disruption of internal elastic lamina. The histopathological picture of the patient discussed here
Figure 5: Photomicrograph showing segmental inflammation of vessel wall and fibrointimal plug with lymphohistiocytic infiltrates, H&E 40x

revealed aforementioned features of both active and healed arteritis.

Occult GCA should always be considered as a differential in a patient above 50 years presenting with visual loss due to anterior ischemic optic neuropathy, central retinal artery occlusion and cilio-retinal artery occlusion. ESR and CRP must be performed on an emergency basis, and the diagnosis should not be disregarded inspite of normal values. A high index of suspicion of GCA should be entertained in elderly patients with CRAO, in absence of obvious contributory cause for the vascular occlusion. The authentication of a precarious diagnosis of GCA mandates a temporal artery biopsy, which substantiates the diagnosis in uncertain cases. Therapy should be commenced immediately with high doses of systemic corticosteroids.

Conclusion

The diagnosis can be elusive in occult GCA. GCA should be considered in the differential diagnosis of CRAO in an elderly patient. CRAO may be the inceptive manifestation of occult GCA, further compounding the diagnostic enigma. Occult GCA constitutes an ophthalmic emergency. The imperative nature of urgent temporal artery biopsy and systemic steroid therapy to prevent visual loss in the other eye cannot be overemphasized.

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