CASE REPORT

Simultaneous Appearance of Arteritic Anterior Ischemic Optic Neuropathy and Central Retinal Artery Occlusion in Giant Cell Arteritis

Say Ting Ooi[1,2], Nor Anita Che Omar[2], Azhany Yaakub[1]

[1] Department of Ophthalmology and Visual Sciences, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia
[2] Ophthalmology Clinic, Hospital Sultanah Nur Zahirah, Jalan Sultan Mahmud, 20400 Kuala Terengganu, Terengganu, Kementerian Kesihatan Malaysia

ABSTRACT

Giant cell arteritis (GCA) is uncommon among Asian population. It is frequently associated with sight threatening complications. Simultaneous bilateral ocular involvement with different pathology is uncommon. We would like to highlight a rare case of GCA that was presented with transient visual loss over the right eye with simultaneous onset of central retinal artery occlusion as well as arteritic anterior ischemic optic neuropathy in both eyes. High dose intravenous methylprednisolone then subsequently maintenance dose of oral steroid and oral aspirin were given. His visual acuity remained the same after treatment. Early diagnosis and treatment of GCA is crucial. Visual outcome can be devastating if treatment is delayed.
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Corresponding Author:
Azhany Yaakub, MMed (Ophthalmology)
Email: azhany@usm.my
Tel: +60199101993

INTRODUCTION

Giant cell arteritis (GCA) refers to an autoimmune disorder. It causes granulomatous inflammatory vasculitis of medium and large arteries. GCA is 20 times more commonly occurs in Caucasians as compared to Asians (1).

GCA may present with a wide variety of ophthalmic, neurologic and systemic complications. Except for occult GCA, systemic and cranial symptoms often precede ophthalmic symptoms, with a headache being the most common presenting symptom (1). Up to 50% of the patients have ophthalmic involvement along the course of the disease. The ocular presentations in GCA are usually attributed to arteritic anterior ischaemic optic neuropathy (AAION) and less commonly, central retinal artery occlusion (CRAO) and posterior ischaemic optic neuropathy (PION) (2-4).

A simultaneous presentation of the different ocular manifestations in bilateral eye is rare. Here we describe a devastating presentation of bilateral giant cell arteritis with simultaneous right eye CRAO and left eye AAION.

CASE REPORT

A 70-year-old Malay man, without any medical problem, experienced simultaneous bilateral visual loss. Premorbidly, his right eye was pseudophakic and had good visual acuity in both eyes. The symptoms started with transient monocular visual loss (TMVL) over left eye. He also experienced jaw claudication, with acute bilateral, frontal and temporal headache, and a "nagging sensation" over his scalp. A week later, he developed sudden and simultaneous bilateral eye blindness.

On examination, there was No-Perception-to-Light in both eyes, both pupils were not reactive to light. On fundus examination, right eye demonstrated acute signs of CRAO with pale posterior pole and "cherry red spot", vessels were attenuated and boxcarring (Fig. 1). Later, the retina re-perfused and optic disc became palish. Left eye demonstrated signs of AAION with initially swollen optic disc and palish especially at inferior disc, which then turned chalky white disc swelling with disc haemorrhage (Fig. 2). Later, the optic disc became palish with large cupping. Bilateral temporal scalps were tender. Temporal arteries on both sides were engorged

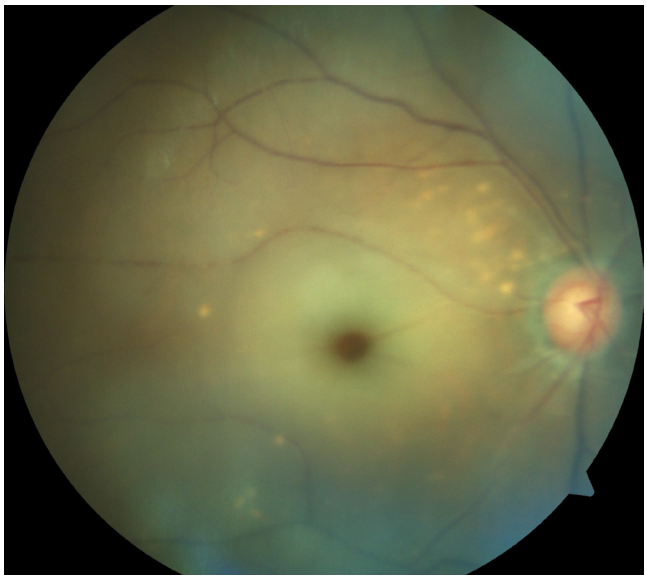

Fig.1: Right eye CRAO, cherry red spot with attenuated and boxcarring vessels.

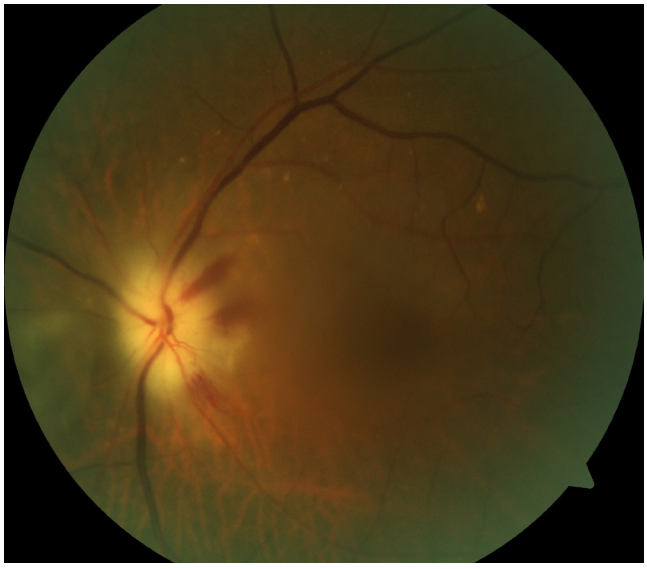

Fig. 2: Left eye AAION, chalky white optic disc swelling with disc haemorrhage.

(Fig. 3). There was an increase in patient's erythrocyte sedimentation rate (ESR) (86). Reading of C-reactive protein (CRP) was significantly elevated too (182.23). Using American College of Rheumatology GCA criteria, GCA was diagnosed clinically, and treatment initiated. Patient was immediately given high dose of intravenous methylprednisolone 250mg QID and aspirin orally. Biopsy of temporal artery showed thickened tunica intima and focal disruption of elastic lamina (Fig. 4A). The vascular wall was infiltrated with inflammatory cells, predominantly lymphocytes (Fig. 4B). No giant cell was seen. These features are in favour of arteritis.

Three days after high dose of methylprednisolone, his headache and jaw claudication were resolved, but there was no improvement in his vision. He was subsequently placed on maintenance dose of oral prednisolone which was slowly tapered. During outpatient follow up, he was

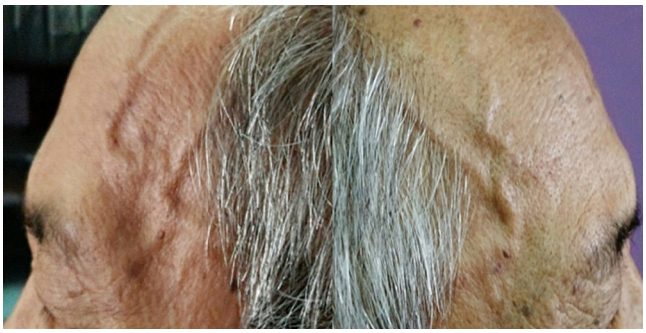

Fig.3: Bilateral prominent and engorged temporal artery.

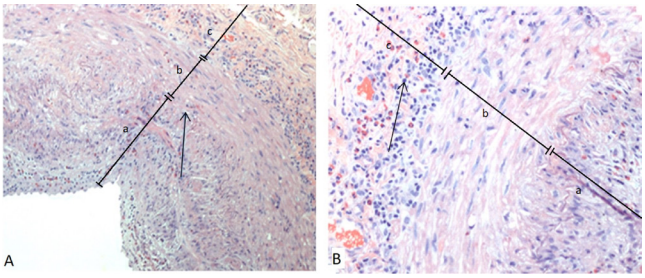

Fig.4: (A) Histopathological examination of temporal artery showed thickened tunica intima with dense infiltrates, fragmentation of internal elastic lamina within vascular wall (arrow). a= tunica intima, b= tunica media, c= tunica adventitia H&E, x4 magnification. (B) Inflammatory cells with lymphocyte predominant (arrow), no giant cell seen. a= tunica intima, b= tunica media, c= tunica adventitia H&E, x20 magnification.

asymptomatic, ESR and CRP levels decreased, however, his visual acuity remained the same.

DISCUSSION

GCA is an uncommon vasculitis disorder. American College of Rheumatology stipulates that to distinguish from other types of vasculitis disorder, GCA must meet at least three of the five criteria listed below: age ≥50 years old, recent headache or a new type of headache, abnormalities of temporal artery such as pain and reduced pulsation, ESR of ≥50 and temporal artery biopsy demonstrate inflammatory changes with mononuclear cells predominance or giant cells detected. Fulfilments of 3 over 5 points yield high sensitivity (93.5%) and specificity (91.2%). This case fulfilled 4 out of 5 criteria in which patient is in the correct age group, new onset headache, temporal artery abnormalities and raised ESR. Although it is crucial to have temporal artery biopsy in all GCA suspected case, treatment should be initiated immediately before the biopsy as this is a potentially blinding ocular emergency (2). There is also evidence which suggested that histopathology of GCA still can be detected up to 1 week after initiation of corticosteroid (3).

A sudden complete blindness of both eyes simultaneously in GCA is uncommon. This condition is even rarer if it involves a different pathology of AAION and CRAO bilaterally (2). There is limited data-to-date that elaborates on such incident. Some studies showed

that bilateral loss of vision in GCA is mostly attribute to a combination of any arteritic condition (AAION, CRAO or PION) that occur in sequence (4). There was a case reported on combined left CRAO and bilateral AAION in GCA, however bilateral eye presenting symptoms was not simultaneous with gapping of 4 days in between eyes. Fundus examination showed bilateral pale disc and left eye cherry red spot, no documented on optic disc edema. This may suggest an older AAION changes (5). In our case report, simultaneous onset of AAION and CRAO in both eyes was based on patient's complaints, evidence of acute appearance of fundus findings at presentation and the evolving changes during follow up. During presentation, his right eye showed acute sign of CRAO which is pale posterior pole, cherry red spot and boxcarring vessels, whereas his left eye showed acute sign of AAION which is chalky white swollen disc and disc haemorrhage. During his follow-up, his right eye's retina re-perfused and the optic disc became palish; his left eye's optic disc became palish with enlarged disc cupping. Adequate treatment with corticosteroid must be initiated immediately even prior to onset of ocular symptoms to prevent subsequent devastating ocular ischemic events.

TMVL or amaurosis fugax can be caused by various conditions such as transient ischemic attack from atherosclerosis, cardiac emboli, migraine attack, hypercoagulability disease and also GCA. The pathogenesis of TMVL is due to transient ischemic process of the retina, choroidal or optic nerve. TMVL occurs in up to 30% of GCA patients prior to visual loss (2,4). When TMVL is presented in GCA, it is an important warning sign of impending permanent vision loss (2-4). Recognition of this warning sign and early treatment is crucial to prevent blindness (2,3).

The visual prognosis in GCA is usually poor. Induction of pulse, high dose of intravenous methylprednisolone (15mg/kg/day) for 3-5 days is recommended for visual threatening patients. Several studies indicated that initiation of methylprednisolone prevents further deterioration of visual acuity and minimizes the risk of fellow eye involvement (2-4). However, there is no evidence to show superiority of intravenous methylprednisolone over oral prednisolone on improvement of visual acuity (2,4). Despite lack of evidence, intravenous methylprednisolone is still the choice of treatment in visual threatening condition of giant cell arteritis as it provides a higher bioavailability (2,4). With regards to our patient, his vision remained unchanged despite being started on the same regime of steroid therapy. We suspect that the pathological process was already underway when he came for medical treatment. Hence, we emphasized on the importance of early recognition of GCA and early treatment with high dose intravenous methylprednisolone.

There are no absolute guidelines that exist, in terms of length of treatment of corticosteroids for GCA. However, it may be reasonable to taper oral corticosteroid slowly and maintain the patient on long term oral corticosteroid. This is to prevent future relapse of disease activity and also progression to blindness or other life-threatening complications like strokes, myocardial infarction and aortic aneurysm (2,3). Patient's symptoms, ESR, CRP and repeat temporal artery biopsy are good indicators to monitor the disease progression.

CONCLUSION

GCA is an ophthalmic emergency, ocular involvement has a devastating visual outcome. Bilateral ocular involvement in GCA usually occurs in sequences. This case demonstrates a rare incidence of concurrent bilateral eye involvement with different pathology (AAION and CRAO) in GCA. As described in this case, clinicians should also be aware that TMVL is an essential indicator of impending visual loss in GCA, which necessitates prompt treatment. Even though GCA is rare among the Asian population, prompt recognition, diagnosis and immediate treatment initiation are crucial to prevent blindness and further complications.

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