

Giant Cell Arteritis with Panocular Involvement in an Indian Male

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Summary

A case of giant cell arteritis with systemic and panocular involvement is reported here. This elderly Indian male presented with symptoms of unilateral temporal headache and intermittent jaw claudication for a month followed by diplopia and blurring of vision and later loss of vision in the right eye. The right eye showed some limitation of ocular movements, presence of relative afferent pupil defect, anterior segment ischaemic changes and anterior ischaemic optic neuropathy. Visual evoked potential showed an absent P1 wave while the left eye with normal 6/6 vision showed a prolonged P1 wave. Fundus fluorescein angiography showed delay in choroidal perfusion. His erythrocyte sedimentation rate (ESR) was 120mm/hr and he was started on oral prednisolone. Superficial temporal artery biopsy obtained one week after starting steroids was positive for giant cell arteritis. Steroids led to the resolution of optic disc swelling, disappearance of anterior segment signs, full recovery of right ocular movements and no further deterioration of the fellow eye. On steroids, he developed insomnia and progressive myopathy which resolved and is now symptom free at lower doses of steroids.

Key Words: GCA, Relative afferent pupil defect, Anterior ischaemic optic neuropathy

Introduction

Giant cell arteritis (GCA) is an idiopathic systemic vasculitis affecting the large and medium sized arteries. Arteries commonly involved are the superficial temporal, occipital, ophthalmic and posterior ciliary arteries. It typically affects the elderly, is more common in whites than non-whites¹ and is more common in women than men. Reports of GCA are rare in this region. This is the first case report in Malaysia. Singapore reported its first case early this year².

Case Report

A 75 year old Indian man presented with a history of blurring of vision, intermittent diplopia, photopsiae and pain in the right eye for one week followed by sudden loss of vision in the same eye. He also gave a history of right temporal headache and pain in the right jaw while chewing, for one month. There was no scalp tenderness or pain on combing his hair. There were no constitutional symptoms and no history suggestive of polymyalgia rheumatica. On general examination he appeared well. His blood

This article was accepted: 28 August 2002

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CASE REPORT

pressure was 150/90 mm Hg. The right superficial temporal vessels were thickened, palpable and non-pulsatile. The muscle power and reflexes were normal. Ocular examination revealed a right pale swollen optic disc with multiple peripapillary cotton wool spots (Figure 1) and vision of no perception of light. The right eye also had anterior segment ischaemic changes with endothelial pigment deposits and keratic precipitates and Descemet's folds due to hypotony. The right intraocular pressure (IOP) was 8mm Hg while the left IOP was 14 mm Hg. There was some limitation of abduction, upgaze, downgaze and intortion of the right eye.

The left eye involvement was detected from the presence of a prolonged P1 wave on visual evoked potential which was absent in the right (Figure 2) and some delay in choroidal perfusion which was obvious in the right on fundus fluoresceine angiography. Otherwise the left vision was 6/6 with a normal looking anterior and posterior segments on examination. His ESR was

120 mm 1st hour. The patient was put on oral prednisolone 1.2mg/kg /day. The dose was titrated against disease activity and ESR and was gradually tapered. The superficial temporal artery biopsy taken 1 week after steroids treatment showed characteristic inflammatory changes. (Figure 3 and 4). The patient responded well to steroids. There was relief of headache, jaw claudication and eye pain. The multiple cotton wool spots and optic disc swelling in the right eye resolved and the disc became pale and cupped. The right intraocular pressure returned to normal with disappearance of the Descemet's folds and anterior chamber activity. There was full recovery of the right ocular movements. The left eye maintained good vision and no further change in visual evoked potential findings. The patient developed insomnia and myopathy while on high dose steroids. These subsided with lower doses and patient is now on 10 mg /day and is symptom free several months after first presentation. His ESR at eight months after presentation is 30 mm / hour.

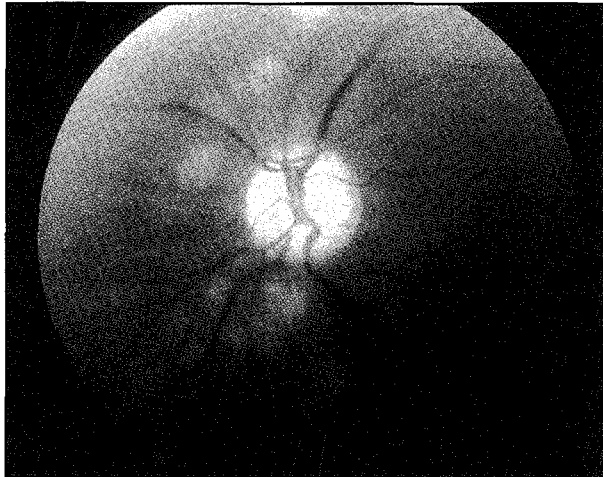


Fig. 1: Right pale optic disc with multiple cotton- wool spots

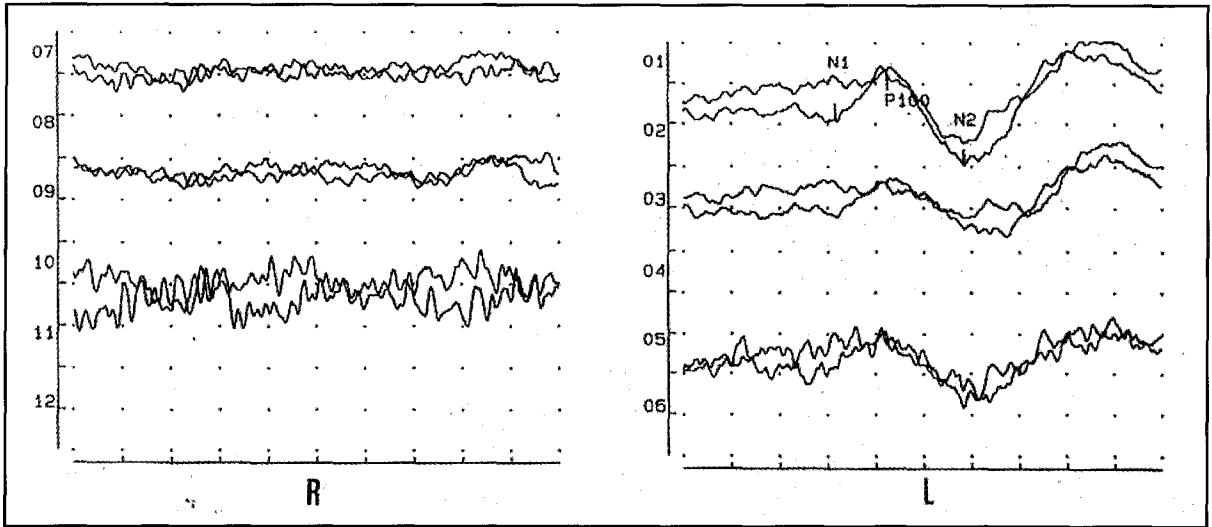


Fig. 2: Visual evoked potentials showing absent P wave in the right eye (R) and prolonged P wave in the left (L).

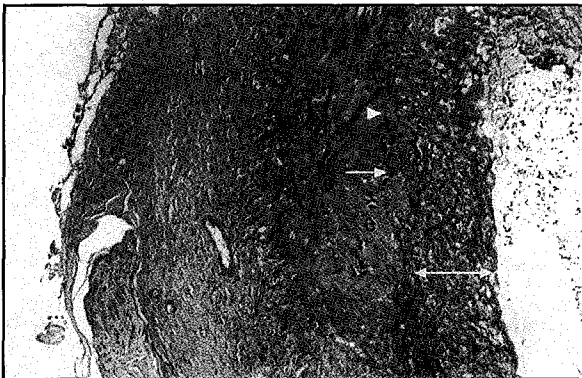


Fig. 3a: Photomicrograph of right temporal artery showing thickening of the intima (), and fragmentation () and partial loss of elastic lamina (). Elastic Van Genson (x100).

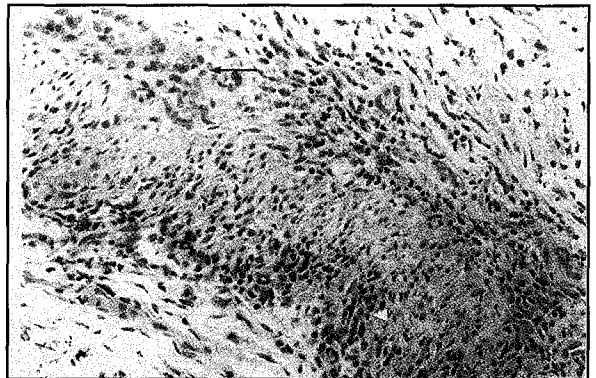


Fig. 3b: Infiltration of vessel wall by plasma cells and lymphocytes (). A cluster of epithelioid cells is also present (). (H&E x150)

Discussion

The ocular manifestations of GCA are varied and include eye pain, anterior segment ischaemia, anterior ischaemic optic neuropathy and ocular motility involvement. Anterior segment ischaemia is rare in GCA and is usually part of the picture of panocular ischaemia. Hayreh noted only one case of anterior segment ischaemia with neovascular glaucoma in his series of 170 biopsy proven patients³. Ocular motility involvement is also rare (2-15%)⁴ and is thought to be due to ischaemia involving the nerves or muscles.

Anterior ischaemic optic neuropathy is the most common ocular manifestation of GCA and cotton-wool spots are an early finding. It is usually unilateral but can be bilateral when one eye is usually more severe than the other as in this case. There is partial, or complete and permanent loss of vision. Blindness can be prevented by immediate treatment with corticosteroids on high

index of suspicion. Biopsy picture of mononuclear cells or granulomatous inflammation can be influenced by steroid therapy but has been shown to be positive even after some time has lapsed steroid treatment has started⁵ as in this case.

A high index of suspicion of giant cell arteritis even in a non-white male with an unusual presentation is necessary to enable early diagnosis. Prompt treatment is effective for some recovery and prevention of progression. The potential side effects of steroids must be borne in mind.

Acknowledgement

We are grateful to Professor L. M. Looi, Pathology Department, University Malaya Medical Center, Kuala Lumpur for the histology slides.

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