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Giant Cell Arteritis Causing Bilateral Sequential Anterior Ischaemic Optic Neuropathy – A Case Report

K Y Goh, T H Lim

ABSTRACT

Giant cell arteritis is a chronic granulomatous inflammation of unknown aetiology involving large and medium size arteries in the elderly. It causes acute visual loss from ischaemia to the optic nerves or central retinal artery occlusion. This is a rare cause of anterior ischaemic optic neuropathy in our local population. We present a patient who had bilateral loss of vision from sequential arteritic ischaemic optic neuropathy. She was treated with intravenous steroids immediately. Diagnosis was based on histopathological studies of temporal artery biopsies.

Keywords: arteritic ischaemic optic neuropathy, biopsy, giant cell arteritis, intravenous steroid, temporal artery

INTRODUCTION

Giant cell arteritis (GCA) is a chronic granulomatous inflammation of the large and medium size arteries, with a predilection for the superficial temporal, ophthalmic, posterior ciliary and vertebral arteries, in the elderly population. Visual loss from GCA in the elderly is an ocular emergency that is potentially reversible with steroids that also prevents involvement of the fellow eye. Histological studies of biopsied tissue from the temporal artery help in confirming the diagnosis.

Case Report

Mdm T S, an 83-year-old Chinese female, presented with rapid painless loss of vision in the left eye followed by the right eye two weeks later. She had vague symptoms of perioral numbness, loss of appetite and dysphagia two months prior to her loss of vision. There was no scalp tenderness, chronic headaches, jaw claudication nor loss of weight.

On examination, she had no light perception in both eyes. Both pupils were amaurotic and eye movements were full. Both optic discs were also swollen. Both superficial temporal arteries were cord-like but non-tender (Fig 1). Full blood counts showed hypochromic, microcytic anaemia. The erythrocyte sedimentation rate was 92 mm/h. Upon admission, she was started on intravenous methylprednisolone 250 mg, 8 hourly. A right temporal artery biopsy was performed under local anaesthesia. Histopathological studies showed infiltration by lymphocytes, macrophages and neutrophils associated with intimal fibrosis (Fig 2). Several multinucleated giant cells were also seen with fragmentation of the elastic lamella (Fig 3). There were neither foci of medial necrosis nor thrombus formation, which are consistent with giant cell arteritis.

The patient's vision improved transiently to counting fingers closely but subsequently deteriorated to no perception of light. She was discharged with 60 mg of oral prednisolone for a week followed by a slow taper over one month.

Although high dosage oral corticosteroids may have serious side effects in the elderly, it is the only available remedy to reverse visual loss. However, in this case, the patient's visual outcome remained eventually unchanged.

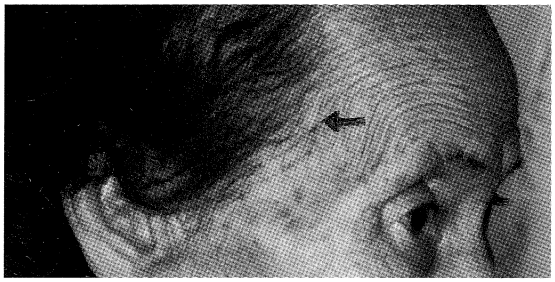

Fig 1 – Thickened right superficial temporal artery (arrow).

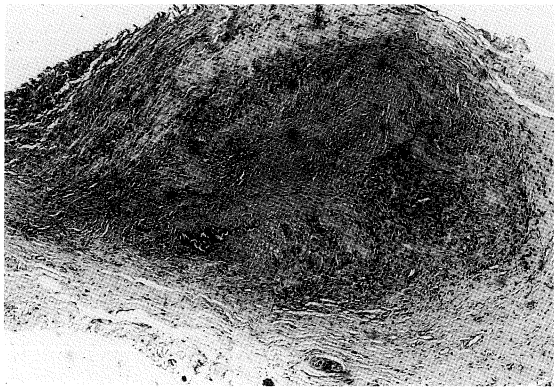

Fig 2 – Low powered view with lymphocytic and histiocytic infiltrates in the artificial wall, with intimal fibrosis.

Division of Ophthalmology
Changi General Hospital
2 Simei St 3
Singapore 529889

K Y Goh, MMed (Ophth), FRCS (Edin), FRCOphth
Consultant

Department of Ophthalmology
Tan Tock Seng Hospital
11 Jalan Tan Tock Seng
Singapore 308433

T H Lim, MMed (Ophth), FRCS (Edin)
Senior Registrar

Correspondence to:
Dr K Y Goh

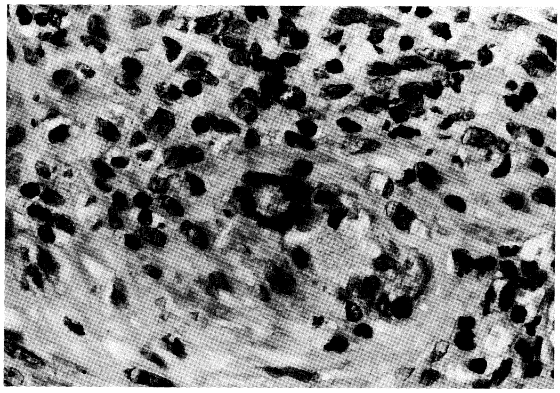

Fig 3 – Multinucleated giant cell within the arterial wall.

DISCUSSION

Acute visual loss in the elderly from arteritic ischaemic optic neuropathy is an ocular emergency that is potentially reversible with early treatment with intravenous steroids[1-3].

Ischaemia arises from inflammation of the posterior ciliary arteries that supply the optic discs. There is initially an acute necrotising phase causing exudation of fibrin into the vessel walls and subsequently in the granulomatous phase, there is inflammatory thickening of the medial muscle cells and internal elastic lamina which later fragments. There is infiltration by histiocytes, lymphocytes, giant cells of foreign body and Langhan's types, as well as neutrophils and eosinophils[4].

In a large retrospective review of 185 patients with giant cell arteritis at Bascom Palmer Eye Institute, Liu et al[5] found that 41 patients (63 eyes) had visual loss: 19 patients (46%) had unilateral defects, 15 (37%) had sequential involvement (mean interval between eyes was 23 days; range 1 to 219 days; median 5 days) and 7 patients (17%) lost their vision in both eyes simultaneously.

Anterior ischaemic optic neuropathy was the most common clinical finding (88%). Initial visual acuity loss was severe: in 44 (70%) of 63 eyes, visual acuity was 20/200 or worse, including 13 (21%) of 63 eyes with no light perception.

After treatment with steroids, 43 (68%) eyes stabilised, 16 (25%) improved and 4 (6%) worsened. Twelve of 13 eyes with no light perception at presentation remained unchanged. Citing the advantages of higher corticosteroid dose and greater bioavailability, several anecdotal reports recommended methylprednisolone over conventional oral therapy for visual loss due to giant cell arteritis[1].

CONCLUSION

Visual loss from GCA in Singapore may become more common due to the increasing numbers of geriatric patients seen.

It is an ocular emergency that requires immediate treatment with intravenous methylprednisolone (20 mg/kg/day for 3 – 5 days), followed by oral prednisone therapy. Diagnosis may be confirmed by performing a temporal artery biopsy.

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