Singapore Med J 2003 Vol 44(6) : 306-308

Giant Cell Arteritis (Temporal Arteritis, Cranial Arteritis) and A Case from Singapore

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ABSTRACT

Giant cell arteritis as the underlying cause of blindness in the elderly is common in the West but is not seen except on rare occasions in South East Asia.

We describe an 86-year-old Chinese man from Singapore who presented with a central retinal artery occlusion. Biopsy of a prominent superficial temporal artery established the underlying cause to be giant cell arteritis which was also the eventual cause of death as the condition resulted in rupture of a dissecting aneurysm of the aorta.

Giant cell arteritis should be considered in all cases of ischaemic eye disease in the elderly. The importance of early diagnosis lies in the very high incidence of second eye involvement within days or at most weeks in untreated patients. A high index of suspicion is required for diagnosis of this condition which is likely under-diagnosed in our local context.

Keywords: Giant cell arteritis, visual loss, central retinal artery occlusion, temporal artery biopsy, dissecting aneurysm of the aorta

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INTRODUCTION

Giant Cell Arteritis (GCA) is an inflammatory disease which mainly involves the cranial branches of the arteries arising from the arch of the aorta and is a common cause of blindness in elderly persons[1]. A study of the population of South-east Scotland gave an annual incidence of 1.3 cases per 100,000 persons[2]. In other studies, rates of up to 23 per 100,000 persons over 50 years have been reported[3].

Visual loss occurs in 50% of GCA patients[2] and is often the only presenting symptom i.e. the so-called "occult GCA"[4]. Anterior Ischaemic Optic Neuropathy (AION) is the common ocular manifestation while a Central Retinal Artery Occlusion (CRAO) accounts for less than 10% of cases[1]. On the other hand, only about 5% of patients with a CRAO have GCA[5]. Visual loss from GCA is not seen except on rare occasions in South-east Asia. In the Singapore population nonarteritic anterior ischaemic optic neuropathy (NAION) is common and is generally associated with diabetes, hypertension, arterio- or atherosclerosis. Only one biopsy-proven case with visual loss due to GCA has been reported to date in the local literature[6]. This was in an 83-year-old Chinese female who lost all vision as a result of bilateral sequential AION.

We now describe the case of an elderly man with biopsy-proven GCA who presented with sudden visual loss in one eye.

CASE REPORT

An 86-year-old Chinese male was admitted to the Medical Department of the Hospital for evaluation of persistent headache and general malaise. He was subsequently referred to the Eye Department because of an additional history of three-week visual loss in his right eye. The patient discovered this incidentally when he covered the other eye. Headaches and generalised muscle pains had been present for three months prior to presentation and there was associated anorexia for one month.

Right vision was recorded as no light perception. The right optic disc was noted to be pale and the retinal vessel attenuated. A diagnosis of a subsiding Central Retinal Artery Occlusion (CRAO) was made. The left eye was normal. At that time, prominent tender temporal vessels were noted by the attending Registrar who recorded the possibility of GCA. Blood pressure was measured at 130/80 mmHg and the only positive findings were an Erythrocyte Sedimentation Rate (ESR) of 72 mm/hour and a reactive Treponema Pallidum Hemagglutination (TPHA) test. The prominent temporal vessels were noted by a Neurologist, but were thought to be insignificant and the diagnosis of GCA not initially accepted. A CT Scan of the brain did not reveal any significant abnormality apart from some cerebral atrophy. The carotid and vertebral arteries were reported as normal. Carotid Doppler imaging was likewise normal.

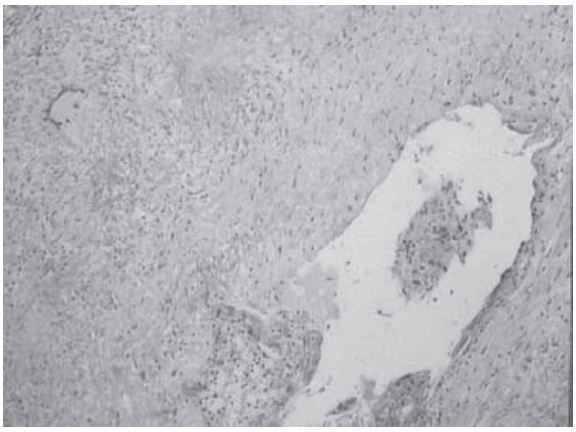

Fig. 1 Artery displaying an infiltrate of lymphocytes and mononuclear cells associated with an occasional multinucleated giant cell. Note the presence of intimal fibrinoid necrosis. *(Haematoxylin and eosin, original magnification x 40)*

After further consideration, it was decided that GCA needed to be excluded and a temporal artery biopsy was then performed.

The histopathological report of the right temporal artery specimen revealed complete occlusion of the lumen with intimal fibrosis, with dystrophic calcification and destruction of the elastic lamina. There was media destruction with fibrosis and necrosis and the presence of multinucleated giant cells. The adventitia was also fibrotic and showed mild to moderate inflammatory infiltrates of lymphocytes mixed with a rare plasma cell and neutrophil. These features were compatible with giant cell arteritis (Fig. 1).

One week following admission, he was started on treatment with oral prednisolone 60 mg daily. Within four days his general symptoms had resolved and the ESR had fallen to 53 mm/hr and two weeks later to 10 mm/hr. He was seen in the Eye Clinic on one further occasion four months after his initial presentation, when the right eye remained blind and the left healthy. The right disc was reported as pale with persistent attenuated retinal vessels. The steroid medication was reduced to 5 mg twice daily and a further appointment made for two months later.

The patient however died suddenly five months after presentation. A report to the Coroner was made and a post mortem examination performed which established the cause of death as "Haemopericardium due to dissecting thoracic aortic aneurysm", this being a common cause of death in patients with GCA *(See Discussion)*.

DISCUSSION

Giant Cell Arteritis first described clinically by Jonathan Hutchinson in 1890[7] is relatively common in the western world and affects both sexes equally[8]. Caucasians especially those of Scandinavian origin are affected more frequently than Afro-Americans, the latter comprising only 10% of clinical cases and 3% of biopsy-proven patients in a series from the Wilmer Institute[9]. The condition has also been reported in Saudi Arabians[10] and in Chinese[11].

The condition is characterised by a combination of fever, anaemia, high ESR, often altered mental state and especially headache in elderly patients. Scalp tenderness with palpable thickened non pulsatile superficial temporal arteries is evident along with jaw claudication and a painful tongue in many incidences. The condition is not infrequently associated with the polymyalgia rheumatica syndrome. Visual loss including total blindness occurring in 50% of untreated cases is the dreaded complication. A significant percentage of GCA patients however have minimal or no systemic symptoms and present with acute visual loss as occult GCA.

Recently Doppler ultrasonography has been used in suspected cases to locate the temporal artery when it cannot be palpated and Colour Doppler flow imaging can also demonstrate reduced velocity in the central retinal and posterior ciliary arteries in some patients.

GCA is associated with a substantially increased risk of the development of aortic aneurysm which may be a late complication and not a uncommon cause of death. Evans et al in a report on 96 patients with GCA found that 11 had thoracic aortic aneurysms, six of whom died suddenly of acute dissection[12]. In a study of the causes of death in 284 biopsy confirmed cases of GCA in Sweden, Nordborg and Bengtsson reported that one year after diagnosis, 21 of their patients had died from vascular disorders (expected number 7.01), 17 having died within four months. The cause of death was cerebrovascular disease in eight cases, myocardial infarction and cardiac failure in three cases each, pulmonary embolism in one case and interestingly two died from rupture of a dissecting aneurysm of the aorta[13].

In the patient reported above, the diagnosis was not considered at the outset because of the rarity of the condition in this part of the world. CRAO as a presenting feature of GCA was also thought to be unlikely, as AION is by far the commonest cause of visual loss in such patients. During the four-year period 1997-2000, there was only one arterial specimen of suspected GCA sent for histopathological examination to the Hospital and this in fact proved positive. We are informed that a similar situation exists in Malaysia (personal communication).

Interestingly, in the past three months during the first author's presence in Singapore, two more biopsy-positive cases, both in Chinese, have been seen in one of the other local eye institutions.

One of these had sequential bilateral AION, the other presented with severe headache and tender swollen temporal arteries without visual symptoms. In view of the recent diagnosis of these two cases (which may have occurred only by virtue of increased awareness of the condition), the possibility of GCA being prevalent in the region has to be borne in mind. This aetiology should therefore be considered in all cases of ischaemic eye disease as is the situation in Europe and the USA.

CONCLUSIONS

Giant Cell Arteritis should be suspected in all cases of visual loss due to Central Retinal Artery Occlusion or Anterior Ischaemic Optic Neuropathy in Singapore and an urgent ESR performed. If this is raised above 40 mm/hr, a temporal artery biopsy should be carried out. The importance of early diagnosis lies in the very high incidence of second eye involvement within days or at most weeks in untreated patients. In cases of aortic aneurysm the possibility of giant cell arteritis should also be considered as an underlying factor.

A high index of suspicion is required for the diagnosis of this condition which is apparently under-diagnosed in our population.

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