



Giant cell arteritis

Presenting as stroke, transient ischaemic attack and dementia



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BACKGROUND

Stroke and multi-infarct dementia are most commonly caused by atherosclerotic disease and are irreversible. Stroke and dementia associated with giant cell arteritis are rare but important to recognise as they may be reversible if diagnosed and treated promptly.

OBJECTIVE

This article describes the case of a patient who presented with a cilioretinal artery occlusion, diagnosed as being due to giant cell arteritis.

DISCUSSION

In retrospect, it seems likely that a stroke suffered 3 years earlier and mental state deterioration over that period may have been associated with the giant cell arteritis. Therefore, features useful in identifying stroke, transient ischaemic attack and dementia caused by giant cell arteritis are described.

Case history – Mr B

Mr B, aged 76 years, presented after sudden onset of right visual disturbance and periocular pain. His corrected right visual acuity was 6/9 and corrected left visual acuity was 6/18. There was a large left central scotoma, and fundoscopic examination revealed an area of retinal oedema and pallor near the disc consistent with a cilioretinal artery occlusion. He was also found to have swollen pulseless temporal arteries. His erythrocyte sedimentation rate (ESR) was 73 mm per hour. Although no formal assessment of his mental state was made, he was clearly confused, and had limited short term memory. He was commenced on 50 mg prednisolone per day. Temporal artery biopsy was performed the next day. At operation, the artery looked like a thick, white cord. Histopathological examination confirmed changes diagnostic of florid giant cell arteritis as shown in *Figure 1, 2*. The diameter measured on microscope slide with an ocular micrometer was 3 mm.

In the years leading up to the diagnosis of giant cell arteritis, Mr B's family had been aware of a fluctuating deterioration in his cognition, particularly regarding memory and visuo-spatial comprehension. One week after commencing prednisolone his visual acuity improved to 6/5 bilaterally and his scalp symptoms had greatly reduced. His family also described a marked improvement in his mental state. His ESR dropped to 5 mm per hour over several weeks. The prednisolone dosage was gradually reduced, and the patient continued to improve.

The spontaneous comment from Mr B's family about his mental state prompted the authors to review his medical history in more detail.

Reviewing the medical history

Three years previously Mr B had presented after sudden onset of right hemiparesis. He was found to be in atrial fibrillation. Computerised tomography demonstrated a corresponding cortical infarct as well as several old cortical and cerebellar infarcts. An echocardiogram performed a week later showed no abnormality. Doppler ultrasound study of the carotid arteries showed minimal wall calcification bilaterally without any

significant stenosis or velocity abnormalities. At the time, Mr B's stroke was thought to be purely the result of atherosclerotic and/or embolic disease.

In retrospect, while we cannot be certain, it seems likely that giant cell arteritis (GCA) was the underlying cause. At the time of the stroke Mr B had also complained of some more common symptoms of GCA including focal headache, temporal scalp tenderness and neck stiffness. Furthermore,

he had complained of intermittent episodes of these symptoms during the 4 years before the stroke; 12 months before the stroke these symptoms had sometimes been associated with impaired right hand coordination and dizziness. Giant cell arteritis was considered as a possible cause on numerous occasions, and on the basis of his age and symptoms alone at that time,¹ Mr B fulfilled the American College of Rheumatology 1990 criteria for the diagnosis of GCA.¹

However, the diagnosis of GCA was not made. The confounding factor may have been his erythrocyte sedimentation rate (ESR) measurements. Mr B's ESR was measured at least twice; the maximum recorded was 4 mm per hour. C-reactive protein was not measured.

Discussion

Common presenting features of GCA are headache, scalp tenderness and systemic

symptoms such as weight loss, fatigue and fever. Polymyalgia rheumatica is frequently associated with GCA, although the relationship remains obscure. Jaw claudication is less common, but considered pathognomonic of GCA. Visual loss occurs in approximately 25% of cases and is the most feared complication as visual recovery is uncommon. Stroke, transient ischaemic attack and dementia are rare complications of GCA.²⁻⁵

Furthermore, it is not always clear when these manifestations are caused by GCA rather than more common causes – chiefly atherosclerotic disease.⁵ The literature suggests that patients with GCA may be at increased risk of stroke during more active phases of their disease, but do not demonstrate an overall increased incidence of stroke.⁵⁻⁸ Intracranial ischaemia in patients with GCA has most frequently been found to be the result of thromboembolism in the absence of local arteritis.^{5,9} This is most likely the result of secondary intrarterial embolism, while the primary site of

thrombus formation is within the inflamed extracranial vessels.^{5,9} Intracranial arteritis caused by GCA has been found to be confined to extradural intracranial vessels and nearly always occurs in association with extracranial vessel disease.^{5,9} Table 1 lists facts useful in identifying strokes, transient ischaemic attacks and dementia caused by GCA.

Isolated intradural vessel disease is rare, and may in fact represent isolated angiitis of the central nervous system rather than GCA.^{5,10,11} This is because the severity and extent of disease caused by GCA is indirectly related to the quantity of elastic tissue in the media and adventitia of the artery concerned, and intracranial arteries possess little elastic tissue.⁹ Intracranial disease caused by GCA has been found to occur up to three times more frequently in the posterior than the anterior circulation.^{5,9,12-14}

Conclusion

Giant cell arteritis is an important disease about which doctors should be

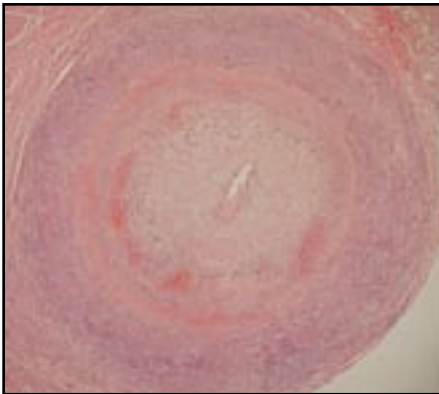


Figure 1. Gross intimal proliferation and fragmentation of the internal elastic lamina

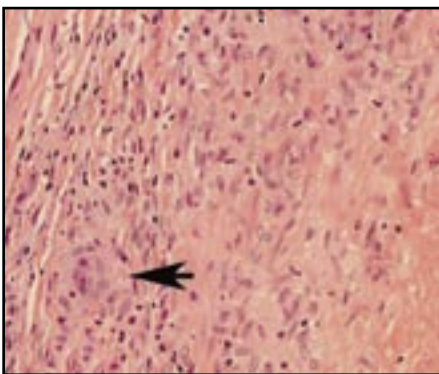


Figure 2. Inflammatory cell infiltrate within the wall of the artery including multinucleate giant cells (arrow)

Table 1. Identifying strokes, transient ischaemic attacks and dementia caused by GCA

- GCA is rare before the age of 50 years; although the incidence increases with age.¹⁵ The overall prevalence among those over 50 years of age is approximately 200 per 100 000 persons¹⁶
- No clear gender predilection exists after adjusting for life expectancy¹⁷
- Stroke, transient ischaemic attack and dementia caused by GCA are usually preceded by other more common manifestations of GCA⁵
- In contrast to atherosclerotic disease, GCA more commonly affects the posterior than the anterior circulation^{5,12-14}
- Intracranial ischaemia is usually the result of extracranial vessel disease and therefore tends to involve large cortical territories
- Manifestations tend to follow a fluctuating course corresponding with fluctuations in disease activity
- ESR is not always elevated in association with GCA.¹⁸ An elevated ESR is not specific for GCA. Combining the results of the ESR and C-reactive protein will increase the sensitivity and specificity of detecting GCA¹⁷
- Temporal artery biopsy remains the gold standard for diagnosis¹⁷
- Patients with GCA associated stroke tend to recover gradually, although not necessarily completely, with prompt administration of corticosteroids^{4,6,8,13,14}

well informed as it is associated with significant mortality and morbidity if not diagnosed and treated promptly. Stroke and dementia associated with GCA are rare but important to recognise as they may be reversible if diagnosed and treated promptly.

Conflict of interest: none.

Acknowledgment

The contribution of Dr Robert Spokes, anatomical pathologist, Pathcare, Geelong, is gratefully acknowledged.

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