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Case report

# Giant cell arteritis presenting as spinal cord infarction

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## 1. Introduction

Giant cell arteritis is easily recognised in older patients presenting with headache, scalp tenderness and raised inflammatory markers. Timely treatment with steroids can prevent serious complications such as stroke and visual loss. As our case demonstrates, giant cell arteritis can lead to less well recognised neurological manifestations that may not prompt immediate consideration of the diagnosis.

## 2. Case report

A 75-year-old Caucasian woman with a background of asthma, presented with a short history of gradual onset neck pain. The pain was felt posteriorly, radiating to both sides of the neck. It became progressively worse, reaching a severity of 8/10, before subsiding after 1 week. As the pain cleared, both her arms became weak (right more than left), progressing over a few days until she was unable to abduct her right shoulder. The weakness was associated with tightness around the upper chest and numbness across her shoulders and neck. There were no symptoms in the legs, no visual disturbance, no speech or swallowing problems, and no bowel or bladder symptoms. There was no history of headache, no jaw claudication and no weight loss.

On examination, she was fully alert and cognitively intact. Cranial nerve examination was normal. Tone was normal in the arms and there was marked bilateral weakness restricted to C5 and C6 innervated muscles. This was worse on the right, with Medical Research Council (MRC) grade 1–2 power of right shoulder abduction, internal rotation and external rotation and grade 2 power of elbow flexion. The left side was only mildly affected with grade 4 power of shoulder abduction and 4+ power of elbow flexion. Her right biceps and supinator jerks were absent and all other upper limb deep tendon reflexes were present but depressed. Tone and power were normal in the legs with normal ankle jerks and downgoing plantars, but the knee jerks were relatively brisk. Sensory examination was normal and there was no ataxia. The temporal artery was easily palpable and non-tender on the right and although the left temporal artery was non-tender, it was noted to be tortuous with a reduced pulse.

On initial presentation, her erythrocyte sedimentation rate (ESR) was 122 mm/h and C-reactive protein (CRP) was 59 mg/L. Magnetic resonance imaging (MRI) of the spine showed a central spinal cord signal abnormality at C6-7 on axial images (Fig. 1). Cerebrospinal fluid was acellular with normal glucose and protein. Autoimmune screen, vitamin B12, folate and myeloma screen were all unremarkable.

A diagnosis of giant cell arteritis was suspected and after the lumbar puncture the patient was commenced on intravenous methylprednisolone (1 g/day for 3 days) and aspirin 75 mg once a day. The patient was then transferred to the regional neurology tertiary centre for further management. Computed tomography (CT) angiography (covering from the aortic arch to the intracranial vessels) showed minor calcific atherosclerotic disease at the

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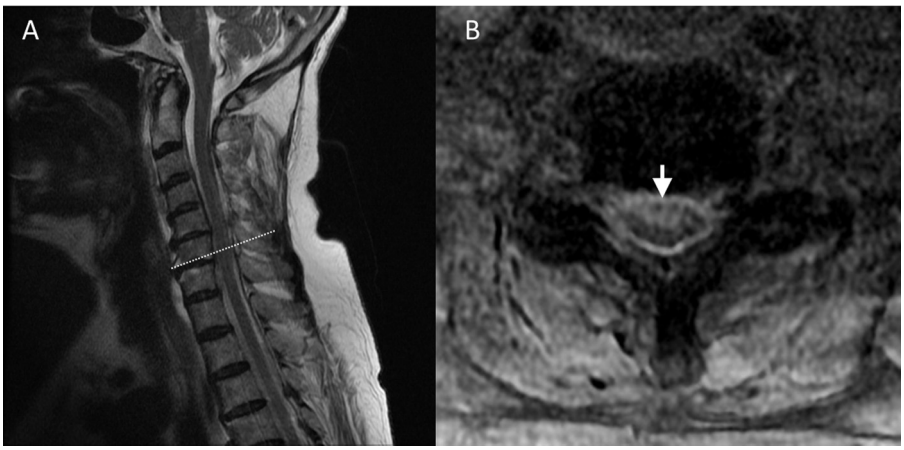


**Fig. 1.** $T_2$-weighted magnetic resonance imaging (MRI) of the spine showing central spinal cord high signal abnormality (arrow) at C6-7 on axial view (B) with sagittal view (A) demonstrating the axial image level (dashed line).

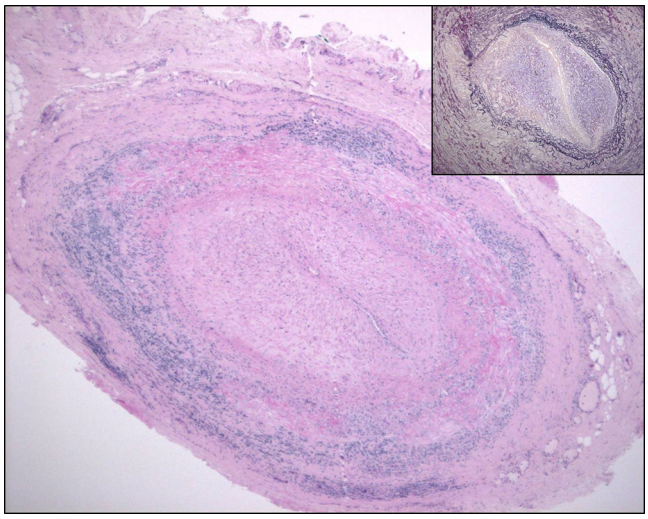

**Fig. 2.** Temporal artery biopsy showing transmural mononuclear inflammation with marked fibrointimal proliferation (original magnification 50×, periodic-acid Schiff stain). Inset demonstrates duplication and focal disruption in the internal elastic lamina (original magnification 100×, elastic von Gieson stain).

carotid bulb bilaterally, but no other significant abnormality and CT imaging of the brain was normal. A temporal artery biopsy was performed (Fig. 2) which showed florid changes consistent with giant cell arteritis, with the majority of the inflammation seen in the adventitia, extending into the media and intima. The inflammatory infiltrate was composed of lymphocytes and histiocytes, rare multinucleated giant cells were present and no clear granulomata were seen. The patient continued on high dose oral prednisolone (1 mg/kg), as per the British Society of Rheumatology guidelines [1]. Her upper limb weakness began to improve and she was discharged home to continue community neurological rehabilitation.

## 3. Discussion

Giant cell arteritis is the most common chronic vasculitis in Caucasians and typically involves the cranial branches of vessels arising from the aortic arch. It occurs almost exclusively in those over the age of 50 and is more common in women (3:1 ratio) [2]. The aetiology of giant cell arteritis has not been established but it is characterised pathologically by transmural inflammation of the arterial wall, with lymphocytes, macrophages and granulocytes in all cases as well as multinucleated giant cells in around half of patients [3]. Perhaps the best known clinical manifestation of giant cell arteritis is temporal and/or occipital headache with associated scalp tenderness. However, there are many other clinical and laboratory manifestations which can occur without the presence of headache, such as jaw claudication, systemic manifestations (fever, fatigue, weight loss), polymyalgia rheumatica, visual loss, stroke, abnormal liver enzymes and normochromic normocytic anaemia [3].

As headache may be absent in up to a third of cases, there is a danger that these less common manifestations are not recognised as giant cell arteritis, with a resultant delay in diagnosis. Left untreated, giant cell arteritis can lead to permanent unilateral or bilateral visual loss thus it is imperative that the condition is promptly recognised and treated. In the majority of cases the ESR will be elevated (>50 mm/h, but often > 100 mm/h) but in 4–15% of biopsy proven cases it is not. When combined with CRP, sensitivity increases to over 99% so it is important to check both [4]. A temporal artery biopsy is recommended to confirm the diagnosis, but steroids and aspirin should be commenced without delay, in line with published guidelines [1].

Although in this case there was no headache, temporal artery tenderness or jaw claudication, the diagnosis was suspected, appropriate treatment was initiated, and the diagnosis was subsequently confirmed histologically. The neurological picture could be localised clinically to the central cervical spinal cord at the C5/6 level and this was confirmed by MRI scanning. We would postulate that the lesion was ischaemic in nature, due to involvement of the medium sized branches of the vertebrobasilar circulation, which supply the anterior spinal artery. The neck pain may have been a direct manifestation of vertebral artery inflammation. The case highlights both the predilection of giant cell arteritis for the vertebral arteries and the importance of the vertebral arteries in the blood supply of the spinal cord. Spinal cord infarction is a rare but recognised complication of giant cell arteritis [5–9], but it serves to illustrate the wide spectrum of neurological presentations associated with the condition.

Neurological manifestations occur in up to a third of patients with giant cell arteritis [10]. In a series of 166 biopsy proven cases, Caselli et al. found neuropathies in 23, transient ischaemic attacks or strokes in 12, neurological syndromes in 11, tremor in 6, neuropsychiatric syndromes in 5, tongue numbness in 3 and a myelopathy in 1 [10]. Thus the neurological presentation of giant cell arteritis can be remarkably varied and the clinician must maintain a high index of suspicion, especially when more widely recognised manifestations are absent. In assessing a patient over 50 years old with unexplained neurological symptoms or signs, we

would recommend specific enquiry regarding the manifestations of giant cell arteritis as well as urgent laboratory measurement of the ESR and CRP. By adopting this strategy, the risk of missing the wider spectrum of this condition will be greatly reduced, minimising the subsequent risk of visual loss or stroke.

## 4. Conclusion

In patients over 50 with unexplained neurology and elevated inflammatory markers, physicians should consider giant cell arteritis in their differential diagnosis.

## Acknowledgements

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