Clinical Medicine 2023 Vol 23, No 1: 94–6

Lessons of the month: Giant cell arteritis with Horner's syndrome and vertebral dissection

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ABSTRACT

We present a case of an 82-year-old woman presenting with left-sided Horner's syndrome and stroke. She also had a 6-week history of intermittent dizziness, reduced appetite, lethargy, muscle stiffness and weight loss. Examination revealed left temporal artery and left posterior auricular artery tenderness. Her ESR showed 62 mm/hr and imaging showed left vertebral artery dissection. Temporal artery biopsy was positive.

The case highlights a rare presentation of giant cell arteritis with Horner's syndrome and left vertebral artery dissection. High clinical suspicion is required to prevent delay in diagnosis and treatment.

KEYWORDS: Giant cell arteritis, vertebral dissection, Horner's syndrome

DOI: 10.7861/clinmed.2022-0479

Introduction

Giant cell arteritis (GCA) is the most prevalent systemic vasculitis affecting medium and large-sized arteries. It is common in women in their fifth decade of life.[1] Vasculitis can be a result of genetics, infections or autoimmune factors.

Patients may present with neurological symptoms such as headache, diplopia or cerebrovascular events.[1] Internal carotid and/or vertebral arteries are frequently involved in cerebrovascular ischemic events in GCA.[1] Ischaemic strokes and transient ischaemic attacks (TIA) due to involvement of intracranial arteries are rare.

Case presentation

An 82-year-old woman presented to the emergency department with complaints of slurred speech, neck pain, left facial droop and occipital headache. Her symptoms had come on over a few hours. She also complained of a tingling sensation in her left hand and left side of her face. Prior to presentation, she had a 6-week history of intermittent dizziness, reduced appetite, lethargy, muscle stiffness and weight loss.

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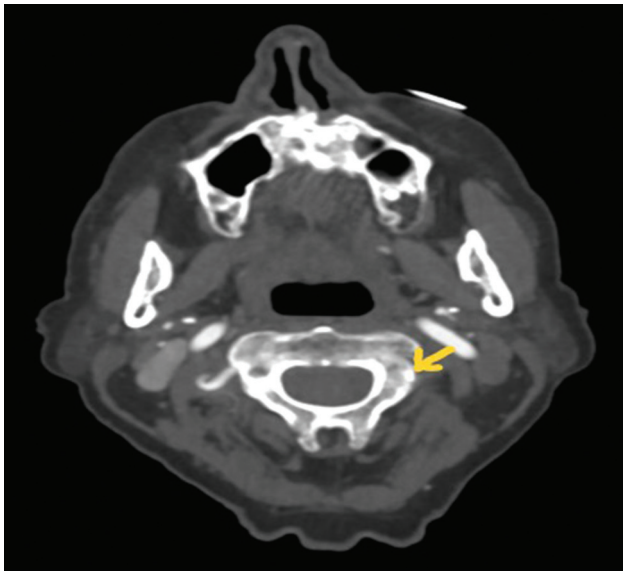

Fig 1. CT Angio of aortic arch and carotid artery both showing appearances of upper left vertebral artery short segment dissection (yellow arrow).

Clinical assessment revealed bilateral shoulder girdle pain and stiffness with left-sided ptosis and miosis. She also displayed cerebellar signs (impaired finger-to-nose test on the left) and a positive Romberg's test. National Institutes of Health Stroke (NIHS) Score was 0 on admission and 1 the next day due to left leg ataxia. Importantly, palpation showed tenderness on left temporal artery and left posterior auricular artery around the mastoid bone.

Biochemistry showed white blood cell count 10.9 $\times$ 10^9/l (reference range (RR): 4–10 $\times$ 10^9/l), neutrophilia (8.73 $\times$ 10^9/l), haemoglobin 119 g/l (RR: 115–165 g/l) and platelets 434 $\times$ 10^9/l (RR:150–400 $\times$ 10^9/l). Inflammatory markers were high with erythrocyte sedimentation rate (ESR) 62 (RR: 0–35), and C-reactive protein (CRP) 61 mg/L (RR: 0–5 mg/l). Antinuclear antibodies (ANA) and anti-nuclear cytoplasmic antibodies (ANCA) were negative.

Computed tomography (CT) head showed no evidence of acute intracranial pathology; however, intracranial CT angiogram revealed appearances of a short dissection in a segment of the upper left vertebral artery (Fig 1). Additionally, brain magnetic resonance angiography showed this dissection was extending to the neck region with little to no flow seen in the proximal intracranial pathway (Fig 2).

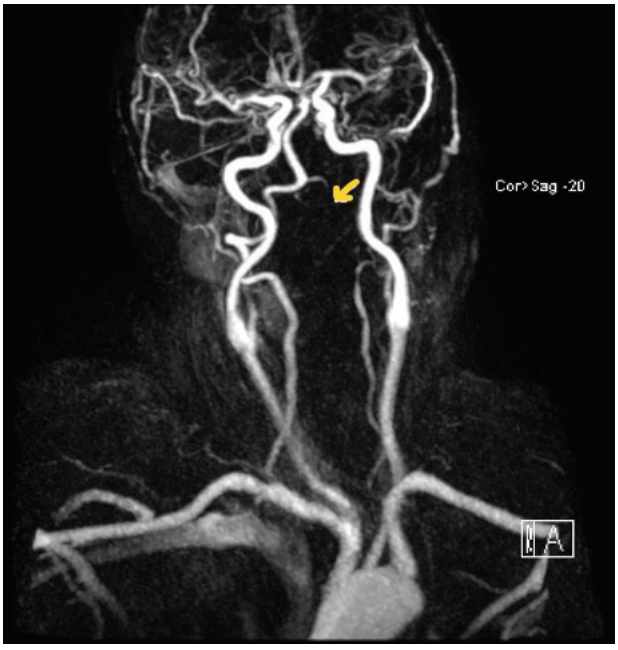

Fig 2. Left vertebral artery dissection on contrast-enhanced MR angiography (CE- MRA).

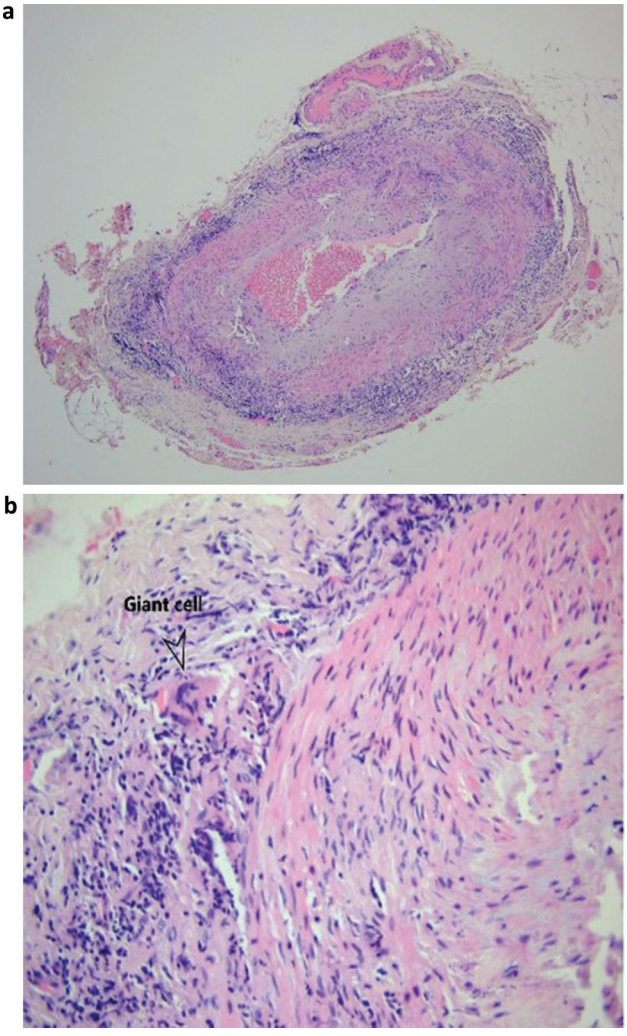

Fig 3. Left temporal artery biopsy showing giant cell arteritis. a) At 10× magnification thickened arterial wall with narrowed lumen. b) At 200× magnification showing disruption of wall architecture and focally dense lymphoid cell collection with multiple multinucleated giant cells in the wall.

She was managed for posterior circulation stroke with possible vertebral artery dissection with aspirin. However, her temporal artery tenderness and raised ESR gave a high clinical suspicion of GCA. Prednisolone 60 mg was started. There was significant improvement in symptoms and a temporal artery biopsy (TAB) was planned.

She was discharged with prednisolone and antiplatelet therapy. Upon outpatient review 2 weeks later, her symptoms had resolved and inflammatory markers normalised.

TAB showed disruption of the architecture and dense collections of lymphoid cells in the wall with multiple multinucleated giant cells, consistent with GCA (Fig 3).

Discussion

Early diagnosis and prompt treatment of GCA is key to avoid irreversible vision loss in patients as GCA is a time-sensitive disease.[1,2]

GCA can present with vague symptoms resulting in a delay in diagnosis and treatment. This patient initially presented with constitutional symptoms of fatigue and weight loss, which are unspecific and often leads the clinician to consider a diagnosis of malignancy. Temporal headache and scalp tenderness, the most common symptoms of GCA, were not present but instead an occipital headache was present.[2]

Atypical presentations such as ptosis, Horner's syndrome or stroke have been associated with GCA. Approximately 2–7% of patients present with stroke in GCA. GCA usually affects vertebrobasilar regions and is mostly seen in elderly men with vascular risk factors. Headache symptoms, neck and pain are present in about 80% of cases with dissection of the cervical segment of the carotid or vertebral arteries.[3] A multicentre case-control study concluded that involvement of vertebrobasilar territory is more common in GCA with recent ophthalmic ischemic symptoms and low inflammatory markers.[4] Intracranial arteries are rarely involved as they have little internal elastic lamina. However, the internal elastic lamina is present in vertebral and carotid arteries from the aorta arch extending up to 5 mm into the dura mater.[2] Our case involved a unilateral vertebral artery extending intracranially. In contrast, our patient had high ESR without any visual symptoms.

A few reported GCA cases present with Horner's syndrome, with or without other symptoms such as internuclear ophthalmoplegia or ipsilateral abduction deficit.[5] It has been suggested that the presence of isolated Horner's syndrome may be the only manifestation of GCA, especially in the elderly.[6] The most likely causal explanation of Horner's syndrome with GCA is the direct involvement of the cervical sympathetic chain enveloping the carotid arteries.[6] TAB is the gold standard for diagnosis, with 39% sensitivity and 100% specificity. Biopsies would show a classic transmural inflammatory involvement of the large arteries.[7]

Corticosteroid is the mainstay of treatment for GCA, with oral prednisolone 60 mg/day given for high-risk neuro-ophthalmic symptoms and impending vision loss.[8] Intravenous methylprednisolone can be considered as initial therapy. Symptoms improve rapidly within hours to days after steroid administration but vision loss may not improve.[9] It is important to follow up patients to monitor relapses. Relapses may require steroid-sparing agents such as methotrexate or tocilizumab.[8,9] GCA with intracranial involvement has been associated with higher complications and mortality rates. Cases where corticosteroids alone did not prevent neurologic complications may necessitate the addition of immunosuppressants.[10] Studies have suggested use of antithrombotics to prevent ischaemic complications in GCA. However, EULAR now recommend that antiplatelets should not be routinely prescribed unless for other reasons such as cerebrovascular disease.[8]

Conclusion

GCA is a systemic vasculitis with serious complications. The diversity of GCA symptoms makes the diagnosis difficult. Anyone presenting with a stroke involving vertebral arteries and/or Horner's syndrome with or without typical symptoms, especially in elderly, should be considered for GCA assessment. ■

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