Giant cell arteritis presenting as multifocal cerebral infarcts

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DESCRIPTION

A 71-year-old woman presented to the emergency department following a witnessed syncopal episode associated with new-onset confusion and psychomotor retardation. She denied any history of headaches, scalp tenderness, visual disturbance, jaw or limb claudication or proximal muscular pain or stiffness. Examination revealed a temperature of 38.1°C and a pronator drift. She was otherwise neurologically intact but unable to perform complex tasks. Witnesses denied any head injury.

MRI (figure 1A) revealed multifocal abnormalities of different ages including acute left frontal (small arrow) and occipital (arrowhead) ischaemic infarcts and an older right parietal infarct with haemorrhagic transformation (large arrow). Raised erythrocyte sedimentation rate of 89 mm in the first hour (reference range; 10 to 30) and C-reactive protein of 268.6 mg per litre (reference range; 0 to 5) were noted.

Figure 1 (A) MRI (FLAIR sequence) revealing multifocal abnormalities of different ages including acute left frontal (small arrow) and occipital (arrowhead) ischaemic infarcts and an older right parietal infarct with haemorrhagic transformation (large arrow). (B) CT angiography showing focal areas of stenosis alternating with dilatations affecting both internal and external carotid branches (arrows).

Figure 2 Positron emission tomography-CT scan demonstrating increased tracer uptake in both ascending and descending thoracic aorta (arrows).

Treatment with intravenous meropenem and acyclovir was commenced to cover for both encephalitis and septic emboli. Lumbar puncture, echocardiography, blood cultures and virology were normal. Cerebral CT angiography was performed (figure 1B) as the patient remained encephalopathic despite treatment with no improvement in inflammatory markers, revealing focal areas of stenosis alternating with dilatations affecting both internal and external carotid branches; this was suggestive of large-vessel vasculitis (LVV). A positron emission tomography was performed, showing increased uptake in the aortic arch, common carotid arteries and thoracic aorta (figure 2).

A diagnosis of giant cell arteritis (GCA) was made and the patient was treated with intravenous methylprednisolone 1 g on alternate days for three doses, followed by 60 mg daily of oral prednisolone. Quick recovery was made with resolution of encephalopathy and inflammatory markers within 1 week of starting treatment. She was transferred to a neurorehabilitation centre 2 weeks later. Oral corticosteroids were slowly tapered with a plan to achieve a dose of 20 mg after 3 months of treatment. Despite improvement however, the patient developed a hospital-acquired pneumonia 2 months later and passed away.

GCA is the most common cause for idiopathic LVV. It almost always occurs after 50 years of age with a steady rise in incidence thereafter, peaking between the ages of 70 to 79.¹ It has a predisposition to affect the aorta and its branches, including the external carotid and the temporal arteries.^{2,3} It typically presents insidiously with constitutional symptoms, headaches, visual disturbances (especially transient monocular visual loss), jaw claudication and elevated inflammatory markers. Rarely, patients present with serious ischaemic complications secondary to LVV such as permanent visual loss and central nervous system involvement including strokes.⁴ Cerebrovascular accidents secondary to active GCA may occur in any vascular territory, but has been described to more commonly effect the vertebrobasilar circulation, with one study reporting an overall rate of 2.8% between symptom onset and the first 4 weeks of glucocorticoid therapy.⁵

Traditional GCA classification criteria were based on presentation and clinical findings, supplemented with histopathological findings on temporal artery biopsy. The use of radiological imaging has grown in recent years owing to advances in imaging techniques and is now recommended to complement clinical criteria for diagnosing GCA. Histological

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Learning points

- ▶ Giant cell arteritis can present acutely with ischaemic complications and in the absence of typical symptoms.
- ▶ Vasculitis is an important differential in patients presenting with multifocal cerebral infarcts affecting more than one arterial territory.
- ▶ Prompt recognition of vasculitis is vital as a delay in treatment initiation could lead to life-threatening complications.

diagnosis is no longer required in cases with high clinical suspicion of GCA together with positive imaging.⁶ As seen in our case, the diagnosis of GCA was based on the findings highlighted by several imaging modalities, together with a dramatic clinical improvement on initiation of immunosuppression.

This case is unique as our patient did not exhibit any traditional GCA signs or symptoms, except for an elevation in inflammatory markers. The presentation was that of a LVV with involvement of the aorta and its main branches, presenting with neurological manifestations secondary to multifocal cerebral ischaemia. The finding of abnormalities affecting several arterial territories on MRI, including both anterior and posterior circulations, was

an early clue to suggest that these lesions were not embolic in nature.

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REFERENCES

- 1 Gonzalez-Gay MA, Vazquez-Rodriguez TR, Lopez-Diaz MJ, *et al.* Epidemiology of giant cell arteritis and polymyalgia rheumatica. *Arthritis Rheum* 2009;61:1454–61.
- 2 Roberts J, Clifford A. Update on the management of giant cell arteritis. *Ther Adv Chronic Dis* 2017;8:69–79.
- 3 Koster MJ, Warrington KJ. Giant cell arteritis: pathogenic mechanisms and new potential therapeutic targets. *BMC Rheumatol* 2017;1:2.
- 4 de Boysson H, Liozon E, Larivière D, *et al.* Giant cell arteritis-related stroke: a retrospective multicenter case-control study. *J Rheumatol* 2017;44:297–303.
- 5 Gonzalez-Gay MA, Vazquez-Rodriguez TR, Gomez-Acebo I, *et al.* Strokes at time of disease diagnosis in a series of 287 patients with biopsy-proven giant cell arteritis. *Medicine* 2009;88:227–35.
- 6 Dejaco C, Ramiro S, Duftner C, *et al.* EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice. *Ann Rheum Dis* 2018;77:636–43.

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