

Multi-territory infarcts caused by intracranial giant cell arteritis

Karim M Mahawish, Pietro Cariga

Giant cell arteritis (GCA) is the most common form of vasculitis in adults. It is characterised by a pan-arteritis of medium to large-sized arteries. Despite major recent advances in the treatment of GCA, the diagnosis is challenging. Symptoms include headache (in two thirds of subjects), scalp tenderness and jaw claudication. As sudden permanent visual loss occurs in 8–30%, and stroke in 3–10% of patients, GCA should be considered a medical emergency.¹

Case report

A 65-year-old woman presented with a one-week history of dysphasia and right hemiparesis. C-reactive protein (72mg/L) and platelet count ($799 \times 10^9/L$) were elevated. Magnetic resonance imaging (MRI) of the brain demonstrated bilateral acute

ischaemic infarcts (Figure 1) and distal left internal carotid critical stenosis (Figure 2A). Other tests including vasculitis auto-antibodies, anti-phospholipid antibodies, syphilis, human immune virus, Janus Kinase 2 mutation and cerebrospinal fluid analysis were unremarkable. Transthoracic echocardiogram and 72 hours of telemetry did not demonstrate any abnormalities.

Three days later, her speech improved and with prompting the patient admitted recent jaw claudication. Temporal arteries were found to be non-tender and non-pulsatile. Ultrasound of the temporal arteries showed low resistive waveform, however no halo sign. Temporal artery biopsy (TAB) demonstrated a heavy lymphocytic infiltrate within the external elastic lamina. GCA was diagnosed and she was commenced on

Figure 1: Diffusion weighted MRI brain with B1000 sequence (left) and apparent diffusion coefficient (right) demonstrating areas of restricted diffusion in the left anterior cerebral artery, left parietal area and in the right hemisphere borderzone middle cerebral/anterior cerebral artery territory.

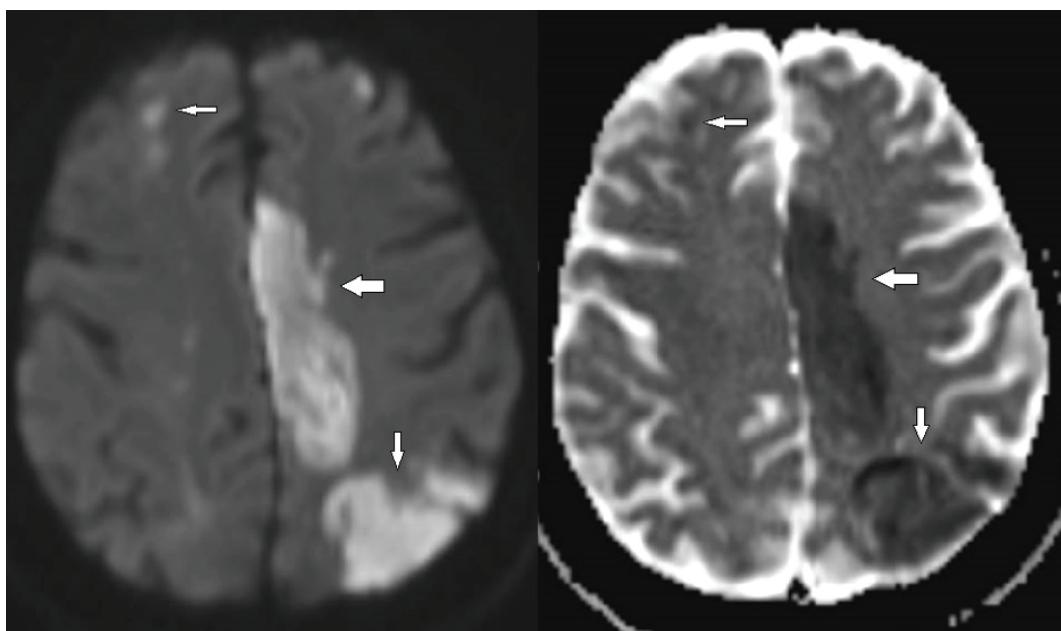
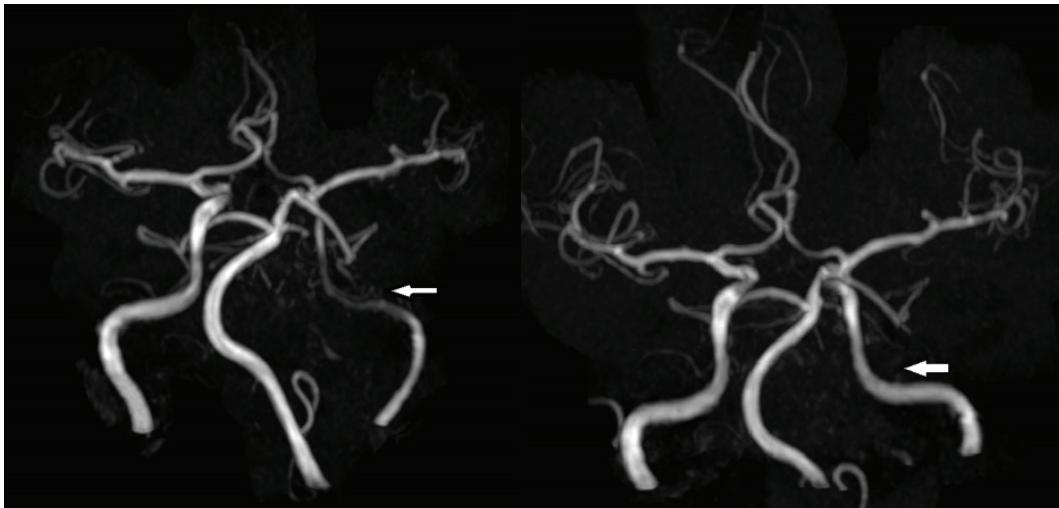


Figure 2A (left): Time of flight MR angiography demonstrating critical stenosis of the distal left internal carotid artery. **2B (right):** Follow-up MR angiography following high dose corticosteroids demonstrating improved caliber of the left internal carotid artery and distal branches of the anterior and middle cerebral arteries.



intravenous methylprednisolone 1g daily for three days, then oral prednisone with tapering dose. Follow-up MRI two weeks later demonstrated improved caliber of the internal carotid artery (Figure 2B) and more prominent distal branches of the middle and anterior cerebral arteries. The C-reactive protein and platelet count normalised within 5 and 50 days of treatment respectively. We speculate that hypoperfusion and a pro-inflammatory state were responsible for the strokes.

Discussion

GCA predominantly affects individuals over 50 years of age, particularly Caucasian females who have a 1% lifetime risk, twice that of men.¹ Previously termed ‘temporal arteritis’, GCA may present with intracranial and/or extra-cranial arterial involvement, or even proximal disease (aorta and associated branches). It is likely that both genetic and environmental factors initiate an inflammatory cascade leading to GCA.²

Constitutional symptoms may be present. Cranial manifestations include temporal cutaneous hyperalgesia, jaw or tongue claudication and prominent, beaded or irregular temporal artery with a decreased pulse. Large vessel manifestations include aortitis, limb claudication and aneurysms. Strokes occur due to the hypercoagulable proinflammatory state, vessel wall inflammation and

co-existing atherosclerosis causing vessel occlusion and/or embolisation of inflammatory thrombi.

The majority of patients have thrombocytopenia and elevated acute phase reactants, however the latter may be normal in 1–10%.¹

Though temporal artery biopsy is considered the gold standard diagnostic test, false-negative rates of up to 40% have been reported.³ This may be due to sampling errors (eg, inadequate biopsy length), the presence of skip lesions and the duration of glucocorticoid treatment.

Non-invasive imaging of involved arteries with colour Duplex Ultrasonography, 3-Tesla MRI or in the case of extra-cranial GCA, 18F-fluorodeoxyglucose positron emission tomography (18F-FDG-PET) are now featuring more prominently as first line investigations in the diagnosis of GCA. These tests demonstrate good sensitivity and specificity for GCA compared with positive TAB.^{4–6}

GCA is treated with high-dose glucocorticoids as first-line therapy; however, long-term toxicity is common. The addition of methotrexate reduces relapse rates and glucocorticoid requirements.¹ Tocilizumab is an IL-6 receptor antagonist which reduces relapse rates and glucocorticoid requirements; however, is not licensed in New Zealand.

In our patient, intracranial GCA progressed insidiously until she experienced hypoperfusion strokes due to poor flow through inflamed vessels. The diagnosis required a high index of suspicion and additional tests to reach the diagnosis. While the predominant left hemisphere infarcts resulted from the severely stenosed left internal carotid, the right hemisphere infarcts were the likely result of stenosed distal branches of the middle and anterior cerebral arteries

resulting in watershed or borderzone infarcts. The follow-up MRI demonstrates improved flow in these vessels.

In summary, features of GCA should be specifically sought from patients presenting with acute ischaemic stroke as symptoms may be insidious and it is associated with significant morbidity. Diagnosis can be challenging, however effective treatments are available.

Competing interests:

Nil.

Author information:

Karim M Mahawish, Consultant in General Medicine and Stroke, Department of Internal Medicine, Midcentral DHB, Palmerston North Hospital; Pietro Cariga, Consultant Neurologist, Department of Neurology, Midcentral DHB, Palmerston North Hospital.

Corresponding author:

Dr Karim M Mahawish, Consultant in General Medicine and Stroke, Department of Internal Medicine, Midcentral DHB, Palmerston North Hospital.

kmahawish@doctors.org.uk

URL:

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