Lesson of the month 2: A stroke of bad luck

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ABSTRACT

Giant cell arteritis is a common cause of headache in patients aged more than 50 years. It is an easy diagnosis to make if classical features, ie temporal headache, jaw claudication, visual symptoms, systemic symptoms of fever or weight loss with high erythrocyte sedimentation rate and anemia, are present. However, it may present atypically and stroke can be the presenting feature. A high index of suspicion is needed in atypical presentations such as stroke. Once a diagnosis is suspected it is imperative to start high dose steroids to prevent visual and neurological complications.

KEYWORDS: Giant cell arteritis, stroke, vasculitis, steroids, aspirin

Case presentation

A 62-year-old woman who was previously fit and well was admitted with a 6-week history of headaches and feeling generally unwell. In the last 8 months she was investigated for unexplained iron deficiency anaemia. Her coeliac serology and upper and lower gastrointestinal endoscopy were normal. Her past medical history included polio of the right leg and agoraphobia. Her only known vascular risk factors included a 30-packs-per-year smoking history. She was not on any medications.

Over the last 3–4 weeks she had noticed difficulty with coordination of her right hand. Three days prior to admission, she developed fever, slurring of speech and increasing weakness of the right hand. On admission she was pyrexial at 38.0°C, blood pressure 130/80 in sinus rhythm and a Glasgow coma score (GCS) of 15; no rash or meningeal signs were noted. However she had right upper and lower limb weakness (4/5) with upper motor neuron facial palsy. Her cardiovascular examination revealed no obvious murmurs.

Investigations

Her initial investigations were as follows: haemoglobin, 9.3 g/dl; white blood cells, 14.6x10^9/l; neutrophils, 11.5x10^9/l, platelets, 594x10^9/l; mean corpuscular volume, 71 fl; erythrocyte sedimentation rate (ESR), 93 mm; C-reactive protein, 159 mg/l; glucose, 6.3 mmol; albumin, 28g/l, alanine transaminase, 11 IU/l, alkaline phosphatase, 126 IU/l, bilirubin, 6 mmol/l, Na, 138 mEq/l; K, 3.4 mEq/l; urea, 3.0 mmol/l, creatinine, 42 mmol/l. Urine analysis showed no proteinuria, haematuria or casts.

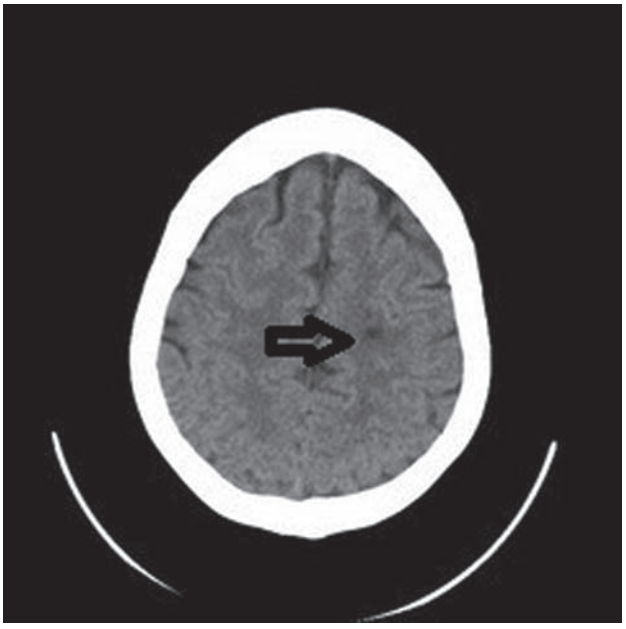

Fig 1. Plain CT scan of the brain shows a watershed zone of ischaemia in the left parietal lobe. CT = computed tomography.

An electrocardiogram showed sinus rhythm and a chest X-ray showed normal lungs and heart size. A computed tomography (CT) scan of her brain (Fig 1) showed a left pre-central gyrus infarct and ischaemia in the border zone of the left anterior cerebral artery (ACA) and the middle cerebral artery (MCA) territories. Unfortunately she suddenly deteriorated on the second day of admission with a GCS score of 8 with dense right sided hemiplegia.

Differential diagnosis

In view of the patient's headache, focal signs and inflammatory response, the differential diagnosis included stroke caused by large vessel vasculitis (giant cell arteritis (GCA) or Takayasu

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Learning points

- GGCA should be suspected with patients aged above 50 years presenting with headaches and raised inflammatory markers.
- There is a need for awareness that GCA can cause strokes.
- High-dose steroids and aspirin should be initiated to reduce chances of neuro-opthalmic complications as soon as GCA is suspected.
- There is a need for urgent discussions in such atypical cases with stroke physicians and neuroradiologists to optimise investigations and treatments.

GCA = giant cell arteritis.

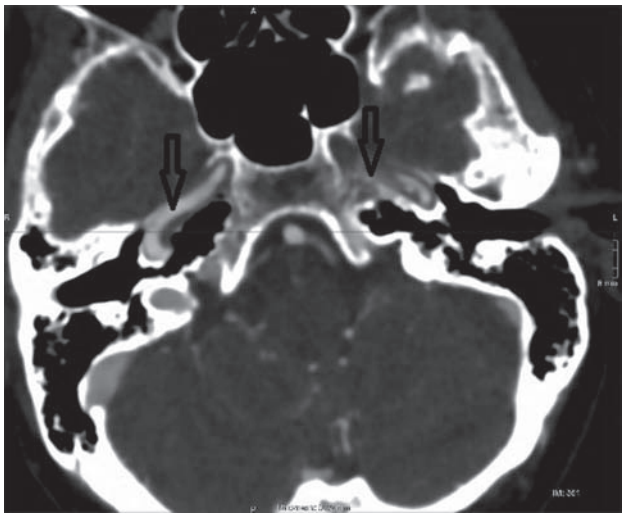

Fig 3. CT angiogram shows a normal filling petrous portion of right ICA but an occluded left ICA (arrows point at ICA – compare right and left lumen sizes). CT = computed tomography; ICA = internal carotid artery.

arteritis), medium vessel vasculitis (polyarteritis nodosa) or small vessel vasculitis (Churg Strauss syndrome, microscopic polyangitis or primary central nervous system (CNS) angitis), and stroke with infective aetiology (occult septic foci with embolic stroke, cerebral abscess or meningitis).

Management

On admission, on consideration of possible sepsis of unknown origin, an appropriate sepsis screen was undertaken without commencement of antibiotics. On the second day, due to the patient's sudden deterioration, an urgent stroke team review was undertaken. A repeat plain CT brain scan (not shown here) revealed no further changes. However, in view of the possibility of sepsis and the underlying iron deficiency anaemia, thrombolysis was considered inappropriate. She was given a stat dose of 1 g intravenous methylprednisolone and broad spectrum benzylpenicillin and gentamicin for possible underlying endocarditis.

A suspicion of GCA by the stroke team prompted an urgent CT angiography to assess cerebral vasculature (Figs 2–4) and temporal artery biopsy (TAB). CT angiography (arch to vertex) showed a normal arch of aorta but a narrowed cervical and obstructed petrous portion of left internal carotid artery (ICA). In the meantime vasculitic antibody screen and trans-thoracic echocardiogram were normal. She was commenced on prednisolone 60 mg with aspirin 300 mg once a day. The TAB performed within 48 hours was consistent with a diagnosis of giant cell arteritis. Antibiotics were stopped after the initial sepsis screen was normal.

Her consciousness improved over a 48–72 hour period but she had residual dense right hemiplegia and mixed receptive and

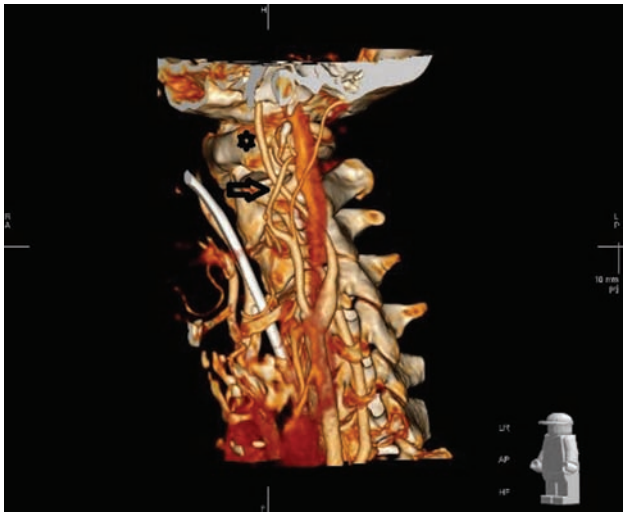

Fig 2. CT angiogram shows a uniformly narrowed cervical portion of left ICA (arrow and star show narrow ICA). CT = computed tomography; ICA = internal carotid artery.

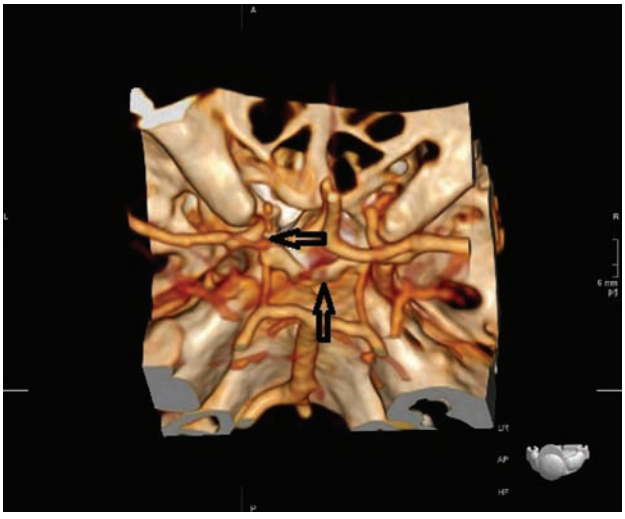

Fig 4. CT angiogram of circle of Willis shows an occluded left ICA and a narrowed right MCA (arrows show narrowed MCA and ICA). CT = computed tomography; ICA = internal carotid artery; MCA = middle cerebral artery.

expressive dysphasia. Her nutrition and hence rehabilitation was impaired as a result of severe depression (compounded by high dose steroids). She needed supplementary naso-gastric tube feeding in the meantime. Her mood improved gradually over the next 3 months after commencement of 40 mg fluoxetine and gradual tapering of steroids by 10 mg every 6 weeks until a dose of 20 mg was reached. She was eventually discharged home but was bed-bound with severe spasticity of the upper limb.

Discussion

Giant cell arteritis is a common cause of headache in patients aged more than 50 years. It affects the cranial branches of the arteries originating from the aortic arch. In 10–15% of cases the extra-cranial branches of the aortic arch are also involved.[1,2] It is an easy diagnosis to make using the American College of Rheumatology criteria,[3] if classical features, ie temporal headache, jaw claudication, visual symptoms, systemic symptoms of fever or weight loss with high ESR and anaemia, are present. However it may present atypically and stroke can be the presenting feature. It is reportedly the cause of first-ever stroke in only 0.11% of patients.[4] In our patient, diagnosis was delayed by the sepsis-like and the subacute stroke-like features. The clinical features in the same distribution supported an ICA pathology as opposed to a cardioembolic source, which usually affects different territories. This prompted discussions with neuroradiologists and hence a CT angiography was performed.

A high index of suspicion is needed in atypical presentations such as stroke. Once a diagnosis is suspected it is imperative to start high dose steroids to prevent neuro-opthalmic complications. Aspirin has antiplatelet and anti-inflammatory properties and is hence important in such cases. It is important to discuss such atypical presentations with stroke teams so that relevant imaging and management be initiated without undue delay. TAB is the cornerstone for diagnosis and remains positive up to six weeks of starting steroids.[5] ■

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