CASE REPORT

Vasculitis revealed by posterior stroke

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ABSTRACT

Posterior ischaemic stroke is relatively uncommon, and its occurrence should alert clinicians to possible uncommon underlying disease.
We report a patient with occipital brain infarction. The combination of age, gender, general malaise and elevated erythrocyte sedimentation rate led to the clinical suspicion of giant cell arteritis. Vertebral artery vasculitis was confirmed by 18-FDG positron emission tomography, combined with CT angiography, and immediate immunosuppressive therapy was started.
Symptoms of stroke should, in a particular clinical context, raise suspicion of giant cell arteritis.

What was known on this topic?
Posterior stroke is a known, but rare, complication of giant cell arteritis.

What does this add?
Combining clinical pattern recognition, i.e. typical signs and symptoms in a particular context, with modern imaging techniques, i.e. 18-FDG PET, can lead to early diagnosis and treatment of large artery vasculitis.

KEYWORDS

CT angiography, giant cell arteritis, positron emission tomography, vertebral artery insufficiency

INTRODUCTION

Giant cell arteritis (GCA) is a vasculitis of predominantly large- and medium-sized arteries, characterised by granulomatous inflammation in the vessel wall.[1] Symptomatic vessel inflammation usually involves cranial branches of arteries originating from the aortic arch, including the superficial temporal artery, the ophthalmic artery and the posterior ciliary arteries.[1,2] The incidence of GCA increases after the age of 50 and peaks between 70 and 80 years of age. Two thirds are women. The disease is associated with polymyalgia rheumatica.[1]
The clinical phenotype of temporal artery involvement in GCA can be quite typical and often includes unilateral headache, jaw claudication and visual loss due to ischaemic optic neuropathy. In more than half of GCA cases, however, arteries other than the temporal artery are involved.[1] In such patients, symptoms are often atypical and may involve arm claudication, signs of regional or global cerebral hypoperfusion, low-grade fever and general malaise. Stroke associated with extracranial involvement in GCA occurs in 3 to 7% of GCA patients.[3,4] Previous studies suggested that 10% of early deaths in GCA were caused by stroke.[5]
In this report we present a patient in whom GCA was revealed by stroke caused by vertebral artery inflammation. The latter was confirmed by positron emission tomography (PET) scintigraphy combined with computer tomography (CT) angiography. Although rare, the combination of cerebral ischaemia with the specific signs and symptoms should raise suspicion of GCA.

CASE PRESENTATION

A 76-year-old woman presented with a fortnight's history of disturbed vision. She also reported complaints of proximal myalgia, general malaise, and headache above the

left eye for two to three months. Neurological examination revealed globally decreased muscle strength. Visual field examination revealed loss of vision on the left side. Laboratory results showed an erythrocyte sedimentation rate (ESR) of 117 mm/h, C-reactive protein (CRP) 137 mg/l, normocytic anaemia (haemoglobin 6.2 mmol/l) and thrombocytosis (630 x 10^9/l).

Brain CT showed abnormalities in the right occipital cortex, compatible with semi-recent infarction and with the characteristics of her visual impairment. The neurologist prescribed aspirin, ended his consultation and requested the internist to analyse potential causes for her seemingly unrelated general symptoms and acute-phase response. The neurological findings, combined with other symptoms, age, gender and sharply elevated ESR, raised our suspicion of GCA causing posterior cerebral ischaemia. However, further questioning revealed no complaints of jaw claudication or scalp tenderness. Physical examination of both temporal arteries showed no abnormalities. Subsequently, an 18-fluorodeoxyglucose (FDG) PET scan was performed which showed clearly enhanced FDG uptake in the vertebrobasillary region (*figures 1* and *2*), as well as some enhanced uptake in part of the aorta and the aortic arch. The other large arteries displayed a normal uptake pattern. CT angiography showed irregular wall thickening and stenoses along the course of both vertebral arteries (*figures 2* and *3*).

To protect the posterior circulation from further ischaemia, treatment with intravenous pulses of methylprednisolone (1 g/day for three days) was commenced, followed by rapid improvement of general complaints and no further deterioration of the neurological deficit. She was discharged on oral prednisone 60 mg/day. At follow-up, her myalgia and malaise had disappeared, and ESR and haemoglobin normalised.

Figure 1. *F6: Coronal 18F-FDG PET slice showing much more intense 18-FDG uptake in both vertebral arteries (red arrows) compared to the ascending aorta (green arrow). 18-FDG uptake in myocardium and brain is physiological*

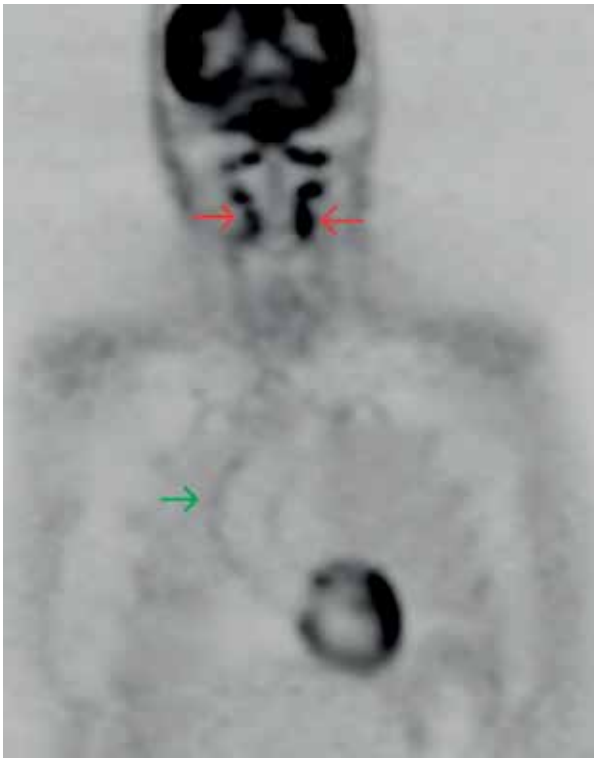


Figure 2. *Transaxial 18-FDG PET (left) showing increased 18-FDG uptake in both vertebral arteries (red arrowheads) with a more intense uptake in the right artery. At the same level CT-angiography (right) shows luminal stenosis and vessel wall thickening (red arrowhead)*

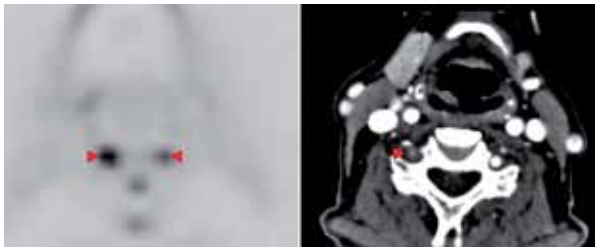
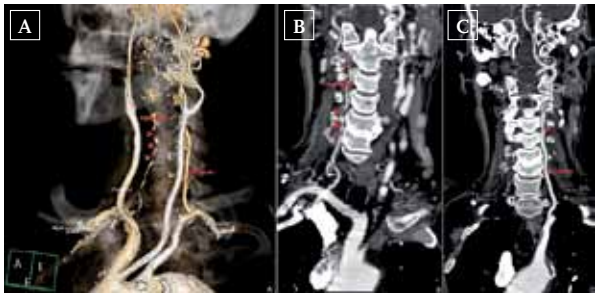


Figure 3. *A: 3D VRT (Volume Rendering Technique)-reconstruction of CT-angiography showing multiple stenosis (red arrowheads) along the course of the right vertebral artery. B and C: oblique-coronal views of curved MPR's (multiplanar reformatting) showing multiple stenoses (red arrowheads) along the course of the right vertebral artery (B) and a mild stenosis (red arrowhead) of the left vertebral artery (C)*



DISCUSSION

The central message of this case report is that pattern recognition of GCA symptoms is important when cerebral ischaemia presents in a non-typical fashion, or when other symptoms are present. Also, 18-FDG-PET images of

vertebral involvement in GCA have appeared only rarely in the medical literature.[6]

Vertebral artery involvement in GCA is usually limited to the extracranial sections, except for the first 5 mm after passing the dura mater.[7] Arteritis of the vertebrobasilar system may result in the full spectrum of associated posterior cerebral ischaemic symptoms and signs.[4] Symptomatic bilateral vertebral artery involvement in GCA is found in only one to two of 1000 patients with ischaemic stroke.[7]

Why did we not perform temporal artery biopsy to support the diagnosis of GCA? Our motivation was that such biopsies, although specific, lack sensitivity. Even if typical signs of temporal artery involvement are present, sensitivity for an abnormal biopsy ranges from 66 to 90%, depending on which signs are present (highest when headache, jaw claudication and scalp tenderness are all present).[8] Limited sensitivity is explained by segmental involvement of the vessel wall.[1] In patients with GCA not associated with specific signs of temporal artery involvement, such as our case, sensitivity drops to less than 50%.[8,9] The generic dilemma here is whether, in the fact of the possibility of having to start long-term, high-risk treatment, one should perform an additional test which is specific, but not sensitive. Obviously, a positive specific test result is reassuring in terms of not exposing the patient to such therapy for no good reason. Admittedly, some patients (and physicians) may benefit from the additional motivation provided by a positive biopsy but, overall, this benefit may be lost by loss of impetus from a negative test result, however irrational this may seem, and however well-explained in advance to patients. In our opinion, the key issue is whether a negative result would change the therapeutic decision. If no alternative additional confirmatory tests are available, the answer to the latter question will often be 'no'. This being the case, we decided that temporal artery biopsy would not have changed diagnosis or management in our case. We do not suggest that performing a biopsy would have been wrong, but rather point to the risks of misinterpretation of a negative test. Unfortunately, we have seen several dramatic cases of GCA with a previously refuted diagnosis based on a negative temporal artery biopsy. Often, as in our case, arteries suspected to be affected by large artery GCA are inaccessible for biopsy, and the impossibility of obtaining histopathological confirmation is thus common.[9]

Large artery GCA is often diagnosed on clinical grounds, complemented with laboratory tests and imaging. The role of 18-FDG-PET in establishing the diagnosis seems essential, and calls for reconsideration of formal diagnostic criteria for GCA.[9,10]

In general, GCA is treated with oral corticosteroids.[1] There are no evidence-based guidelines for treatment of GCA associated with vertebrobasillary ischaemia. As we feared progression of posterior cerebral ischaemia, we instituted immediate high-dose immunosuppression, i.e. intravenous methylprednisolone, followed by oral prednisone.[11]

In conclusion, in patients with cerebral ischaemia, specific symptoms, combined with an elevated ESR, should raise the suspicion of GCA. Clinical judgement (i.e. pattern recognition) and modern imaging (i.e. 18-FDG-PET) go hand in hand, as they should.

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