A confusing presentation of giant cell arteritis

Introduction

The unusual presentation of this patient's giant cell arteritis caused some diagnostic delay. Her severe disease course, with leg vessel involvement, is a good illustration of the spectrum of illness this condition can cause.

Discussion

Although often thought of as a disease affecting mainly intracranial arteries – hence its alternative name 'temporal' arteritis – this case demonstrates that giant cell arteritis can affect vessels throughout the body. Involvement of extracranial vessels is thought to occur in about 15% of patients, with the aorta and its upper limb branches most frequently affected (Salvarani et al, 2002).

Whereas the most feared complications of 'classical' giant cell arteritis are stroke and blindness, extracranial disease can lead to problems including aortic dissection and progressive vessel stenosis causing limb claudication (Nuenninghoff et al, 2003). Vasculitis (including giant cell arteritis) should always be considered in the differential diagnosis for a chronically ischaemic limb.

Presentation with an acutely ischaemic limb is much more unusual, and led to some diagnostic confusion in this case. There are isolated reports of acute leg ischaemia in giant cell arteritis (Rodriguez et al, 1995), but the authors have been unable to find any previous reports of the disease presenting with acute arm ischaemia, as it did in this patient.

This patient subsequently developed lower limb involvement which, although well recognized, is less common than upper limb disease. In one series of 168 patients, only one suffered ilio-femoral disease (Nuenninghoff et al, 2003). Interestingly, the pattern of vessel involvement seen in the authors' patient – with distal disease, but sparing of proximal vessels – was shared by four patients with lower limb giant cell arteritis reported by Tatò and Hoffman (2006), suggesting that giant cell arteritis of the legs may preferentially affect distal vessels. Two of their patients – and the one discussed in this article – had positive temporal artery biopsies, demonstrating that biopsy can aid diagnosis even in predominantly peripheral disease.

Conclusions

This case is a valuable reminder that giant cell arteritis can affect arm and leg arteries, and shows that the condition can present gradually – with claudication – or more acutely. Clinicians should consider giant cell arteritis in their differential for both acute and chronic peripheral arterial insufficiency. **BJHM**

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Salvarani C, Cantini F, Boiardi L, Hunder GG (2002) Polymyalgia rheumatica and giant-cell arteritis. *N Engl J Med* **347**(4): 261–71

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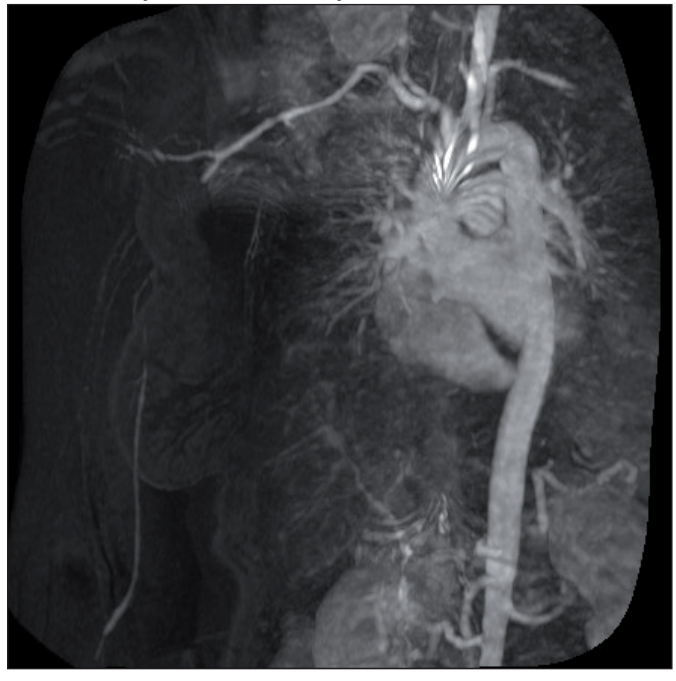
Figure 1. Magnetic resonance angiogram of right arm vessels, showing occlusion of the proximal brachial artery.



Case Report

A 56-year-old woman presented with a pale, painful, paraesthetic right arm. Examination revealed a cold right hand, with absent right radial, ulnar and brachial pulses. A provisional diagnosis was made of thrombotic or embolic arterial occlusion; the patient was anticoagulated and referred to the vascular surgical service.

Magnetic resonance angiography showed an occluded right brachial artery (*Figure 1*). Ultrasonography confirmed this, but also demonstrated widespread narrowing of the vessel, with surrounding hypoechoic tissue and fluid within the connective tissue suggesting an inflammatory process.

On questioning, the patient admitted to occipital headache and scalp tenderness. The erythrocyte sedimentation rate was 63 mm/hr, and the C-reactive protein level 155.1 mg/litre. A temporal artery biopsy showed changes typical of giant cell arteritis.

Prednisolone 60 mg daily was commenced with good effect. However, the patient's symptoms recurred when steroids were tapered below 50 mg daily. Moreover, 2 months later she developed bilateral calf claudication at 100 yards. Magnetic resonance angiography of the legs demonstrated narrow, diseased anterior and posterior tibial arteries bilaterally, but normal peroneal and proximal vessels.

Cyclophosphamide treatment was started, with resolution of the claudication and other symptoms.