Case report

Atypical giant cell arteritis resulting in arm amputation

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Giant cell arteritis is a chronic inflammatory disorder targeting large and medium sized arteries. It predominantly affects elderly women. In giant cell arteritis, an immune insult in the vascular wall initiates a reaction in the artery that leads to structural changes, intimal hyperplasia and luminal occlusion.¹ It most often affects the superficial temporal artery, although territories such as the facial, carotid, myometrial and upper and lower limb arteries may be affected.² We present a case of atypical giant cell arteritis which affected the subclavian artery. Unfortunately, the patient eventually lost her arm.

Case report

A 69-year-old woman presented to a general surgical admissions ward with an acutely ischaemic right arm. On examination, she had a pale, cool right hand and arm. The brachial and radial pulse were noted to be weak. Duplex ultrasonography of the arm showed a localised occlusion of the right axillary/proximal brachial artery approximately 3.5 cm long compatible with local thrombosis or embolus and with damped flow beyond. Full blood count, renal and liver profiles were normal, the ESR was 26, and her chest X-ray unremarkable. There was no cervical rib identified. It was decided to treat her conservatively with heparin. Unfortunately, she deteriorated over the next few days with first the radial pulse and then the brachial pulse becoming impalpable. It was, therefore, decided to proceed to a right arm embolectomy. At operation, a lot of debris and clot were withdrawn from the brachial artery and this was sent for histology along with a biopsy of the axillary artery. However,

Figure 1 The aortogram demonstrates occlusion of the subclavian artery under the proximal third of the clavicle.

despite this there was no pulsatile inflow and the patient was transferred under general anaesthetic to the X-ray department for an aortogram which demonstrated an

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Figure 2 Retrograde injection of contrast via the brachial artery demonstrates occlusion of a long segment of subclavian artery.

occlusion of the subclavian artery beginning under the proximal third of the clavicle continuing for several centimetres beyond (Fig. 1). This was confirmed by retrograde injection of contrast via the brachial artery through a catheter left in the arteriotomy (Fig. 2). An axillo-axillary cross-over graft with end-to-side anastomoses was performed.

Histology of the biopsies showed giant cell arteritis (Fig. 3) with marked endothelial oedema, fibrosis and active inflammation of the arterial wall which was focally transmural, and marked elastic layer fragmentation with giant cell formation.

Postoperatively, the axillo-axillary graft occluded within hours requiring re-exploration the same day. Subsequently,

Figure 4 Dry gangrene of the arm.

Figure 3 Histology of the biopsies of the arterial wall shows marked endothelial oedema, fibrosis and active inflammation of the arterial wall with giant cell formation.

it gradually re-occluded. She was reviewed by the rheumatology team who commenced treatment with pulsed intravenous methyl prednisolone converting to high-dose oral prednisolone (60 mg). She developed dry gangrene of the arm which demarcated to the level of the elbow (Fig. 4). An above-elbow amputation was performed.

Discussion

Giant cell arteritis typically presents in patients over 50 years of age with symptoms which include unilateral headache, scalp tenderness, and jaw or tongue claudication. There may be tenderness of one or both temporal arteries and systemic manifestations including malaise, fatigue and fever. Investigations usually reveal an elevated ESR (50–120 mm/h) and CRP. Temporal artery biopsy from the affected side if positive provides the definitive diagnosis, but may be negative in up to 30% of cases due to patchy arterial involvement and 'skip' lesions.³ Unfortunately, many cases are not classical and giant cell arteritis is notorious for its diversity of presentations and atypical cases.

There are occasional reports of giant cell arteritis affecting extracranial sites such as upper limb arteries, visceral arteries, aorta, spinal and even superficial femoral arteries. Mickley *et al.*⁴ reported a case series of three women presenting with arm claudication found to be due to stenosis of upper limb arteries caused by giant cell arteritis. Skopinski *et al.*⁵ published another series of six patients with upper limb involvement. Duplex ultrasonography demonstrated a hypo-echogenous halo of the arterial wall and acceleration of flow in four cases, and

arteriography showed long regular stenoses in five of the six. Klein *et al.*⁶ analysed a series of 248 patients presenting to a rheumatology clinic; 23 (9%) had definite large artery involvement, and four of these had upper limb claudication and absent arm pulses as initial manifestations of their arteritis. Angiography was helpful in indicating arteritis rather than atherosclerosis. There are common features between these cases and our patient who presented with arm claudication and an on-table angiogram which demonstrated occlusion of the subclavian artery. Retrospectively, we think that a pre-operative angiogram should have been done for our patient as she presented with a sudden ischaemia of her arm and there was no evidence of atrial fibrillation.

Giant cell arteritis can affect virtually any large arteries. Disseminated visceral giant cell arteritis was observed in four autopsied men. Giant cell arteritis was seen in the arterioles of three or more of the following organs: heart, lungs, kidneys, liver, pancreas and stomach. None had collagen disease, sarcoidosis, hepatitis or other infections, and vasculitis was only diagnosed after death.⁷ Giant cell arteritis can affect the aorta, a rare but potentially lethal event. A case has been reported with giant cell arteritis independent of atheromatous lesions in the aorta. The patient later died of aortic dissection causing haemopericardium. Temporal arteritis had been diagnosed prior to onset of the aortic complications.⁸ A case of giant cell arteritis manifesting as myelopathy and bilateral visual loss has been reported. Extensive necrotising granulomatous arteritis was found in the spinal arteries.⁹ A 30-year-old white male with severe bilateral leg claudication is presented as a case by Tamoff *et al.*¹⁰ Both superficial femoral arteries were explored, and a reversed saphenous vein bypass graft to the right leg inserted. Histology of the superficial femoral arteries postoperatively provided the diagnosis, but a temporal artery biopsy was negative.¹⁰ Similarly, in our case, the diagnosis was provided by histology of the axillary artery rather than temporal artery biopsy.

Giant cell arteritis can rarely present with a normal ESR, as it did in our patient. Zweegman *et al.* estimated the incidence as 5–30%; 34 such cases were identified, and the

conclusion reached was that a normal ESR cannot be used to exclude the diagnosis of giant cell arteritis.¹¹

Asymptomatic giant cell arteritis is another recognised, rare entity. Except for symptoms arising from the ischaemic arm, our patient was also asymptomatic.

This case illustrates both some of the difficulties in diagnosing giant cell arteritis, and the dangers of delayed or missed diagnosis. It also appears to be the first case reported as asymptomatic in an extracranial location with a normal ESR. Physicians and surgeons need to maintain a high index of suspicion of giant cell arteritis in order to identify and treat the condition, and thus avert potential catastrophic, irreversible sequelae.

References

1. Weyand CM, Goronzy JJ. Pathogenic principles in giant cell arteritis. *Int J Cardiol* 2000; **75** (Suppl. 1): S9–15, discussion S17–9.
2. Garcia Vazquez JM, Carriera JM, Seoane C, Vidal JJ. Superior and inferior limb ischemia in giant cell arteritis: angiography follow-up. *Clin Rheumatol* 1999; **18**: 61–5.
3. Klein GE, Campbell RS, Munder GG, Camey JA. Skip lesions in temporal arteritis. *Mayo Clin Proc* 1976; **51**: 504–10.
4. Mickley V, Friedrich JM, Sunder-Plassmann L. Angiographic diagnosis in giant cell arteritis of the arm arteries [in German]. *Rofo Fortschr Geb Rontgenstr Neuen Bildgeb Verfahr* 1992; **157**: 579–83.
5. Skopinski S, Constans J, Cherifi H, Midy D, Jarnier P, LeMetayer P *et al.* Inflammatory arteriopathy of the arms in the course of Horton disease. Report of six cases [in French]. *J Mal Vasc* 1999; **24**: 45–8.
6. Klein RG, Hunder GG. Large artery involvement in giant cell (temporal) arteritis. *Ann Intern Med* 1983; **83**: 806–12.
7. Lie JT. Disseminated visceral giant cell arteritis: histopathological description and differentiation from other granulomatous vasculitides. *Am J Clin Pathol* 1978; **69**: 299–305.
8. Manigand G, Taillandier J, Benhamou A, Barbagelatta M, Lemaigre G. Aortic dissection and Horton arteritis [in French]. *J Mal Vasc* 1982; **7**: 217–20.
9. Galetta SL, Baicer LJ, Lieberman AP, Syed NA, Lee JM, Oberholtzer JC. Refractory giant cell arteritis with spinal cord infarction. *Neurology* 1997; **49**: 1720–3.
10. Tarnoff JF, Pearlman E, Balkoura MH, Venkatraman M. Bilateral superficial femoral giant cell arteritis. *J Cardiovasc Surg* 1990; **31**: 375–9.
11. Zweegman S, Makkink B, Stehouwer CD. Giant cell arteritis with normal erythrocyte sedimentation rate: case report and review of the literature. *Neth J Med* 1993; **42**: 128–31.