CASE REPORT

Bilateral axillary arterial obstruction

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SUMMARY

A 74-year-old patient with longstanding diabetes mellitus and hypertension presented with obstruction in both axillary arteries. Although atherosclerotic obstructive disease would have been the most likely diagnosis in this patient, clinical findings were highly suggestive of extracranial giant-cell arteritis. Prompt treatment with systemic corticosteroids resulted in an excellent clinical response, avoiding the risks of endovascular or surgical intervention.

BACKGROUND

Giant-cell arteritis is the most common form of large artery vasculitis, with a predilection for the cranial branches of the carotid artery. Well-known symptoms include new onset headache, polymyalgia, jaw claudication and blindness. The latter constitutes a feared complication of this systemic disease, which manifests outside the cranium occasionally. The subclavian, axillary and mesenteric arteries are most frequently involved in the extracranial cases.¹ Failure to recognise a less typical presentation, thereby awaiting prompt corticosteroid treatment, bares the risk of progressive ischaemic vascular disease.

CASE PRESENTATION

A 74-year-old Caucasian woman, with a history of insulin-dependent diabetes mellitus, hypothyroidism and hypertension, was referred to the outpatient clinic. She had experienced a sudden onset of pain in her left arm and weakness in her left hand. Moreover, she felt fatigue and had non-specific chest pain. On physical examination a difference in blood pressure was found between the left and right arm, 128/80 and 166/85 mm Hg, respectively. Temporal arteries were non-tender. Ophthalmological examination was normal.

INVESTIGATIONS

Laboratory data showed an elevated erythrocyte sedimentation rate, 102 mm in the first hour ($n < 20$), a normocytic anaemia, haemoglobin level 5.6 mmol/L (n 8.5–10.0) and serum ferritin 549 mg/L ($n < 150$). Renal function, liver enzymes and C reactive protein were unremarkable. Despite statin treatment, total cholesterol level was slightly elevated at 5.7 mmol/L. The data, suggesting severe inflammatory response, had been normal on previous visits. Arterial obstruction in the upper extremities was suspected. On echo Doppler a monophasic signal and decreased pressures were seen bilaterally, suggestive of occlusion. Complementary, the upper chest MR angiography (MRA) confirmed obstruction of both axillary arteries (figure 1). On arteriography, stenosis of the left distal subclavian artery and occlusion of the left

axillary artery, length of obstruction at least 5 cm, was seen (figure 2). The brachial artery was filled via collaterals. The contralateral right axillary artery showed an occlusion as well. Temporal artery biopsy, which remained the diagnostic standard, was denied by the patient.

DIFFERENTIAL DIAGNOSIS

We present a case of a woman with diabetes in her 70s with multiple risk factors for atherosclerotic disease and a new onset arterial problem, suggested advanced atherosclerosis. Other causes of arterial obstruction, such as cardiac emboli were excluded. The patient had no vascular prostheses as a focus for infection. The clinical findings prompted us to a diagnosis of systemic arteritis. The arteriography findings, with a smooth 5 cm occlusion rather than a short distance atherosclerotic lesion, fitted in with the diagnosis. Peripheral atherosclerotic obstructive disease, which was our initial thought, will usually be treated locally, either by endovascular stent placement or bypass surgery. When considering autoimmune disease, such as giant-cell arteritis or Takayasu's disease, systemic treatment will prevail. Revascularisation of upper limb arteries then is seldom required because of exuberant collateral circulation that occurs.

TREATMENT

On the basis of the clinical presentation, laboratory investigations, bilateral MRA and arteriography results, extracranial giant-cell arteritis was diagnosed. Takayasu's arteritis was considered less likely in a patient in her 70s. Prednisone 60 mg once daily was started, after which the clinical condition improved markedly. The pain and weakness diminished and the function of the arm was regained.

Figure 1 MRI of the aortic arch and upper branches, showing narrowing of the left subclavian and occlusion of the axillary artery (arrows).

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Figure 2 Arteriographic detail of the left axillary occlusion.

To reduce the increased risk of cardiovascular events² the patient received aspirin. She had already been treated with a statin for several years because of dyslipidemia.

OUTCOME AND FOLLOW-UP

Since the start of treatment, more than 4 years ago, the course of the disease has been uneventful, although mild differences in blood pressures between left and right arm persisted. Prednisone therapy was gradually tapered to 2.5 mg daily. Laboratory values normalised gradually: after 3 months of treatment with prednisone, erythrocyte sedimentation rate was 12 mm in the first hour and haemoglobin level returned to 7.9 mmol/L.

DISCUSSION

The criteria to diagnose giant-cell arteritis were developed more than 20 years ago by the American College of Rheumatology. The classic signs are: age at disease onset >50 years, new headache, temporal artery abnormality, elevated erythrocyte sedimentation rate and abnormal artery biopsy. The presence of any three of these five criteria establishes the diagnosis with a sensitivity of 93.5% and a specificity of 91.2%.³ Recognising giant-cell arteritis, however, might be challenging when symptoms are incomplete or less typical, which will occur in about 20–40% of patients. Moreover, approximately 66% of patients with biopsy-proven giant-cell arteritis show large vessel involvement outside the cranium and 10% develop aortic involvement despite corticosteroid treatment.⁴ This will usually occur 1–5 years after the onset of the disease. Although upper or lower limb obstruction as first clinical manifestation is very unusual, atypical clinical presentations, including the aforementioned, have been described.^{5 6}

Peripheral arterial disease in giant-cell arteritis is located mainly at the level of the ascending aorta and its branches. Obstructive disease of primary and secondary branches as well as aneurysms, due to inflammation of the aorta, has been reported. It is recommended to perform a yearly screen in order to detect thoracic and even abdominal aneurysms, in patients with a high-risk cardiovascular profile.^{4 7}

As to the definite diagnosis, one could argue that temporal artery biopsy should be performed in every patient with atypical

signs of giant-cell arteritis. Negative findings up to 42%, however, have been reported in patients with extracranial large vessel manifestation.⁸ This along with imaging tools, such as ultrasound, MRA, arteriography and positron emission tomography (PET), have become the diagnostic tools of choice. MRA could provide accurate information on the involvement of the aortic arch and its branches and is able to demonstrate typical changes of inflammation.^{9–11} However, the sensitivity of duplex and MRA will decrease rapidly, once corticosteroids have been prescribed.¹² PET imaging has the advantage of revealing inflammation at different sites.

The patient's course of disease shows that large vessel obstruction might be the first presentation of giant-cell arteritis, even in patients with a high-risk profile for atherosclerotic disease.

Learning points

- ▶ Large vessel obstruction might be the first manifestation of giant-cell arteritis.
- ▶ Vasculitis is part of the differential diagnosis in large artery obstruction, even in patients with multiple risk factors for atherosclerotic disease.
- ▶ Extracranial giant-cell arteritis is preferentially treated with corticosteroids.
- ▶ Large vessel presentation in giant-cell arteritis should prompt the clinician to follow-up, in order to detect aortic aneurysms specifically.

Contributors All the authors have equally contributed to the preparation of the manuscript. BvdM wrote the first draft. AAMZ and AD prepared the final manuscript, including learning points and literature data.

Competing interests None.

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