A 69-year-old woman with intermittent claudication and elevated ESR

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A 69-year-old woman was referred to our out-patient clinic because of long-term hypertension and stable chronic renal failure (creatinine 132 $\mu\text{mol/l}$), which had been attributed to nephroangiosclerosis. She presented with a one-year history of anorexia, asthenia and loss of 6 kg of weight. In addition, she complained of intermittent claudication of the left arm and both legs lasting 2 months. This provoked important functional limitation of the three limbs, and impaired ambulation. She did not complain of headache or symptoms suggestive of polymyalgia rheumatica.

Her blood pressure was 160/95 mmHg on the right arm and undetectable on the left arm and lower limbs. Both temporal arteries were palpable and not painful. Auscultation of both carotid arteries was normal, without murmurs. Cardiac and pulmonary auscultation were normal. Murmurs were audible on both subclavian arteries. Neither the humeral nor the radial pulse were detectable on the left upper limb. Murmurs were also audible on both femoral arteries, and neither popliteal nor distal pulses were palpable. Her feet were cold although they had no ischaemic lesions. Funduscopic examination was essentially normal. The most relevant laboratory data were: erythrocyte sedimentation rate (ESR) 120 mm/h, C-reactive protein 4.4 mg/dl (normal range <1.5); antinuclear and antineutrophil cytoplasmic antibodies were negative.

A digital subtraction angiography (DIVAS) of both upper limbs was performed as part of her intermittent claudication syndrome evaluation (figures 1 and 2). All carotid, vertebral, renal arteries and aorta were normal and there were no atheroma plaques at any level. A DIVAS of the lower limbs showed stenosis of 50–80% in both femoral arteries and complete proximal obliteration of both popliteal arteries (figure 3).

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Figure 1 Arteriography disclosed a stenosis greater than 50% in the right subclavian artery and a long filiform stenosis greater than 80% in the left (arrows)

Figure 2 Arteriography of the left upper limb showing a filiform and nearly total occlusion of the axillary artery (arrows)

Figure 3 Arteriography of the left lower limb showing a complete stop in the popliteal artery (arrow). The same image appeared in the right lower limb.

Questions

- 1 What diagnosis is suggested by the images obtained with angiography?
- 2 What further investigation would you perform?

Answers

QUESTION 1

The first differential diagnosis to be taken into account in an elderly patient with intermittent claudication is arteriosclerosis. However, angiographic examination disclosed several findings that could exclude it in this patient: the absence of atheroma plaques, the unusual localisation of the lesions (the subclavian and axillary arteries), and the long and filiform morphology of the stenosis. The angiographic pattern therefore suggested large vessel vasculitis.

QUESTION 2

There are two large vessel vasculitides: Takayasu arteritis and giant cell arteritis (GCA). The table gives the criteria of the American College of Rheumatologists for distinguishing between them.^{1,2} Since the patient in this case was over 50 years of age, GCA was the most likely diagnosis. For this reason a biopsy of the temporal artery was performed, in spite of the absence of headache and polymyalgia rheumatica. The biopsy revealed a significant lymphoplasmacytic inflammatory infiltrate in the medium layer with giant multinucleated cells (figure 4). This established a definite diagnosis of GCA.

Treatment

Treatment with steroids (prednisone 1 mg/kg daily) was started with significant and rapid clinical improvement. Painless mobility of the affected limbs was restored, making surgical repermeabilisation unnecessary. On discharge, her ESR was 3 mm/h and the chronic anaemia had completely resolved. Six months later, her clinical improvement persists, and the patient has a nearly normal lifestyle, although recuperation of the absent pulses has not been achieved.

Discussion

The first differential diagnosis to be borne in mind in an elderly patient with involvement of large vessels is arteriosclerosis. The angiographic pattern in this case rules out arteriosclerosis and clearly suggests a large vessel vasculitis. Both Takayasu arteritis and GCA can present

Figure 4 Biopsy of the temporal artery (Haematoxylin-eosin stain; original magnification $\times 40$). Note the marked inflammatory infiltrate formed by lymphocytes, epithelioid and giant multinucleated cells

with giant cell granulomas and both have an excellent response to corticosteroids. However, temporal artery involvement is extremely unusual in Takayasu. Further, the last international conference on vasculitides nomenclature held in Chapel Hill,³ considered age (over 50 years in GCA) a useful parameter to distinguish between these entities. The present case was therefore diagnosed as GCA.

Few studies in the literature give the frequency of involvement of large vessels in GCA and most of these are based on necropsies. The largest study on this subject included 248 patients with confirmed GCA; in 23 patients (9.2%) the aorta or its branches were involved.⁴ The frequency in other series ranges from 7.6%⁵ to 15%.⁶ This variability is probably due to different patient selection criteria and different diagnostic methods used to evaluate arterial lesions.

Usually, large vessel involvement in GCA becomes apparent in two situations: either at the beginning, coinciding with cephalic and polymyalgia rheumatica-related symptoms, or when steroids are tapered off in a previously diagnosed case. In the first situation the diagnosis is suggested by the clinical manifestations and can be confirmed by temporal artery biopsy.⁴⁻⁶ Occasionally, however, the presenting symptom of this vasculitis can be isolated intermittent claudication or ischaemia of the extremities, as in four of the 248 patients with GCA reported by Klein *et al*,⁴ and two of the 91 patients studied by Ninet *et al*.⁵

Table The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis and Takayasu arteritis

Takayasu arteritis

- age ≤ 40 years
 - claudication of an extremity
 - decreased brachial artery pulse
 - >10 mmHg difference in systolic blood pressure between arms
 - bruit over the subclavian arteries
 - arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities
- The presence of ≥ 3 criteria demonstrated a sensitivity of 90.5% and a specificity of 97.8%

Giant cell arteritis

- age ≥ 50 years
- new onset of localised headache
- temporal artery tenderness or decreased temporal artery pulse
- elevated ESR ≥ 50 mm/h
- biopsy sample showing necrotising arteritis characterised by a predominance of mononuclear cell infiltrates or a granulomatous process with multinucleated giant cells.

The presence of ≥ 3 criteria was associated with a sensitivity of 93.5% and a specificity of 91.2%

2 In order to normalise the ESR and stabilise the arterial lesions, these patients were given daily doses of steroids higher than the standard for GCA (≥ 1 mg/kg). Most of them showed significant improvements in claudication and pulses (60%). Although the pulses were not recovered in cases with total vascular thrombosis, amputation of the involved limb was avoided.⁴ The mean response time was 2–4 weeks.

Finally, this case emphasises the need to exclude GCA when ischaemia of the limbs occurs within a toxic inflammatory picture. It also emphasises the importance of excluding this type of complications in a patient with a known GCA, before attributing them to polymyalgia rheumatica.

Learning points

- intermittent claudication may be the first symptom of a giant cell (temporal) arteritis
- giant cell arteritis must be excluded in a patient with intermittent claudication associated with a toxic syndrome and/or elevated ESR which cannot be attributed to other causes

Final diagnosis

Giant cell arteritis.

Keywords: intermittent claudication; giant cell arteritis

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Self-mutilation and behavioural disorder

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A 26-year-old man presented with history of disinterest in performing household chores, hearing voices conspiring against himself, and poor personal hygiene. In addition, his relatives were extremely worried over the fact that the patient often pinched and scratched his nose, to the extent that he had torn off a portion of the left ala of the nose, exposing his nasal septum. The above complaints were of 13 years duration.

There was no history of head injury, epilepsy, substance abuse, prolonged elation or depression of mood or any other neurotic disorders. There was no past or family history of similar behaviour. His birth and initial developmental milestones were normal, and general physical examination was within normal limits. No abnormalities were found during systemic examinations including a detailed neurological examination. A complete haemogram, blood sugar urea, serum uric acid level, liver function tests and serum electrolytes were within normal limits. Electroencephalogram and computed tomographic scan did not reveal any abnormalities. Examination of the nose revealed chronic inflammatory changes of the left nasal septum with a portion of the left ala of the nose missing (figure). Detailed mental status examination revealed referential ideas, persecutory delusions of victimisation, and third-person auditory hallucinations, ie, voices discussing and threatening him.

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Figure

Questions

- 1 What is the diagnosis?
- 2 What is the aetiopathology of the condition shown?
- 3 What are the differential diagnoses?