Case report

Giant cell arteritis of myometrial and axillary arteries and polymyalgia rheumatica

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SUMMARY Giant cell arteritis (GCA) of the uterus and adnexa is extremely rare—only nine cases have been reported. We report a further case, in which the patient was found to have GCA of the myometrial arteries on routine hysterectomy. She then developed another rare manifestation of GCA, involvement of the axillary arteries causing arm claudication and chronically ischaemic hands.

Key words: myometrial arteritis, aortic arch arteritis, arm claudication.

Polymyalgia rheumatica (PMR) and giant cell arteritis (GCA) are well recognised as different manifestations of the same disease. Patients typically present with general malaise, proximal muscle pain and stiffness, bitemporal headache, and visual blurring. Although arteries of the head and neck are most often involved, GCA may occasionally affect the aorta and branches and very rarely affects pelvic arteries. We report a patient with GCA of the myometrial arteries who developed arm ischaemia because of axillary artery involvement.

Case report

A 73 year old woman was referred for gynaecological consultation because of bleeding from a cervical polyp. She was otherwise fit. The gynaecologists found prolapse in addition to the cervical polyp. Haemoglobin preoperatively was 9.3 g/dl (93 g/l). She had an uneventful anterior repair and vaginal hysterectomy. Microscopic examination of the specimen showed a benign polyp and an unremarkable endometrium. The myometrium included a benign leiomyoma. The myometrial arteries showed the changes of florid giant cell arteritis characterised by a predominating mononuclear and giant cell inflammatory infiltrate of the vessel wall associated with extensive destruction of the vascular elastic laminae. Many of the vessels showed fibrous intimal proliferation with obstruction of their lumina, and in some there was fibrinoid necrosis (Fig. 1).

Shortly after discharge the patient complained of a numb, cold right hand and pain in the right forearm on exertion. The left arm was less severely affected. She felt tired, and on further questioning admitted to stiffness in the buttocks and groin, especially in the morning. She had lost weight.

On examination both hands were cold and the right hand was blue. The radial and ulnar pulses were impalpable on the right and reduced on the left. The right brachial artery pulse was reduced. There was a short midsystolic apical murmur but no other bruits. Blood pressure in the right arm was not recordable and in the left arm was 150/75 mmHg. The temporal arteries and all other pulses were normal. Hip abduction caused discomfort.

The following investigations were carried out: Hb 11.3 g/dl (113 g/l), white blood cell count 8.4×109/L, erythrocyte sedimentation rate (ESR) 115 mm/h (Westergren). Alkaline phosphatase 163 U/l (normal range (NR) 30–135 U/l). C reactive protein 3.6 mg/dl (34.9 mg/l) (normal <0.6 mg/dl (6 mg/l)), Albumin 30 g/l (NR 30–44 g/l). IgG 16.9 g/l (NR 8–16 g/l), IgA 1.8 g/l (NR 0.7–3.7 g/l), IgM 0.9 g/l (NR 0.1–1.2 g/l), C3 65 mg/dl (NR 80–180 mg/dl), C4 17 mg/dl (NR 15–50 mg/dl). Rheumatoid factor negative, antinuclear factor negative.

Accepted for publication 18 September 1986.
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Fig. 1 Giant cell arteritis of a myometrial artery. Mononuclear cells infiltrate the wall, and the vessel is ringed by giant cells. The lumen is compromised by intimal proliferation. (Haematoxylin and eosin.)

Fig. 2 Arteriogram showing occlusion of the right axillary artery.

6–13 g/l, IgM 2.6 g/l (NR 0.4–2.2 g/l), immune complexes 38% (NR 0–24%). Urea and electrolytes, liver tests, Rose-Waaler test, creatine phosphokinase, autoantibodies, thyroxine, thyroid stimulating hormone, complement, chest and hip x rays, and electrocardiogram were normal or negative.

Arch aortography was carried out. This showed unfolding of the aortic arch and some dilatation and tortuosity of the innominate and both subclavian arteries, with occlusion of the right axillary artery over 4 cm (Fig. 2) and almost complete occlusion of the left axillary artery. There was a moderately good collateral supply with good distal run off bilaterally and the brachial, radial, and ulnar arteries were of reasonable calibre. These changes were felt to represent atheromatous dilatation and tortuosity proximally, but the blocks and stenosis in the axillary arteries were felt to be due to arteritis.

A diagnosis of PMR and GCA affecting myometrial and axillary arteries was made, and treatment with prednisolone 60 mg/day started. Her left arm returned to normal and she has only occasional claudication in the right arm. The radial pulse is now palpable but reduced and her blood pressure is recordable in the right arm. She has full, pain free movement in both hips. Her ESR is 7 mm/h and prednisolone dose 14 months after starting treatment is 7.5 mg/day.

Discussion

This woman had an extremely rare presentation of PMR/GCA. There are only seven reports in the literature of nine cases of arteritis of uterus, ovaries, or fallopian tubes. In none of these cases did GCA cause gynaecological symptoms. Six patients had features of PMR/GCA. Our patient conformed to this pattern, in that the GCA was an
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incidental finding on hysterectomy, though on further questioning she did have features of PMR.

She then developed another unusual manifestation of GCA, arm ischaemia secondary to arteritis of the axillary arteries. Aortic arch involvement was first described in 1938. The typical findings are arm claudication, reduced or absent radial and ulnar pulsation, and on arteriography long segments of smooth stenosis affecting subclavian, axillary, or brachial arteries. Although symptomatic improvement occurs rapidly after treatment with steroids, it takes a mean of 23 months for pulses to return and surgery is not helpful.

The features of classical PMR/GCA present in almost all the reported cases of gynaecological PMR/GCA suggest that this is not an atypical arteritis but GCA in an unusual site, and it should be treated with oral corticosteroids in the standard way.

References