Case report

Successful treatment of dissecting aortic aneurysm due to giant cell arteritis

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SUMMARY A 72-year-old woman with polymyalgia rheumatica clinically controlled on maintenance steroid therapy presented with symptoms of chest pain and numbness in the right arm. A diagnosis of dissecting aortic aneurysm was confirmed at thoracotomy and the aorta was successfully resected. Histology revealed active giant cell aortitis. We suggest that a normal erythrocyte sedimentation rate in patients with treated temporal arteritis does not preclude large vessel involvement.

Giant cell aortitis is a rare cause of dissecting aortic aneurysm. The combination of temporal arteritis and dissecting aneurysm has previously been found only at necropsy.¹ We describe the case of a 72-year-old woman with well controlled temporal arteritis who was successfully treated for a dissecting aortic aneurysm due to giant cell arteritis. We believe this to be the first recorded case.

Case history

In November 1978 a 70-year-old retired nurse was referred by her general practitioner with a 2-week history of sudden onset of severe, right-sided temporal headache and scalp tenderness. She had had no visual disturbance, joint pains, night sweats, or polymyalgia, though over the previous 4 months she had felt lethargic and unwell. Her past history was unremarkable.

General examination revealed tenderness over the right parietal region, but both temporal arteries were pulsatile and not swollen. The pulse was 84/min, regular. All peripheral pulses were present. No bruits were heard. The blood pressure was 170/110 mmHg. Examination was otherwise normal.

Laboratory investigations (normal ranges in brackets) were as follows: haemoglobin 11 g/dl; white cell count and platelets normal; erythrocyte sedimentation rate (ESR) 94 mm/hour; Rose-Waaler test negative; urea, electrolytes, and liver function tests normal, apart from alkaline phosphatase 127 U/l (30-92). Protein electrophoresis showed marked acute phase reaction. Immune complexes

were elevated at 61% (0-24). Syphilis serology negative. Chest x-ray showed slight left ventricular enlargement, and the aorta was unfolded, tortuous, with some calcification in the arch. Temporal artery biopsy gave a normal result.

A diagnosis of temporal arteritis was made despite the normal biopsy and she was started on high-dose steroids.

In December 1979 she was asymptomatic on prednisone 5 mg daily. Blood pressure was 150/80 mmHg, controlled with atenolol and a thiazide diuretic, and her ESR was 21 mm/hour. Immune complexes were normal.

One month later she was admitted as an emergency complaining of severe interscapular and right shoulder pain with coldness and numbness in the right arm. She was in pain at rest. The right arm was noticeably colder than the left, and the brachial and radial pulses were absent. The remainder of the peripheral pulses were equal, and there were no bruits or radial-femoral delay. The pulse was 84 regular, of normal volume (left radial), blood pressure 140/80 mmHg (left brachial), apex beat at the 5th intercostal space in the anterior axillary line. Heart sounds were normal, with no murmurs. There was no evidence of heart failure.

Chest x-ray showed widening of the mediastinum. The ring of calcification in the aorta was separate from the edge of the aortic shadow by 6 mm (Fig. 1). The features were of aortic dissection. ESR was 26 mm/hour.

Aortography within 36 hours of admission revealed the presence of an extensive dissection involving the whole of the ascending aorta and passing into the arch to involve the innominate artery and descending

Fig. 1 Chest x-ray showing widening of the mediastinum and separation of the ring of aortic calcification from the aortic shadow.

aorta. These findings were confirmed at emergency operation with cardiopulmonary bypass. The ascending aorta was resected with a Dacron graft after the dissecting layers of the aorta had been reconstituted.

Postoperative recovery was uneventful, and six months later she remained well. Histology of the aorta showed giant cell aortitis (Fig. 2) with necrotic foci.

Fig. 2 Section of the ascending aorta showing an area of giant cell infiltration. (Haematoxylin and eosin, $\times 270$).

Discussion

Giant cell aortitis was first reported in 1937 by Sproul and Hawthorne.² In 1950 Magarey³ reported the post-mortem findings of dissecting aneurysm due to giant cell aortitis without features of temporal arteritis. Subsequently temporal arteritis and aortic dissection have been found at necropsy.¹ Klein *et al.*⁴ in a review of 248 patients with giant cell arteritis found 34 with 'definite' or 'possible' large artery involvement. The clinical findings were of upper extremity claudication, bruits over the carotid, subclavian, axillary, and brachial arteries, and absence or diminution of pulses in the neck or arms. Raynaud's phenomenon was also common in these patients. Dissecting aneurysm was the cause of death in 3 patients, all with clinical evidence of aortic disease. They found that large vessel involvement was invariably associated with raised ESR. Jones and Hazleman⁵ described 3 patients with well controlled disease who developed sudden blindness despite treatment. Prior to dissection this patient was asymptomatic and ESR was normal. Interestingly, her original temporal artery biopsy was negative, and this underlines the nature of the disease and the importance of diagnosing temporal arteritis despite a normal biopsy. Harrison *et al.* recommends that patients with temporal arteritis should routinely have the blood pressure measured in both arms and pulses in both arms and legs palpated and auscultated. Symptoms of intermittent claudication and Raynaud's phenomenon should also be sought.⁶ We suggest that a normal ESR in patients with treated temporal arteritis does not preclude large vessel involvement and that, when there is such involvement, higher doses of steroid therapy may be required to prevent large vessel rupture.

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