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Aortic valve replacement for severe aortic regurgitation caused by idiopathic giant cell aortitis

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ABSTRACT Giant cell aortitis occurred in a 25-year-old woman, with absent pulses in the left arm and severe aortic regurgitation from dilatation of the valvar annulus. The aortic valve was replaced by a Starr-Edwards prosthesis, and the patient was treated with steroids. Five years later, she continues asymptomatic and haemodynamically stable. The left brachial and radial pulses have returned.

Giant cell aortitis can be a cause of aortic regurgitation, but it is rarely of such severity as to warrant valve replacement. A review of published reports showed only four such cases.[1-3] Two of these patients also had aneurysmal dilatation of the ascending aorta.[1 2] We report a case of giant cell aortitis in a 25-year-old woman who had absent pulses in the left arm and severe aortic regurgitation from dilatation of the valve ring. The patient was successfully treated by aortic valve replacement and steroids.

Case report

A 25-year-old woman was admitted to hospital in January 1974 with a two-year history of increasing tightness in the chest, dyspnoea on exertion or excitement, and frequent temporal headaches. She gave no history of rheumatic fever, chorea, arthritis, tuberculosis, venereal disease, or polymyositis, and there was no family history of heart disease. Her right eye had been enucleated for retinoblastoma at the age of 2 years. Three years before her present admission she had an ovarian cystectomy that was followed by pulmonary embolism, necessitating anticoagulation. Six months later, on a routine medical examination, she was found to have a precordial aortic diastolic murmur.

She was pale and had absent left brachial and radial pulses and feeble pulses in the right arm. The pulse was regular. The blood pressure in the right arm was 100/65 mmHg, and 260/60 mmHg in the legs. It was unrecordable in the left arm. The jugular venous pressure was normal, but there was prominent carotid pulsation. The apex beat was diffuse and heaving, with a diastolic thrill over the left parasternal border. Both heart sounds were audible with a harsh, ejection systolic murmur, grade 3/6 in the aortic area, loudest over both the carotid arteries and a grade 3/6 early diastolic murmur along the left parasternal border. There was no hepatomegaly, ascites, or oedema.

A chest radiograph showed moderate cardiomegaly with pulmonary venous congestion in the upper lobes. The electrocardiogram showed sinus rhythm, and left ventricular hypertrophy. The haemoglobin was 10·7 g/dl, the haematocrit 33%, and the erythrocytes normochromic and normocytic. The white cell count was $5{\cdot}4\times10^9$/l and the ESR was 58 mm in one hour (Westergren). Blood urea, serum electrolytes, and liver function tests were all normal. C-reactive protein was positive. Antistreptolysin titre, antinuclear factor, and serology for rheumatoid factor were all negative. No LE cells were seen. Cardiac catheterisation data (right and left) are shown in the table, confirming severe left ventricular dysfunction. Aortography showed gross aortic regurgitation, greatly increased left ventricular volume, and poor contractions (ejection fraction 45%). No mitral regurgitation was seen.

At operation, under standard cardiopulmonary bypass with coronary perfusion, the ascending aorta was found to be oedematous and grossly thickened (fig 1), which gave rise to considerable difficulty with aortic cannulation. The intima

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Table *Cardiac catheterisation data (mean pressure in parentheses)*

Cardiac chamber	*Pressure (mmHg)*	
Right atrium	x −4, v 4, y 2	(1)
Right ventricle	36/4	
Pulmonary artery	38/18	(25)
Pulmonary artery wedge	a 20, x 16, v 22, y 12	(18)
Aorta	140/40	(65)
Left ventricle	140/15 a 30	
Cardiac index (l/min/m²)	2·8	
Left ventricle ejection fraction	45%	

looked scarred, and was covered with pale opaque areas. The aortic cusps showed some shortening, but the basic lesion was dilatation of the valve ring causing aortic incompetence. The valve was excised and replaced by a cloth-covered, No 2320, size 11-A Starr-Edwards prosthesis, and a biopsy specimen of the aortic wall was taken.

Histologically there was a severe inflammatory infiltration that affected the outer half of the media and the adventitia (fig 2). The inflammatory cells were mainly lymphocytes, with a few histiocytes and plasma cells. Scattered multinucleated giant cells were noted in the media (fig 3). There was much adventitial fibrosis and endarteritis of occasional vasa vasorum. The picture was that of a florid giant cell aortitis associated with extensive medial destruction. The aortic cusps were thin and normal in structure on histological examination.

Recovery was complicated by atrial fibrillation that spontaneously reverted to sinus rhythm. The patient was given prednisone 15 mg thrice daily in addition to anticoagulants and digoxin. After six months the left brachial and radial pulses were palpable, the ESR had returned to normal, and the C-reactive protein was consistently negative. On her last follow-up, five years after operation, she was asymptomatic and haemodynamically stable with a blood pressure of 130/90 mmHg in both arms. The heart had returned to normal size, but the ECG still showed evidence of slight left ventricular hypertrophy. At present she takes a small maintenance dose of prednisone.

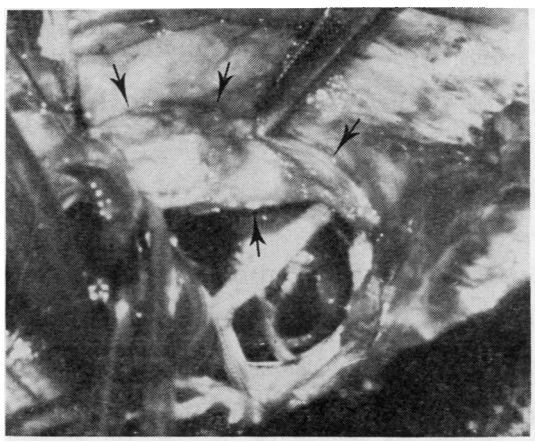

Fig 1 *Grossly thickened and oedematous ascending aorta (arrows).*

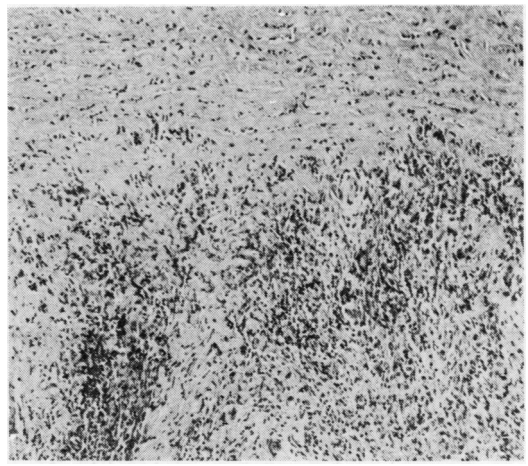

Fig 2 *Aortic wall, showing heavy inflammatory infiltration in outer media and adventitia. Haematoxylin and eosin, original magnification ×80.*

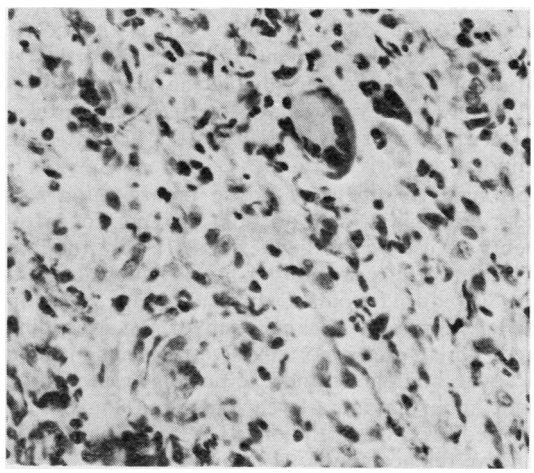

Fig 3 *Giant cells admixed with lymphocytes and histiocytes in aortic media. H & E, original magnification ×250.*

Discussion

The aetiology of giant cell aortitis is unknown. The panarteritis that occurs may be indistinguishable from that recorded in Takayasu's disease, and the differential diagnosis is based almost entirely on the distinctive age incidence. Nevertheless, Takayasu's disease, the onset of which is usually between the ages of 10 and 20 years, has occasionally been diagnosed in the elderly, and giant cell arteritis is not exclusively a disease of the older age groups. There is the same female preponderance in the aortic arch syndrome from giant cell arteritis as in classical Takayasu's disease.[4] The suggestion by Cairns and Oleesky[5] that the two diseases represent a single entity and form components of a continuous spectrum of inflammatory arterial disease has much to recommend it. The proposal by Hall,[6] that the term "idiopathic arteritis" should be applied to cranial arteritis, polymyalgia rheumatica, and Takayasu's disease, since they possibly represent different manifestations of one underlying disorder, is rational.

Current opinion favours an autoimmune aetiology. Antiarterial antibodies have been shown in some cases,[7 8] but whether they are primarily implicated in the arterial damage is uncertain.

The aortic medial inflammation and necrosis that characterise the disease may extend into or cause obstruction of the major vessels arising from the arch of the aorta, resulting in inequality or absence of the pulses, as observed in this case. Stenosis or dilatation of the aortic lumen may occur with aneurysm formation[9 10] and dissection may develop, especially in the presence of hypertension.[11] The dilatation may affect the aortic valve ring causing varying degrees of aortic regurgitation. On the other hand, the aortic involvement may be asymptomatic, as has been proved at necropsy in some patients who present with temporal arteritis.

Surgical treatment of aortic regurgitation secondary to giant cell aortitis is reported infrequently. Austen and Blennerhassett[1] were the first to treat aortic regurgitation by valve replacement in a 30-year-old woman who had absent pulses in the left arm and an aneurysm of the ascending aorta. Gula *et al*[2] reported valve replacement with an aortic homograft and excision of an ascending aortic aneurysm in a 61-year-old man. Honig *et al*[3] reported two cases with aortic regurgitation treated by valve replacement.

The course of giant cell arteritis varies in different patients. In some it becomes arrested with or without treatment, whereas in others it leads to death. Steroids have been effective in some patients, and in the present case there was a dramatic improvement after a course of prednisone, the brachial and radial pulses returning and the ESR becoming normal, suggesting an arrest of the disease. The patient is asymptomatic five years after valve replacement while continuing to take a small dose of prednisone. This is the longest reported follow-up after operation. Treatment with steroids, therefore, appears to be justified after aortic valve replacement in these cases. None of the other patients treated surgically was given steroids after operation.

The clinical diagnosis of giant cell aortitis as a cause of rapidly progressive aortic regurgitation is extremely difficult. Nevertheless, it should be suspected in a relatively young patient with no history of rheumatic fever but with polymyalgia, headaches, pyrexia, malaise, or weight loss. In addition some patients may have absent or diminished pulses, considerably raised ESR, and normochromic, normocytic anaemia. Final confirmation can only be achieved by biopsy.

We thank Dr D Boyle who referred this patient for operation.

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