Case reports

British Heart Journal, 1978, **40**, 1052-1054

Aortic regurgitation as a manifestation of giant cell arteritis

J. HOW¹ AND R. W. STRACHAN

From the Department of Medicine, Dumfries and Galloway Royal Infirmary, Dumfries

SUMMARY The detailed clinical findings of a 65-year-old woman who developed aortic regurgitation caused by giant cell aortitis are presented. The initial phase of the disease was dominated by severe non-specific constitutional symptomatology suggesting infective endocarditis or a malignancy. Aortic regurgitation as a manifestation of giant cell arteritis has hitherto received scant attention in the published reports. The clinical and therapeutic relevance of this masquerade is discussed.

Although it has long been established that giant cell arteritis is a generalised vascular disease (Gilmour, 1941; Cooke *et al.*, 1946), major involvement of the aorta or its branches has been poorly recognised by clinicians (Klein *et al.*, 1975). In this paper, we wish to draw attention to the occurrence of aortic regurgitation as a manifestation of giant cell aortitis and to discuss the therapeutic importance of recognising this association.

Case report

The patient, a 65-year-old white woman, was first seen by one of the authors in March 1972. She had been admitted to a surgical ward a few weeks earlier having been unwell for 5 months, with increasing tiredness, anorexia, a gradual loss of 19 kg in weight and fairly constant aching discomfort in the central abdomen and right iliac fossa. In 1965 and 1971 a urethral carbuncle had required treatment by diathermy and during this period recurrent frequency of micturition had proved repeatedly to be caused by infection with *Esch. coli*. On this occasion though intermittent mild fever was noted, urine cultures, blood urea, and an intravenous pyelogram were all normal. The erythrocyte sedimentation rate (ESR), however, was repeatedly over 100 mm in the first hour and the haemoglobin level persistently low at 9.7 g/dl (66%). The peripheral blood film showed only mild anisocytosis and occasional hypochro-

masia with normal white cell morphology and distribution.

Physical examination was unhelpful, there being no definite abnormality in the abdomen and no clinical features suggesting myelomatosis or leukaemia. The blood pressure was 130/80 mmHg and there were no cardiac murmurs. The temporal arteries were easily located, being non-tender and pulsatile. No bruits were heard over the neck or abdomen. Chest x-ray film, studies of liver function, tests for faecal occult blood, cholecystogram, blood cultures, barium meal, sternal marrow aspirate, electrocardiogram, and cultures of urine and sputum for tubercle bacilli all showed no abnormality. A barium enema disclosed no lesions other than mild diverticular disease. The serum iron level was low at 40 µg/100 ml (7.1 µmol/l), but stainable iron was present in the marrow smears. Serum protein electrophoresis showed a diffusely increased gamma-globulin of 1.93 g/100 ml and a search for Bence Jones protein was negative. Her Wassermann reaction and tests for rheumatoid factor and lupus erythematosus cells were negative. Red cells were identified on urinary microscopy on several occasions.

Since she remained unwell a month after admission to hospital, with persistence of pronounced anaemia and gross rise in the ESR, a laparotomy was carried out but no abnormality was shown. She was transferred to the medical unit where further observation and studies failed to establish a diagnosis. She was finally discharged nearly a month later, there having been moderate improvement

¹Present address: Department of Therapeutics and Clinical Pharmacology, University of Aberdeen, Foresterhill, Aberdeen.

Fig. 1 Chest x-ray film showing dilatation of aorta.

with no specific treatment. At that time the haemoglobin was 11.0 g/dl (75%), the ESR 6 mm in the first hour, and her weight was 70.7 kg. Five months later she had gained 12.7 kg in weight, though she was still tired. The ESR was 8 mm in the first hour and the haemoglobin 13.7 g/dl (94%).

Two years later in 1974 she was again seen complaining of tiredness, unpleasant throbbing in the neck, and intermittent heavy retrosternal discomfort. Her weight had risen further to 90.7 kg, the

ESR being normal and the haemoglobin 14.1 g/dl (97%). There was now, however, a grade 2/6 mid-systolic murmur at the base and an immediate soft diastolic murmur at the left sternal edge, indicating mild aortic regurgitation. The blood pressure was 160/85 mmHg and chest film showed dilatation of the aorta (Fig. 1). The temporal arteries still appeared normal to clinical examination, but biopsy of the right artery confirmed a diagnosis of giant cell arteritis. X-ray films of the spine and sacroiliac joints showed only minor osteoarthritic changes. Observation over the next 5 months showed a fall of 12.7 kg in weight and suggested that, despite the normal ESR, the aortic regurgitation was increasing in severity. She continued to complain of aching discomfort in the front and back of the chest and an archaortogram was carried out to assess the severity and extent of the aortic disease. This showed distinct dilatation of the whole of the thoracic aorta and its branches (Fig. 2). Treatment with prednisolone 3 mg t.d.s. was initiated and within a few weeks she reported clearing of the fatigue and the pains in the chest which had troubled her for over a year. At the last assessment in December 1976 she remained well and there were neither clinical nor electrocardiographic features to suggest deterioration of the aortic regurgitation.

Discussion

Giant cell arteritis which primarily affects elderly patients is not a rare disease but the many guises under which it may present is undoubtedly the major factor responsible for diagnostic failures (Paulley and Hughes 1960; Hamilton *et al.*, 1971). In addition, Paulley and Hughes (1960) have emphasised that, 'in remission it may only sleep' and 'if untreated it may progress slowly leading to serious complications or sometimes more rapidly to a fatal termination'. The combination of anaemia, weight loss, and rapid ESR in the elderly raises apprehensions of occult malignancy, and, as in the present case, the patient is often subjected fruitlessly to a wide range of investigations including exploratory laparotomy. Despite our interest in the various arteritic syndromes (Strachan, 1964, 1966) it was more than two years after the initial presentation that the finding of aortic regurgitation prompted the diagnosis of an underlying giant cell arteritis. Though the temporal arteries were 'normal' clinically, the histological features on biopsy were typical of active giant cell arteritis, emphasising the importance of the asymptomatic temporal artery as a source of histological and arteriographical confirmation of the diagnosis in possible cases (Gillanders *et al.*, 1969; Bonventre, 1974). Un-

Fig. 2 Aortogram showing dilatation of the whole of the thoracic aorta and its branches.

fortunately, there is still great reluctance on the part of clinicians to undertake these investigations in the absence of tenderness or pulselessness of a temporal artery.

Aortic regurgitation as a manifestation of giant cell arteritis has received very scant attention (Paulley and Hughes, 1960; Austen and Blennerhassett, 1965; Castleman and McNeely, 1967, 1971). Textbooks of cardiology and comprehensive reviews (Roberts *et al.*, 1973) list extremely rare and untreatable causes of aortic regurgitation but fail to mention giant cell arteritis. It is, therefore, not surprising that this association may be completely overlooked during life resulting in years of ill health with confirmation of the diagnosis occurring only at necropsy (Castleman and McNeely, 1971). It probably occurs more frequently than previously suspected, accounting for some of the cases of 'negative culture' endocarditis. Fauchald *et al.* (1972) found 3 instances of aortic regurgitation in the course of observing 35 biopsy proven cases of giant cell arteritis, though these authors did not present any clinical data.

The pathogenesis of the aortic regurgitation is probably dilatation of the aortic valve ring but necropsy reports are very limited (Castleman and McNeely, 1971). The radiological finding of pronounced dilatation of the ascending aorta in our patient suggests that a similar mechanism is responsible for the aortic regurgitation. Consequently, in patients with suspected or proven giant cell arteritis, careful and regular auscultation is recommended to identify not only vascular bruits but also the aortic regurgitant murmur.

The recognition of aortic involvement as a manifestation of giant cell arteritis has therapeutic implications. Early diagnosis and treatment with corticosteroids may prevent aortic rupture and death or occlusions of its major branches (Klein *et al.*, 1975). Ill-defined constitutional symptoms in association with established or developing aortic regurgitation may, despite negative blood cultures, be interpreted as indicating infective endocarditis and the patient may be subjected to at least 6 weeks of intensive antibiotic therapy (Oakley, 1974). Moreover, delay or failure to reach the correct diagnosis leaves the patient at great risk of developing serious complications, such as blindness and stroke.

The present case, together with the limited number of previous reports, emphasize that the possibility of giant cell arteritis should be entertained in patients presenting with vague symptoms and aortic regurgitation. The latter manifestation is a facet of the disease which is of undoubted relevance to the cardiologist.

References

- Austen, W. G., and Blennerhassett, J. B. (1965). Giant-cell aortitis causing an aneurysm of the ascending aorta and aortic regurgitation. *New England Journal of Medicine*, **272**, 80-83.
- Bonventre, M. V. (1974). Temporal arteritis. *New York State Journal of Medicine*, **74**, 483-488.
- Castleman, B., and McNeely, B. U. (1967). Case records of the Massachusetts General Hospital. *New England Journal of Medicine*, **276**, 106-114.
- Castleman, B., and McNeely, B. U. (1971). Case records of the Massachusetts General Hospital. *New England Journal of Medicine*, **284**, 1201-1210.
- Cooke, W. T., Cloake, P. C. P., Govan, A. D. T., and Colbeck, J. C. (1946). Temporal arteritis: a generalised vascular disease. *Quarterly Journal of Medicine*, **15**, 47-76.
- Fauchald, P., Rygvold, O., and Øystese, B. (1972). Temporal arteritis and polymyalgia rheumatica. Clinical and biopsy findings. *Annals of Internal Medicine*, **77**, 845-852.
- Gillanders, L. A., Strachan, R. W., and Blair, D. W. (1969). Temporal arteriography. A new technique for the investigation of giant-cell arteritis and polymyalgia rheumatica. *Annals of the Rheumatic Diseases*, **28**, 267-269.
- Gilmour, J. R. (1941). Giant-cell chronic arteritis. *Journal of Pathology and Bacteriology*, **53**, 263-277.
- Hamilton, C. R., Shelley, W. M., and Tumulty, P. A. (1971). Giant-cell arteritis: including temporal arteritis and polymyalgia rheumatica. *Medicine*, **50**, 1-27.
- Klein, R. G., Hunder, G. G., Stanson, A. W., and Sheps, S. G. (1975). Large artery involvement in giant cell (temporal) arteritis. *Annals of Internal Medicine*, **83**, 806-812.
- Oakley, C. M. (1974). Infective endocarditis. *British Journal of Hospital Medicine*, **11**, 101-108.
- Paulley, J. W., and Hughes, J. P. (1960). Giant-cell arteritis, or arteritis of the aged. *British Medical Journal*, **2**, 1562-1567.
- Roberts, W. C., Dangel, J. C., and Bulkley, B. H. (1973). Nonrheumatic valvular cardiac disease: a clinicopathologic survey of 27 different conditions causing valvular dysfunction. *Cardiovascular Clinics*, **5**, 333-446.
- Strachan, R. W. (1964). The natural history of Takayasu's arteriopathy. *Quarterly Journal of Medicine*, **33**, 57-69.
- Strachan, R. W. (1966). Pre-pulseless and pulseless Takayasu's arteritis. *Postgraduate Medical Journal*, **42**, 464-468.

Requests for reprints to Dr J. How, Department of Therapeutics and Clinical Pharmacology, University of Aberdeen, Foresterhill, Aberdeen AB9 2ZD.