

## Case report

# Aorto-atrial fistula in rheumatoid arthritis

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**SUMMARY** A patient with severe deforming rheumatoid arthritis presented with a short history of chest pain. The clinical signs were of an unusual diastolic murmur and profound shock unresponsive to therapy. Post-mortem examination disclosed the unexpected finding of a large aorto-atrial fistula.

Large intravascular communications are common in congenital heart disease where the aorta may be joined to the atria, ventricles, or pulmonary artery. Fistulae from the aorta also occur in adult life and present usually, if large, as catastrophic events or, if small, with slowly developing physical signs of a shunt. They may be preceded by an infective illness, trauma, or surgery involving the aorta or aortic valve. We present a patient with no features of intravascular shunting in whom a large aorto-atrial fistula had obviously been present for some time.

### Case report

A 65-year-old female was admitted with severe lower chest pain. The patient was known to have seropositive, nodular rheumatoid arthritis and had developed numerous complications of the disease. These included anaemia, keratoconjunctivitis, hypopyon, Felty's syndrome, pulmonary fibrosis, and pleural and pericardial effusions. She eventually became bedridden, when she required an amputation for persistent infection in a deformed foot. She had never received steroid therapy, as a gastric ulcer had precluded this form of treatment. In the 12 months prior to this episode she had suffered 2 similar bouts of chest pain requiring hospital admission. On both occasions she was noted to have a pansystolic apical murmur; serial cardiac enzymes remained normal, and apart from a short period of asymptomatic atrial tachycardia her electrocardiograms were unchanged. She was treated with digoxin and verapamil for this arrhythmia.

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On admission she was nauseated, short of breath, and in pain. Her therapy consisted of simple analgesics plus the above-mentioned antiarrhythmic drugs. On examination, apart from her obvious nodular rheumatoid disease, she was pale, sweaty, and tachypnoeic. Her pulse was slow, with unrecordable blood pressure. The venous pressure was grossly elevated, and on auscultation, in addition to the previously noted soft apical systolic bruit, there was a soft early decrescendo diastolic bruit audible only in the right mid clavicular line at the fourth interspace. Her abdomen was soft, and all pulses were present and synchronous. Electrocardiography showed slow atrial fibrillation and no changes from her previous recordings. The heart was enlarged on chest x-ray, but the lung fields showed no evidence of left ventricular failure. The patient's condition deteriorated rapidly, and despite ventilation and treatment with inotropic drugs and steroids she died shortly after admission.

Post-mortem examination revealed the previously mentioned multisystem lesions associated with the rheumatoid arthritis. In the thorax there were pleural and pericardial adhesions. The pulmonary arteries were normal, the aorta contained a number of small areas of atherosclerosis, and the coronary arteries were normal. On the posterolateral aspect of the ascending aorta 3 cm above the aortic valve a large, smooth-edged defect 1 cm in diameter was found. This circular defect (Fig. 1) joined the aorta to the superior vena cava and right atrial junction. A large laminated thrombus occupied most of the dilated right atrium and almost entirely occluded the venous side of the fistula. All the cardiac valves were normal, no ventricular hypertrophy was present, and no other abnormalities were found. On histological examination the endocardium and myocardium appeared



Fig. 1 This section of the aorta shows the aortic valve below, with the ascending aorta extending upwards. The large aorto-atrial defect (arrowed) may be clearly seen in the centre of the illustration. (A suture has been inserted where the specimen was inadvertently sectioned.)

normal. The pericardium showed nonspecific fibrous adhesions but no definite evidence of rheumatoid change. Sections from the edge of the aorto-atrial fistula showed normal tissues, with no evidence of inflammation of granuloma formation. The aorta and aortic valve were similarly normal.

The cause of death was not myocardial infarction or pulmonary embolism but probably tricuspid valve occlusion by the intra-atrial thrombus leading to shock, with raised venous pressure.

### Discussion

The haemodynamic features of arteriovenous fistulae depend on the size of the communicating vessels and pressure within them, the size of the fistula, and the resistance to flow in the low-pressure system.<sup>1</sup> Fistulae between large central vessels

present early with serious haemodynamic effects, whereas smaller communications in peripheral vessels usually present with localised signs.

Congenital fistulae may communicate between the aorta and the atria, ventricles, or pulmonary arteries.<sup>1,2</sup> However, apart from this fistulae having a smooth edge there is little to suggest that it could have been present since birth. The site of the defect, the absence of any aneurysmal formation, the patient's age, and the absence of any right ventricular hypertrophy secondary to left-to-right shunting make it unlikely that this fistulae was congenital in origin.

All the usual causes of acquired aorto-atrial fistulae may be excluded in our patient.<sup>3</sup> There was no history of blunt or penetrating trauma. The aortic valve and root showed no evidence of existing or previous infection. There was also no evidence of aortic dissection, which can erode into the vena cava.<sup>4,5</sup>

The association of rheumatoid arthritis and cardiovascular disease is well recognised, with pericarditis, valvular abnormalities, arteries, and nodular formation being the commonest manifestations.<sup>6,7</sup> These intracardiac granulomata may degenerate after infection, necrosis, or haemorrhage,<sup>8,9</sup> and a coronary sinus fistulae has been reported following necrotic degeneration.<sup>10</sup> One possible cause of our patient's fistulae could be the regression, either spontaneous (as occurs with peripheral nodules of similar histology) or secondary, of a granuloma in the aortic wall. Another possibility is spontaneous aortic rupture, as has been reported previously in rheumatoid arthritis,<sup>11,12</sup> and subsequent erosion into the right atrium.

Aortic bruits in rheumatoid arthritis are usually due to valvular abnormalities. We describe a rare but lethal cardiac complication of severe rheumatoid arthritis the only clue to whose diagnosis in life was an unusually sited murmur.

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