

Editorial

Cardiac sarcoidosis

Sarcoidosis is characterised by multisystem disease with granulomatous infiltration of the lungs, reticulo-endothelial system and skin in particular, though almost any organ may be affected. Pulmonary granulomas are the most common manifestation, and these may lead to diffuse fibrosis, pulmonary hypertension, and right heart failure. Primary sarcoid disease of the heart is uncommon clinically but may be found at necropsy in up to a third of cases, in most of which non-caseating granulomas are present in many organs.¹

Myocardial sarcoidosis is notoriously hard to diagnose; it may remain occult even when extensive and may present with sudden death.² If the diagnosis is not to be missed sarcoid heart disease should be considered in any patient (particularly if young) with cardiac arrhythmia, conduction defects, or unexplained heart failure. Sarcoid infiltration is one of the few causes of focal left ventricular abnormality in the absence of disease of the major coronary arteries. The distribution is typically in the proximal part of the left ventricle and upper septum,^{3,4} where it produces akinesia of the affected region; with healing the previously thick akinetic wall becomes thin or even aneurysmal. The appearance of proximal akinesia and dilatation but a vigorously contracting apex of the left ventricle contrasts with the much more common anteroapical akinesia seen after an extensive infarct in the territory of the anterior descending artery when the proximal ventricle contracts well. It differs also from the distribution of infarction following occlusion of any combination of major coronary artery branches. In cardiac sarcoidosis a left ventricular aneurysm may develop adjacent to the mitral annulus or mitral regurgitation may result from infiltration of papillary muscles.⁵ Mitral stenosis from infiltration of the mitral leaflets has also been reported.⁶ Microscopic granulomas may cause arrhythmias, conduction defects, or sudden unexplained death, before which myocardial function may have appeared to be entirely normal.

In this issue Wait and Movahed describe typical angina or atypical chest pain in 12 out of 43 black Americans with sarcoidosis.⁷ They suggest that anginal chest pain is common in black male patients with sarcoidosis and that it may signal sarcoid infiltration of the myocardium.

Fleming⁸ recently reported a series of 138 fatal cases in 300 patients with sarcoid heart disease, who differed from those studied by Wait and Movahed in that nearly all were white. Cardiovascular features at presentation in those reported by Fleming were ventricular or supraventricular arrhythmias in 73%, complete heart block in 26%, and "cardiomyopathy" in 24%. A further 61% had right or left bundle branch block or partial heart block. Only 15 patients (5%) had a presentation that simulated myocardial infarction, and presumably therefore had chest pain.

Anginal pain is not mentioned, even though the average age at presentation was 50 years and more than half of the patients were male, so that some at least are likely also to have had atheromatous coronary artery disease. Sudden death occurred in 77 cases in Fleming's series, in 49 of which cardiac sarcoidosis had been previously undiagnosed; this is acknowledged to be the most common manifestation of cardiac sarcoidosis, though many patients experience antecedent arrhythmias or atrioventricular block.

All of the American patients with sarcoidosis who had chest pain and a matched control group with sarcoidosis but without chest pain plus a control group of normal subjects underwent cardiopulmonary exercise studies. None had electrocardiographic evidence of ischaemia, abnormalities of rhythm, or conduction disturbances but the patients with angina had lower maximum heart rates and double products than those without chest pain. Both those with chest pain and those without had significantly lower anaerobic thresholds than normal control subjects but did not differ from each other. Ten of the patients with chest pain had abnormal thallium scans, of whom six had reversible perfusion defects suggesting coronary artery disease, but angiography showed normal coronary arteries. All had normal pulmonary artery pressure, left ventricular ejection fraction, and valvular function. Thallium scans carried out in nine control

patients with sarcoidosis but without chest pain were all normal.

Infiltration of small coronary artery branches may be found⁹ in cardiac sarcoidosis and in some cases there may be considerable vasculitis.⁸ This is an important part of the pathological process, and suggests that such patients should respond to corticosteroids. As with hypercalcaemia and sarcoid disease of the eye, infiltration of the myocardium in sarcoidosis is accepted as an indication for steroid treatment but the response is hard to judge. Fleming suggests that these drugs should be used more aggressively and for longer than is commonly done.

Myocardial ischaemia in patients with vasculitis could well explain the anginal pain and the perfusion defects seen on the thallium scan in the patients of Wait and Movahed. Such perfusion defects have been described previously in patients with sarcoidosis¹⁰ but chest pain has not been a prominent feature in other series or linked to abnormal thallium scans. Similar perfusion defects have been described in scleroderma.¹¹ Microvascular spasm has been postulated both in dilated cardiomyopathy¹² and in scleroderma¹¹ and Wait and Movahed suggest that this may be responsible for the chest pain and the defects on the thallium scan in sarcoidosis. Spasm of the extramural coronary arteries was thought to be unlikely. Some support for the concept of microvascular spasm was found in the response to nitrates seen in most of the patients but this would not distinguish between ischaemia due to sarcoid disease and ischaemia due to other causes. Arteritis, as was shown so well in the papers of James⁹ and Fleming,⁸ provides a more convincing explanation.

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