Myocardial sarcoidosis

A case report

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Summary

A 63-year-old man with sarcoidosis developed recurrent ventricular tachyarrhythmias, intermittent heart block and congestive cardiac failure. Transvenous endomyocardial biopsy demonstrated sarcoid infiltration of the myocardium. Ventricular tachyarrhythmias were abolished with amiodarone and prednisone.


Sarcoidosis is a systemic granulomatous disease which may affect the heart. Myocardial sarcoidosis, which may cause life-threatening rhythm or conduction disturbances, congestive cardiac failure and sudden death, is now considered an underdiagnosed and potentially treatable cardiac disease, but whether steroid therapy improves the poor prognosis of myocardial sarcoidosis is undetermined. Myocardial sarcoidosis is often difficult to diagnose with certainty during life. In the case reported here histological proof of the diagnosis was obtained by endomyocardial biopsy. Amiodarone and prednisone abolished the ventricular tachyarrhythmias.

Case report

A 63-year-old Black man gave a 2-year history of lower retrosternal chest pain unrelated to effort but associated with bouts of palpitations, swelling of the legs and breathlessness. He had been treated intermittently for biventricular failure. In December 1979 a chest radiograph (Fig. 1) showed mediastinal widening and tomography confirmed hilar lymphadenopathy. Scalene node biopsy (Fig. 2) showed extensive granulomatous infiltration with numerous giant cells and no evidence of necrosis. Stains for acid-fast bacilli and fungi were negative and appearances were compatible with sarcoidosis.

In October 1980 he was admitted to hospital with further chest pain and severe palpitations. The ECG showed ventricular tachycardia which was terminated with intravenous lignocaine. On subsequent examination he was in sinus rhythm, with a blood pressure of 145/80 mmHg and with 10-15 mmHg pulsus paradoxus. The jugular venous pulse, which was clearly palpable, was grossly elevated at 15 cm with a dominant 'a' wave. The apex beat was displaced laterally to the anterior axillary line and there was a prominent right ventricular systolic lift. The pulmonary component of the second heart sound was accentuated and an apical third heart sound was audible, but no murmurs were present. There was moderate leg oedema.

Laboratory investigation showed a haemoglobin value of 13.8 g/dl, a WCC of 8.3 x 10^9/l and an ESR of 44 mm/1st h. The following were normal: serum electrolytes, calcium, creatine kinase, lactate dehydrogenase, alkaline phosphatase, oxalo-acetic and pyruvate transaminase levels. The ECG (Fig. 3) showed sinus rhythm, a PR interval of 180 ms, a QRS axis of -45° due to left anterior fascicular block, left atrial and left ventricular hypertrophy and nonspecific T-wave changes. The chest radiograph was unchanged. Cardiac catheterization revealed the following intracardiac pressures: right atrium — mean 13 mmHg; right ventricle — 51/4-16 mmHg; pulmonary wedge — mean 23 mmHg; and left ventricle — 134/4-20 mmHg.

The moderate elevation of the pulmonary arterial pressure together with elevation of the diastolic pressures in both ventricles was consistent with congestive cardiac failure. There was no valvular disease. Cine angiography revealed dilated,
poorly contracting right and left ventricles. Selective coronary angiography was normal. Right ventricular endomyocardial biopsy (Fig. 4) showed a small, interstitially situated epithelioid cell granuloma with no evidence of necrosis. Staining for acid-fast bacilli and fungi was negative.

A diagnosis of ventricular tachycardia related to myocardial sarcoidosis was made. Persistent frequent ventricular ectopic activity including runs of self-limited ventricular tachycardia (Fig. 5) persisted despite treatment with lignocaine and mexiletine, but was abolished by amiodarone hydrochloride 400 mg 8-hourly for 5 days and then 200 mg/d as a maintenance dose.

Discussion

The criteria for the diagnosis of myocardial sarcoidosis in our patient were the demonstration of hilar lymphadenopathy and characteristic non-caseating epithelial cell granulomas in lymph nodes and myocardium. These conform to the definition of sarcoidosis given by Mitchell et al.² The clinical diagnosis of myocardial sarcoidosis may be extremely difficult and is often not established during life.³ The coexistence of systemic sarcoidosis and myocardial dysfunction is insufficient evidence and the validity of some reported cases has recently been questioned on these grounds.⁴ Silverman et al.,⁵ in a clinicopathological study of 84 unselected patients with systemic
sarcoidosis, found that cardiac symptoms, particularly congestive cardiac failure and rhythm or conduction disturbances, were present in 44%. Less than half of these had any demonstrable granulomas in their hearts and the symptoms could be attributed to systemic disease or a cardiac disorder other than myocardial sarcoidosis. Furthermore, patients with sarcoid infiltration of the heart extensive enough to cause cardiac dysfunction rarely have evidence of dysfunction of another organ system; conversely, sarcoid infiltration of the myocardium sufficient to produce symptoms is unusual in patients with symptomatic involvement of a non-cardiac organ. In our patient the possibility of myocardial sarcoidosis was considered because he was known to have systemic sarcoidosis; if myocardial sarcoidosis had not been positively looked for it is likely that a diagnosis of idiopathic dilated cardiomyopathy would have been made, with consequent failure to give appropriate steroid treatment.

The incidence of myocardial sarcoidosis is difficult to assess. Cardiac involvement in systemic sarcoidosis was first described by Bernstein et al. in 1929. In 1971 Gozo et al. reviewed the 70 cases reported in the world literature and stated that clinically recognizable myocardial involvement was rare. Fleming, however, was able to collect a total of 50 cases and felt that the condition was not rare but underdiagnosed, a view substantiated by the demonstration of myocardial granulomas in 27% of patients with sarcoidosis at autopsy. Nevertheless, the studies of Silverman et al. and Roberts et al. showed that not all patients with myocardial granulomas have clinical evidence of cardiac sarcoidosis and that the difference in clinical presentation correlated with the extent of sarcoid infiltration. Clinical manifestations of cardiac sarcoidosis include ventricular arrhythmias, conduction blocks, congestive cardiac failure, sudden death, papillary muscle dysfunction, recurrent pericardial effusion, ventricular aneurysm and electrocardiographic signs of myocardial infarction. Arrhythmias and conduction disturbances are the most predictive of cardiac sarcoidosis, mild or severe, and patients with major cardiac dysfunction have massive sarcoid infiltration, affecting principally the left ventricular free wall and interventricular septum.

The diagnosis of myocardial sarcoidosis is of clinical importance for several reasons. In the series of Roberts et al. sudden death was the most common mode of death in patients with clinical cardiac dysfunction, and the duration of symptoms before death was generally less than 12 months. Of 50 patients who died suddenly, ventricular arrhythmias or complete heart block had been documented on the ECG in 76%. Since sarcoidosis often affects young adults, cardiac involvement may be an important but neglected cause of sudden death in this group. Corticosteroid therapy is known to cause healing of myocardial sarcoid granulomas and replacement by fibrous tissue. Clinically, steroids have been reported to reduce or abolish arrhythmias and to reverse myocardial dysfunction. The principal aims of treatment are to reduce the risk of sudden death, to improve impaired myocardial function, and possibly to prevent the progression of mild, microscopic granulomatous involvement into massive infiltration, which carries the highest risk of sudden death and congestive cardiac failure. Whether steroids do alter the natural history of cardiac sarcoidosis has not, however, been definitely established.

Few patients have been reported in whom a diagnosis of myocardial sarcoidosis has been established by means of percutaneous endocardial biopsy. In the present case this simple technique established the diagnosis of cardiac sarcoidosis, allowing long-term steroid therapy to be commenced on a rational basis.

Since cardiac sarcoidosis is almost certainly an underdiagnosed and potentially treatable cause of sudden death and cardiac disability, endomyocardial biopsy should be mandatory in the investigation of cardiomyopathy, especially in younger patients.

REFERENCES