We present the case of a 55-year-old male who presented with symptoms of dyspnoea and pre-syncope. A 12 lead electrocardiogram demonstrated extensive conduction abnormalities with 2:1 heart block, right bundle branch block and a small Q wave in lead V1. This indicated significant myocardial and septal involvement. Echocardiography confirmed the presence of right ventricular infiltration and dysfunction whilst cardiac magnetic resonance showed the infiltration to be nodular in nature. Although a diagnosis of cardiac sarcoidosis and lymphoma were initially considered, sarcoidosis was eventually confirmed following a cervical lymph node biopsy. This case firstly demonstrates the usefulness of the 12-lead electrocardiogram in determining the likely anatomical locality of significant bradyarrhythmias. Secondly it highlights the difficulties in diagnosing cardiac sarcoidosis when cardiac dysfunction is the sole manifestation of the disease.

A 55-year-old gentleman presented to our institute with a 2-week history of increasing exertional dyspnoea and pre-syncope. He was a normally fit and well individual with no significant medical co-morbidities. Although a chest X-ray demonstrated non-specific abnormalities at both hila (Figure 1A), the 12 lead electrocardiogram was markedly abnormal. There was advanced second degree AV block (2:1), a prolonged PR interval, right axis deviation, right bundle branch block and a small Q wave in lead V1 (Figure 1B). These abnormalities suggested extensive myocardial and septal involvement and the patient was referred for urgent echocardiography.

This demonstrated pronounced discrete areas of cardiac infiltration within the right ventricular outflow tract (Figure 2A). There was marked right ventricular hypertrophy (1.6 cm) (Figure 2B) and an associated reduction in systolic function. The peak right ventricular free wall systolic shortening velocity on tissue Doppler imaging was 7 cm/s (Figure 2C) and the tricuspid annular plane systolic excursion was measured at 10 mm (Figure 2D). There were no significant abnormalities identified of the left ventricle where both systolic and diastolic function was preserved. Based upon the above findings a differential diagnosis of cardiac lymphoma or sarcoidosis was made.

Further investigations included an immunoglobulin and autoimmune profile (normal), serum calcium (2.4 mmol/l), angiotensin converting enzyme levels (73 units/mL), lactate dehydrogenase levels (312 units/L) and an urgent cardiac magnetic resonance (CMR) imaging scan.

CMR (Figure 3) demonstrated a globally hypokinetic right ventricular free wall and dyskinesis of the interventricular septum. Upon gadolinium enhancement there was florid enhancement of these territories with gross nodular plaque-like infiltration and extension to the cardiac apex. Note was also made of concomitant infiltration of the left ventricle (ejection fraction 58%). Since these findings were associated with the presence of symmetrical bilateral hilar and mediastinal lymphadenopathy the differential diagnosis of either lymphoma or sarcoidosis was maintained.

The patient remained haemodynamically stable and underwent permanent pacemaker implantation with a Guidant ALTRUA DDDR pacemaker. This was to permit the availability of high quality diagnostic intracardiac electrograms to monitor for ventricular arrhythmias.

Given the uncertainty of the diagnosis an endomyocardial biopsy was performed, which was inconclusive, and thereafter a computed tomography scan of the neck, thorax and abdomen. This demonstrated evidence of widespread cervical and mediastinal lymphadenopathy. Later histological analysis of an excised lymph node confirmed the presence of non-necrotising ‘naked’ granulomas characteristic of sarcoid.

The patient was commenced on corticosteroids and after stabilization discharged from hospital. At 3 months the patient’s symptoms had improved, the echocardiographic findings had regressed and the patient had been fitted with an internal cardiac defibrillator (ICD).

Symptomatic cardiac involvement is estimated to occur in 2–7% of patients with sarcoidosis, although up to 20% of patients have demonstrable myocardial involvement at necropsy.1,2 Sudden death due to unsuspected myocardial involvement occurs in as many as 35% of affected individuals2 although the 5 year mortality rate for patients with symptomatic cardiac sarcoidosis is estimated to be in excess of 50%.
Conduction abnormalities may range from first degree heart block to complete heart block (25–30%). Sustained or non-sustained ventricular tachycardia has been reported in 23% of patients with cardiac sarcoidosis. Ventricular fibrillation is rarely reported but, along with complete heart block, is felt to account for the majority of cases of sudden cardiac death in this patient group. Atrial arrhythmias are less common, with an estimated incidence of 19%. Cardiac sarcoidosis may also precipitate systolic or diastolic heart failure, which when present, carries a poor prognosis.

The diagnosis of cardiac sarcoidosis may precede, follow or occur concurrently with involvement of the lungs or other organs. Corticosteroid therapy is the mainstay of treatment of patients although there remains a lack of randomized controlled trials to confirm efficacy. In one review of 95 Japanese patients, overall survival rates were shown to be 60% at 5 years and 44% at 10 years. In those

**Figure 1** (A) Erect chest X-ray demonstrating the presence of bilateral hilar opacification and an absence of concomitant lung disease. (B) 12-lead electrocardiogram demonstrating a PR interval >200 ms, advanced second degree AV block (2:1), right axis deviation and right bundle branch block. The Q wave in V1 suggests extensive myocardial and septal involvement.
patients treated with corticosteroids the 5 year survival rate increased to 75%, and 89% where the ejection fraction was >50%. This compared with only a 29% 5 year survival rate in those patients not on corticosteroids and with an ejection fraction <50%. Multivariate analysis demonstrated the independent predictors of mortality to be the left ventricular end diastolic diameter, NYHA class and the presence of sustained ventricular tachycardia. Since cardiac sarcoidosis is potentially lethal, corticosteroids are generally advised for patients with confirmed or suspected cardiac involvement and in particular when there are cardiac symptoms and evidence of electrocardiographic abnormalities.

For patients who develop heart failure, pharmacological treatment should follow standard guidelines. In those patients with documented tachyarrhythmias medical treatment is problematic with there being no prospective trials of either beta-blockers or amiodarone. Although permanent pacemakers should be implanted as indicated for documented conduction abnormalities, the role of ICD’s has not been well defined.

In the current case the diagnosis of cardiac sarcoidosis was suspected by the presence of conduction abnormalities in an otherwise well individual. This indicated conduction block at the His-Purkinje system and abnormal depolarization of the right ventricle. Subsequent imaging confirmed myocardial infiltration within these areas. Although a number of investigations were required to reach an eventual diagnosis, this was confounded by the lymphadenopathy on the CT scan and a non-diagnostic endomyocardial biopsy. Had a CT scan confirmed the presence of lymphadenopathy earlier on, this would have been the preferred sampling site for obtaining a tissue specimen. This would have avoided the low diagnostic yield of an endomyocardial biopsy, because of patchy involvement of the disease process, and also the potential for procedure related complications.

Figure 2 Transthoracic echocardiogram showing discrete areas of cardiac infiltration within the right ventricular outflow tract (A, parasternal short axis plane), right ventricular hypertrophy (B, apical 4-chamber plane) and an impairment of right ventricular systolic function both by tissue Doppler imaging (C) and tricuspid annular plane systolic excursion by M-Mode (D). RA denotes right atrium, RV, right ventricle, LA, left atrium and LV, left ventricle.
This case firstly illustrates that patients with cardiac sarcoidosis and heart block may present earlier than patients with idiopathic heart block. Secondly, it demonstrates that conduction system disease may alert clinicians to the anatomical location of cardiac involvement. Thirdly, this case reiterates the difficulties in obtaining a diagnosis of cardiac sarcoidosis when cardiac dysfunction is the sole manifestation of this disease.

**Figure 3** Cardiac magnetic resonance scan. (A–C) 4-Chamber plane without (A) and with early (B), and late gadolinium contrast (C) demonstrating the presence of an irregular and hypertrophied right ventricular free wall that enhances with late Gadolinium. (D–F) Short axis plane without (D) and with late gadolinium contrast (E and F) demonstrating gross nodular plaque like infiltration to the apex of the right ventricle and partial involvement of the left ventricle.
Conflicts of interest: None to declare.

References