Case report

A dramatic presentation of cardiac sarcoidosis

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A 40-year-old male pilot presented to the emergency department of his local hospital after experiencing palpitations. His symptoms had started 6 h earlier, while playing cricket for the village team. He reported feeling generally unwell and light-headed, but denied chest pain or breathlessness. He had no chronic medical problems, was a non-smoker, denied recreational drug use and had no family history of cardiac disease. On assessment, he was found to be cool and clammy with a heart rate of 200/min and a blood pressure of 88/60 mmHg. Examination of his cardiovascular system was normal, but there were bibasal crepitations at his lung bases. The initial electrocardiogram (ECG) revealed a broad complex tachycardia, felt to be ventricular in origin and he was promptly electrically cardioverted to sinus rhythm.

Initial investigations revealed his resting ECG to be abnormal with widespread T-wave abnormalities (Figure 1). His Troponin I was elevated at 1.77 ng/ml, while his echocardiogram showed a structurally normal heart with good left ventricular function. Coronary angiography demonstrated unobstructed coronary arteries. A cardiac magnetic resonance (CMR) study was performed showing delayed hyperenhancement in the basal portion of the left ventricular cavity wall, a finding commonly associated with inflammation, oedema or fibrosis (Figure 2a). The CMR also identified enlarged lymph nodes in the paratracheal distribution. A high-resolution computed tomography (CT) scan of the chest was performed, which confirmed calcified hilar lymphadenopathy (Figure 2b) and lung parenchymal changes consistent with sarcoid.

A diagnosis was made of cardiac sarcoidosis with pulmonary involvement. Subsequent endomyocardial biopsy demonstrated only focal minor fibrosis. The gentleman was given oral corticosteroids and received an implantable cardioverter defibrillator.

Discussion

Sarcoidosis is a multisystem disease of unknown aetiology characterized by the accumulation of non-caseating granuloma in affected tissue. Myocardial granulomata are present in 20% of cases of sarcoidosis at post-mortem, but only 5% of patients with sarcoidosis have clinical manifestations of cardiac involvement. The typical signs of cardiac sarcoidosis include myocardial dysfunction, conduction abnormalities and occasionally ventricular arrhythmias. These life-threatening dysrhythmias are caused by re-entry circuits within the ventricular myocardium precipitated by inflammation and ultimately scarring.

The diagnosis of cardiac sarcoidosis is often challenging, as symptoms often mimic other more common cardiological conditions and investigations may be falsely reassuring. The 12-lead ECG may show signs of myocardial scarring such as pathological Q-waves or conduction abnormalities but equally can be normal. Transthoracic echocardiography, the usual first-line investigation, may show abnormalities in ventricular performance, but its overall sensitivity is low (14%). Endomyocardial biopsy may yield crucial histological evidence, but
due to the patchy nature of the disease there is a high false negative rate. More recently, CMR has emerged as a useful tool as it is able to demonstrate areas of myocardial oedema associated with active inflammation as well as areas of scarring and wall thinning.\(^3\)

The treatment of cardiac sarcoidosis is dependent on the clinical picture and consists of immunosuppressive therapy to modify disease activity and cardiac devices. Systemic corticosteroids are indicated if there is active cardiac disease and appear to halt disease progression while improving prognosis.\(^4\) Disease within the intrinsic conduction system of the heart should be managed along conventional guidelines by pacemaker insertion. Implantable cardioverter defibrillator therapy should be considered for those who present with significant ventricular arrhythmias regardless of their ventricular function, as in this case.

This case highlights the challenges encountered in the diagnosis of cardiac sarcoidosis and emphasizes the importance of multi-modality imaging. CMR should be considered in all patients presenting with unexplained ventricular arrhythmias.

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References


