Isolated cardiac sarcoidosis: critical role of cardiac MRI for diagnosis and management

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A 49-year-old female was referred for further evaluation of exercise-induced symptomatic 2nd degree 2:1 atrioventricular (AV) block (Mobitz type 2, Panel A). Several days of telemetric surveillance confirmed intermittent AV block but did not reveal evidence of any other arrhythmias.

 Coronary angiography showed normal coronary arteries. However, despite overall normal left ventricular ejection fraction, laevography showed an unusually deformed left ventricular endocardial border due to several protuberances of the ventricular wall, which were particularly obvious in systole (Panel B, arrows). Cardiac magnetic resonance imaging (MRI; delayed enhancement imaging) revealed multiple large, discrete areas of hyperenhancement throughout the left ventricular myocardium and not confined to a coronary territory (Panel C, arrows). In addition, T2-weighted imaging showed diffuse left ventricular oedema (Panel D, arrows). Clinical and MRI findings raised the suspicion of cardiac sarcoidosis. However, an extensive work-up did not reveal evidence of extracardiac sarcoidosis, and right ventricular endomyocardial biopsy could not establish a pathological diagnosis of sarcoidosis either.

 Given the highly suggestive MRI findings, the extensive left ventricular involvement, the well-known high sampling error of right ventricular endomyocardial biopsy, and the high risk of ventricular arrhythmia associated with cardiac sarcoidosis an implantable cardiac defibrillator (ICD) was implanted after extensive discussion with the patient.

 One month after hospital discharge the patient fainted, which on ICD interrogation was found to have been due to ventricular fibrillation successfully terminated by an appropriate ICD shock (Panel E, ICD strip).

 The present case highlights the pivotal role of cardiac MRI for the diagnosis of cardiac sarcoidosis.

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