Reminder of important clinical lesson

Undiagnosed myocardial sarcoidosis presenting as life threatening cardiac dysrhythmia

Stacy A Davis, Walter J Moore, Alyce M Oliver

Section of Rheumatology, Department of Medicine, Medical College of Georgia, Augusta, Georgia, USA

Correspondence to Dr Alyce Marie Oliver, aloliver@mcg.edu**Summary**

Sarcoidosis is a systemic disorder of unknown aetiology characterised by its pathological hallmark of non-caseating granuloma. Definitive diagnosis requires compatible clinical and imaging features as well as pathogenic identification of non-caseating granulomas in at least one organ. The disease has a wide variety of clinical and radiological manifestations but is associated with low mortality. However, cardiac involvement which is clinically only identified in 5% significantly worsens prognosis due to complications such as congestive heart failure, ventricular tachyarrhythmia, pulmonary hypertension or conduction disturbance leading to sudden death. Cardiac involvement is implicated in 77–85% of deaths directly related to sarcoidosis. Autopsy series in sarcoidosis patients show cardiac involvement with sarcoidosis in up to 79% cases. This case details the clinical course of a 56-year-old female who had experienced refractory cardiac dysrhythmias for many years in the context of also having biopsy proven pulmonary sarcoidosis. She had failed multiple antiarrhythmics as well as pacemaker and implantable cardioverter defibrillator placement. It was not until she presented as a potentially fatal ventricular tachycardia that the possibility of cardiac involvement from her sarcoidosis was entertained as the aetiology of her cardiac problems. Confirmation of myocardial sarcoidosis with PET CT imaging and subsequent treatment with prednisone resulted in her clinical improvement.

BACKGROUND

We present this case to first alert physicians to the clinical presentation of cardiac sarcoidosis and highlight that early diagnosis has been shown to decrease morbidity and mortality. Second, we further show that in recent years PET CT imaging has emerged as a non-invasive study with increased sensitivity over other imaging modalities.

CASE PRESENTATION

A 56-year-old female presented with a 6-year history of biopsy proven pulmonary sarcoidosis, previously only treated with an average low-dose prednisone of 10 mg daily, who experienced refractory tachyarrhythmias. She had difficulty to treat atrial fibrillation-flutter with New York Heart Association Functional Class III heart failure and was status postatrioventricular ablation with pacemaker and implantable cardioverter defibrillator (ICD) placement. She presented to the outpatient cardiology service for possible repeat ablation but was noted to be in ventricular tachycardia. Her symptoms included shortness of breath, palpitations, faint feelings, fatigue and night sweats. She denied any other organ involvement. Physical examination revealed regular rhythm at a rate of 119, blood pressure 116/76, respiratory rate of 20 and she was afebrile. Her jugular venous pulsations were flat, no carotid bruits present, heart sounds 1 and 2 heard without murmurs and peripheral pulses present. Lung fields were clear.

INVESTIGATIONS

Laboratory studies revealed an unremarkable complete blood count and comprehensive metabolic panel. Her INR was 5.3. Sedimentation rate was 35, C reactive protein 1.39 and angiotensin-converting enzyme level <5.

EKG demonstrated a wide complex tachycardia with an inferiorly directed axis and left bundle branch block morphology. Interrogation of her ICD revealed an underlying atrial fibrillation with underlying ventricular sensing with a cycle length of about 500 ms, consistent with a ventricular tachycardia. Echocardiogram showed severe global hypokinesis, left ventricular ejection fraction of 20–25%, moderate mitral regurgitation and a pulmonary artery pressure of 42 mm Hg. To determine if these tachyarrhythmias could be due to cardiac involvement, PET CT imaging was performed due to the availability of a PET CT system at our institution. It demonstrated increased metabolic activity involving the myocardium, bone, muscles and lymph nodes. This finding was consistent with disseminated active sarcoidosis in those organs (figure 1).

TREATMENT

Given this finding by PET/CT imaging, high dose prednisone was given as treatment.

OUTCOME AND FOLLOW-UP

Repeat PET/CT scan performed 1 month after high dose prednisone therapy demonstrated good response at all previous sites of active sarcoidosis by showing decreased FDG uptake. This improvement in imaging correlated well with her clinical cardiac improvement (figure 2). We believe that her dysrhythmias improved in response to high-dose prednisone given her history over the years of unresponsiveness to medical and surgical interventions.

DISCUSSION

Cardiac sarcoidosis may present as an infiltrative cardiomyopathy. ECG reveals ventricular arrhythmias in

Figure 1 PET image highlighting significant myocardial uptake.

Figure 2 Disseminated and post treatment images.

as many as 22% of patients with sarcoidosis.¹ The most common presentation of cardiac sarcoid is high degree atrioventricular (AV) conduction blocks, leading to complete AV block.² ECG findings are also suggestive of a higher incidence of right bundle branch block, supraventricular/ventricular premature contractions and ST-T abnormalities as compared with findings in an age- and sex-matched healthy control group.² The intraventricular septum, often with involvement of the conducting system is the second most common location for granulomas and scars at autopsy.³ Early diagnosis has been found to positively impact on the degree of morbidity and mortality associated with this condition. We present this case as a reminder that FDG PET imaging has emerged in recent years as a non-invasive cardiognostic modality which may facilitate early diagnosis based on its improved sensitivity approaching 100% compared traditional nuclear imaging studies such as ^{99m}Tc-MIBI SPECT and ⁶⁷Ga scintigraphy which have sensitivities of 63.6% and 36.3% respectively.⁴

This case highlights a probable delay in the diagnosis of cardiac sarcoidosis for many years, resulting in this patient presenting with a life threatening ventricular dysrhythmia. This delay in diagnosis and treatment may have been prevented if her physicians had a heightened awareness of various presentations of cardiac sarcoidosis and a high index of suspicion in this particular case. Cardiac sarcoidosis may present as an infiltrative cardiomyopathy with resultant dysrhythmias and conduction abnormalities. Early diagnosis has been found to decrease morbidity and mortality associated with this condition. Current data indicates that approximately 47% of patients with sarcoidosis-associated cardiomyopathy require pacemaker or ICD placement at some time during their disease course.² The risk of death from myocardial sarcoidosis is estimated to be 20% at 1.7 years of follow-up.²

Routine cardiac investigation for suspected sarcoidosis should include 12-lead ECG, transthoracic echocardiography and nuclear imaging or Gadolinium-enhanced cardiac MRI.² Though endomyocardial biopsy continues to be the most specific diagnostic tool, the yield is very low approximating 20% in some series. FDG PET imaging has emerged in recent years as a non-invasive cardiognostic modality with improved sensitivity approaching 100% compared with nuclear imaging studies such as ^{99m}Tc-MIBI SPECT and ⁶⁷Ga scintigraphy which have sensitivities of 63.6% and 36.3%.⁴ Cardiac MRI is which also a recommended diagnostic modality with sensitivity of 75–90%, and is more affordable and more widely available than FDG PET imaging. One study suggested cardiac MRI has superior specificity of 76% compared to that of FDG PET imaging at 38.5%.² However, other series have estimated the specificity of

FDG PET imaging for cardiac sarcoidosis to range from 81.5% to 90.9%.^{4 5}

The novel utility of ¹⁸F-FDG PET for cardiac sarcoidosis involves using standardised uptake value (SUV) measurements of ¹⁸F-FDG. For the analysis, myocardial accumulation of ¹⁸F-FDG in regions of interest for 13 myocardial segments is measured, with segmental uptake of the tracer being graded by visual assessment using a 4-point scoring system whereby 0=normal, 1=mild reduction, 2=moderate reduction and 3=severe reduction or absence of tracer uptake.⁴ Of note, PET CT imaging is believed to provide the same nuclear imaging information as plain FDG PET. A PET CT system is a combination of a FDG PET and a CT scanner with a single patient table. It uses low-dose CT for anatomical co-localisation of PET findings. A low-dose CT is not intended for radiological diagnosis.⁶

The sequelae of cardiac sarcoidosis is preventable to some extent if the disease is diagnosed promptly. FDG PET imaging facilitates early diagnosis as this modality provides a non-invasive study with improved diagnostic sensitivity.

Learning points

- Cardiac sarcoidosis is associated with high mortality risk.
- The sequelae of cardiac sarcoidosis is preventable to some extent if the disease is diagnosed promptly.
- FDG PET CT may facilitate early diagnosis of cardiac sarcoidosis as this modality provides a non-invasive study with clinically useful diagnostic sensitivity and specificity.

Competing interests None.

Patient consent Obtained.

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