**CLINICAL VIGNETTE**

# Cardiac Sarcoidosis: The Great Mimicker

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***Case***

A 64-year-old male presented to ambulatory cardiology with chest pain for several weeks. He described the pain as a pressure sensation over his left chest. Exertion, including hiking, his main hobby, exacerbated the pain which was accompanied by shortness of breath. His symptoms would usually subside with rest, but he noted onset of symptoms at rest for the past two weeks and sought medical evaluation.

His past medical history includes moderate obstructive sleep apnea, hypertension, and dyslipidemia. He has screening colonoscopy ten years ago. He is a lifetime non-smoker but drinks 5-6 cans of beer per week. His mother had an abdominal aortic aneurysm and dementia. His medications included losartan 100mg per day, metoprolol succinate 25mg per day, and simvastatin 40mg per day. He denied lightheadedness, palpitations, fevers, vision changes, or weight loss.

Physical exam included blood pressure of 137/80 mmHg, pulse of 73, and oxygen saturation of 95%. The cardiovascular, pulmonary, and abdominal examinations were unremarkable. Laboratory studies included normal complete blood count, comprehensive and metabolic panel and thyroid showed diffuse T wave inversions (Figure 1). Hemoglobin A1C was 6.8. Electrocardiogram (ECG) demonstrated moderate concentric left ventricular (LV) hypertrophy with estimated LV ejection fraction (EF) of 65%. There were no significant valvular or aortic abnormalities.

A stress treadmill echocardiogram was performed with good exercise tolerance and no ECG or echocardiographic evidence of coronary ischemia. Coronary computed tomography (CT) angiography showed mild coronary calcium in the left circumflex artery but no significant coronary obstruction. An unusual pattern of circumferential thickening of the myocardium up to 2 cm at the LV infero-septal wall was observed on CT and a cardiac magnetic resonance imaging (MRI) scan was recommended.

Cardiac MRI showed multiple abnormal areas of patchy delayed myocardial hyperenhancement in the LV. The sub-epicardium, mid-myocardial basal septal wall, and mid-chamber lateral wall were involved in a pattern highly suggestive of sarcoidosis (Figure 2).

A fluorodeoxyglucose (FDG)-positron emission tomography (PET) CT scan of the chest showed normal hilar lymph nodes and pulmonary parenchyma without evidence of extra-cardiac sarcoidosis. Hypermetabolic uptake of FGD was noted in the LV septal-inferior wall confirming active inflammation (Figure 3). Autoimmune blood tests including anti-nuclear antibodies and rheumatoid factor were within normal limits. Isolated cardiac sarcoidosis was diagnosed, and the patient was started on systemic steroids with improvement in symptoms.

***Discussion***

Isolated cardiac sarcoidosis (CS) is rare as compared to systemic CS with cardiac involvement.[1,2] Symptoms of cardiac CS range from no symptoms to life threatening ventricular arrhythmias and cardiac arrest. Cardiac biopsy showing non-caseating granulomas is the gold standard for diagnosis, but yield of biopsy is low with approximate sensitivity of 25%, due to patchy distribution of CS in the heart. Therefore, cardiac imaging remains the most practical modality to diagnose CS. Imaging modalities to assess for CS include MRI, FDG-PET, and Gallium 67-scan. Gallium-67 scan has low sensitivity and has been largely replaced by FDG-PET. Echocardiography does not have the tissue resolution or functional assessment of MRI or FDG-PET, but may suggest underlying pathology. Thinning of the septum on echocardiography may be present in more advanced CS as fibrosis develops at the final stages of unabated inflammation. Electrocardiographic features of CS range from asymptomatic abnormal T waves to advanced heart block and bradycardia due to inflammation/fibrosis of the conduction system.

Advanced cardiac imaging is important in the diagnosis or exclusion of CS. Cardiac MRI has a higher sensitivity than FDG PET for the diagnosis of CS and is used as the initial advanced imaging modality for CS in most cases.[3] A negative cardiac MRI is 95% accurate to exclude CS. Cardiac MRI and FDG PET perform similarly regarding specificity in the range of 80-85% to diagnose CS.[3] Combined cardiac FDG PET-MRI may be useful but may not be readily available.

Left ventricular function remains an important prognostic factor for CS. Patients with low LV function are at high risk for sudden cardiac arrest and heart failure. However, the presence of

normal LV function does not imply low risk as a significant portion of patients with normal LV function, but abnormal cardiac MRI develop ventricular arrhythmias. A study of over 300 patients with CS, reported up to 24% experienced sudden cardiac arrest or sustained ventricular arrhythmia over 5-years span.[4] Anti-inflammatory therapies may not improve LV function after onset of the fibrotic stage. Therefore, clinical suspicion and early detection are important to prevent progressive fibrosis and complications of CS.

Treatment strategies are aimed at reducing the inflammatory process. They usually involve steroids with dosing dependent on the clinical scenario. For example, in a patient with life threatening ventricular arrhythmias due to inflammation, intravenous pulse dose steroids may be appropriate hospitalized patients. Chronic anti-inflammatory therapy includes biological modification with oral or intravenous medications. Implantable cardiac defibrillators (ICD) may be useful in high-risk patients including those with known ventricular arrhythmias. Unlike other cardiomyopathic processes where LV function is a major determinant of ICD implantation, CS patients with normal LV function may benefit from an ICD. Antiarrhythmic therapies include medications, sympathetic denervation surgery, and catheter ablation to prevent recurrent ventricular arrhythmias.

***Figures***

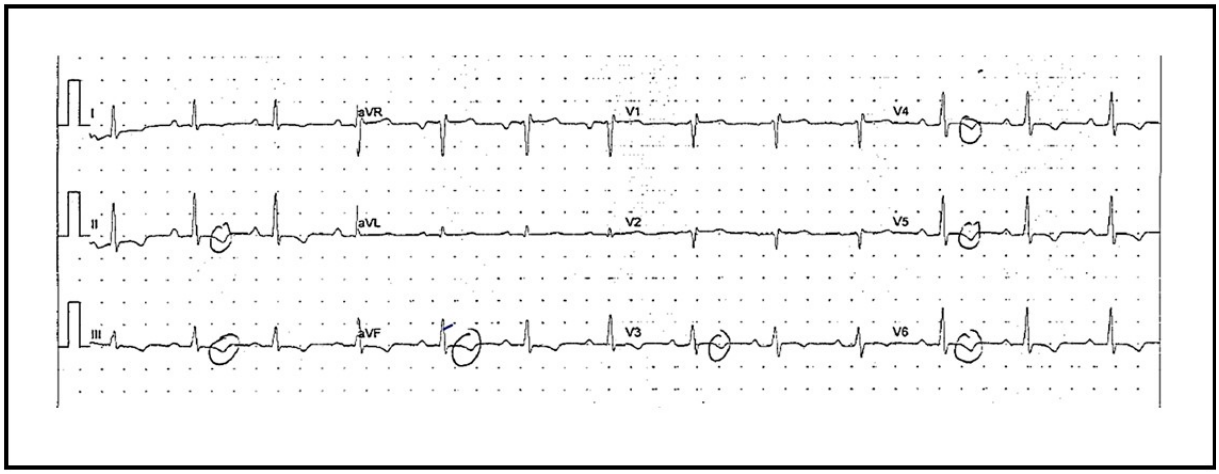


Figure 1: Baseline 12-lead ECG showing sinus rhythm with diffuse T wave inversions (circles).

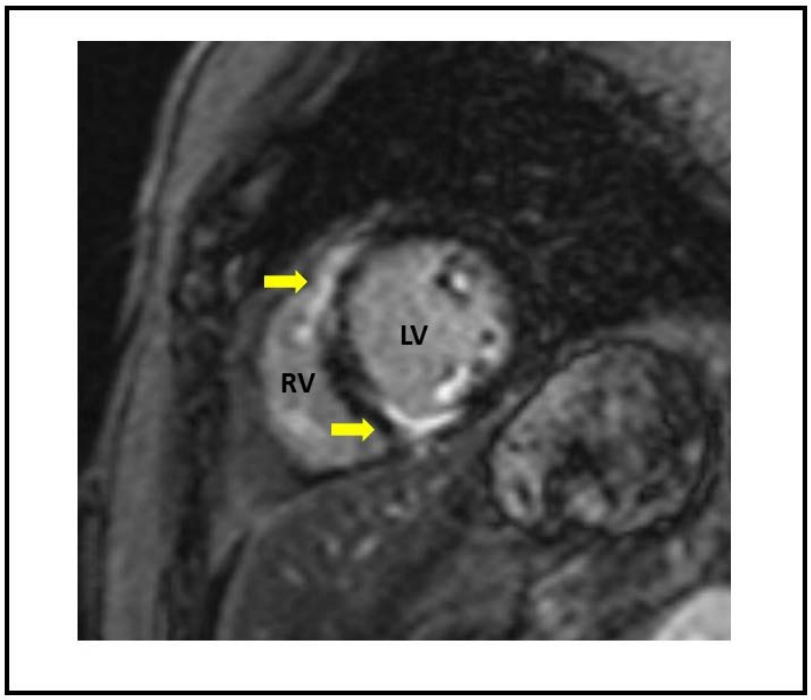


Figure 2: Cardiac MRI. Short axis view of the LV with late gadolinium enhancement (yellow arrows) in a pattern consistent with cardiac sarcoidosis. LV = left ventricle; RV = right ventricle.

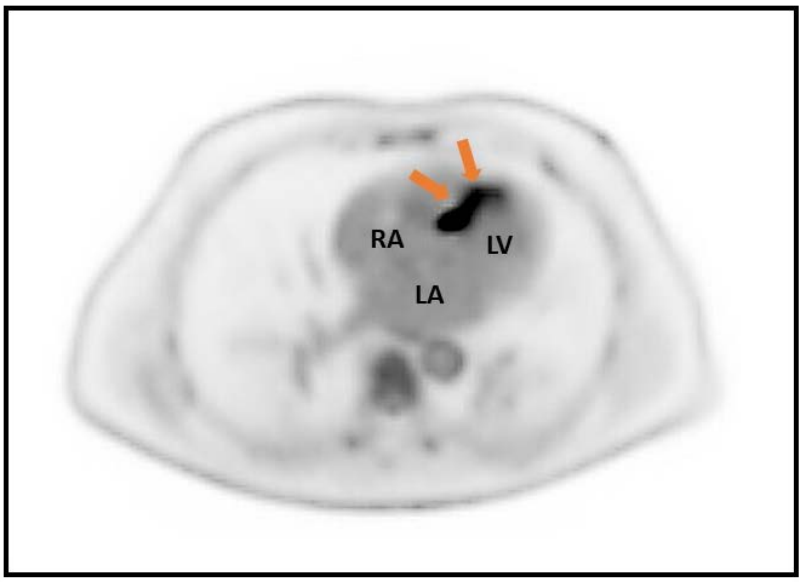

Figure 3: FDG-PET Chest CT. FDG uptake and hypermetabolism in the LV septum (orange arrows) consistent with active inflammation. RA = right atrium; LA = left atrium.

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