

Rapidly progressive cardiac sarcoidosis: Initial presentation with sinus node dysfunction and right bundle branch block



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Introduction

Complete heart block is the most common electrocardiogram (ECG) finding in patients with cardiac sarcoidosis because of the predilection of granulomatous infiltration of the basal septum or involvement of the atrioventricular nodal artery.¹⁻⁵ We report a cardiac sarcoid case presenting with sinus node dysfunction and describe the utility of fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET-CT) scans in making the initial diagnosis.

Case report

A 44-year-old male endurance athlete with no significant medical history was referred by his primary physician for cardiac electrophysiological evaluation because of asymptomatic resting bradycardia (Figure 1A). Right bundle branch block was present with QRS duration 130 milliseconds and conducted PR interval of 200 milliseconds. The patient denied any symptoms such as dizziness, lightheadedness, presyncope, syncope, or exercise intolerance. He was an avid athlete and had recently been distance running and cycling without limitation.

Results of an initial medical evaluation, including thyroid stimulating hormone, erythrocyte sedimentation rate, and Lyme titers, were normal. Exercise stress testing revealed excellent functional capacity (16 metabolic equivalents) and baseline bradycardia with maximal sinus rate of 130 beats

per minute (74% maximum predicted heart rate). No arrhythmias were noted during exercise. A transthoracic echocardiogram showed a left ventricular (LV) ejection fraction of 55% and normal LV systolic function. Because of concern for infiltrative disease, cardiovascular magnetic resonance imaging (CMR) was performed; the results revealed normal biventricular systolic function without evidence of late gadolinium enhancement. The patient was scheduled for outpatient follow-up.

Two months after his initial evaluation, the patient presented with progressive fatigue and exercise intolerance. An ECG revealed sinus bradycardia at 34 beats per minute, a conducted PR interval of 422 milliseconds, and a new left anterior hemiblock in addition to his right bundle branch block (Figure 1B). Repeat CMR results showed a new LV basal anteroseptal and mid-posterolateral late gadolinium enhancement (Figure 2). A high-resolution chest computed tomography scan showed no enlarged mediastinal lymphadenopathy.

Because of high clinical suspicion of cardiac sarcoidosis, an 18F-FDG PET-CT scan was then performed following greater than 18 hours of a high-fat, high-protein, low-carbohydrate diet and injection of intravenous unfractionated heparin to suppress myocardial 18F-FDG uptake. Results of myocardial perfusion imaging with intravenous technetium-99m sestamibi were normal (Figure 3A); however, focal uptake of 18F-FDG was noted in the right atrium, basal LV anteroseptum, and basal LV posterolateral wall (Figure 3B). Focal 18F-FDG uptake in the setting of normal perfusion imaging is consistent in this case with early disease.⁶ Multiple foci of 18F-FDG uptake were also noted in the hilar and paratracheal lymph nodes. Mediastinoscopy was performed, and results of a lymph node biopsy confirmed noncaseating granulomatous disease, establishing the diagnosis of cardiac sarcoidosis. The patient was initiated on high-dose steroids and underwent implantation of a dual-chamber, single-coil primary prevention defibrillator.

A repeat 18F-FDG PET-CT scan was performed 3 months following the initiation of steroid therapy, and the

KEYWORDS Cardiac sarcoidosis; Computed tomography (CT); Fluorine-18-fluorodeoxyglucose (18F-FDG); Positron emission tomography (PET); Sinus node dysfunction

ABBREVIATIONS 18F-FDG = fluorine-18-fluorodeoxyglucose; CMR = cardiovascular magnetic resonance imaging; ECG = electrocardiogram; LV = left ventricular; PET-CT = positron emission tomography-computed tomography (Heart Rhythm Case Reports 2016;2:57-59)

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KEY TEACHING POINTS

- Sinus node dysfunction is an uncommon presentation for cardiac sarcoidosis.
- Cardiac magnetic resonance imaging, despite proven utility in evaluation for cardiac sarcoidosis, may not be sufficiently sensitive to detect the early stages of the disease.
- Results of a fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography scan may highlight areas of cardiac inflammation that correlate with clinical disease activity, identify nonenlarged mediastinal lymph nodes suitable for biopsy, and help determine response to steroid therapy.

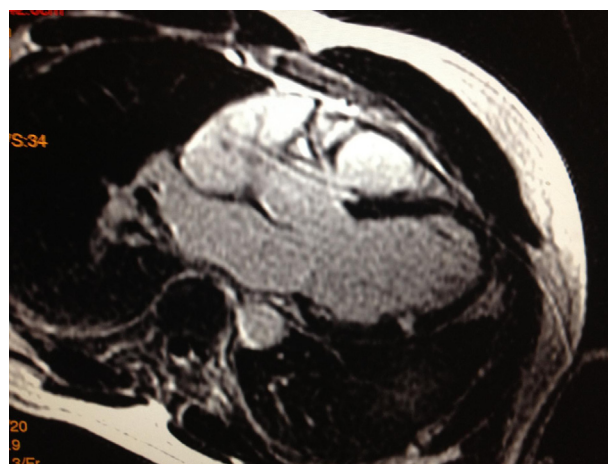


Figure 2 A repeat cardiac magnetic resonance image, which was taken 2 months after cardiac magnetic resonance imaging with normal results, shows a small inferior delayed hyperenhancement.

results showed significant improvement of 18F-FDG uptake in the mediastinal lymph nodes, right atrium, and lateral left

ventricle (**Figure 3C**). The patient had clinically improved and returned to near baseline exercise capacity.

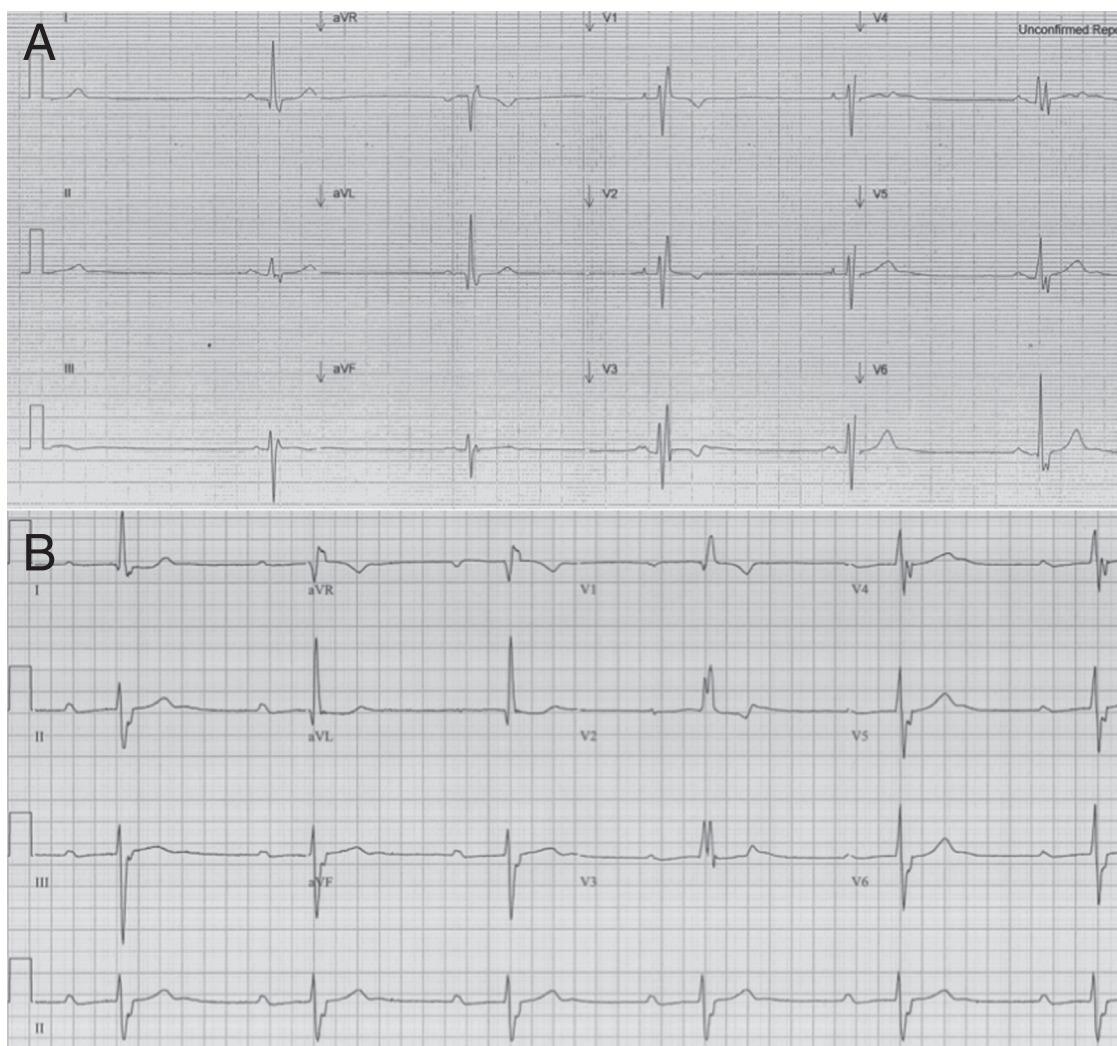


Figure 1 **A:** An electrocardiogram recorded at presentation. **B:** An electrocardiogram recorded 2 months after initial presentation shows progressive conduction system disease with PR prolongation and new left anterior hemiblock.

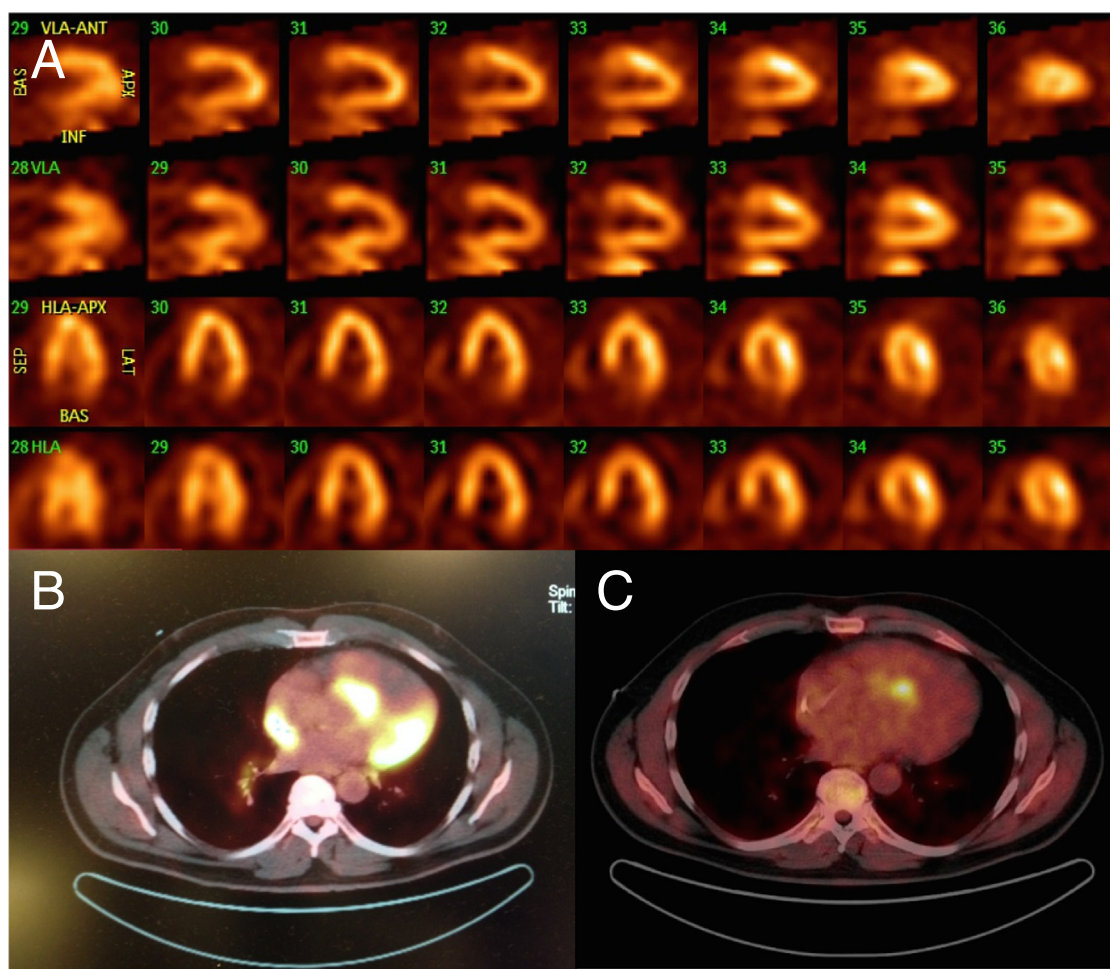


Figure 3 **A:** Normal technetium-99m sestamibi rest-and-stress perfusion image. **B:** Result of a cardiac fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography scan. Fluorine-18-fluorodeoxyglucose uptake is present in the right atrium, septum, and lateral left ventricle. **C:** Repeat image that was taken after 3 months of steroid therapy. VLA = vertical long axis; HLA = horizontal long axis; APX = apex; INF = inferior; LAT = lateral; BAS = basal; SEP = septal.

Discussion

Despite a predilection for granulomatous involvement of the cardiac conduction system, marked sinus nodal inflammation and dysfunction is an uncommon initial presentation for cardiac sarcoidosis. There are limited case reports of extensive granulomatous lesions of the sinus node found during autopsies of cardiac sarcoid patients who experienced sudden death, and even one case of an obstructive granulomatous angiitis of the sinus node artery.^{7,8}

The potentially wide clinical expression of cardiac sarcoidosis, including asymptomatic ECG abnormalities as in this case, makes initial clinical suspicion and use of advanced imaging important for early diagnosis. CMR, despite proven utility in evaluation for cardiac sarcoidosis, may sometimes not be sufficiently sensitive in the early stages of disease. In our patient, 18F-FDG PET-CT images revealed extensive right atrial inflammation as well as extensive ventricular inflammation. In addition to identifying areas of cardiac inflammation that correlate with clinical disease activity, 18F-FDG PET-CT images may also

highlight nonenlarged mediastinal lymph nodes suitable for biopsy and help determine response to medical therapy.

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