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A Case of Stress-Related Heart Block in Cardiac Sarcoidosis

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A Case of Stress-Related Heart Block in Cardiac Sarcoidosis

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Abstract

There are often remarkably long delays in the diagnosis of cardiac sarcoidosis. We present a case of a 61-year-old man with exertional dyspnea and light-headedness. A resting ECG showed a 1st-degree AV block. A complete heart block was unmasked by an exercise stress test. The coronary angiogram was normal as were the cardiac troponins and Lyme antibody titer. He received a pacemaker on an emergent basis. Cardiac PET scan confirmed sarcoidosis. Computed tomography of the chest was significant for hilar and mediastinal adenopathy. He was treated with methotrexate with good symptom relief. This case demonstrates the importance of prompt clinical suspicion and immediate stress testing to unmask higher degree heart blocks in patients with cardiac sarcoidosis, which would have been missed otherwise. Treatment with a pacemaker, an ICD, and an immunosuppressant is recommended in such patients.

Keywords

cardiac sarcoidosis, atrioventricular block, positron emission tomography

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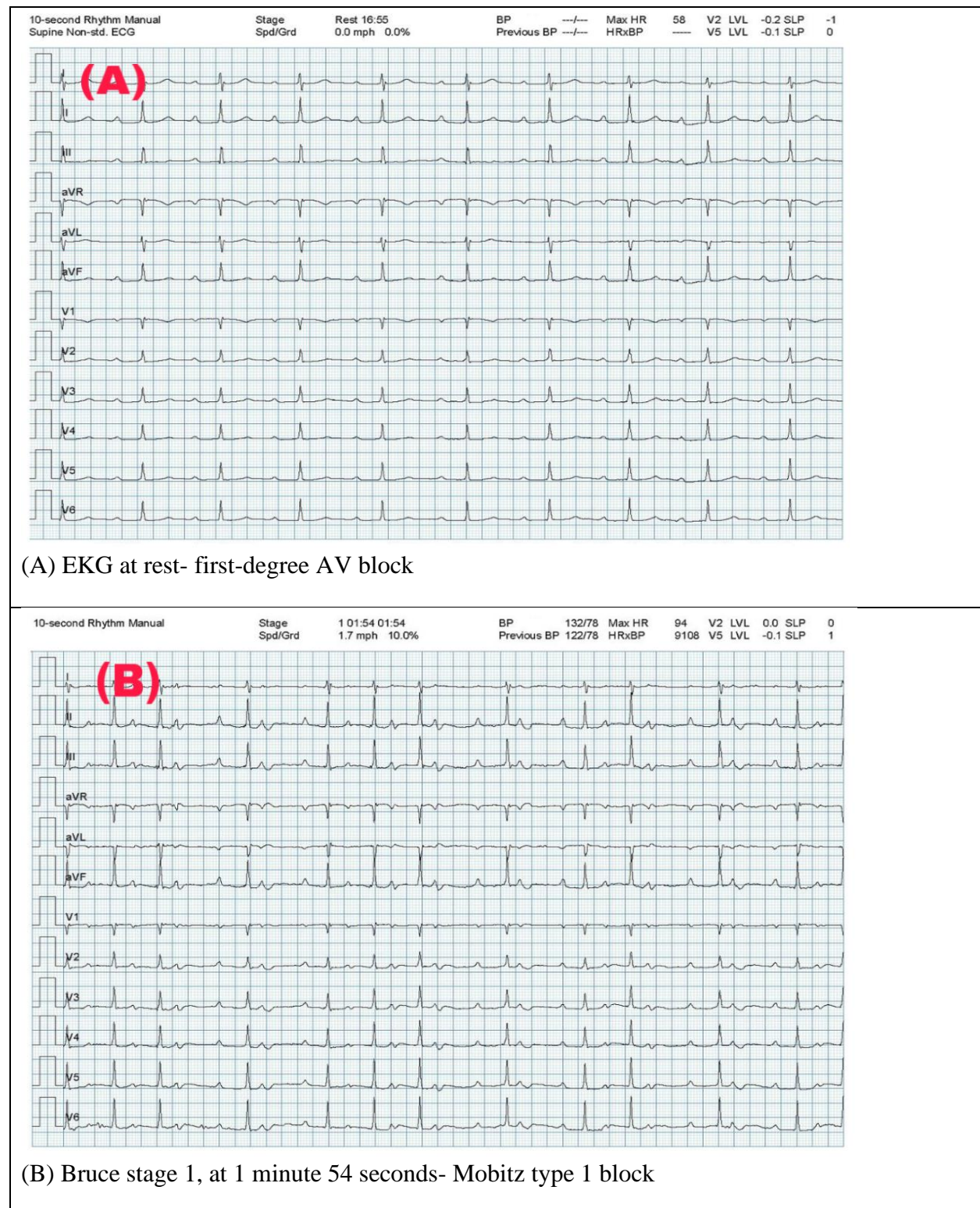
Introduction:

Sarcoidosis is a systemic autoimmune disease that can have isolated organ manifestations in some cases. Patients with cardiac sarcoidosis often have delays in the diagnosis of this condition as it can remain asymptomatic for long periods or can have varied presentations, including heart blocks, ventricular arrhythmias, and heart failure. Although extensive cardiac imaging is not recommended for every patient with first-degree heart block, from our case experience, it would be worth considering stress testing in such patients who experience exertional lightheadedness. Stress testing can unmask a high-grade heart block, which then requires further investigation.

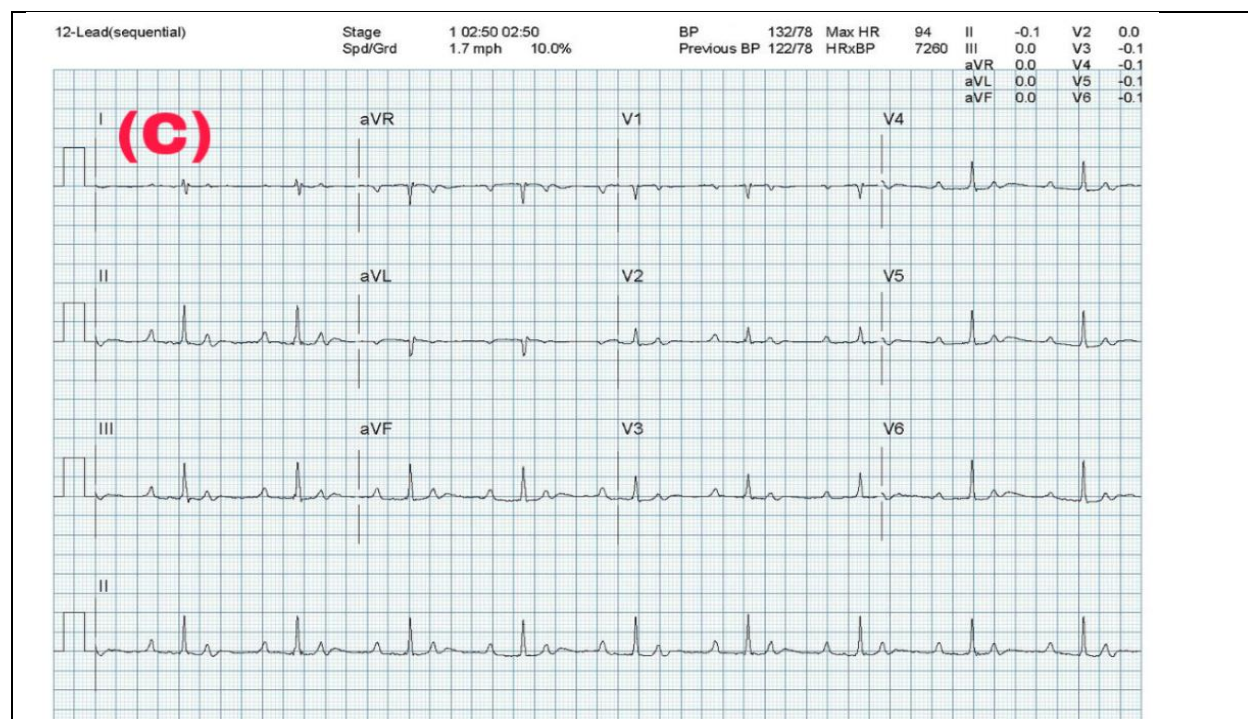
Case presentation:

A 61-year-old Caucasian man presented to the office with a 6-month history of exertional shortness of breath, lightheadedness, palpitations, and atypical chest pain. His symptoms had been occurring consistently with even minimal exertion over the past month and were relieved by rest. He also noted near syncopal events while walking. The patient noted a tick bite a year prior to his presentation. On examination, his heart rate was 62 beats per minute and regular. Blood pressure was 120/80mmHg without any orthostatic hypotension. His oxygen saturation was 97% on room air, and his Body Mass Index (BMI) was 27.16 kg/m². Examination revealed normal heart and lung sounds on auscultation. A late presentation of congenital heart disease, hypertrophic obstructive cardiomyopathy, coronary artery disease, and restrictive cardiomyopathy were considered among the differentials. Blood work showed normal hemoglobin-14.5gm/dl, creatinine-1.0 mg/dl, total bilirubin-0.4mg/dl, troponin I-<0.01ng/ml. NT-proBNP was mildly elevated at 149 pg/ml (normal range 0-124pg/mL). Lyme antibody titer was negative, and angiotensin-converting enzyme (ACE) level was 16. Electrocardiogram (ECG) at rest showed sinus bradycardia with first-degree atrioventricular (AV) block (**figure 1A**). A standardized treadmill exercise stress test by Bruce protocol was performed. During Bruce stage 1, at 1 minute and 54 seconds, he developed a second-degree Mobitz type 1 heart block (**figure 1B**). At 2 minutes and 50 seconds, he developed a second-degree Mobitz type 2 block with 2:1 conduction (**figure 1C**). When entering Bruce stage 2, at 3 minutes and 20 seconds, he developed a complete heart block (**figure 1D**). Transthoracic 2D echocardiogram revealed a normal left ventricular function with no evidence regional wall motion abnormality suggesting structural heart disease. A coronary angiogram was normal. Following this negative ischemic workup, a dual-chamber pacemaker was placed. Cardiac positron emission tomography (PET) scan was done, revealing intense heterogeneous fluorodeoxyglucose (FDG) uptake in the right ventricle, septum, and splotchy "string of pearls" uptake in the inferior, lateral, and apical wall regions consistent with active inflammatory myocardial sarcoidosis. Computed tomography of the chest was significant for mediastinal and bilateral hilar lymph nodes with maximal short axis 1.2 cm, some with partial calcification, consistent with sarcoidosis. In addition, a 6mm granuloma was noted in the left lung base. He was started on a steroid-sparing regimen with methotrexate 20mg weekly along with folic acid. The patient remained symptom-free after starting methotrexate and continued to be paced >90% of the time.

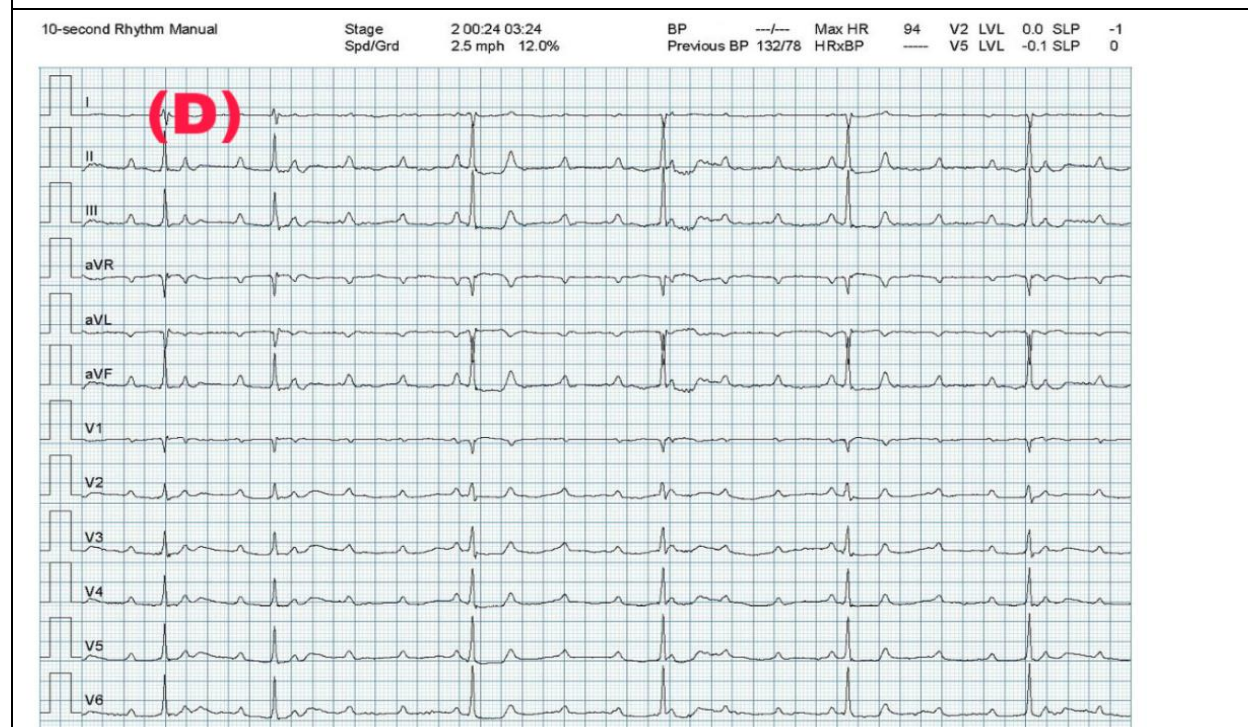
Figure 1:



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(C) Bruce stage 1, at 2 minutes 50 seconds- Mobitz type 2 block with 2:1 conduction



(D) Bruce stage 2, at 3 minutes 20 seconds- Complete heart block

Discussion:

Sarcoidosis is a chronic systemic disease with the characteristic finding of non-necrotizing granulomas on tissue biopsy in multiple organs. A higher prevalence is noted among the African-American population. Systemic sarcoidosis typically affects the lungs but can involve any organ [1,2]. Based on a review by Okada et al, the rate of isolated cardiac sarcoidosis (CS) is seen in up to 25% of patients [3]. CS is secondary to the deposition of the granulomas in the myocardium and subsequent scarring. The common presentations of CS include heart failure, arrhythmias, and sudden cardiac death. The occurrence of a lone atrioventricular block as the initial clinical manifestation of CS is frequently seen; however, there are often remarkably long delays in its diagnosis. A thorough history and high clinical suspicion are required, and alternative etiologies of complete heart block like coronary artery disease and Lyme disease must be ruled out early. Patients presenting initially with AV blocks are at increased risk of experiencing life-threatening ventricular arrhythmias. The 5-year mortality rate of CS presenting with non-serious arrhythmias like low-grade AV blocks can be up to 10% [4].

The diagnosis of CS is based on the expert consensus from the Heart Rhythm Society published in 2014 [5]. CS is classically a pathologic diagnosis, but in clinical practice, endomyocardial biopsies can be both difficult to obtain and unreliable owing to its patchy involvement of the heart muscle. Hence the diagnosis of CS can be quite challenging. Clinical findings that suggest CS include unexplained reduced left ventricular ejection fraction (<40%), unexplained sustained ventricular tachycardia, or unexplained AV block (second degree or higher). Using Heart Rhythm Society (HRS) criteria, extra-cardiac tissue diagnosis of sarcoidosis is required in addition to the clinical manifestations listed above to diagnose probable CS. Although our patient did have findings suggestive of extra-cardiac sarcoidosis (hilar and mediastinal lymphadenopathy), he did not have a biopsy-proven diagnosis of sarcoidosis. In such cases, advanced cardiac imaging such as cardiac magnetic resonance (CMR) and FDG-PET can help in confirming the diagnosis. According to the Society of Nuclear Medicine and Molecular Imaging and the American Society of Nuclear Cardiology, in the absence of histologic evidence of extra-cardiac sarcoidosis, a cardiac PET scan should be considered only if a patient has a sustained second or third-degree heart block. In our patient, the higher degree heart block was unmasked only on exertion, as shown on the stress test. Thus, even though a stress test is not part of the usual diagnostic algorithm, it might be considered when a higher degree of AV block is suspected in patients presenting with symptoms exclusively on exertion. Generally, on PET-CT, the active necrotizing granulomatous disease shows up as increased 18-FDG uptake in the myocardium with no perivascular distribution as was demonstrated in our patient [6].

High-grade AV block in CS is managed with a permanent pacemaker. ICD implantation may be considered in patients undergoing permanent pacemaker implantation (Class IIA recommendation). The indications for ICD placement in patients with CS include spontaneous ventricular arrhythmias which are sustained in nature, cardiac arrest and/or if the left ventricular ejection fraction is $\leq 35\%$ despite optimal medical therapy and a trial of immunosuppression [5]. A recent systematic review by Sadek et al suggests that immunosuppression with corticosteroids can improve AV conduction. Steroids are usually started at higher doses (60mg/day) and gradually tapered over one year. Steroid sparing agents like methotrexate, mycophenolate mofetil, and azathioprine can be used if there is a concern for steroid-related side effects [7].

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Conclusion:

This case demonstrates how a good clinical exam and history-taking skills, and the right approach of investigations are necessary for prompt diagnosis of this complex autoimmune condition. Although extensive investigation with PET and CMR is not recommended for every patient with first-degree heart block, we recommend considering stress testing in such patients with exertional lightheadedness. A pacemaker is recommended in patients presenting with high-grade heart blocks, and an ICD implant should be considered as these patients are more prone to life-threatening ventricular arrhythmia associated with CS.

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