Resolution of new left bundle branch block and ventricular tachycardia with immunosuppressive therapy in a patient with cardiac sarcoidosis

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Introduction

About 5% of patients with sarcoidosis have symptomatic cardiac involvement while 20%–25% of the patients have cardiac involvement in the absence of symptoms.[1,2] These patients may present with conduction system abnormalities, ventricular arrhythmias, or heart failure symptoms. Patients with cardiac sarcoidosis (CS) are known to have a worse prognosis compared to those without cardiac involvement.[3] We report a case of left bundle branch block and ventricular tachycardia (VT) in a patient with CS that resolved with immunosuppressive therapy.

Case report

A 42-year-old man with history of hypertension and bicuspid aortic valve presented to the emergency room with gradually worsening headache and blurry vision. Head magnetic resonance imaging (MRI) revealed enhancing extraaxial masses along the anterior right tentorium and left parietal/temporal convexity with a differential diagnosis including meningioma and neurosarcoidosis. Chest computed tomography showed hilar lymphadenopathy. Angiotensin-converting enzyme level was <15 U/L (normal 14–82 U/L), but C-reactive protein was elevated at 1.7 mg/dL (normal 0–0.5 mg/dL). An endobronchial ultrasound-guided lymph node biopsy revealed noncaseating granulomas, confirming the diagnosis of sarcoidosis. The patient was treated with high-dose prednisone taper (initial dose 100 mg daily) that resulted in shrinking and stabilization of the intracranial masses.

In addition, the patient complained of intermittent palpitations but denied presyncope or syncope. Based on the HRS screening algorithm,[4] cardiac magnetic resonance was performed within 1 month of tissue diagnosis and showed

KEY TEACHING POINTS

- All patients with extracardiac sarcoidosis should be screened for cardiac involvement, which can cause heart block, ventricular arrhythmias, and heart failure.
- Patients with cardiac sarcoidosis should be monitored closely for progressive conduction system disease and ventricular arrhythmias that may indicate active disease or disease progression.
- Prompt treatment of cardiac sarcoidosis with immunosuppressive therapy may lead to resolution of cardiac rhythm abnormalities.

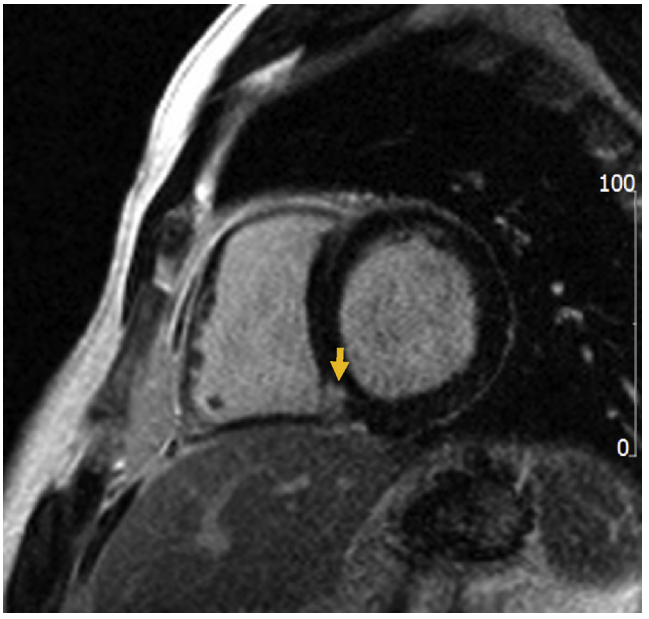

Figure 1 Cardiac magnetic resonance imaging showing delayed gadolinium enhancement of the basal right ventricular septum (*arrow*).

KEYWORDS Bundle branch block; Cardiac sarcoidosis; Immunosuppressive therapy; Implantable cardiac defibrillators; Ventricular tachycardia
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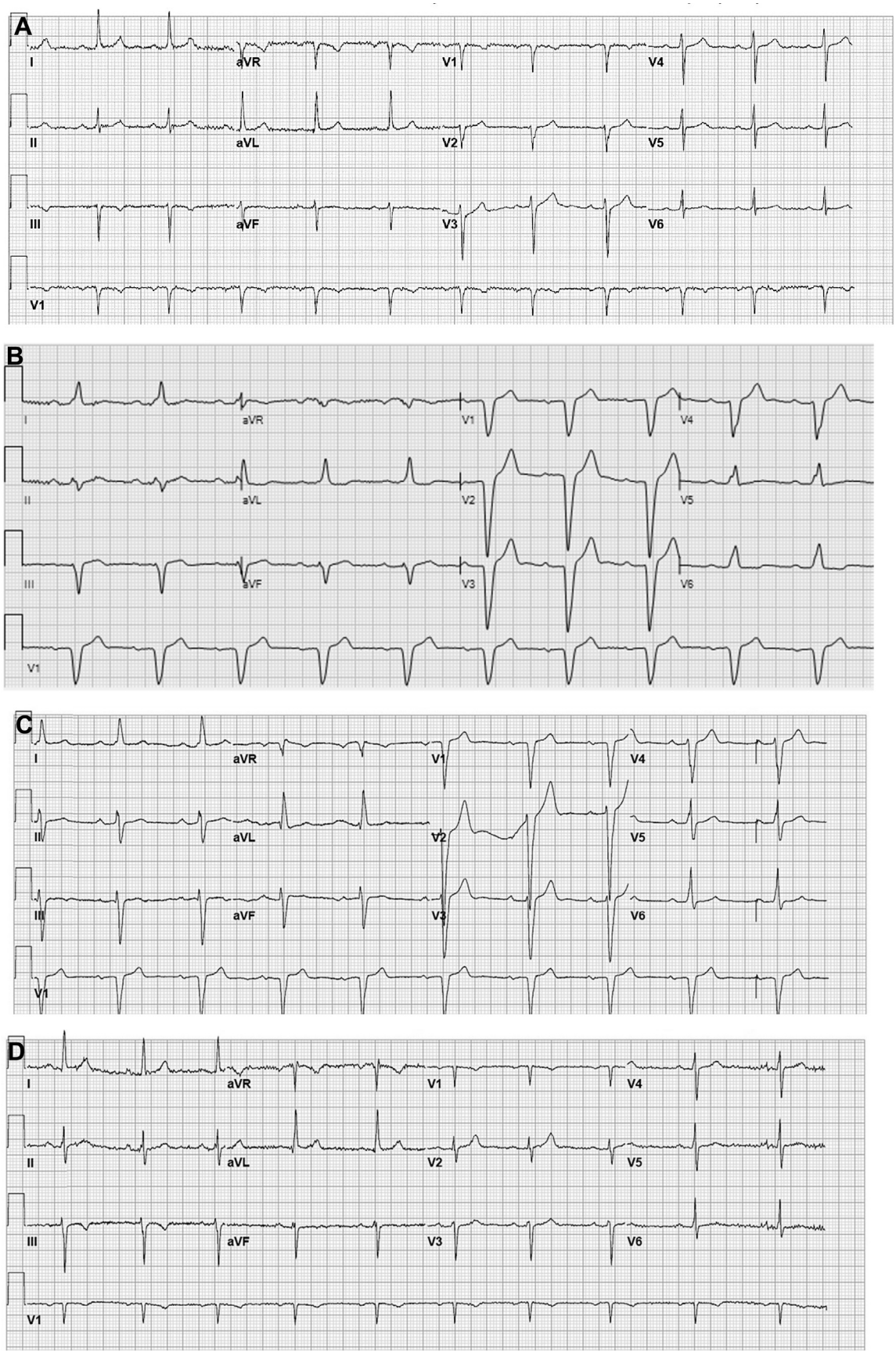

Figure 2 **A:** Baseline electrocardiogram (ECG) showing sinus rhythm with normal PR interval and QRS duration 88 ms. **B:** ECG showing development of first-degree atrioventricular block with PR interval 248 ms and left bundle branch block with QRS 146 ms. **C:** ECG after 3 weeks of steroids showing PR interval 228 ms and interventricular conduction delay with QRS 136 ms. **D:** ECG 5 months later shows PR interval 204 ms and narrow QRS 88 ms.

normal ejection fraction with delayed enhancement of the basal inferior septum along the right ventricular aspect, suggesting likely cardiac involvement of sarcoidosis (Figure 1). Baseline electrocardiogram (ECG) showed normal sinus rhythm with normal PR interval and narrow QRS (Figure 2A). He was started on methotrexate 15 mg weekly as a steroid-sparing agent for multiorgan system sarcoidosis along with metoprolol. He underwent a dual-chamber implantable cardioverter-defibrillator implantation for primary prevention for a class IIA indication based on 2017 AHA/ACC/HRS guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death.[5] The patient was eventually tapered off steroids.

During subsequent clinic visits, device interrogation revealed 2 episodes of VT at 190 beats per minute, terminated by antitachycardia pacing. ECG revealed a new first-degree atrioventricular (AV) block and left bundle branch block with a QRS duration of 146 ms (Figure 2B). These findings were suggestive of worsening CS. Methotrexate dose was increased; however, it had to be stopped owing to elevation of liver enzymes. He was started on prednisone 40 mg daily for 5 days, followed by a slow taper. After 3 weeks of prednisone taper, ECG revealed shortening of the PR interval and narrowing of the QRS duration to 136 ms with an interventricular conduction delay (Figure 2C). After 2 months of steroids, the patient was weaned off prednisone and started on infusions of infliximab, a monoclonal antibody to tumor necrosis factor-α. ECG showed normalization of QRS duration to 88 ms (Figure 2D). No further VT episodes were noted on subsequent device interrogations at follow-up visits during 6 months from the presentation of VT. Methotrexate was not reinitiated owing to liver toxicity. The patient did not receive any antiarrhythmic therapy.

Discussion

CS can cause abnormalities at any site of the conduction system, including sinus node dysfunction, various degrees of heart block, bundle branch block, or fascicular block, as well as ventricular arrhythmias. Right bundle branch block is more common than left bundle branch block in CS.[6] ECG abnormalities in CS can also include QRS fragmentation, ST-T wave changes, pathologic Q waves, and epsilon waves. In patients with AV nodal conduction system disease owing to CS, about half of those treated with corticosteroids have improvement in AV conduction.[7–11] Meanwhile, studies have shown inconclusive evidence for suppression of VT with immunosuppression in CS patients.[7,12–15] Tung and colleagues[12] evaluated 103 patients with unexplained cardiomyopathy and ventricular arrhythmia and found 49% had focal fluorodeoxyglucose (FDG) uptake on positron emission tomography(PET). During 3-year follow-up, 81% of patients treated with immunosuppressive therapy were free from recurrent VT; however, 38% of these patients were concomitantly treated with antiarrhythmic therapy. Naruse and colleagues[13] treated patients with ventricular arrhythmias with antiarrhythmic drugs, with the majority (34/37) also receiving immunosuppressive therapy. During the 39-month follow-up, 62% remained free from VT with medical therapy. A 2013 systematic literature review identified only 2 studies evaluating the effect of corticosteroids on ventricular arrhythmia.[6] In 1 study, 6 of 7 patients treated with corticosteroids had no recurrence of VT over a mean follow-up of 48.8 $\pm$ 38.7 months; however, 5 of the patients were concomitantly started on amiodarone.[14] In another study of 31 CS patients, there was no significant difference in the number of premature ventricular contractions or nonsustained VT before and after steroid therapy; however, patients with less advanced LV dysfunction (ejection fraction $\geq$ 35%, n = 17) showed a significant reduction in premature ventricular contraction burden and nonsustained VT.[13] In 68 CS patients in Japan, VT and electrical storm occurred frequently in the first 12 months after initiation of corticosteroid therapy, presumably because of cardiac inflammation, and positive gallium scintigraphy was a significant and independent predictor of VT.[15]

Patients with sarcoidosis should be monitored closely for progressive conduction system disease and ventricular arrhythmias that may indicate active disease or disease progression. In addition to histologic diagnosis, guidelines recommend using either cardiac MRI or FDG-PET to assess for cardiac involvement in patients with sarcoidosis. Serial FDG-PET examinations showing change in inflammation can be used for monitoring these patients.[4,16] Early identification and prompt initiation of immunosuppressive therapy may lead to the resolution of cardiac rhythm abnormalities before tissue fibrosis causes irreversible conduction system damage.[11] This case highlights the need for early screening for cardiac involvement in patients with extracardiac sarcoidosis and rigorous monitoring of CS patients for electrical abnormalities, which can suggest active cardiac inflammation requiring immunosuppressive therapy.

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