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CASE REPORT

BEGINNER

CLINICAL CASE

Cardiac Sarcoidosis Initially Diagnosed as Spontaneous Coronary Artery Dissection

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ABSTRACT

We present the case of a woman who developed presumed spontaneous coronary artery dissection of a septal branch. She later developed high-grade atrioventricular block that led to a diagnosis of cardiac sarcoidosis involving the interventricular septum. This case illustrates a rare and challenging presentation of cardiac sarcoidosis. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2021;3:1656–1660) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 51-year-old active White woman with psoriasis/psoriatic arthritis and hypertension presented with lightheadedness and fatigue. The day of presentation, she experienced dizziness and palpitations several hours after climbing a hill. Her wearable heart monitor showed a heart rate of 138 beats/min that suddenly dropped to 40 beats/min. She otherwise reported 1 week of upper respiratory symptoms (pre-SARS-CoV-2) without chest discomfort. At the hospital, she was bradycardic with occasional nonconducted p waves and had an elevated troponin I level (1.3 ng/mL). She was transferred to a tertiary care center for further evaluation. Her medications included aspirin, amlodipine, and occasional prednisone for psoriatic arthritis flares. Her latest flare was 15 months before this presentation. Her physician had recently stopped her beta-blocker because of fatigue and bradycardia. On admission, she had sinus bradycardia at 54 beats/min with otherwise stable vital signs and unremarkable physical examination.

LEARNING OBJECTIVES

- To recognize that patients with CS may present with a wide range of symptoms and findings.
- To evaluate patients with suspected CS, advanced imaging modalities, such as cardiac magnetic resonance and 18F-fluorodeoxyglucose positron emission tomography, are extremely useful.
- Coronary granulomatous arteritis from active CS can mimic spontaneous coronary artery dissection.

MEDICAL HISTORY

The patient developed premature atrial contractions 15 months before this presentation. She received a diagnosis of spontaneous coronary artery dissection (SCAD) on coronary angiography (CAG) following an episode of atypical chest pain with elevated troponin I level (2.0 ng/mL) 13 months before this presentation

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(Figure 1). At the time, her electrocardiogram (ECG) and transthoracic echocardiogram (TTE) findings were normal. She was started on clopidogrel, a beta blocker, and aspirin and she was enrolled in cardiac rehabilitation but continued to experience fatigue, dyspnea, and palpitations. Exercise tolerance testing 4 months before the current presentation revealed excellent exercise capacity without ischemia. She then presented to the emergency department with atypical chest discomfort 1 month before this presentation. Her troponin I level was again elevated (1.7 to 1.9 ng/mL). TTE showed a septal wall motion abnormality corresponding to the territory of her presumed SCAD. Subsequent CAG revealed a stable septal dissection without other obstructive coronary artery disease (CAD).

DIFFERENTIAL DIAGNOSIS

Given evidence of myocardial injury and recent upper respiratory symptoms, myocarditis (eg, viral, Lyme disease) was high on the differential. Her elevated troponin and atrioventricular block (AVB) raised concern for an infiltrative process, such as cardiac sarcoidosis (CS). In the absence of typical angina, ECG changes, and new obstructive CAD, acute coronary syndrome was unlikely. Finally, senile conduction disease would be uncommon at her age.

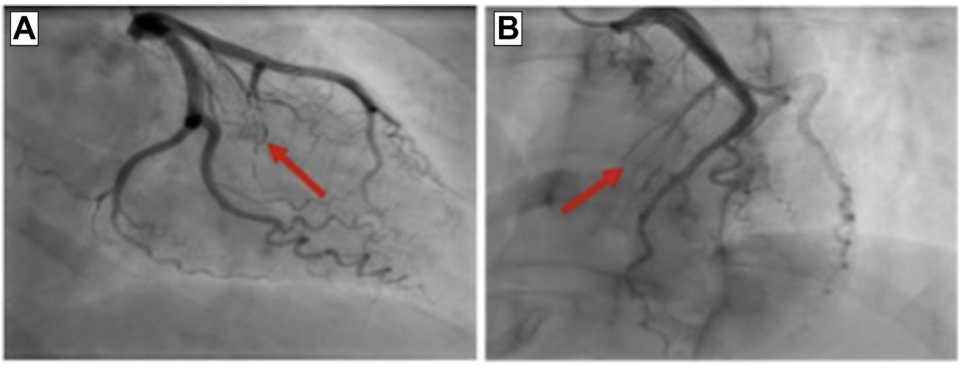
INVESTIGATIONS

During her admission, cardiac telemetry showed sinus bradycardia (40 to 50 beats/min) with first-degree and occasional second-degree type II AVB with a narrow QRS interval. Laboratory test results showed negative Lyme serologies and normal serum angiotensin-converting enzyme level. Noncontrast chest computed tomography (CT) findings were negative for pulmonary disease and lymphadenopathy. TTE revealed a small left ventricular cavity and asymmetric left ventricular hypertrophy involving the basal to mid-interventricular septum (IVS) with basal IVS hypokinesis and mid-IVS akinesis; her IVS and posterior wall measured 14 mm and 10 mm, respectively. CT coronary angiography showed a chronic septal branch dissection with transmural hypoenhancement of the basal to mid-IVS and borderline focally increased wall thickness, raising suspicion for infiltrative disease. Accordingly, cardiac magnetic resonance was performed and revealed a discrete region of transmural late gadolinium enhancement in the basal to mid-IVS with increased myocardial edema and wall thickness in the IVS and inferior wall (Figure 2). This was most consistent with CS, but the lack of involvement of other organ systems was unusual. Accordingly, the patient underwent myocardial 18F-fluorodeoxyglucose positron emission tomography (FDG-PET)/CT with dietary suppression of myocardial uptake to assess for active CS and determine her need for a pacemaker alone or a pacemaker/implantable cardioverter-defibrillator to mitigate her risk of CS-associated ventricular arrhythmias. FDG-PET/CT showed intense FDG uptake in the IVS and inferior wall in addition to multiple mediastinal and hilar lymph nodes (Figure 3). These findings were highly suspicious for active CS. After placement of a

ABBREVIATIONS AND ACRONYMS

AVB = atrioventricular block
CAD = coronary artery disease
CAG = coronary angiography
CS = cardiac sarcoidosis
CT = computed tomography
ECG = electrocardiogram
FDG = 18F-fluorodeoxyglucose
IVS = interventricular septum
PET = positron emission tomography
SCAD = spontaneous coronary artery dissection
TTE = transthoracic echocardiogram

FIGURE 1 Coronary Angiography



Apparent spontaneous coronary artery dissection of a large first septal branch **(arrows)**. **(A)** Right anterior oblique caudal view. **(B)** Left anterior oblique cranial view.

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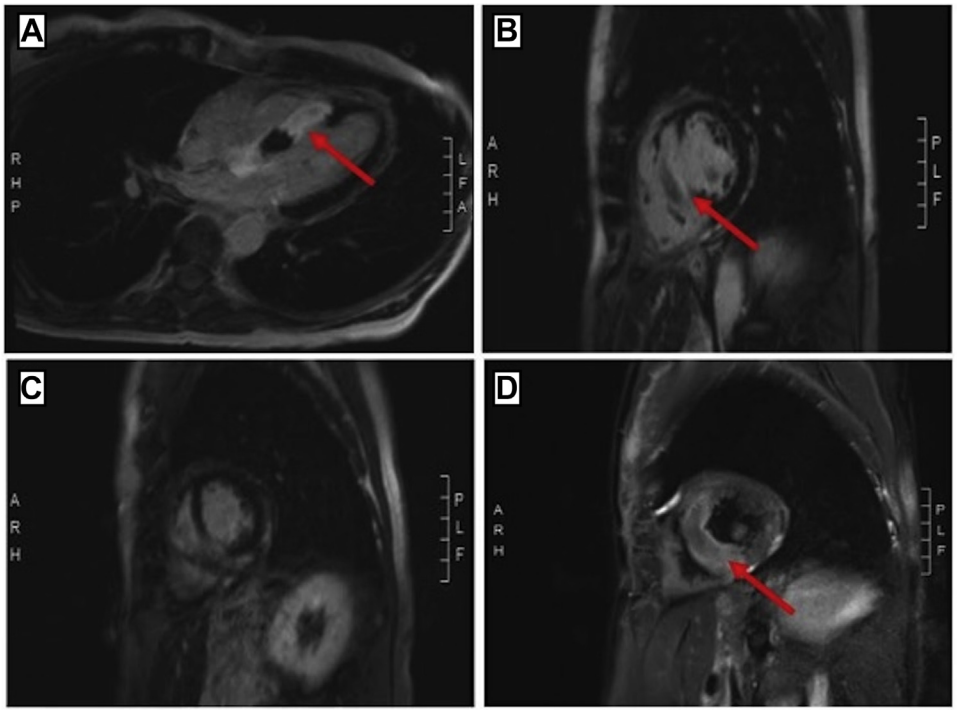
FIGURE 2 Cardiac Magnetic Resonance Showing Evidence of Active Cardiac Sarcoidosis



Discrete region of transmural late gadolinium enhancement (ie, fibrosis) in the basal to mid-interventricular septum **(arrows)**: **(A)** radial projection, **(B)** short-axis basal projection, and **(C)** short-axis mid projection. **(D)** T_2 imaging shows evidence of increased myocardial edema and wall thickness in the interventricular septum and inferior wall.

temporary transvenous pacemaker, an ultrasound-guided transbronchial needle aspiration of several mediastinal lymph nodes was performed. Pathology analysis demonstrated noncaseating granulomas (Figure 4), confirming CS.

MANAGEMENT

A dual-chamber pacemaker/implantable cardioverter-defibrillator was implanted, and the patient was discharged on high-dose prednisone (40 mg daily).

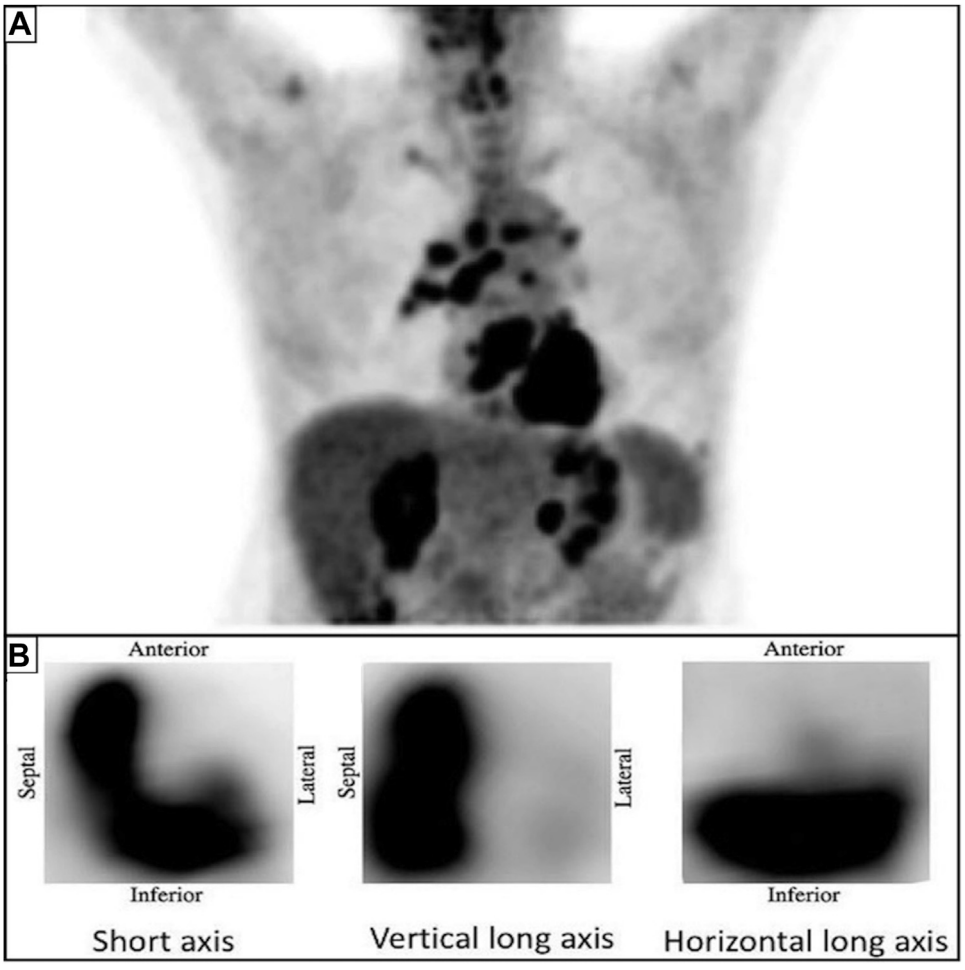
DISCUSSION

Sarcoidosis is a multisystem disease characterized by noncaseating granulomas that involve various organs including the lung, heart, liver, spleen, skin, and eyes (1). Symptomatic CS occurs in only 5% of patients with sarcoidosis, but cardiac involvement is actually present in 25% to 28% and is being increasingly identified using advanced cardiovascular imaging techniques (1).

Cardiac conduction abnormalities are common in CS; however, SCAD has only rarely been described in the context of CS (1). In fact, several findings suggest that the initial diagnosis may have been misleading. First, the patient's cardiac magnetic resonance findings were not typical of prior SCAD, which would usually show subendocardial or transmural late gadolinium enhancement without ongoing edema. Second, her persistently elevated troponin level without new obstructive coronary lesions or evidence of increased demand suggested ongoing myocardial injury from a process other than SCAD. Third, the appearance and location of her presumed SCAD were not typical. Accordingly, what might have initially appeared to be SCAD may have been coronary granulomatous arteritis associated with CS or, less likely, vascular compression from septal inflammation. Although rare, coronary granulomatous arteritis in the context of sarcoidosis has been previously reported; these cases presented with myocardial infarction (with coronary stenosis) (2,3) or compromised left ventricular function (4,5).

Although the suspicion of coronary granulomatous vasculitis was not biopsy proven, this entity provides a more plausible explanation for the

FIGURE 3 Myocardial 18F-Fluorodeoxyglucose Positron Emission Tomography/Computed Tomography



(A) Whole-body maximal-intensity projection demonstrating exuberant focal tracer uptake in the heart and multiple mediastinal and hilar lymph nodes. **(B)** Cardiac projections show focal tracer uptake in the interventricular septum and inferior wall.

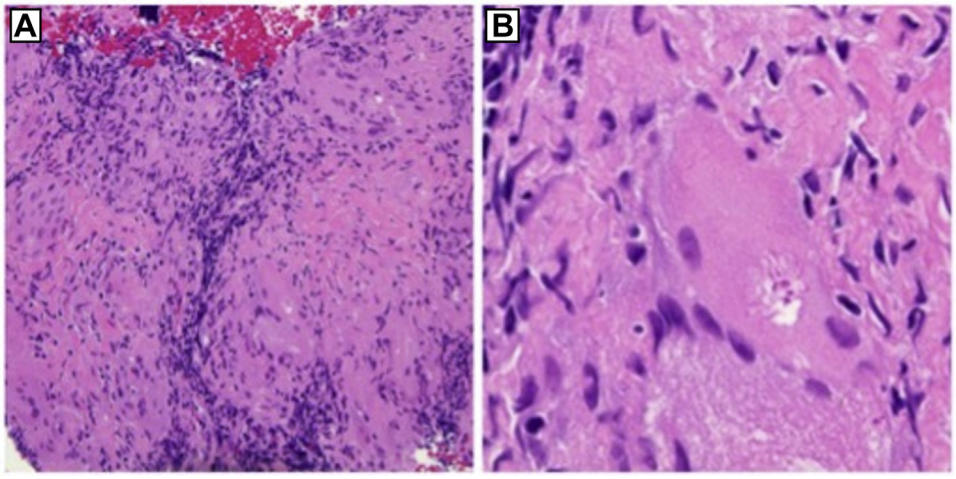
patient's CAG findings than SCAD. If she did indeed have SCAD, then this is the first known antemortem report of a patient with previously undiagnosed CS who presented with SCAD; the only other antemortem case is that of a patient with an established CS diagnosis (6).

This case highlights the importance of suspecting CS when assessing patients with conduction abnormalities, bradycardia, or unexplained myocardial injury. This is especially important in the absence of other clinical manifestations of sarcoidosis; the only extracardiac involvement in this patient involved hilar and mediastinal lymph nodes. This case also demonstrates the extent of possible presentations with which CS can manifest with coronary granulomatous arteritis being a rare but important entity to consider, especially in patients with myocardial injury but minimal traditional cardiovascular risk factors. Finally, although biopsy remains the gold standard for diagnosis, advanced imaging has emerged as a valuable, noninvasive tool to aid in CS diagnosis.

FOLLOW-UP

After discharge, the patient continued to experience occasional palpitations and developed jitteriness and anxiety while on high-dose prednisone. Ventricular pacing frequency dropped from approximately 25% to <0.1% within days. Her most recent ECG revealed first-degree AVB with normal QRS interval. Moreover, subsequent FDG-PET/CT demonstrated resolution of myocardial and lymph node FDG uptake. She

FIGURE 4 Histologic Slides



Noncaseating granulomas on hematoxylin-eosin–stained lymph node tissue histologic sections: magnifications of **(A)** ×100 and **(B)** ×400.

remained on prednisone 40 mg daily for 2 months and then was tapered to 20 mg by reducing by 10 mg/month over 2 months. She remained on 20 mg for 3.5 months, then reduced the dose by 5 mg every 1.5 months to the current maintenance dose of 5 mg with future plans to consider a steroid-sparing agent.

CONCLUSIONS

This case illustrates the importance of maintaining a suspicion for CS in unusual clinical presentations given a wide range of possible disease manifestations.

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