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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Cardiac Sarcoidosis Causing Ventricular Tachycardia After Myocardial Infarction

A Shocking Diagnosis

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ABSTRACT

Scar-mediated ventricular tachycardia (VT) commonly results from ischemic heart disease. We present a case of recurrent VT, which was initially attributed to ischemic disease; however, the scar location pointed to an alternate pathology. This case demonstrates the utility of multimodality imaging in diagnosing sarcoidosis as a cause of VT. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1056–61) © 2020 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/).

HISTORY OF PRESENTATION

A 57-year-old man with a history of presumed infarct-related cardiomyopathy, ventricular tachycardia (VT), and implantable cardioverter-defibrillator (ICD) presented with 2 ICD shocks. He was using the bathroom while at work and was shocked by his ICD. He then walked out of the bathroom and once again was shocked, prompting a call to emergency services. He presented to the emergency room with normal vital signs and without distress; interrogation of his ICD revealed appropriately treated episodes of polymorphic VT (**Figure 1**).

LEARNING OBJECTIVES

- To appreciate that, even when coronary artery disease is present, not all VT is due to ischemia or infarction.
- To understand the benefits of CMR in evaluating scar-related VT.
- To understand the benefits of FDG PET in diagnosing and monitoring cardiac sarcoidosis.

MEDICAL HISTORY

One year prior to the current presentation, the patient presented to his local hospital with an episode of syncope while driving. In the hospital, the patient had recurrent ventricular arrythmias on cardiac monitoring. Work-up at that time had revealed a significant stenosis in the left anterior descending artery (LAD), and it was presumed that LAD territory ischemia was responsible for the VT and syncope. The patient received 2 drug-eluting stents and was discharged on amiodarone and with an external defibrillator—ejection fraction (EF) was noted to be 35% with an inferior wall motion abnormality on echocardiography.

A few months later, the patient's echocardiogram revealed normal chamber size, grade I diastolic dysfunction, and EF of 30% despite revascularization and medical therapy. Electrocardiography revealed

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T-wave inversions in the inferior leads. An ICD was implanted, and amiodarone was discontinued to prevent long-term adverse effects.

Three months after implantation, he experienced an ICD shock but did not seek medical attention. Two months later, he experienced a second shock and presented to his local hospital for evaluation. Device interrogation showed multiple episodes of nonsustained VT and polymorphic VT, with polymorphic VT episodes being treated by his device. It was presumed that the VT was scar mediated from a prior infarct, with premature ventricular contractions (PVCs) being the trigger based on device interrogation. Electrophysiology study and VT ablation was performed, with PVCs in the mid and apical inferoseptum mapped as triggers for polymorphic VT. The mid inferoseptal PVCs were ablated; however, the apical PVCs were not completely ablated, as they were associated with hemodynamically unstable VF; the facility did not have capability to complete the ablation with hemodynamic support.

Two months after ablation, he had recurrent VT and ventricular fibrillation and once again presented to his local hospital. Cardiac catheterization was performed at that time, which revealed patent LAD stents and no significant coronary artery disease. He was started on antiarrhythmic therapy with mexiletine 200 mg 3 times a day but 2 months later presented to our hospital with recurrent ICD shocks for VT and ventricular fibrillation. Notably, the patient has a family history of sudden cardiac death in his mother when she was in her 60s, as well as in 2 siblings who were in their 50s.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for recurrent VT includes ischemic heart disease, end-stage heart failure, and congenital heart disease including arrhythmogenic right ventricular dysplasia, hypertrophic cardiomyopathy, and infiltrative heart disease.

INVESTIGATION

Electrocardiography on most recent presentation showed sinus bradycardia with inferior T-wave inversions (Figure 2). Given the recurrence of VT despite revascularization, VT ablation, and antiarrhythmic therapy, cardiac magnetic resonance (CMR) with an ICD protocol was performed to investigate for infiltrative disease and scar burden (1).

The CMR showed normal left ventricle size with reduced function and a left ventricular EF of 32%, with mid to distal inferior, inferoseptal, and inferolateral thinning with akinesis. Late gadolinium enhancement was performed and was markedly abnormal, with full-thickness transmural scar extending throughout the inferior, inferoseptal, and inferolateral walls (Figure 3).

Given that the scar location did not correspond to the patient's treated coronary artery disease, infiltrative disease was suspected, with sarcoidosis being high on the differential. A cardiac fluorodeoxyglucose (FDG) positron emission tomography (PET) study was ordered to evaluate for inflammation, and showed increased FDG uptake in

ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

EF = ejection fraction

FDG = fluorodeoxyglucose

ICD = implantable cardioverter-defibrillator

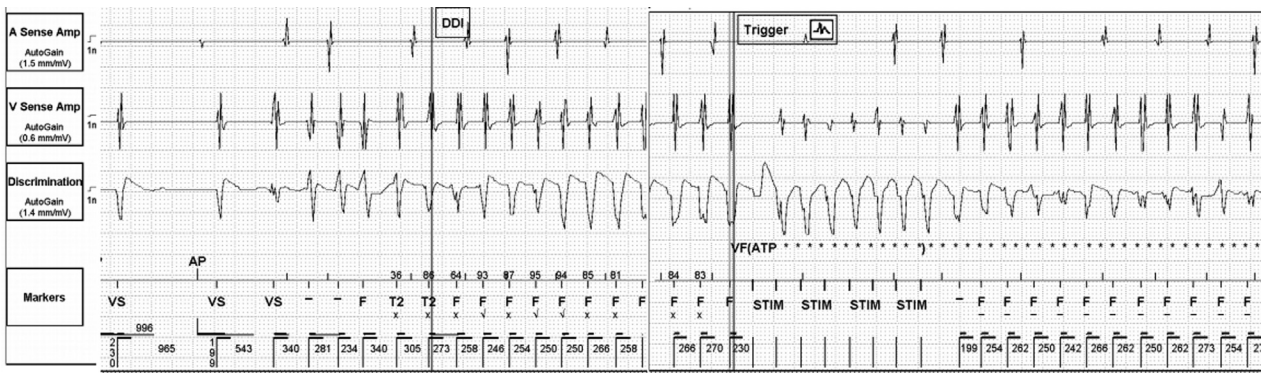
LAD = left anterior descending artery

PET = positron emission tomography

PVC = premature ventricular contraction

VT = ventricular tachycardia

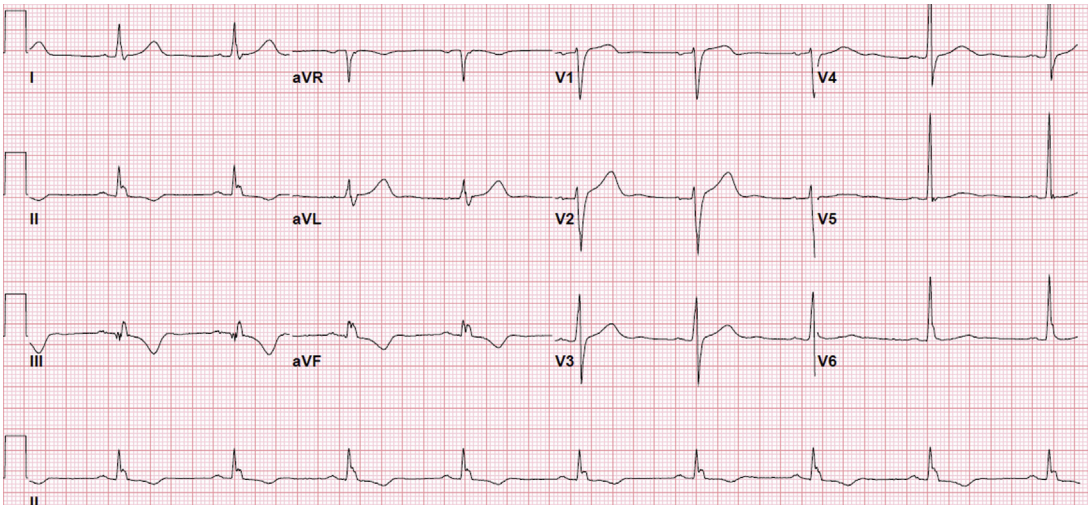
FIGURE 1 Device Interrogation



Intracardiac electrocardiogram's from the patient's implantable cardioverter-defibrillator showed multiple examples of premature ventricular contraction–triggered ventricular arrhythmias. In this representative example, a premature ventricular contraction triggers polymorphic ventricular tachycardia, and there is an attempt to break the rhythm with antitachycardia pacing. The rhythm eventually self-terminated.

JACC: CASE REPORTS, VOL. 2, NO. 7, 2020
JUNE 17, 2020:1056–61

FIGURE 2 Electrocardiography



The patient's electrocardiography showed sinus bradycardia with inferior T-wave inversions.

the basal inferior, inferoseptal, and inferolateral walls. There was also hilar lymph node uptake along with mid to basal inferior and inferoseptal resting perfusion defects on the rubidium perfusion images, indicative of scar and inflammation—findings consistent with sarcoidosis (Figures 4 to 6). Maximum standardized uptake values were 21.8 in the inferoseptum and 5.1 in the hilar nodes. The patient underwent bronchoscopy with lymph node biopsy, which confirmed the diagnosis of sarcoidosis.

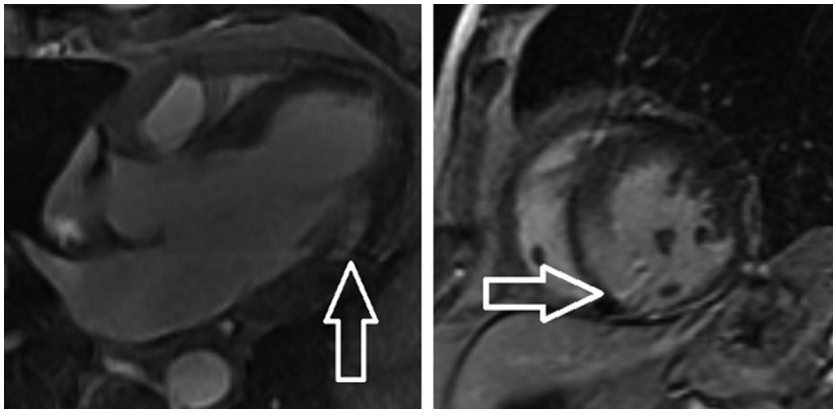
MANAGEMENT

The patient continued on mexiletine in the short term for VT suppression; meanwhile, the underlying cause of his VT, sarcoidosis, was treated with oral prednisone.

DISCUSSION

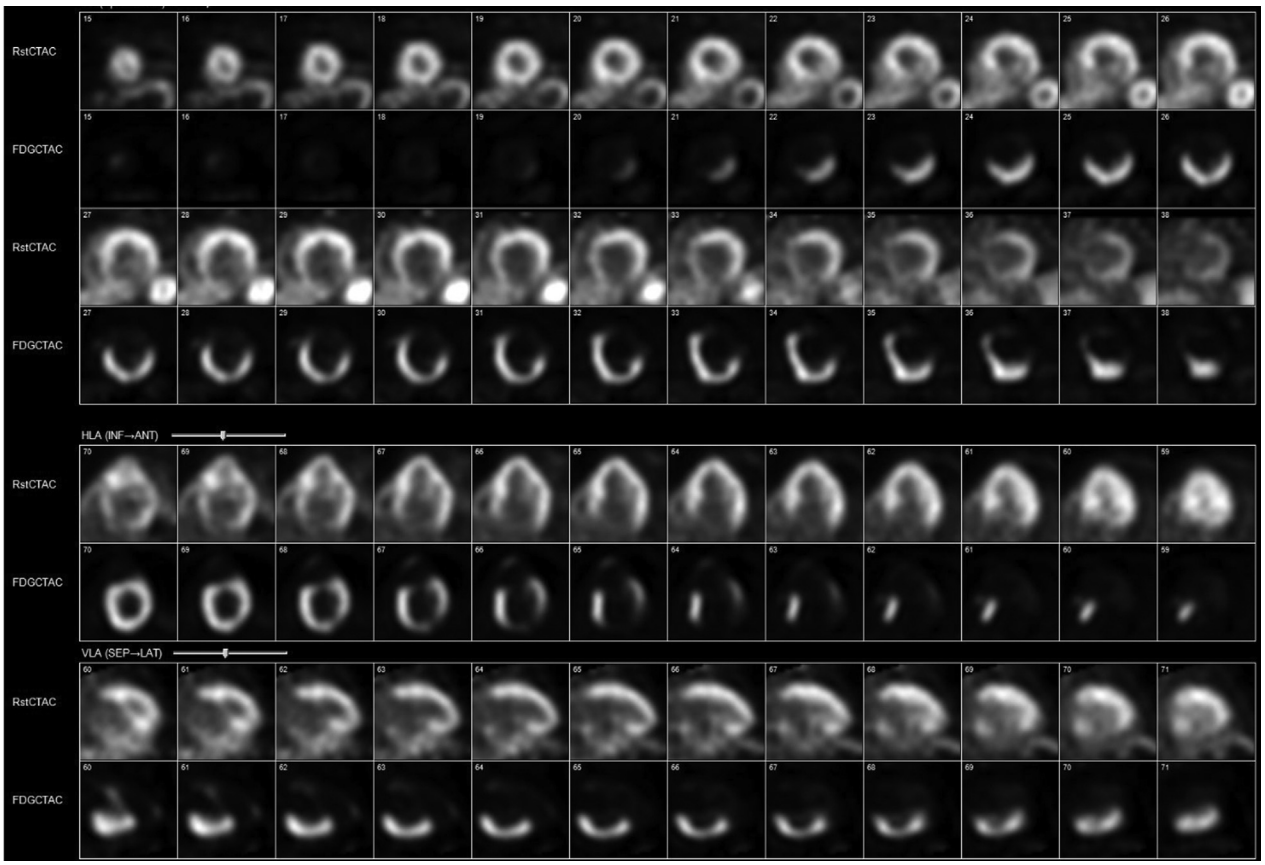
Sarcoidosis is a granulomatous disease that can affect any organ. Among patients with sarcoidosis, rates of

FIGURE 3 Cardiac Magnetic Resonance



The patient's cardiac magnetic resonance showed thinning of the inferior and inferolateral wall. Late gadolinium enhancement in this area indicates scar. **Arrows** indicate areas of late gadolinium enhancement.

FIGURE 4 **Cardiac FDG PET Splash Images**



The fluorodeoxyglucose (FDG) positron emission tomography showed a resting perfusion defect in the inferior, inferoseptal, and inferolateral walls, and increased amount of FDG uptake in the matched area, consistent with sarcoidosis-mediated scar and inflammation.

cardiac involvement have been reported to be as low as 5% to as high as 27% in autopsy studies (2,3).

Making the diagnosis of cardiac sarcoidosis is challenging, but perhaps even more difficult is initially suspecting cardiac sarcoidosis and knowing when to screen for it. The 2014 Heart Rhythm Expert Consensus Statement on the Diagnosis and Management of Arrhythmias Associated With Cardiac Sarcoidosis provides some guidance (4). Based on that statement, individuals with diagnosed extracardiac sarcoidosis should be screened with at least a 12-lead electrocardiogram, a history with emphasis on syncope and palpitations, and an echocardiogram. More advanced imaging modalities are noted to be appropriate only when those initial tests are abnormal.

The consensus is less clear when there is no prior diagnosis of extracardiac sarcoidosis. This is relevant not only because there are cases of isolated cardiac sarcoidosis, but also because even when extracardiac sarcoidosis is present, the initial presentation, as demonstrated by our case, may still be cardiac (5). Small studies and case series have described sarcoidosis patients who had an initial presentation of VT (6,7). This is still thought to be rare, leading to discord among the experts; a majority of the writing group for the 2014 Heart Rhythm Society Expert Statement recommended screening for sarcoidosis in patients with unexplained monomorphic VT, but it did not meet the threshold to be included as a formal recommendation.

Even if that recommendation had been made a standard of care, our patient would have been missed because it was assumed that ischemic heart disease was the explanation for VT. Nonetheless, several clues were present that led to the further work-up of our patient's VT. Notably, review of echocardiography from his initial presentation revealed inferior

JACC: CASE REPORTS, VOL. 2, NO. 7, 2020
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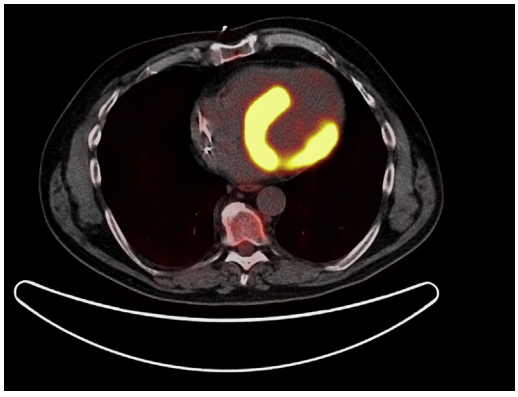
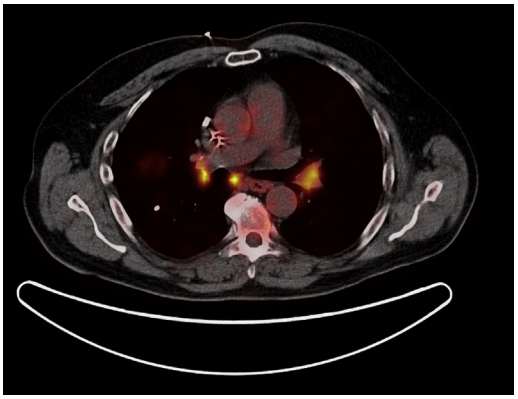
FIGURE 5 Whole-Body Transverse Image of Cardiac Fluorodeoxyglucose Positron Emission Tomography



Positron emission tomography revealed increased fluorodeoxyglucose uptake in the inferior, inferoseptal, and inferolateral walls. This is consistent with cardiac sarcoidosis.

FIGURE 6 Fluorodeoxyglucose Positron Emission Tomography



This also revealed increased fluorodeoxyglucose in the hilar lymph nodes, consistent with systemic sarcoidosis.

wall hypokinesis. This should have raised a question about his initial syncopal event, as his coronary disease was only significant in the LAD territory, suggesting an alternate etiology for the inferior wall hypokinesis. The question could have been raised again during his VT ablation when he was found to have isolated scar in the inferior wall, which was ablated.

The lack of correlation between the location of scar and coronary disease, combined with his incessant polymorphic VT despite a lack of active ischemia, appropriate revascularization, and antiarrhythmic therapy led to the further work-up of his VT. CMR is a useful modality not only to show the scar location, but also to provide the scar burden, tissue characteristics, and a reproducible EF.

Although endomyocardial biopsy is the gold standard for diagnosing cardiac sarcoidosis, and could have been considered after our CMR, the patchy nature of sarcoidosis gives the test a low sensitivity. In the right clinical scenario, FDG-PET imaging has high diagnostic yield in cardiac sarcoidosis, and can also be used for prognostication. A metanalysis of 7 studies with a total of 164 patients calculated a pooled sensitivity of 89% and a pooled specificity of 78% (8). When combined with resting perfusion imaging, cardiac PET has the ability to colocalize scar and inflammatory burden in the myocardium. An important consideration before performing cardiac PET for sarcoidosis is to exclude significant coronary artery disease, prior myocardial infarction, resting ischemia, or hibernating myocardium. The presence of both scar and inflammation is considered an adverse prognostic sign associated with a higher risk of death or VT. FDG-PET can also identify extracardiac sarcoid and in fact, the Heart Rhythm Society criteria prefer extracardiac tissue biopsy over endomyocardial biopsy because of the safety and higher yield of the former to confirm the diagnosis. In addition, it is possible to quantify inflammatory disease activity using standardized uptake values, and these values can be used to monitor response to treatment (9,10). Our patient's FDG-PET showed not only cardiac inflammation and scar, but also typical hilar lymphadenopathy, and confirmatory diagnosis was thus made by lymph node biopsy.

FOLLOW-UP

The patient continued on oral steroids with a gradual taper toward a maintenance dose. At 3-month follow-up, device interrogation revealed no episodes of sustained ventricular arrhythmias. A repeat PET scan showed less inflammation than the initial diagnostic scan, with a maximum cardiac SUV of 8.4 (prior 21.8) and maximum hilar SUV of 2.9 (decreased from 5.1). He will continue on oral steroids and have his dose adjusted based on symptoms and serial PET scanning.

CONCLUSIONS

Sarcoidosis can be a hidden cause of VT, even in patients with coronary artery disease. Presence of scar

on CMR or resting perfusion imaging may not always represent myocardial infarction. An infiltrative process can be the culprit and the cause of recurrent ventricular arrhythmias, and should be suspected whenever there is a discrepancy between scar location and angiographic coronary artery disease.

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