

CLASSIC IMAGING IN CARDIAC SARCOIDOSIS

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CASE PRESENTATION

A 52-year-old man presented to an outside hospital after the advice of his cardiologist due to concerns of recurrent ventricular tachycardia seen on device checks. The patient was overall asymptomatic despite his ventricular arrhythmias. His medical history was pertinent for type II diabetes mellitus, hypertension, and hyperlipidemia. He also had a history of complete heart block (CHB) for which he underwent dual chamber pacemaker placement in March 2021. His echocardiogram at that time had normal left ventricular ejection fraction (LVEF). Imaging at pacemaker placement had evidence of prominent hilar adenopathy. Notably, the patient also had a history of recurrent respiratory infections for which he was frequently treated with antibiotic and steroid therapies. In the interim, the patient continued follow-up with cardiology. His LVEF dropped to 25% then moderately improved to 40% after goal directed medical therapy. He was lost to follow-up until current presentation. In the outside hospital, patient underwent heart catheterization which showed non-obstructive coronary disease as well as repeat echocardiogram with LVEF of 26%. Patient was

discharged home with Life Vest with plan for outpatient upgrade of pacemaker to implantable cardiac defibrillator (ICD). He was evaluated by his cardiologist who became suspicious for underlying cardiac sarcoidosis (CS) leading to expedited admission and workup. Inpatient workup included advanced cardiac imaging. Cardiac positron emission tomography (PET) showed myocardial ¹⁸F-fluorodeoxyglucose (¹⁸F-FDG) uptake (focal, 3 to 5 segments) with resting perfusion defects in the basal septum consistent with perfusion-metabolism mismatch concerning for active inflammation (Figure 1).

DISCUSSION

The advent of advanced cardiac imaging including cardiac PET and CMR has helped immensely in bolstering a diagnosis of CS without need for endomyocardial biopsy which is both invasive and has low sensitivity given the patchy, focal nature of CS. Cardiac PET findings that are classically associated with CS are focal areas of ¹⁸F-FDG uptake. If concurrent FDG is noted in the same territory as perfusion defect (perfusion-metabolism mismatch) then the perfusion defect likely is secondary to inflammation. The presence of perfusion defect without any associated ¹⁸F-FDG uptake, is consistent with scarring (2, 3). It is therefore vital to compare rest myocardial perfusion images with FDG uptake when evaluating for CS. In our case, the cardiac PET showed moderate inflamed myocardium (focal, 3 to 5 segments), resting perfusion defect in the basal septum with metabolism mismatch with

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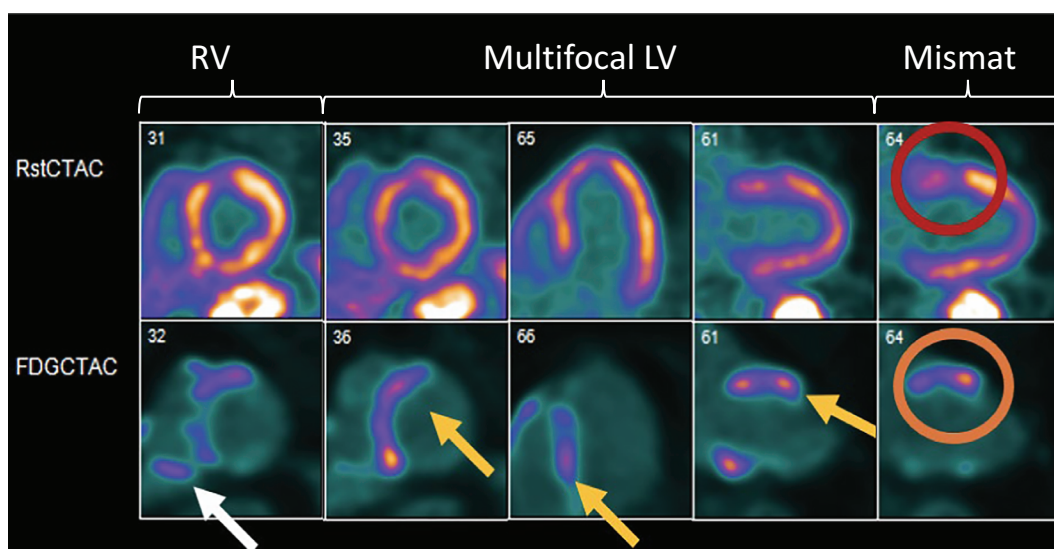


Figure 1. Perfusion metabolism PET-CT with ^{82}Rb and ^{18}F -FDG shows patchy areas of mismatch perfusion metabolism in the basal septal, anterior and inferior walls consistent with inflammation. Yellow arrows indicate ^{18}F -FDG uptake (basal septum, basal to mid anterior). White arrow indicates ^{18}F -FDG uptake in the right ventricle. The circles show evidence of mismatch pattern with red circle indicating perfusion defect and orange circle showing corresponding ^{18}F -FDG uptake. Notably there was prominent ^{18}F -FDG uptake in hilar lymph nodes. Cardiac magnetic resonance (CMR) imaging showed multifocal and patchy late gadolinium enhancement (LGE) in the basal antero-septum (mid-myocardial), mid anterior wall (transmural), and superior/inferior RV insertion points (sub-epicardial). (Figure 2). There was also a small component of subendocardial to mid wall LGE in the distal inferoseptum. Patient underwent bronchoscopy with endobronchial ultrasound (EBUS) transbronchial needle aspirations (TBNA) of left and right hilar lymph nodes which were positive for non-necrotizing granulomas. He was initiated on prednisone and leflunomide therapies and upgraded to implantable cardiac resynchronization therapy defibrillator. This illustrates a classic case of cardiac sarcoidosis in a middle-aged man who had delay in diagnosis from the outset. A diagnosis of complete heart block in an otherwise healthy man should prompt further evaluation for inflammatory conditions such as sarcoidosis. Studies suggest that up to 5% of sarcoidosis have symptomatic cardiac involvement, and that as much as 25% of sarcoidosis have asymptomatic cardiac involvement (1). It is essential to have high index of suspicion for cardiac sarcoidosis given potentially fatal outcomes including heart failure, conduction abnormalities, ventricular tachycardia, and sudden cardiac death. The diagnosis of CS is multi-faceted and often, multi-disciplinary.

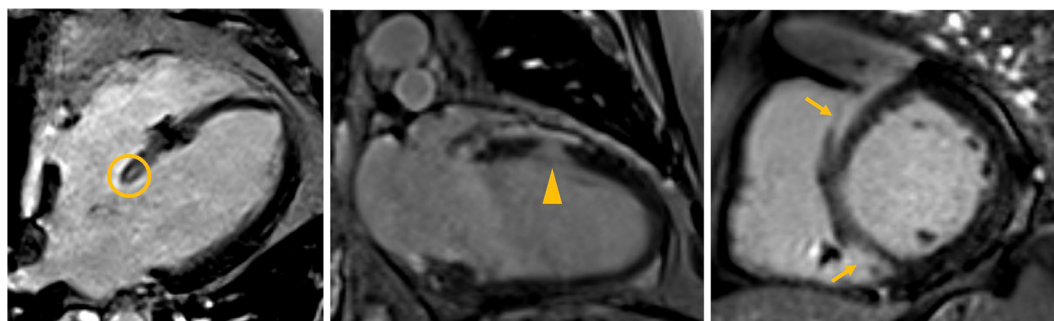


Figure 2. Late gadolinium enhancement imaging shows patchy multifocal enhancement in the basal antero-septum (mid-myocardial, circle), mid anterior wall (transmural, arrowhead), and superior/inferior RV insertion points (sub-epicardial, arrow).

FDG uptake, consistent with active inflammation (Figure 1). CMR findings that are typical for CS include LV subepicardial involvement, which is multifocal, septal, and often involves the RV free

wall (4), all of which our patient exhibits (Figure 2). These typical findings have been associated with a high risk of ventricular arrhythmias independent of LVEF and extent of LGE (5).

CONCLUSION

This case highlights the importance of having a high index of suspicion for CS. Patients may have subclinical features making this diagnosis even more challenging. Advanced cardiac imaging techniques and specific sarcoidosis protocols have allowed clinicians to diagnose and treat CS more readily and forgo invasive endomyocardial biopsies. However, there is still work to be done in the realm of identifying imaging characteristics and patterns which may clue us into the various spectrum of severities seen in CS.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

Informed Consent: Informed consent was telephonically obtained from all participants following the principles outlined in the Declaration of Helsinki.

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