Cardiac Sarcoidosis: A Resonating Diagnosis

University of Colorado Anschutz Medical Campus

Shea Claflin, MS2[1] and Elizabeth Gillespie, MD[2]

[1]*University of Colorado School of Medicine, Denver, CO.*
[2]*Department of Medicine-Hospital Medicine, University of Colorado, Denver, CO.*

Introduction

Sarcoidosis is a granulomatous disease of unknown etiology that has various clinical presentations dependent upon its systemic involvement.[1] Approximately 5% of patients diagnosed with sarcoidosis also meet clinical criteria for cardiac sarcoidosis (CS), whereas subclinical cardiac involvement is observed in 20-50% of cases.[2,3]

Common clinical presentations include:[2]
- Syncope, palpitations, chest pain
- AV block – Mobitz I and II, 3rd degree AV block
- Ventricular tachycardia and fibrillation
- Sudden cardiac death
- Less common: heart failure, valvular abnormalities[4]

Though the true prevalence of CS is unknown, there is evidence that is associated with a poorer prognosis.[5]

Case Description

History:
Mr. D is a 46 yo male who presents for evaluation of palpitations, syncopal episodes, and shortness of breath on exertion. During transfer to Denver Health for chest pain and abnormal EKG findings of 1st degree AV block and SVT, he experienced a pre-syncopal episode preceded by light- headedness and dizziness. Providers at the transferring facility had recently observed 2nd degree Mobitz I AV block (while on a beta blocker). Mr. D reports 5 years of intermittent chest pain and syncope. He has taken metoprolol for the past 14 years, for nonspecific tachyarrhythmia and palpitations.

Studies:
- EKG: 2nd degree AV block Mobitz II, prolonged QTc, PVCs
- Telemetry: Mobitz I and II, 3rd degree AV block
- CXR: perihilar prominence, streaky opacities
- Echocardiogram: thinner basal antero-septum
- CMR: late gadolinium enhancement
- Chest CT: scattered bilateral pulmonary nodules
- Transbronchial biopsy – no granulomas identified

Management:
- Dual chamber pacemaker with ICD
- Ophthalmology exam normal
- Discharged on metoprolol for continued palpations and escitalopram for anxiety surrounding newly diagnosed heart condition

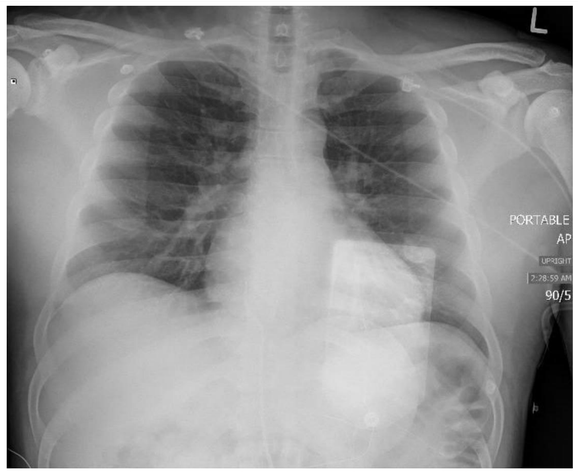


Figure 1. (left)
Initial Chest X-Ray demonstrated perihilar prominence and streaky retrocardiac opacities.

Figure 2. (right)
Cardiac MRI demonstrated late gadolinium enhancement with epicardial predominance, involving basilar anterior and posterior insertion of RV walls into IV septum, and inferior insertion extending to midventricle and involving inferior RV free wall.

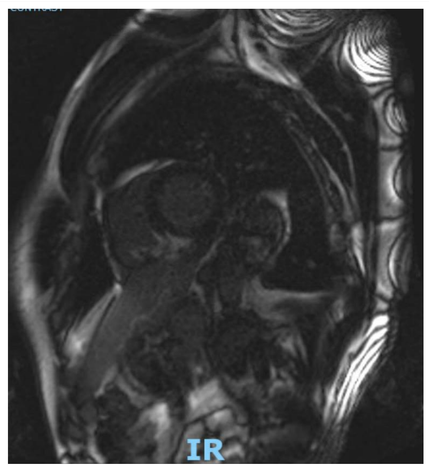
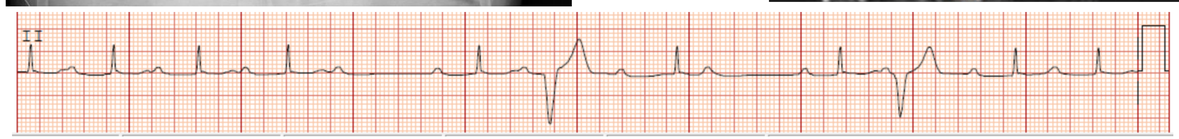


Figure 3. (below)
ED EKG revealed Mobitz II, prolonged qTc interval, and PVC.



Discussion

Cardiac sarcoidosis often presents with nonspecific symptoms and findings on studies like EKG and echocardiogram. The gold standard for diagnosis is endomyocardial biopsy, but this has a low sensitivity of 20-30% in exchange for a high risk of complications. Thus, the diagnosis can be challenging to make, so a high index of clinical suspicion is of utmost importance. Suspect CS in patients with new onset conduction abnormalities, unexplained syncope, unexplained cardiomyopathy, unexplained sudden cardiac death, and sustained ventricular tachycardia.[2,6]

There are currently three sets of clinical criteria that can be used to make the diagnosis of CS: the Japanese Ministry of Health and Welfare (JMHW), the Heart Rhythm Society (HRS), and the World Association for Sarcoidosis and Other Granulomatous Disorders.[7]
A generalized diagnostic approach that most closely resembles the HRS guidelines is shown below:[8]

H&P w/ 12-lead ECG (and echocardiogram)

Evaluation for patients with previously confirmed extracardiac sarcoidosis or those under clinical suspicion for CS.

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CMR or FDG-PET

Advanced Cardiac Imaging Criteria:
With extracardiac sarcoidosis
- Palpitations, presyncrope, syncope
- ECG abnormalities
- Echo abnormalities

Without extracardiac sarcoidosis:
- Unexplained Mobitz II or 3rd degree block, age <60
- Sustained monomorphic ventricular tachycardia

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Histologic Confirmation

Biopsy is required for definitive diagnosis, whether endomyocardial biopsy or extracardiac.

CS management consists of immunosuppressive therapies, medical or surgical management of conduction abnormalities, and goal-directed therapy for heart failure.

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Disclosures

We have no disclosures.