

# Introduction

Cardiac sarcoidosis (CS) can classically cause infiltrative nonischemic cardiomyopathy. This is a case of severe sarcoidosis leading to cardiogenic shock.

### Case

- 46-year-old AA female with PMH of HTN presented with DOE, orthopnea, and PND suggestive of HF exacerbation.
  - BNP 1500
  - 2D Echo: LA/LV dilation, EF 15%. Global hypokinesis.
  - LHC negative. RHC moderate pulmonary HTN.
- Patient was discharged but ultimately unable to follow up with advanced HF in outpatient setting.
- She was readmitted for HF exacerbation several times over the course of the next year without inpatient workup for NICCM. She suffers from PCA stroke during this time.
- Her next admission was to CICU for cardiogenic shock, where she was stabilized with inotropic support. Cardiac MRI was ordered for evaluation for LVAD implementation.
  - MRI showed pulmonary sarcoid with abnormal myocardial enhancement (Fig. 1).
  - Core biopsy during LVAD placement showed numerous non-necrotizing granulomas.
- Over the next 2 years, she experienced several infections of the LVAD drive line, most recently with *Pseudomonas*. Repeat 2D echo showed recovered EF to 55% and the decision was made to explant the device.
- She was readmitted for HF exacerbation with echo showing interval decrease in EF to 25-30%. PET CT showed extracardiac sarcoid. Patient was started on methotrexate and steroid taper.

### References

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# **A Noncaseating Case of Elusive Sarcoidosis**

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### Figure 1: MRI Cardiac

Delayed myocardial hyperenhancement in the interventricular septum and in portions of the lateral wall.



# Figure 2: Cardiac PET/CT

Showing FDG activity in the anterior wall without perfusion defect. New uptake in inferior wall more extensive than prior. Pattern may represent early recurrent cardiac sarcoidosis.

# **Case continued**

- She was readmitted months later with adrenal insufficiency secondary to difficulty with steroid taper instructions. She was also found to have adherence issues with methotrexate. PET/CT during this admission showed active cardiac sarcoidosis (Fig. 2).
- She was started on CellCept and inpatient steroids. Since starting CellCept, she is following closely with HF clinic and rheumatology without further readmission and has an EF recovered to 45%. Most recent PET/CT without active sarcoidosis.

## Discussion

- Sarcoidosis is an inflammatory condition that is classically found in the lungs. It causes a spectrum of disease ranging from mild dyspnea to pulmonary hypertension. The incidence in the AA population is 34 in 100k.<sup>1</sup>
- CS is a rare manifestation of sarcoidosis, happening in less than 10% of cases. In these cases, the most common manifestation is conduction abnormalities including AV nodal blockade and ventricular arrythmias.
- CS can cause a restrictive type CM in up to 80% of patients, but clinically significant HF is seen in less than 30% of patients.<sup>2</sup>
- This case is exemplary in that new NICCM an infiltrative evaluation should be warranted in absence of other common factors given the significant risk of the disease, such as sudden cardiac death as the first manifestation.
- First line treatment of CS is often steroids with repeat PET imaging in 3 months to evaluate resolution of disease. Presence of disease after reevaluation prompts the need for immunosuppressive therapies such as methotrexate or mycophenolate mofetil.<sup>3</sup>



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