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A Peculiar Presentation of Cardiac Sarcoidosis As Third-Degree Atrioventricular Heart Block Complicated By Right Ventricle Perforation

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BACKGROUND

- Cardiac sarcoidosis is a rare, inflammatory multisystem disorder that manifests as noncaseating granulomas of multiple organs.
- The clinical presentation of sarcoidosis is variable and may be underrecognized by clinicians.
- Individuals may be asymptomatic during their lifetimes and may be incidentally diagnosed with the disease for the first-time during autopsy.
- Patients with cardiac involvement of sarcoidosis may initially present with complications including arrhythmias, heart block, bundle branch block, congestive heart failure, pericardial effusion, pulmonary hypertension, and/or sudden cardiac death.

CASE DESCRIPTION

- A 59-year-old African American female with past medical history of untreated hypertension and hyperlipidemia presented following multiple episodes of syncope and near syncope.
- On initial exam, patient was found to be in 3rd degree atrioventricular heart block with a ventricular escape rhythm of 40 beats per minute. **(Figure 1)**
- A temporary transvenous pacemaker was placed through the right Internal Jugular Vein with good capture.
- The patient was transferred to a tertiary care center for a permanent pacemaker placement.
- Upon arrival, the patient was noted to have regained intrinsic rhythm with EKG demonstrating normal sinus rhythm with a first-degree atrioventricular heart block and right bundle branch block at 70 bpm. **(Figure 2)**
- Later that day, the temporary pacemaker malfunctioned and stopped capturing despite increasing the pacing threshold to the maximum setting.
- CT scan of the chest demonstrated that patient's transvenous pacemaker lead had perforated through the free wall of the right ventricle with the tip of the pacing catheter extending approximately 3-4 cm into the pericardium. **(Figures 5 & 6)**
- The original temporary pacemaker was then removed under transesophageal echocardiogram guidance and a separate temporary pacemaker was floated through the right femoral vein under fluoroscopic guidance.
- A chest x–ray performed initially showed bilateral hilar adenopathy and the CT scan of the chest verified the presence of mediastinal, subcarinal and hilar adenopathy concerning for pulmonary sarcoidosis. **(Figures 3 & 4)**

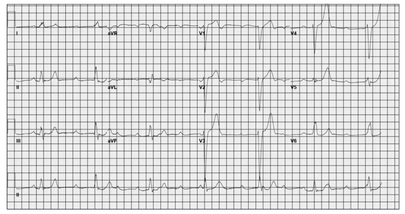

Figure 1
EKG demonstrating 3rd degree atrioventricular heart block with a ventricular escape rhythm of 40 bpm.

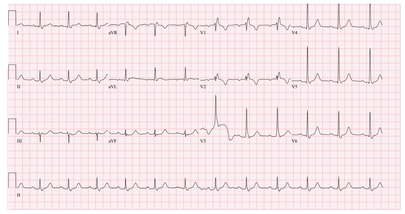

Figure 2
EKG demonstrating normal sinus rhythm with first degree atrioventricular heart block and right bundle branch block.

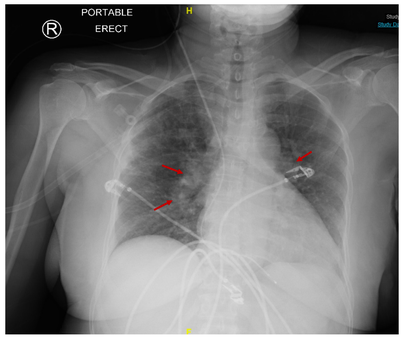

Figure 3
CXR showing bilateral peri-hilar lymphadenopathy.

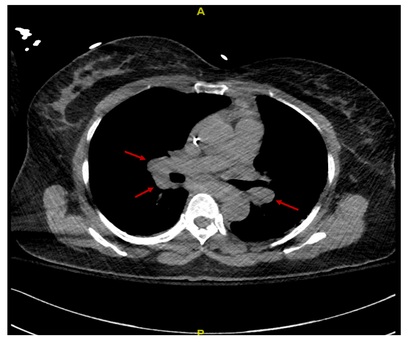

Figure 4
Chest CT scan showing bilateral peri-hilar lymphadenopathy.

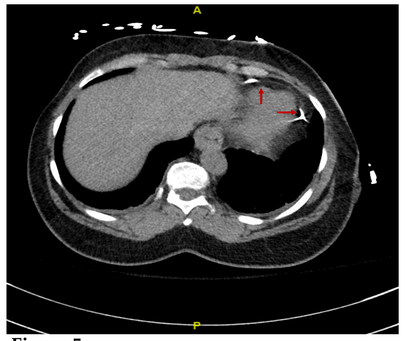

Figure 5
Chest CT showing perforation of right ventricle by temporary transvenous pacemaker lead.

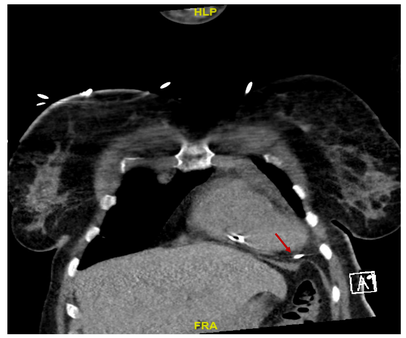

Figure 6
Chest CT coronal slice showing perforation of right ventricle by temporary transvenous pacemaker lead.

CASE OUTCOME

- A dual chamber implantable cardioverter defibrillator was placed in accordance with the 2017 American College of Cardiology/American Heart Association/ Heart Rhythm Society (AHA/ACC/HRS) guidelines.
- Patient was discharged in a stable condition to follow up with a cardiologist and a pulmonologist for further confirmatory testing.

DISCUSSION

- **The patient's clinical presentation with episodes of heart block and pulmonary lymphadenopathy lead to the presumptive diagnosis of cardiac sarcoidosis.**
- Cardiac Sarcoidosis is associated with a poor prognosis due to the many lethal complications arising from the disease, particularly high degree atrioventricular blocks, ventricular arrhythmias, and sudden cardiac death.
- A diagnosis of cardiac sarcoidosis can be made by non-invasive testing such as a cardiac MRI or a cardiac FDG-PET scan if a patient has histologic or clinical evidence of extracardiac sarcoidosis and signs and symptoms of palpitations, syncope/near syncope, conduction abnormalities such as heart block, ventricular tachycardia, or even echocardiographic abnormalities such as regional wall motion abnormality, ventricular aneurysm, basal septal thinning, or depressed LV ejection fraction.
- It is recommended that patients with extracardiac sarcoidosis with or without cardiac symptoms should be evaluated for subclinical or clinical cardiac involvement.

CONCLUSION

- This case highlights a severe, insidious clinical manifestation of cardiac sarcoidosis and the importance of early recognition and treatment of the condition.

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