Through the looking glass: ECG as a window to diagnosing Cardiac Sarcoidosis in incident heart failure

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Background

Diagnosing Cardiac Sarcoidosis (CS) is challenging, requiring multi-modal imaging and/or histopathology. Yet, some of the most common manifestations can be detected on simple point of care testing and can clue clinicians in to the diagnosis early.

History of Present Illness

A 68 year old male presented with multiple, new episodes of acute dyspnea when walking that resolved with rest. He denied orthopnea, paroxysmal nocturnal dyspnea, palpitations, or chest pain. Until recently, he went to the gym multiple times per week without limitations.

Past History

Past Medical History: Hypertension, hyperlipidemia, HCV antibody positive (viral load undetectable)
Medications: Tamsulosin, multivitamin
Family History: Mother with sarcoidosis and COPD
Social History: Retired nurse. Independent in activities of daily living. Drank 1 beer daily, 50 pack-year tobacco smoking history, active intranasal heroin and cocaine use

Exam and Initial Workup

Physical Exam

VS: Heart Rate: 102, BP: 114/77, Resp: 18, SpO2: 98% on room air
General: Well appearing, laying flat in no acute distress.
Cardiovascular: Tachycardic, regular rhythm, normal S1, S2, no murmurs, rubs, or gallops. JVP 8cm.
Pulmonary: Breathing unlabored, lungs clear to auscultation
Abdomen: Soft, nontender, no organomegaly
Extremities: Warm and well perfused. 1+ bilateral pedal edema.

Labs

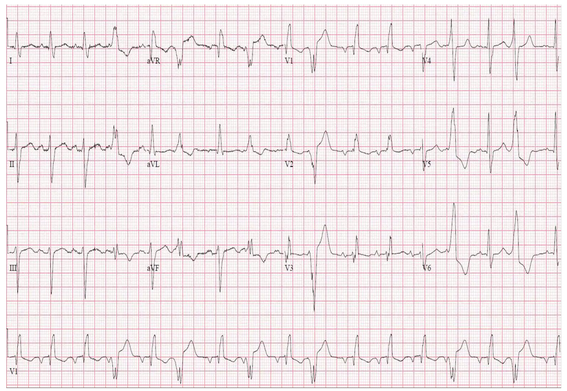
CBC, BMP within normal limits. Liver enzymes mildly elevated at AST 51 U/L, ALT 56 U/L. Urine toxicology + cocaine, fentanyl.
Troponin I 0.07 ng/mL > 0.06 > 0.05. Pro-BNP 2,917 pg/mL.
D-dimer 0.98 mg/L FEU.

Initial imaging in Emergency Department

Chest X-ray: small bilateral pleural effusions, cardiomegaly, mild vascular congestion.

CTA Chest: no pulmonary embolism, small (right greater than left) pleural effusions and dependent atelectasis, emphysema, mediastinal and hilar lymphadenopathy

ECG and Echo



Admission ECG: first degree atrioventricular (AV) block, right bundle branch block (RBBB), left anterior fascicular block (LAFB) and premature ventricular contractions (PVCs).

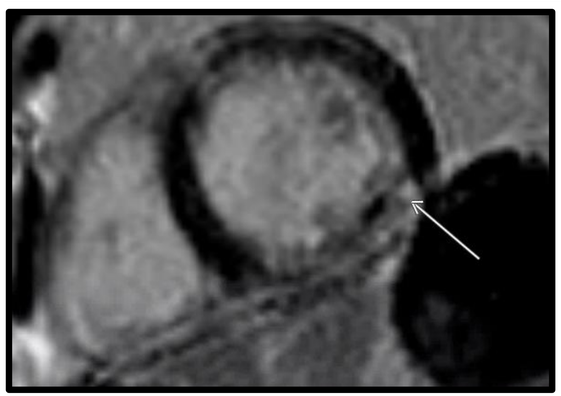
Echocardiogram: Left ventricule ejection fraction (LVEF) 30-35%. Inferior and inferoseptal wall akinesis. Moderate hypokinesis of the other LV walls. Grade II diastolic dysfunction. Moderately decreased right ventricular systolic function.

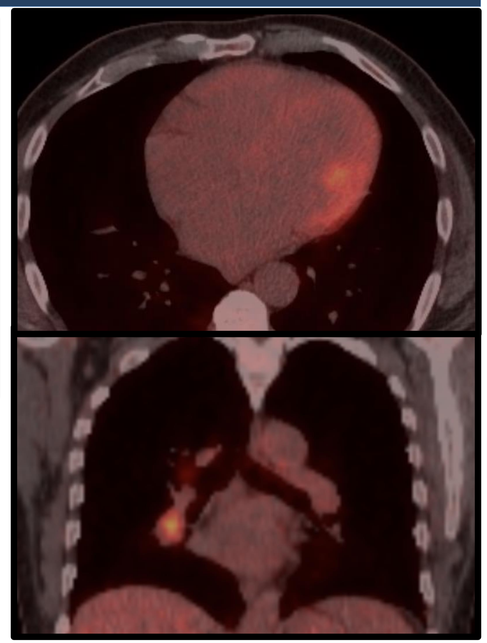
His ECG findings, in combination with hilar lymphadenopathy and a family history of sarcoidosis, raised suspicion for CS.

Further diagnostic testing for other etiologies of cardiomyopathy (thyroid function tests, ferritin, coronary CTA, serum and urine protein electrophoresis, and technetium-99 m pyrophosphate scan) were all unremarkable.

Cardiac MRI and CS protocol ^{18}F-fluorodeoxyglucose (FDG)-PET scan were then pursued to investigate for CS.

Cardiac MRI and FDG-PET





Cardiac MRI (above): Midcavity short axis with subepicardial and mid myocardial late gadolinium enhancement in inferolateral wall.

FDG-PET scan (right, upper): Four chamber view of inferolateral FDG activity.

FDG-PET scan (right, lower): Representative images of FDG-avid mediastinal lymphadenopathy.

Hospital Course

- The patient underwent endobronchial ultrasound-guided mediastinal lymph node biopsy, which showed non-caseating granulomas. Given his imaging findings, this was consistent with a clinical diagnosis of CS.
- He was diuresed with symptom improvement and initiated on heart failure (HF) guideline directed medical therapies.
- For sarcoidosis, he was started on oral prednisone with planned taper and mycophenolate mofetil.
- A 14 day event monitor was placed on discharge. This showed sinus rhythm with first degree AV block, Mobitz 1 block, RBBB, 7.7% PVC burden, 11 runs of non-sustained ventricular tachycardia (longest 11 beats) and 1 run of supraventricular tachycardia.
- Follow up PET scan demonstrated resolution of cardiac FDG uptake. Echocardiogram showed improvement in LVEF to 40-45%.

Teaching Points

- Although alternative HF etiologies were evaluated, the initial ECG showing conduction abnormalities (RBBB and LAFB) in addition to PVCs led to immediate clinical suspicion of CS. Unexplained conduction disease and HF, particularly with extracardiac findings such as mediastinal lymphadenopathy, should prompt further investigation for CS.
  - The following ECG findings are diagnostic criteria per the Japanese Circulation Society 2016 Guideline on Diagnosis and Treatment of Cardiac Sarcoidosis: high grade AV block, ventricular tachycardia, frequent PVCs, complete RBBB, axis deviation, or abnormal Q waves
- Diagnosis is imperative given the increased risk of sudden cardiac death and need for tailored therapies in CS.
  - Treatment includes corticosteroids, steroid-sparing immunomodulating agents, HF medications and antiarrhythmics.
  - Appropriately selected patients may undergo implanted cardiac devices such as pacemaker or defibrillator.

References

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