CASE REPORTS

Recurrent hemopericardium with cardiac tamponade as an initial presentation of cardiac sarcoidosis

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ABSTRACT

This case report illustrates an unusual presentation of recurrent hemopericardium with cardiac tamponade secondary to pulmonary sarcoidosis with extra-pulmonary cardiac involvement. It also demonstrates the usefulness of cardiac magnetic resonance (CMR) imaging as a non-invasive modality for the diagnosis of cardiac sarcoidosis.

Key Words: Sarcoidosis, Cardiac sarcoidosis, Hemorrhagic pericardial effusion, Cardiac magnetic resonance imaging

1. INTRODUCTION

Sarcoidosis is a multi-systemic granulomatous disease of unknown etiology which typically affects young adults. It can involve almost all organ systems to varying degrees. The primary pathology is formation of non-caseating granulomas in the involved organ systems. Cardiac involvement in sarcoidosis can be asymptomatic, or can manifest as arrhythmias, heart block, pericardial involvement, heart failure or valvular dysfunction. It can also lead to sudden cardiac death.

2. CASE REPORT

2.1 Case history

A 48 year old African American man was referred to the emergency department for sudden onset left-sided chest pain, acutely worsening shortness of breath and easy fatigability. His past medical history was remarkable for hypertension and pulmonary sarcoidosis diagnosed 5 years ago. On initial presentation, the heart rate was 120 beats per minute, blood pressure 101/65 and pulse oximetry was 87% on room air. He had coarse breath sounds bilaterally on lung auscultation and an erythema nodosum rash on bilateral lower extremities.

2.2 Methods

Electrocardiogram (ECG) showed sinus tachycardia with ST segment elevations and electrical alternans (see Figure 1). Initial cardiac markers were within normal range. Initial presentation was concerning for a pulmonary embolism given the sudden chest pain, shortness of breath, tachycardia and hypoxia, so he underwent a computed tomography (CT) angiogram of thorax which revealed no pulmonary embolism, but hilar lymphadenopathy and end-stage bullous sarcoid with fibrosis and peri-lymphatic nodules consistent with stage IV pulmonary sarcoidosis (see Figure 2). Additionally, it revealed a complex moderate sized pericardial effusion with reflux of contrast into the inferior vena cava and azygos vein, suspicious for a cardiac tamponade. He underwent an emergent bedside transthoracic echocardiography (TTE), which confirmed a moderate pericardial effusion with right sided collapse of heart chambers consistent with tamponade physiology (see Figure 3). He underwent emergent

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pericardiocentesis, with drainage of 400 cc of bloody fluid. A repeat TTE 3 hours later showed no new pericardial effusion and he remained stable. Four hours later, he had clinical deterioration with hypotension and tachycardia. A bedside TTE again showed recurrent pericardial effusion with tamponade. He emergently had another pericardiocentesis of 400 cc bloody fluid and a pericardial drain was left in place. Pericardial fluid was exudative with increased red blood cells. He continued to have bloody drainage from the pericardial drain. Cardiac magnetic resonance imaging (CMR) showed concentric left ventricular hypertrophy with preserved left and right ventricular systolic function and post-gadolinium contrast images showed an area of mid-wall enhancement in the mid inferior wall consistent with cardiac sarcoid (see Figure 4). Respiratory viral polymerase chain reaction was negative. Anti-neutrophil antibody, rheumatoid factor, double stranded DNA antibody, C and P anti-neutrophil cytoplasmic antibody and anti-myeloperoxidase/Proteinase 3 antibody were all negative. Pleural fluid adenosine deaminase was within normal limits. Pericardial fluid cytology was negative for malignancy and acid fast bacilli.

3. Results

Given his history of pulmonary sarcoidosis, the presentation was likely consistent with extra-pulmonary involvement of cardiac sarcoidosis. Medical therapy was initiated with oral prednisone. Patient had symptomatic improvement after steroid therapy and output from pericardial drain subsequently tapered off and the drain was removed. Pericardial cultures grew propionibacterium acnes, a common skin contaminant, which was treated with doxycycline on discharge. A repeat TTE 5 months later showed complete resolution of the pericardial effusion after prolonged treatment with prednisone 20 mg daily.

4. Discussion

Sarcoidosis can affect multiple organ systems, including the cardiovascular system, and involvement can range from asymptomatic cardiac involvement to sudden cardiac death. Cardiac sarcoidosis affects 2%-7% of patients with sarcoidosis,[1] but multiple studies have shown much higher prevalence of asymptomatic cardiac involvement.[2,3] The clinical manifestations of cardiac sarcoid include cardiac arrhythmias, heart block, heart failure, valvular dysfunction, and rarely, pericardial disease, as seen in this case. Usually, pericardial involvement presents as small, clinically asymptomatic pericardial effusions.[4] Significant pericardial effusions with hemorrhage and cardiac tamponade with hemodynamic instability is unusual.[5]

Cardiac sarcoid should be suspected in patients less than 55 years old with new unexplained cardiac symptoms, ECG changes or unexplained heart block, and with sustained monomorphic ventricular tachycardia (VT) or cardiomyopathy.[6] Patients with extra-cardiac sarcoidosis should also always be worked up for cardiac involvement.

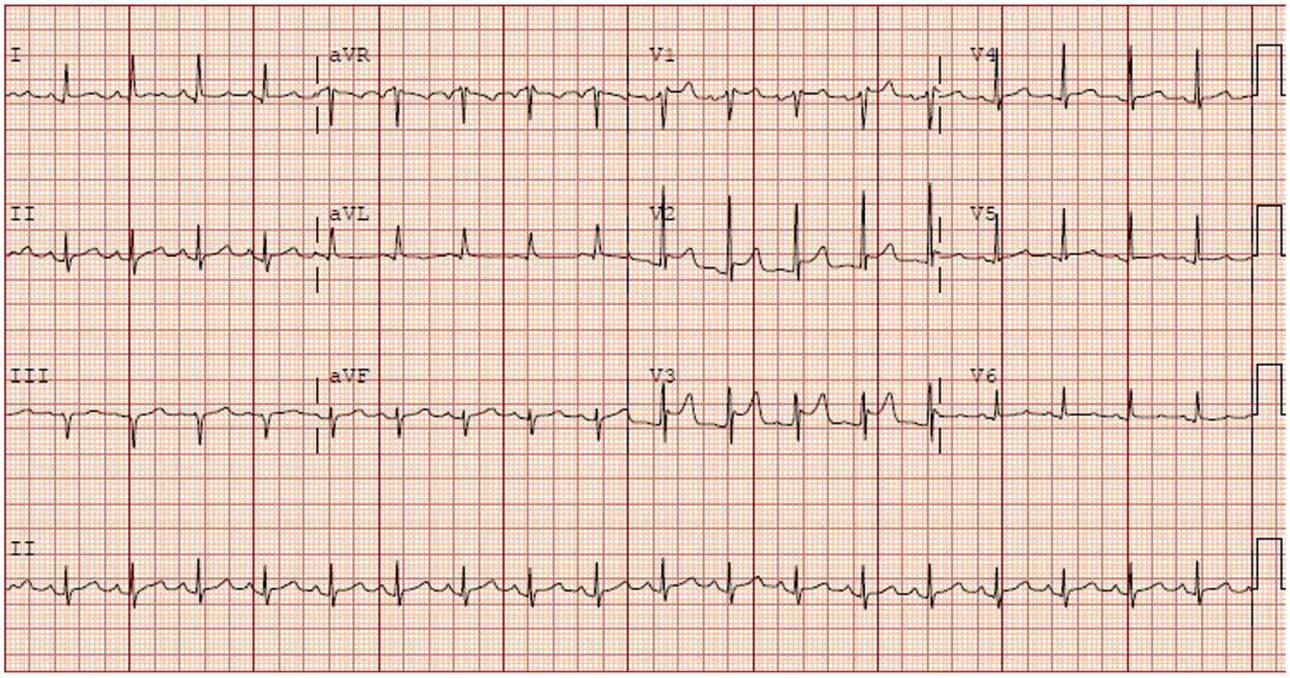

Figure 1. ECG with sinus tachycardia, ST segment elevations and electrical alternans suggestive of pericarditis

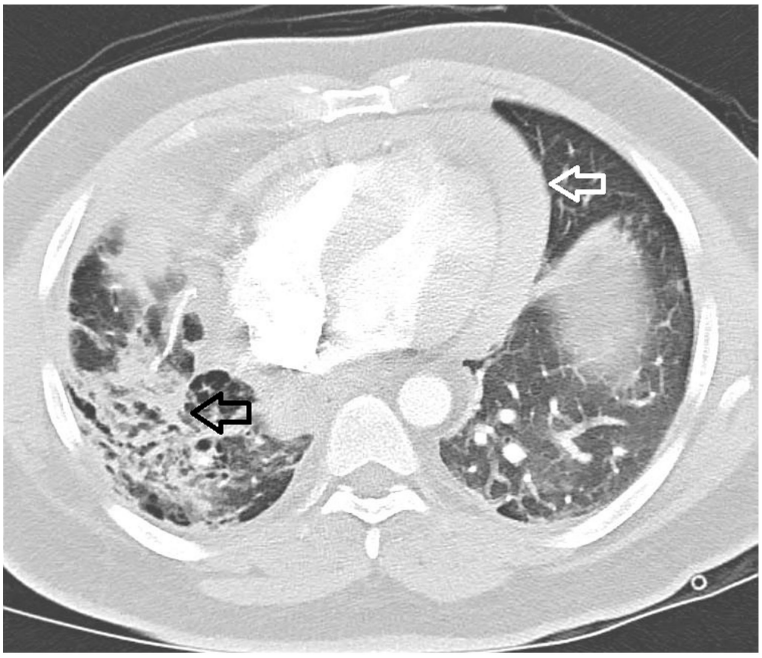

Figure 2. CT Thorax with moderate pericardial effusion (white arrow) and pulmonary sarcoid with fibrosis (black arrow)

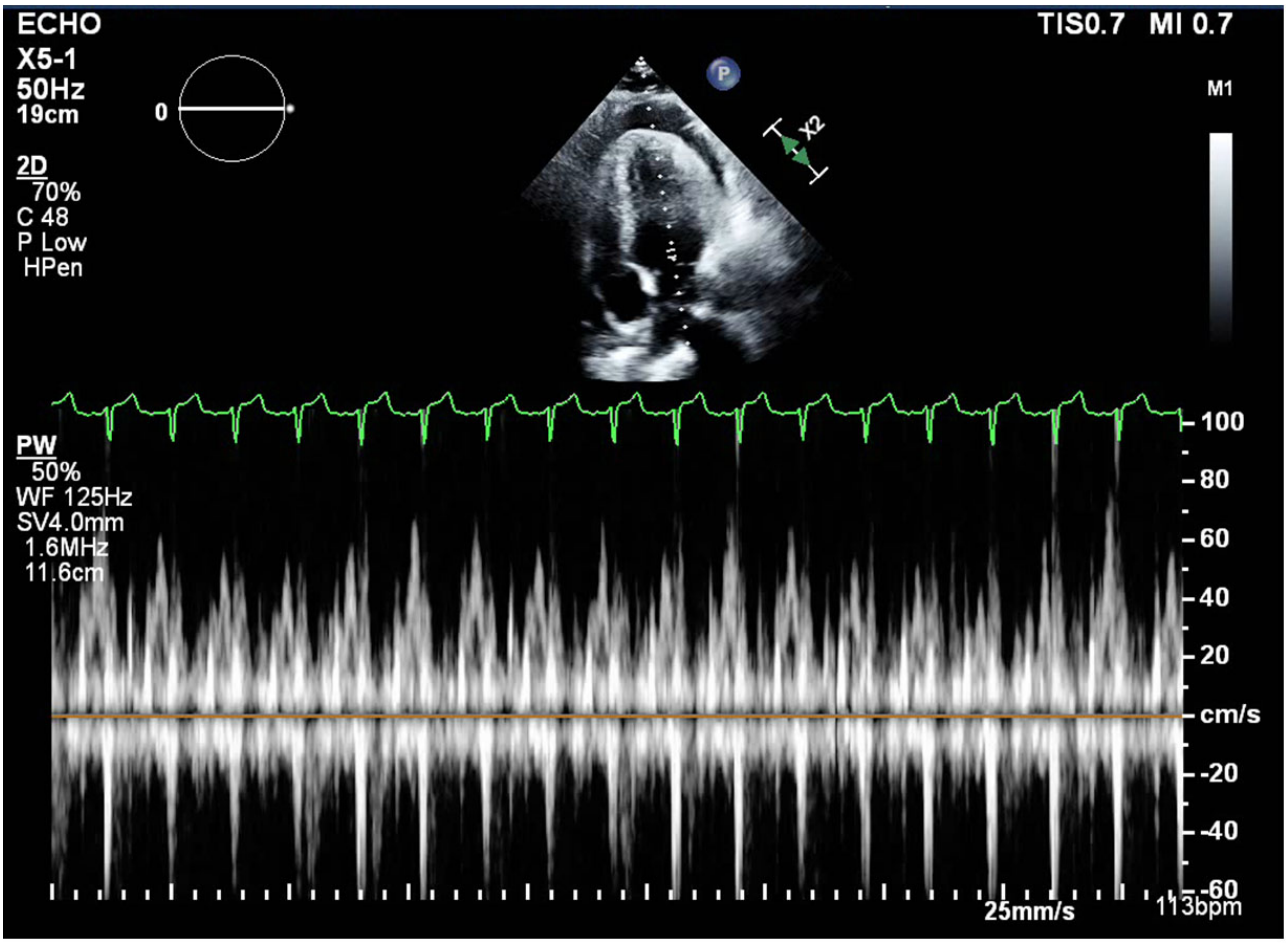

Figure 3. TTE with moderate pericardial effusion; and respiratory variation of mitral valve inflow velocities, consistent with tamponade

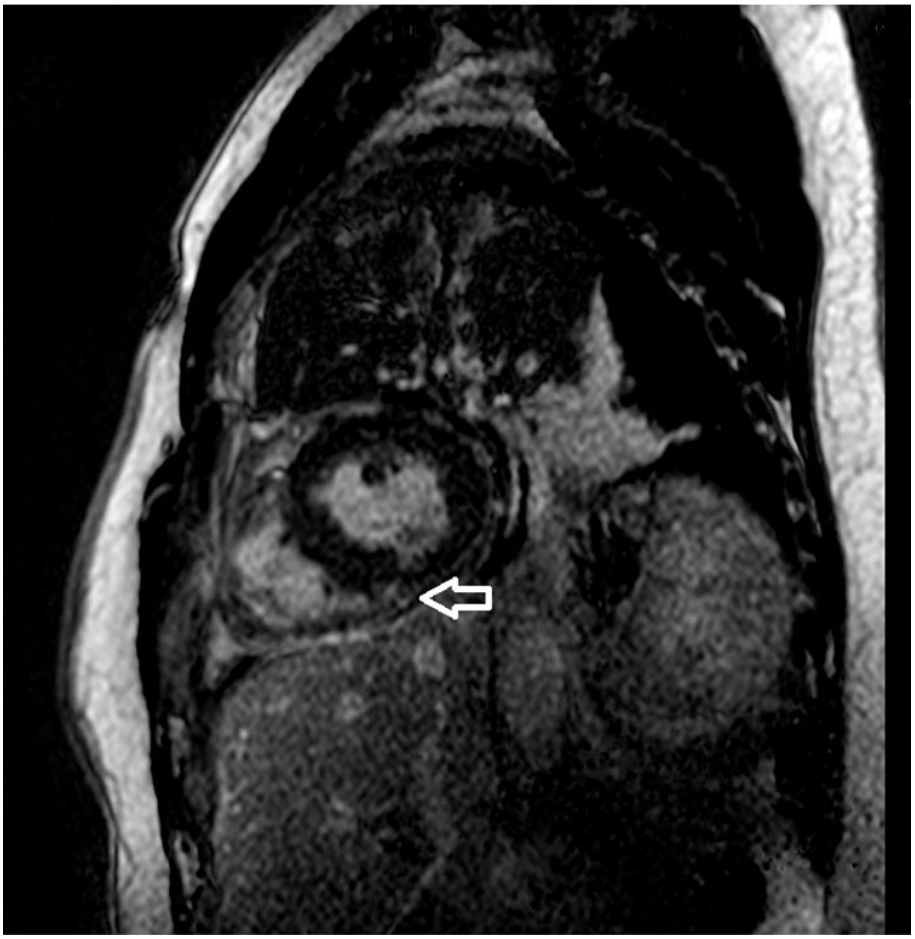

Figure 4. Cardiac MRI with LGE in inferior wall consistent with cardiac sarcoid

Initial evaluation involves an ECG to assess for conduction defects, echocardiogram to assess for structural defects and a 24-hour Holter monitoring to detect arrhythmias.

Subsequent imaging usually involves CMR. In patients with sarcoidosis, delayed enhancement CMR imaging is extremely sensitive and accurate for detecting cardiac involvement.[7] CMR involves using T2 weighted images and early gadolinium images to detect early inflammation, while late gadolinium enhancement (LGE) detects chronic disease. The presence of myocardial scar by LGE is the best independent predictor of potential fatal events.[8] An alternative diagnostic test is fluorodeoxyglucose-positron emission tomography (FDG-PET), which is more sensitive than CMR, but less specific, as it can be positive in other inflammatory diseases of myocardium or ischemia. The gold standard is for detection on cardiac sarcoid is detection of non-caseating granulomas by endomyocardial biopsy, however sensitivity is as low as 20%.

5. Learning points

Establishing the correct diagnosis of pericardial involvement of sarcoid is crucial, as appropriate treatment with anti-inflammatory medications led to the resolution of this patient's pericardial effusion and nearly fatal cardiac tamponade. Delayed enhancement CMR imaging is extremely sensitive and accurate for detecting cardiac involvement in sarcoidosis.

Conflicts of Interest Disclosure

The authors have no conflict of interest related to this publication.

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