Recurrent Hemopericardium With Cardiac Tamponade as an Initial Presentation of Cardiac Sarcoidosis

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Cardiac sarcoidosis can be asymptomatic or can manifest as arrhythmias, heart block, pericardial involvement, heart failure, valvular dysfunction or sudden cardiac death. Hemopericardium with cardiac tamponade is extremely rare, especially as an initial presentation.

A 48 year old African American male presented with acutely worsening shortness of breath, chest pain and fatigue. His history was remarkable for pulmonary sarcoidosis diagnosed 5 years ago but he was not on any systemic steroids or immunosuppressants.

Physical exam: He was tachycardic, hypoxic, had coarse breath sounds and an erythema nodosum rash on lower extremities.

Initial workup: Electrocardiogram showed sinus tachycardia. Cardiac markers were normal.

Imaging: Initial presentation was concerning for pulmonary embolism (PE); a CT-angiogram thorax revealed no PE, but showed hilar lymphadenopathy and bullous sarcoid with fibrosis and peri-lymphatic nodules consistent with sarcoidosis. Additionally, it revealed a moderate-sized pericardial effusion with contrast reflux into the IVC and axygos vein suspicious for tamponade.

Emergent transthoracic-echocardiography (TTE) confirmed a pericardial effusion with right-sided heart chamber collapse consistent with tamponade physiology. Emergent pericardiocentesis was done with drainage of 400cc bloody fluid.

Repeat TTE 3 hours later showed no effusion and he remained stable. However, four hours later he clinically deteriorated with hypotension and tachycardia. TTE showed recurrent effusion with tamponade, requiring repeat pericardiocentesis with pericardial drain placement. Pericardial fluid was exudative with increased RBCs; cultures were negative. He continued to have bloody drainage from the drain.

Other workup: Respiratory viral PCR panel was negative. ANA, RF, dsDNA, c and p-ANCA, anti-MPO/PR3 antibody were all negative. Pleural fluid ADA was normal. Pericardial fluid cytology was negative for malignancy and AFB.

Cardiac magnetic resonance (CMR) imaging showed concentric LVM and post-gadolinium contrast images showed inferior wall enhancement consistent with cardiac sarcoidosis.

Presentation was consistent with extra-pulmonary involvement of cardiac sarcoidosis. Medical therapy was initiated with oral prednisone. He had symptomatic improvement after steroid therapy and pericardial drain output subsequently tapered off and it was removed. He was stable and discharged to home.

On out-patient follow up, he reported significant symptomatic improvement with no recurrence of symptoms and was started on mycophenolate mofetil as a steroid sparing agent.

Pericardial involvement in sarcoidosis is rare and usually seen as small asymptomatic effusions. Significant hemorrhagic pericardial effusion and tamponade with hemodynamic instability is extremely rare.

This case illustrates an unusual initial presentation of extra-pulmonary cardiac sarcoidosis with recurrent hemopericardium and tamponade. It demonstrates use of CMR imaging as a sensitive, accurate and non-invasive modality for diagnosis of cardiac sarcoidosis.

Establishing the correct diagnosis of pericardial involvement is crucial, as appropriate treatment with anti-inflammatory medications led to resolution of this patient’s pericardial effusion and nearly fatal cardiac tamponade.
