CLINICAL VIGNETTE

Hearts Aflame: Myopericarditis with Cardiac Tamponade, a Rare Initial Presentation of Systemic Lupus Erythematosus

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Case Presentation

A 54-year-old African-American female with a history of hypertension, hypertensive retinopathy, and tobacco dependence presented to the emergency department (ED) with progressive shortness of breath. She noted two weeks of worsening dyspnea, initially on exertion, which rapidly progressed to dyspnea at rest. She denied any orthopnea, paroxysmal nocturnal dyspnea, dizziness, or lower extremity edema. She did report mild pleuritic chest pain as well as mild patchy hair loss which she attributed to age. She had no family history of cardiac or pulmonary disease. Her mother died in her 70s from an unknown autoimmune condition.

Upon presentation to the ED, she was afebrile with normal oxygen saturation on room air. Her blood pressure was elevated to 210/110 mm Hg. Physical exam revealed muffled heart sounds, JVP elevation of approximately 10cm, bilateral crackles in the lower lung fields and a hypopigmented patch of alopecia above the left ear. She did not have any significant lower extremity edema.

Laboratory testing revealed low hemoglobin of 7.6 g/dL, creatinine 1.3 mg/dL, mildly elevated BNP 140 pg/mL (reference range (RR): <100pg/mL), elevated ESR 110 mm/hr (RR: <30mm/hr) and CRP 11.0 mg/L (RR: <5mg/L). Serial troponins were negative and electrocardiogram was without acute ischemic changes. Thyroid stimulating hormone (TSH) was within normal limits, and infectious evaluation was negative including Covid-19 / Influenza PCR, blood cultures, HIV, and tuberculosis testing. Initial chest x-ray revealed a significantly enlarged cardiac silhouette without pulmonary infiltrates or significant effusions (Figure 1). She then underwent further imaging with transthoracic echocardiogram (TTE) that demonstrated a large pericardial effusion with evidence of cardiac tamponade (Figure 2).

The patient was admitted to the cardiac intensive care unit, where she underwent emergent pericardiocentesis, draining 1.4 liters of fluid. Repeat TTE showed resolution of tamponade. Pericardial fluid cultures returned negative for infection, and fluid cytology was negative for malignancy. Cardiac MRI revealed a diffuse inflammatory process suggestive of myopericarditis (Figures 3 and 4). Additional laboratory tests resulted with an ANA titer 1:640 (RR: <40), C3: 54 mg/dL (RR: 83-193mg/dL), C4: 9 mg/dL (RR: 15-57mg/dL), anti-Smith Ab 165 units (RR: <20 units). Clinical presentation, laboratory testing, and imaging met criteria for a new diagnosis of myopericarditis with cardiac tamponade due to Systemic Lupus Erythematosus (SLE).

The patient was initiated on pulse dose steroids with methylprednisolone, resulting in substantial improvement in her symptoms and without further increase in pericardial effusion. She was transitioned to prolonged oral prednisone taper as well as hydroxychloroquine and colchicine on discharge, with rheumatology and cardiology follow up.

Discussion

SLE is an autoimmune disorder whose pathophysiology is mediated by antibody formation and immune-complex deposition resulting in an inflammatory cascade. It may affect almost every organ system, resulting in a heterogeneous disease presentation.[1] Myopericarditis with cardiac tamponade as a first-time presentation of SLE is quite rare.

SLE can be diagnosed using the Systemic Lupus International Collaborating Clinics (SLICC) criteria. Patients need to have at least 4 out of 17 total points, including at least one clinical criterion and one immunologic criterion. Our patient had five points: nonscarring alopecia, serositis (pericarditis), low C3/C4, high titer ANA, and elevated anti-Smith Ab. Importantly, there was also no evidence for infection for malignancy.

The development of SLE in patients is not clearly understood, but is believed to be multifunctional influenced by genetic, environmental, and hormonal factors.[1] Given the increased female incidence of SLE, a potential link to an estrogen hormonal effect has been postulated, with increased rates of SLE in populations with increased estrogen exposure.[2] In addition, current cigarette smoking (as in our patient) has been associated as a risk factor for developing SLE, potentially via oxidative stress, epigenetic modification, cytokine-driven inflammation, and/or impaired T- and B-cell function.[3] African American populations also have greater disease prevalence and severity in SLE.

Cardiac involvement in SLE is a significant complication that may be associated with high morbidity and mortality. SLE can affect the cardiovascular system in a multitude of ways including arrhythmias, valvular disease, accelerated coronary

artery disease, pericardial disease, myocardial involvement, and ultimately cardiomyopathy with heart failure. Pericarditis is the most common cardiac manifestation, with up to 40% of patients having concurrent pericardial effusion.[4] Myocardial involvement and cardiac tamponade in SLE are rare, especially as the initial presentation.

It is postulated that myocarditis in SLE is an immune complex mediated disease. Immunofluorescence studies demonstrate fine granular immune complexes and complement deposition in the walls and perivascular tissues of myocardial blood vessels.[4] Clinically, the signs and symptoms are similar to myocarditis from other causes and may include dyspnea, chest pain, arrhythmias and may present subacutely. Troponin levels are often elevated. Left untreated, myocarditis may progress to cardiomyopathy and heart failure.

Diagnosis of lupus myocarditis is challenging. Cardiac MRI is an effective, accurate, and increasingly used modality for diagnosis. MRI may show late gadolinium enhancement in regions where cardiac inflammation is present.[5] Echocardiography has lower diagnostic utility, and may demonstrate nonspecific findings in patients with myocarditis.[6] While endomyocardial biopsy remains the gold standard for diagnosis,[7] our patient opted not to undergo biopsy following a risk/benefit discussion.

After confirming the diagnosis of SLE-induced myopericarditis, prompt medical treatment is indicated, as lupus myocarditis may result in significant morbidity and mortality. A study of 29 patients with lupus myocarditis reported 3 deaths after 37-month follow-up.[8] Cardiac tamponade causing hemodynamic instability warrants emergent pericardiocentesis, as was performed in our patient. First-line medical treatment for myopericarditis due to SLE includes immunosuppression with systemic steroids. In conjunction with steroids, other immunosuppressants including cyclophosphamide, azathioprine, and mycophenolate mofetil have been administered with success. IVIG has also been used.[8] Treatment strategies are currently based on expert opinion as opposed to randomized trials. Our patient had substantial improvement in her symptoms after pulse dose methylprednisolone, followed by a prolonged oral prednisone taper and hydroxychloroquine.

Conclusion

Clinical presentation, laboratory workup and imaging conferred a rare diagnosis of myopericarditis with cardiac tamponade secondary to newly diagnosed SLE. Prompt recognition is essential given high complication rates if left untreated. Pericardiocentesis followed by steroids and immunomodulators are the treatment of choice.

Figures

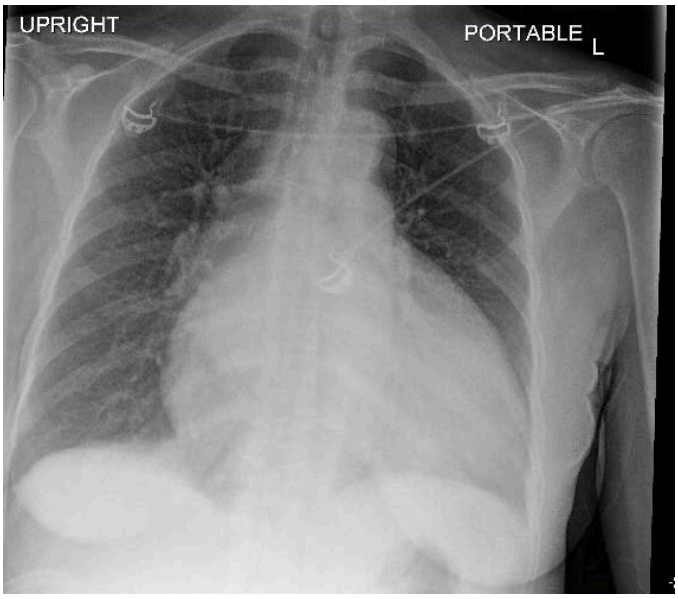

Figure 1: Chest x-ray on presentation with marked cardiomegaly

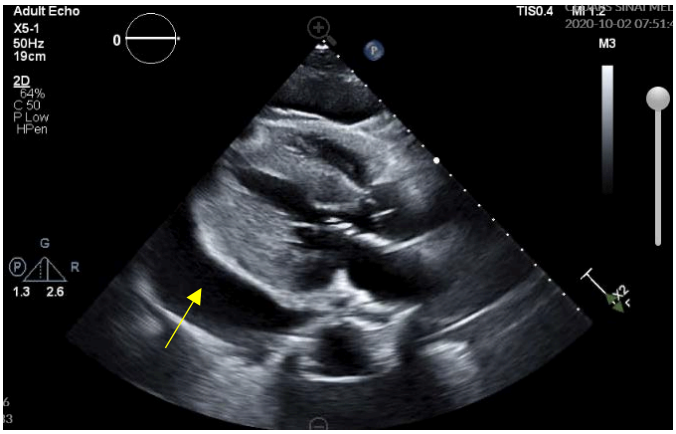

Figure 2: Echocardiogram parasternal long axis view demonstrating presence of large pericardial effusion (yellow arrow)

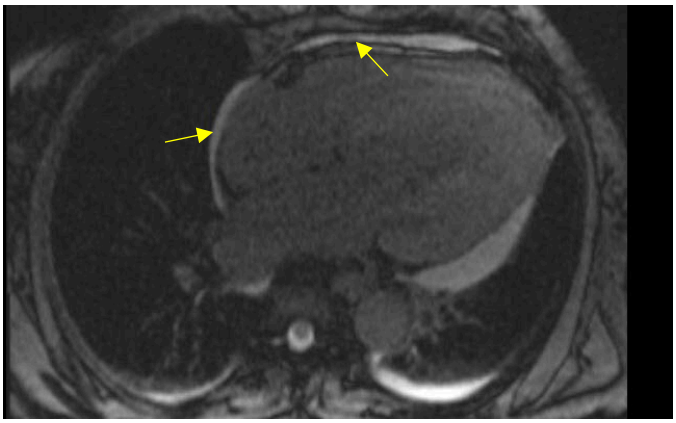
Figure 3: Cardiac MRI demonstrating enhancement of the pericardium (yellow arrows), indicating evidence of pericarditis

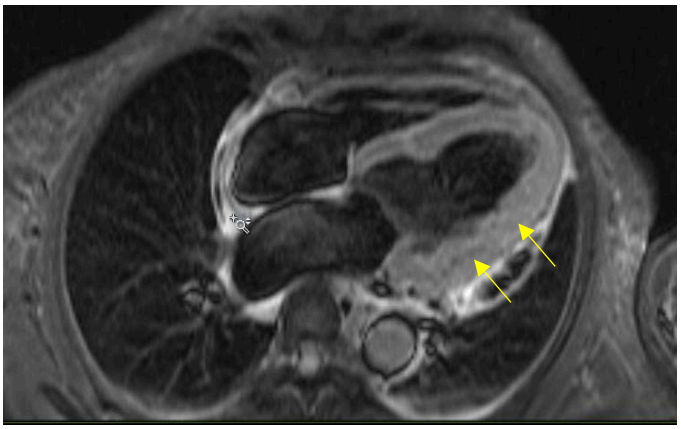

Figure 4: Cardiac MRI demonstrating myocardial enhancement and edema (yellow arrows), indicating evidence of myocarditis

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