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CLINICAL VIGNETTE

Echocardiographic Evidence of Cardiac Tamponade as an Initial Manifestation of Systemic Lupus Erythematosus

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Introduction

Systemic lupus erythematosus (SLE) is a systemic chronic autoimmune disease process that manifests in a variety of organ systems. Although pleurisy and pericardial effusions present commonly in patients with SLE, cardiac tamponade is a rare occurrence in less than 1% of patients¹. It is especially uncommon as an initial manifestation of the disease. We present the rare case of a patient who presented initially with echocardiographic evidence of cardiac tamponade that later was determined to be an element of previously undiagnosed SLE.

Case Presentation

A 27-year-old African American female presented to our Emergency Room with 2 to 3 weeks of progressive dyspnea on exertion, with decreased exercise tolerance, paroxysmal nocturnal dyspnea as well as sharp chest pain for 2 to 3 days. She also reported generalized swelling in her face, arms and legs, and a bilateral lower extremity rash for the past month.

The patient denied any flu-like illnesses, hemoptysis, dysuria, hematuria, fevers, nausea, vomiting or diarrhea. She also denied a history of malar rash, discoid lesions, aphthous oral ulcers, headaches or photosensitivity, but did report pain in her fingertips whenever she smoked or when there was cold weather. She complained of nonspecific knee pain at times, but no deformities, effusions, or swelling in her joints.

There was no significant past medical history. Her only medication was over-the-counter 8-bromotheophylline, given for swelling for 2 weeks prior to admission. She reported an allergy to penicillin. There was no family history of cardiac disease, or lupus, or any other autoimmune diseases. She smoked tobacco, drank socially, and denied any history of intravenous drug use. She worked as a teller at a bank, had no travel history, no history of tuberculosis contacts, no history of incarceration, homelessness, or any other risk factors. She had no pregnancies.

On physical examination, the patient was afebrile, pulse 105 beats/min, blood pressure 120/65 mmHg, respiration rate 20 breaths/min, and oxygen saturation at 97% on room air. She was resting comfortably, in no respiratory distress, and was alert and oriented to name, place and time. Pupils were equal and reactive bilaterally, with no periorbital edema present. No oral ulcers were noted. Her jugular venous pressure was elevated to approximately 15 cm H₂O, no carotid bruits, cervical or supraclavicular lymphadenopathy, parotid tenderness, or goiter was present. Her heart sounds were audible, tachycardic, and a pericardial knock was heard. No murmurs, rubs, or gallops were auscultated. A pulsus of 6 mm was present. Her lung examination was clear. Her abdominal examination was unremarkable, and negative for ascites. Examination of extremities revealed a non-tender, diffuse macular rash extending to below her knees bilaterally. No anasarca was noted. Her knee, elbow, wrist, and hand examinations were unremarkable for nodules, effusions, erythema, or deformities. Her neurologic examination revealed no cranial nerve deficits, and she had +5/5 upper and lower extremity strength.

The patient's complete blood count showed a white blood cell count (WBC) of 8100/cu mm, a hemoglobin of 8.1 gm/dL with an MCV of 59.4 fL, and an RDW of 19.1%, hematocrit of 19.1%, platelet count of 407,000/cu mm. A chemistry panel showed a sodium of 134 mmol/L, potassium of 4.3 mmol/L, chloride of 111 mmol/L, bicarbonate of 19 mmol/L, BUN of 16 mg/dL, creatinine of 0.9 mg/dL, and a glucose of 89 mg/dL. Her serum total

protein was 7.7 gm/dL and a serum albumin was low at 2.3 gm/dL. Complement levels C3 and C4 were decreased at 30 and 2 mg/dL, respectively. TSH was normal. An electrocardiogram (ECG) showed low voltage in the frontal and precordial leads, with right axis deviation present with sinus rhythm (**Image 1**). A chest radiograph revealed

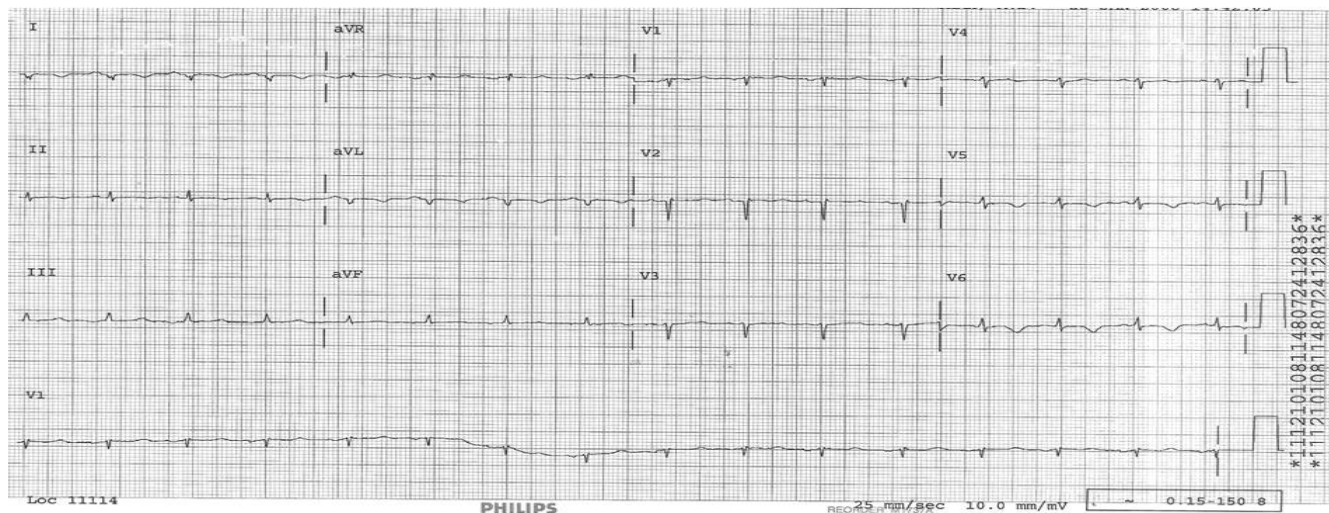


Image 1. Electrocardiogram done on admission showing low voltages in the frontal and precordial leads, sinus tachycardia, right axis deviation, and T-wave inversions in the lateral leads.

significant cardiomegaly suspicious for pericardial effusion (**Image 2**). A urine pregnancy test was negative. Urinalysis showed +2 protein, trace leukocytes, +3 blood, specific gravity of 1.011, trace ketones, pH of 5.5, with 8 white blood cells, and 28 red blood cells. Spot urine protein to creatinine ratio yielded approximately 600 mg/day of protein.

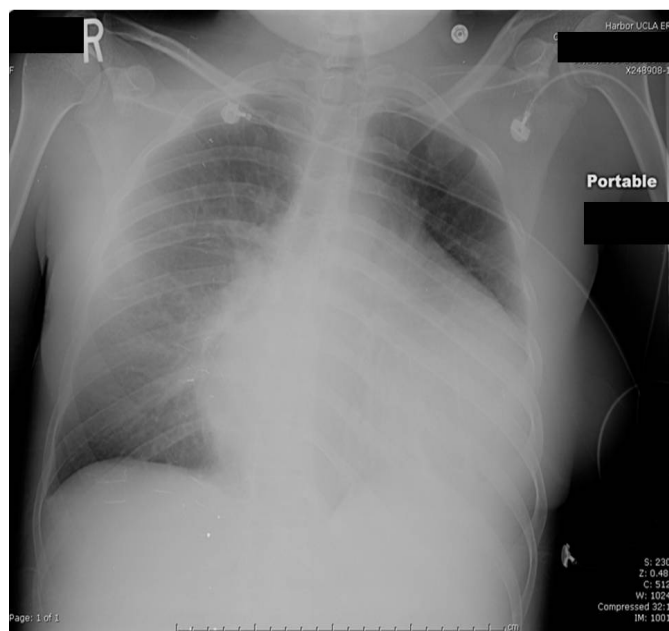


Image 2. Portable X-ray of patient on admission, showing cardiomegaly and possible left sided pleural effusion.

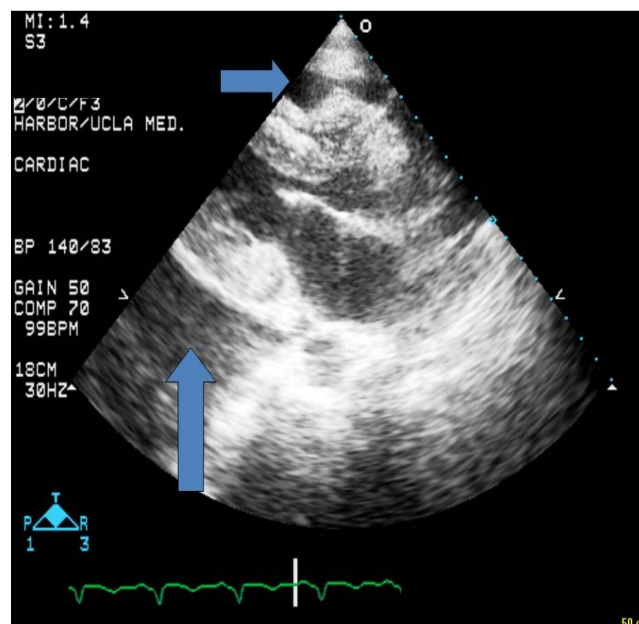


Image 3. 2-Dimensional Transthoracic Echocardiography, long-axis parasternal view demonstrating a large pericardial effusion (up arrow) and evidence of right ventricular diastolic collapse (right arrow).

The patient was admitted to the Cardiology Service and a transthoracic echocardiogram was performed which revealed mild concentric left ventricular hypertrophy, a normal left ventricular ejection, and grossly normal left and right ventricle. A large pericardial effusion was seen with diastolic right ventricular collapse, meeting echocardiographic evidence of tamponade (**Image 3**). The patient was taken to the catheterization laboratory and a peri-

cardial drain was inserted, initially withdrawing 950 milliliters of yellow, straw colored fluid. The cell count of the pericardial fluid revealed 2290 red blood cells/cu mm, 650 white blood cells/cu mm, with a differential of 45% neutrophils, 14% lymphocytes, 6% monocytes, 2% eosinophils, 21% histiocytes, and 11% mesothelial cells. A total protein count was 4.6 g/dl and glucose was 92 mg/dl. Bacterial and acid-fast bacillus cultures were negative, and cytology was negative for malignancy. A right atrial pressure was measured at 7, right ventricular pressure of 31/5, pulmonary artery pressure at 43/14, and pulmonary capillary wedge pressure of 17. No end diastolic equalization of pressures nor was there a prominent X-descent seen. After drainage of the pericardial fluid, pericardial pressure decreased from 17 to 12. A repeat CXR showed resolution of the large pericardial effusion and repeat radiographs showed improvement in the cardiac silhouette (**Image 4**).

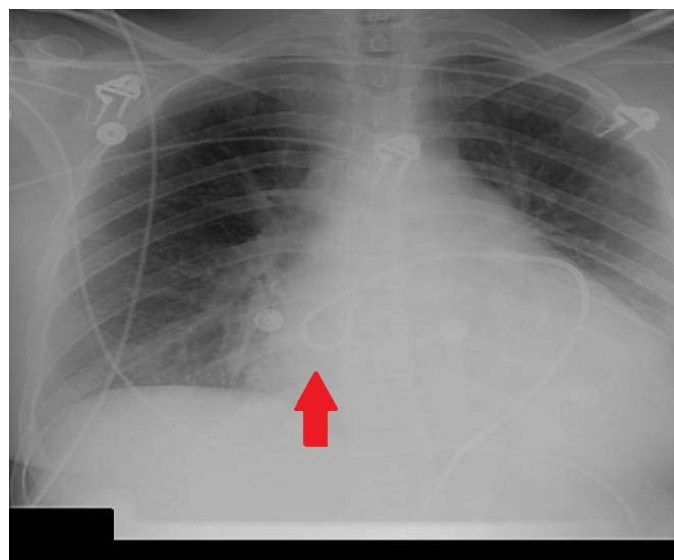


Image 4. Portable X-ray of patient after placement of pericardial drain (arrow), with reduction in size of cardiac silhouette.

The patient continued to have persistent drainage from the pericardial effusion. Later in the hospital course, the patient's antinuclear antibody (ANA) titer returned positive at 1:640 with a speckled pattern, with an anti-dsDNA antibody at 1:80. IgG Anti-Smith antibodies were positive at 1:25600. Based on the positive ANA titer, presence of anti-Smith/dsDNA antibodies, serositis, and proteinuria greater than 500 mg/day, the patient fulfilled American College of Rheumatology classification criteria for the diagnosis of SLE².

The Nephrology Service was consulted and a renal biopsy was performed on the fourth hospital day which revealed the presence of diffuse proliferative lupus nephritis, with 5% crescents, World Health Organization (WHO) Class IV-G(A), indicating active disease. The patient was started on steroids and plaquenil for treatment of lupus nephritis. After steroids were started, the pericardial drainage decreased and other symptoms improved. The tachycardia resolved and serial pulsuses measured throughout the patient's hospital course did not increase. On the seventh hospital day, the pericardial drain was removed. The patient was discharged on the tenth hospital day, with Rheumatology follow-up and monthly infusions of cyclophosphamide for lupus nephritis.

Discussion

Systemic lupus erythematosus (SLE) is a connective tissue disease that can affect multiple organ systems due to its formation of autoantibodies and immune complexes. Cardiac manifestations vary widely, involving the pericardium, myocardium, valves, coronary arteries, and the conduction system. Pericarditis in SLE patients has been reported in 12% to 48%, with echocardiographic studies demonstrating effusion in 21% to 46% of nonselected SLE patients³. Clinically evident pericarditis has been shown to be present in 25% of patients with SLE, and in an autopsy series pericardial involvement was found to be in 62% of patients with SLE. A controlled, prospective study looking at echocardiographic findings in 75 patients with SLE in an outpatient setting were found to have pericardial effusion and/or thickening in up to 37% of patients. There was also a significant association with pericardial pain ($p < 0.05$), and active disease ($p < 0.001$), as well as left ventricular hypertrophy with systemic hypertension⁴ ($p < 0.05$). Despite its relatively common occurrence in patients with SLE, cardiac tamponade is rare. In a combined series of more than 1,332 patients with SLE related pericarditis, tamponade was found¹ in 0.8%. Symptoms of pericarditis with SLE patients can include substernal or precordial chest discomfort, which also can be positional in nature. Fever, tachycardia, and decreased heart sounds are also present. When cardiac tamponade or constrictive pericarditis is present, the jugular pulse is prominent, and the jugular venous pressure is elevated. The X-descent, which is due to atrial relaxation, the downward displacement of the tricuspid valve during right

ventricular systole, and the ejection of blood from both the ventricles, is prominent, and the Y-descent, which is due to the tricuspid valve opening with the rapid flow of blood into the right ventricle, is absent. Patients may demonstrate tachycardia and hypotension, muffled heart sounds, pulsus paradoxus, and progressive dyspnea.

Echocardiographic evidence of tamponade involves visualization of a large pericardial effusion, decreased total transverse dimensions, decreased right ventricular diameter, early collapse of the right ventricular outflow tract, and indentation of the left atrial free wall³. In order to validate the collapse of the right atrium and ventricle as useful diagnostic signs of cardiac tamponade, Singh et al. correlated hemodynamics with right ventricular and atrial collapse in 16 patients with pericardial effusion. Most had elevated jugular venous pressures (88%) and 75% had a pulsus paradoxus greater than 10 mm Hg. Right heart catheterization was performed at the same time echocardiography was performed before and during pericardiocentesis. The echocardiographic readings were compared to catheterization hemodynamic measurements, where cardiac tamponade was noted to be present if there was equalization of the right atrial, pulmonary capillary wedge, and intrapericardial pressures and elevation of these pressures of greater than 100 mmHg. The sensitivity and specificity of right ventricular collapse as a marker of cardiac tamponade was 92% and 100%, respectively, with a predictive value of 100%. The sensitivity and specificity of right atrial collapse was 64% and 100%, respectively, with a 100% predictive value⁵.

A later study looked prospectively at the correlation between clinical and Doppler echocardiographic signs in the diagnosis of cardiac tamponade in a larger series of patients (n=110) with a newly diagnosed moderate or large pericardial effusion from various causes. They compared cardiac chamber collapse and venous flow patterns with clinical criteria for tamponade (arterial hypotension, venous hypertension, pulsus paradoxus). Of interest, despite our echocardiography findings of diastolic collapse, thought to be highly likely of tamponade, our hemodynamic measurements did not show evidence of equalization of pressures. They found that by using clinical features of tamponade as the diagnostic standard, sensitivity and specificity for any cardiac chamber collapse was 90% and 65%, 68% and 66% for right atrial collapse, 60% and 90% for right ventricular collapse, and 45% and 92% for simultaneous collapse of both chambers, respectively. For venous flow, the sensitivity and specificity was 75% and 91%, respectively. Thus, there was a good correlation noted between absence of collapse and absence of tamponade, but correlation was poor between collapse and tamponade⁶. In another prospective study, 50 patients with echocardiographic evidence of pericardial effusion and right atrial or ventricular diastolic chamber collapse underwent right-sided cardiac catheterization and percutaneous pericardiocentesis. While all patients had elevated pericardial pressures, 94% had little to no hemodynamic instability, and while pericardiocentesis resulted in hemodynamic improvement, other factors such as dyspnea, or tachycardia did not improve. In addition, the presence of right ventricular diastolic and atrial collapse did not identify a more hemodynamically decompensated group as opposed to patients who did not display these findings on echocardiography⁷.

Our patient, although displaying elevated jugular venous pressure, tachycardia, dyspnea, did not have arterial hypotension or a pulsus paradoxus on examination but displayed right ventricular collapse and echocardiographic criteria for tamponade. In addition, on hemodynamic measurements with a pulmonary artery catheter, she did not meet criteria for tamponade, given there was no equalization of pressures. This illustrates the crucial role of clinical assessment in determining the urgency of the need for pericardiocentesis. This is a gray area of diagnosis of "impending tamponade" that needs continual assessment and prognostic studies. Characteristics of the pericardial fluid from SLE patients usually shows exudative fluid with an increased leukocyte count and predominantly polymorphonuclear cells. Although hemorrhagic fluid can occur, it is rare. It can be also markedly acidic (pH < 7) and pH may be used to differentiate from other types of effusions. Pericardial fluid protein concentration is increased, and the glucose can be normal to low³. In a review of 8 cases of patients with large pericardial effusions with SLE, common associated clinical findings included lupus nephritis, (n=5), arthralgia, (n=4) and Raynaud's phenomenon⁸ (n=3). Positive dsDNA and positive Smith antibodies were also present in most patients, and the nature of the fluid was exudative in all but one case. In another review of the pericardial fluid profile of 11 patients with SLE who underwent 12 pericardiocenteses, median leukocyte levels in the pericardial effusion was 6,785/mm³, with polymorphonuclear leukocytes (>70%) being present in 8 of 12 samples. Average pericardial glucose levels were 68

mg/dL, and serum anti-dsDNA antibodies were positive in 5 of 10 patients, and serum ANA titers positive in 8 out of 10 patients⁹.

Treatments usually consist of nonsteroidal anti-inflammatory drugs (NSAIDs), or oral steroids with good results. In a study conducted of 310 SLE patients with lupus-related serositis, 26% had pericardial effusions, NSAIDs were used in initially 35% of patients, and oral prednisolone was used in 76% for both serositis and multiorgan related complications from lupus. While pericardiocentesis was performed in selected patients, it is not mentioned the exact number of how many underwent this procedure. All episodes of serositis resolved completely within 2 months. Over a mean period of 46 months, 9 patients had 18 relapses of serositis, which were responsive to either NSAIDs or increasing prednisolone dosage¹⁰.

Conclusion

In conclusion, we present a very interesting case of a young woman presenting with echocardiographic evidence of tamponade as her initial manifestation of lupus, which has been shown to be a rare initial manifestation of this systemic disease. Her pericardial effusion was drained and the patient responded well to steroid therapy.

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