Systemic Lupus Erythematosus Presenting as Cardiac Tamponade and Pleural Effusion: A Case Report

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Abstract

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that can affect the heart, lungs, and other organs. We describe a case of a 36-year-old female patient who first presented with nonspecific symptoms before receiving a diagnosis of SLE along with initial evidence of pleural effusion and cardiac tamponade. Heart tamponade, which is characterized by fluid accumulation in the pericardial space, is an unusual but serious side effect of SLE. Pleural effusion, or an accumulation of fluid in the pleural cavity, is a typical hallmark of SLE, however, it rarely manifests as the disease’s initial symptom. The early identification and diagnosis of these cardiovascular symptoms of SLE is critical for timely intervention and improved patient outcomes.

This case report highlights the significance of considering SLE when performing a differential diagnosis for patients who have cardiovascular symptoms, particularly when pleural effusion and cardiac tamponade are present. To increase awareness and knowledge of these uncommon presentations of SLE, more investigation and comprehension of the underlying pathophysiology are required.

Categories: Cardiology
Keywords: impending pericardial effusion, cardiac complications, plural effusion, pericardial tamponde, systemic lupus erythromatosus

Introduction

Systemic lupus erythematosus (SLE) is an inflammatory chronic condition defined as an increase of autoantibodies that may impact several body parts like the skin, joints, lungs, neurological system, serous membranes, and/or other body organs [1].

One of the most commonly affected organs in SLE is the heart. Pericardial effusion, pericarditis, Libman-Sacks endocarditis, myocarditis, and, most seriously, coronary heart disease (CHD) are all examples of cardiac involvement, leading to significant cardiac morbidity and mortality [2]. Pericardial effusion is the most common cardiac manifestation of SLE the progression of it to cardiac tamponade is very rare the incidence of it is between 1-3% [3].

The involvement of the heart as an early manifestation of SLE has been documented in the literature. Cardiac tamponade is a rare manifestation of SLE, and when it comes as the first presentation in SLE patients, it is even rarer [4]. Lupus pleuritis stands as the preeminent pulmonary manifestation of systemic lupus erythematosus (SLE), Pleural effusion alone may appear as an extremely rare initial sign of SLE [5].

In this case report, we describe a young female who came to our medical world with non-specific symptoms, found to have massive pericardial effusion and bilateral pleural effusion, mild at the right and moderate at the left, as a first presentation of systemic lupus erythematosus (SLE). This paper intends to elaborate on the pathophysiological underlying cardiac tamponade, delineate the infrequent occurrence of this presentation as a manifestation of systemic lupus erythromatosus (SLE), and attempt to heighten the awareness regarding SLE’s potential presence within the spectrum of differential diagnoses for cardiac tamponade and pleural effusion within the context of relevant clinical scenarios.

Case Presentation

A 36-year-old, 10-day postpartum female came to us with complaints of chest pain, fatigue, dyspnea, and fever over the past month. Her chest pain was at the left side described as “stabbing,” radiated to the left
shoulder, and was related to increased exertion. It was not associated with palpitations. She had an unremarkable past medical or surgical history. During her stay in the hospital, the patient was febrile and became increasingly fatigued. Lately, she became diaphoretic, developed dizziness, and her dyspnea significantly worsened. The cervical swab was negative. She denied flu-like symptoms recently or intravenous abuse. She denied dental caries, hair loss, oral ulcers, a rash, joint pain, or chest trauma. No known autoimmune disease in her family history. She doesn’t drink alcohol or smoke cigarettes.

When a physical examination was done it showed that she had a heart rate of 108 bpm, other vital signs were normal except for the temperature which was measured at 36.3°C. She had pale skin and pale conjunctiva. Head and neck examinations were normal, she had no goiter or cervical lymphadenopathy, but it has been noticed that she had axillary lymphadenopathy. Her abdomen was soft and lax, but significant guarding at the site of the uterus was recognized. The respiratory examination determines the absence of breath sounds and dull on percussion on the left lower hemithorax. On cardiovascular examination, Cardiac auscultation revealed attenuated heart sounds, accompanied by a diffuse and inferiorly displaced apex beat. Jugular venous distension was clinically apparent, while no manifestations of joint inflammation were observed, DVT, peripheral edema, or clubbing. Neurological, psychological, and musculoskeletal exams were normal.

Lab and imaging studies were done. Electrocardiography showed sinus tachycardia. The chest X-ray shows an enlarged cardiac silhouette with a moderate left-sided pleural effusion (Figure 1). Transthoracic echocardiography (TEE) revealed features of moderate-to-severe cardiac tamponade circumferential pericardial effusion inferioposteriorly, with mobile fibrinous bands of the intrapericardial and exudative pleural effusion. The left and right ventricles were of normal size and contracted well. There were no regional wall motion abnormalities (RWMA). Both atria are normal. Mild tricuspid regurgitation was detected. The Inferior vena cava was dilated, not collapsed. Chest CT showed pericardial effusion, and pleural effusion as shown in (Figure 2).

**FIGURE 1:** Chest X-ray shows enlarged cardiac silhouette and moderate left pleural effusion.

Blue arrows show cardiac silhouette sign
Laboratory investigations revealed normochromic normocytic anemia, no increased schistocytes, and no nucleated red blood cells (NRBCs). Other investigations are as shown in Table 1. Stool occult blood was positive. Iron studies for anemia and upper and lower endoscopy were done to rule out malignancy or connective tissue disease. A manual breast exam and gynecological consultation were done. Breast U/S showed axillary lymph node enlargement. Whole body CT was refused by the patient. The patient’s serum tested strongly positive for Anti Sjogren’s Syndrome A (SS-A) and Anti Sjogren’s Syndrome B (SS-B) antibodies and was also positive for anti-double stranded DNA antibodies (anti-dsDNA), also tests shows elevation of the serum complements level and Cancer Antigen 125 (CA-125), but other malignant markers are negative. Pleural fluid cytology showed cells of an inflammatory response, but there was no evidence of malignancy. The plural culture showed no evidence of growth.

Her Potassium was decreased, so she was given KCL orally and in a slow IV fluid infusion. The iron profile showed iron deficiency anemia. COOMB’s test, Serologic tests for hepatitis C virus (HCV), Hepatitis B surface antigen (HbsAg), human immunodeficiency virus (HIV) and a tuberculin skin test were negative. Cardiac biomarkers were all normal (troponin is negative).
**Blood work**

<table>
<thead>
<tr>
<th>Blood test</th>
<th>Patient test result</th>
<th>Normal range</th>
<th>Interpretation of the test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin (HB)</td>
<td>7.5 mg/dl</td>
<td>13.5 - 17.5 mg/dl</td>
<td>Decreased</td>
</tr>
<tr>
<td>Hematocrit (HCT)</td>
<td>22.70%</td>
<td>36 - 48%</td>
<td>Decreased</td>
</tr>
<tr>
<td>MCV</td>
<td>78.3 fl</td>
<td>80 - 100 fl</td>
<td>Decreased</td>
</tr>
<tr>
<td>WBC</td>
<td>4,200/mm³</td>
<td>4,500 - 11,000/mm³</td>
<td>Mild decreased</td>
</tr>
<tr>
<td>Lymphocytes</td>
<td>15.30%</td>
<td>25 - 45%</td>
<td>Decreased</td>
</tr>
<tr>
<td>Neutrophils</td>
<td>78.50%</td>
<td>45 - 65%</td>
<td>Elevated</td>
</tr>
<tr>
<td>Platelet</td>
<td>226,000/mm³</td>
<td>150,000 - 450,000/mm³</td>
<td>Normal</td>
</tr>
<tr>
<td>INR</td>
<td>1.5</td>
<td>0.8 - 1.1</td>
<td>Elevated</td>
</tr>
<tr>
<td>Iron</td>
<td>14 µg/dL</td>
<td>37 - 145 µg/dL</td>
<td>Decreased</td>
</tr>
<tr>
<td>LDH - Serum</td>
<td>220</td>
<td>250 - 450</td>
<td>Decreased</td>
</tr>
<tr>
<td>C3</td>
<td>243 mg/dL</td>
<td>75 - 175 mg/dL</td>
<td>Elevated</td>
</tr>
<tr>
<td>C4</td>
<td>52 mg/dL</td>
<td>15 - 45 mg/dL</td>
<td>Elevated</td>
</tr>
<tr>
<td>Creatinine</td>
<td>0.8 mg/dL</td>
<td>0.7 - 1.3 mg/dL</td>
<td>Elevated</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>147 IU/L</td>
<td>44 - 147 IU/L</td>
<td>Normal</td>
</tr>
<tr>
<td>Calcium</td>
<td>8.5 mg/dL</td>
<td>8.5 - 10.5</td>
<td>Decreased</td>
</tr>
<tr>
<td>Triglyceride</td>
<td>214 mg/dL</td>
<td>less than 150 mg/dL</td>
<td>Elevated</td>
</tr>
<tr>
<td>ESR</td>
<td>150 mm/hour</td>
<td>0 - 20 mm/hour</td>
<td>Elevated</td>
</tr>
<tr>
<td>CRP</td>
<td>84 mg/L</td>
<td>0 - 6 mg/L</td>
<td>Elevated</td>
</tr>
<tr>
<td>Sodium - Serum</td>
<td>138 mmol/L</td>
<td>133 - 146 mmol/L</td>
<td>Normal</td>
</tr>
<tr>
<td>Potassium - Serum</td>
<td>2.4 mmol/L</td>
<td>3.5 - 5.5 mmol/L</td>
<td>Decreased</td>
</tr>
<tr>
<td>Chloride - Serum</td>
<td>104 mmol/L</td>
<td>95 - 110 mmol/L</td>
<td>Normal</td>
</tr>
</tbody>
</table>


The diagnosis of Systemic Lupus Erythematosus (SLE) was ascertained through affirmative clinical and immunologic parameters. The patient met criteria established by the Systemic Lupus International Collaborating Clinics (SLICC), fulfilling four of the 17 specified criteria for SLE classification. These included serositis, diminished serum complements levels, positive serum antinuclear antibodies (ANA), affirmative anti-double-stranded DNA (anti-dsDNA) antibodies, as well as positive anti-SSA and anti-SSB antibodies. Additional support for the diagnosis was derived from the patient’s reported history of fatigue, generalized lymphadenopathy, and an elevated erythrocyte sedimentation rate (ESR). Subsequently, corticosteroid therapy was initiated, Colchicine 0.5 mg 1*2 PO, Trufen 600 mg 1*3 PO for 2 weeks, Nexium 40 mg 1*1 PO, Rocephin 2g 1*1 IV, Slow K 600 mg 2*3, and Folic acid 5mg 1*1. Repeated echocardiography tests every day showed very good progressive improvement the effusion became milder every day till day five of admission the echocardiography showed mild circumferential pericardial effusion with very mild left-sided pleural effusion. At the follow-up visit four months after discharge, She maintained a favorable clinical course and was enrolled in the clinic for the purpose of long-term management of Systemic Lupus Erythematosus (SLE).

**Discussion**

Our patient presented with symptoms of left-sided chest pain radiating from her shoulder, fatigue, fever, and dyspnea on exertion. Examination showed tachycardia, tachypnea, hypotension, jugular venous pressure elevation, and muffled heart sounds, leading to a cardiac tamponade diagnosis. A moderate left-sided pleural effusion was found on a chest CT. Electrocardiography revealed sinus tachycardia and electrical
alternations. Electrical alternans, which cause the alteration of the amplitudes of the QRS complexes with every other beat, are an unusual finding in cases of cardiac tamponade, with reported sensitivities of only 8% to 21% [6]. However, it is highly indicative of pericardial effusion, with a specificity of 89% and a positive predictive value of 82% [6]. These features are highly suggestive of pericardial effusion and tamponade. Chest radiography demonstrated an enlarged cardiac silhouette with an infiltrate on the left side, an integration recorded to have a positive predictive value for pericardial effusion. The initial presentation of systemic lupus erythematosus in our patient was a cardiac tamponade. This was confirmed by autopsy, which showed high anti-nuclear antibody titers, anti-double-stranded DNA antibodies, and anti-SSA antibodies. A conventional physical examination finding, such as Beck’s triad, is an undependable pericardial effusion or tamponade diagnosis [6]. Our patient had normochromic, normocytic anemia.

The most common cardiac manifestation of SLE is pericardial effusion, which occurs in up to 50% of patients somewhere in the course of the illness. It is common and potentially life-threatening, but it can be treated. It is usually mild and unusually seen early in the course of the disease; in rare cases, it leads to cardiac tamponade. This pleural effusion can be due to SLE polyserositis [7, 8]. In some cases, the pericardial effusion can be due to renal failure rather than lupus serositis [9].

Cardiac tamponade in SLE is actually exceedingly rare [6, 10], but it can be the initial presentation of SLE or can happen throughout the course of the disease, and after emergent treatment for it is done, workup for SLE is needed when the underlying cause is not easily explained by any disease process. It is a medical emergency that needs early diagnosis and treatment to reduce morbidity and mortality [7]. It must be well known that cardiac tamponade in SLE can be from both circumferential and loculated effusions; the latter one gives an atypical picture and is called regional tamponade. The progression to tamponade in these patients is affected by a group of factors that can vary from patient to patient, which complicates the identification of tamponade predictors [8]. A significant predominance of females was noted [8, 6]. If an adolescent female presents with hemodynamic compromise with cardiac tamponade, we should consider SLE in the differential diagnosis, especially if features and findings are suggestive of a rheumatic process [10]. Knowing that tamponade can occur at any point in the course, this life-threatening diagnosis should be considered in patients who have unexplained venous congestion [6].

Pericardial involvement is also somehow common in other connective tissue diseases like systemic sclerosis (>60%), mixed connective tissue disease (10-30%), rheumatoid arthritis (10-30%), Sjögren’s syndrome (30%), adult Still’s disease (30%), polymyalgia, and dermatomyositis (<10%). It occurs rarely in sarcoidosis [6].

The size of the pericardial effusion is an important determinant of the development of tamponade. However, in patients with lupus, tamponade can occur in moderate and even small sized effusions [11]. This size is a good indicator of tamponade, but there is a wide variability in the volumes of effusion that are drained on pericardiocentesis. This ensures that although patients are more likely to decompensate with these effusions, because of the subacute accumulation in SLE, the critical volume at which tamponade will develop differs from case to case [8]. However, echocardiography can detect small and silent pericardial effusions [6].

The diagnosis of tamponade is made when there is evidence of right-sided cardiac chamber collapse, dyspnea, hypotension, tachycardia, and elevated jugular pressure, along with pericardial effusion or paradoxical pulse. The majority of studies deport on low serum C4 levels below normal, and 50% or more of tamponade cases showed this finding. It was found that distinct case reports of SLE patients who suffered from tamponade had normal levels of serum C4. Generally, the clinical exacerbations might be able to be predicted by the serial monitor of C4 levels in serum. Another study was done to see if low C3 and C4 levels corresponded to the latest flare of SLE. A relationship between low C4 and SLE flare was unable to be established [12].

When the serum complement C4 level is low, this indicates the progress of cardiac tamponade in SLE [6]. Our patient had high levels of it. Cardiopulmonary features of lupus like myocarditis and myositis are associated with anti-ribonucleoprotein (RNP) antibodies and engage features like myositis and Raynaud’s phenomenon. The mitral valve was most commonly involved [9].
Echocardiography should be done if feasible, to grade and localize the pericardial effusion, reveal pericardial thickening, and see intrapericardial adhesions, in addition to being a standard for confirmation of the diagnosis [6]. In our case, echocardiography obviously visualized a great circumferential pericardial effusion with distinctive signs of tamponade and a left-sided pleural effusion. Within the pericardial effusion, there were partially attached dynamic fibrinous bands traversing the visceral and parietal layers of the pericardium. This feature has been seen in parallel SLE cases and might be a sign of hard pericardiocentesis [6]. The pericardial fluid is mainly described as hemorrhagic in appearance. Other markers in it that can help establish the diagnosis are antinuclear antibodies, the presence of immune complexes, and complement levels. Its protein concentration varies, being high in exudates and low in transudates. Glucose levels are within the normal range [7].

The job of CRP in SLE activity diagnosis is controversial. A recent study showed that there was a logical link with the activity of disease up to a level of 50 mg/L; values above this level were more suggestive of a related infection. The Complement has been outlined to correlate well with lupus activity. There is a significant correlation between a drop in C3 and C4 and a flare of lupus nephritis and a hematological flare, but there are no associated flares in other organ systems [9].

Recent systematic reviews and meta-analyses concluded that anti-nucleosome antibodies have equal specificity but a higher sensitivity and prognostic value than anti-dsDNA antibodies when we diagnose SLE. There is an importance of concomitant pleuritis, anti-nucleosome antibodies, and the size of pericardial effusion in predicting the development of tamponade. We believe that immunosuppression by using methylprednisolone and IV cyclophosphamide is critical, especially to decrease the risk of reaccumulating and surgery [11].

Previous Reports have shown that anti-RNP antibodies are associated with pulmonary disease, specifically pleuritis, in SLE and in other connective tissue diseases. In the serologic profile, anti-Sm antibodies are reported as they are linked with an increased risk of CNS disease, renal disease, and mortality in SLE. To explain this serologic observation, we depend on potential pathophysiologic mechanisms. Anti-RNP antibodies can play a role in the process of tissue injury and inflammation through the up-regulation of inflammatory cytokine production and adhesion molecules at the level of endothelial cells (endothelial leukocyte adhesion molecule-1, and intercellular adhesion molecule-1), which is done through the expression of MHC class II on endothelial cells, and also via the interaction with Toll-like receptors [13].

Myocardial infarction due to coronary arteritis may be a significant cause of death in young adults with SLE [11]. The exact cause of it is probably multifactorial, involving immune myocarditis, small vessel vasculitis, and hypertension due to steroids [9].

A misclassification of pleuritis is probable. Serositis, according to ACR criteria, involves both pericardial and pleural inflammation; thus, the prevalence of pleural inflammation might be overestimated. However, our definition of pleuritis does not differ from that of other huge cohorts. Finally, medical record errors, physician or patient recall bias, and the identification of more prevalent than incident cases of SLE might have had an effect on how serositis was recorded.

Concomitant seropositivity for RNP and Sm antibodies, in addition to greater cumulative organ damage, a longer disease period, and the onset of disease at a younger age, were each associated with an elevated risk for prevalent lupus pleuritis. Constructing factors linked to pleuritis provides valuable information. We suggest that seropositivity for both RNP and Sm antibodies is linked with a higher likelihood of developing pleuritis. At the end, we need to recognize the clinical relevance of pleuritis beyond its serologic and clinical correlates, and we could not do this directly in this cross-sectional study. Previous reports demonstrated a link between short-term mortality and pleuritis. Identifying SLE patients at risk for pleuritis and its consequences has been hampered in the past by a lack of predictors of pleuritis [13].
Using the SLICC criteria, our patient was diagnosed with SLE. However, our patient was unfit to satisfy four of the 11 ACR criteria for classifying SLE. As seen here, low complement levels have not been reported. The SLICC criteria have introduced anti-dsDNA antibodies, anti-Sm antibodies, and antiphospholipid antibodies to contribute in the diagnosis. By permitting a higher weighting of immunologic criteria, the use of the SLICC criteria could be more sensitive to diagnose such cases, possibly leading to earlier diagnosis and treatment [6].

To treat cardiac tamponade immediately, we have to withdraw the pericardial fluid by pericardiocentesis, which is the treatment of choice [7]. The echocardiography guided pericardiocentesis has been associated with lower morbidity and mortality rates, compared to surgical pericardiectomy or placement of a pericardial drain, which are less commonly used. The medical management involves anti-inflammatory medications like NSAIDs, high-dose corticosteroids to reduce the frequency of pericardiocentesis and antimalarials [6]. Pericardiocentesis was not done for this patient. She became well, and her symptoms somehow regressed with the medical treatment as mentioned in the case presentation.

Follow-up is mandatory to exclude recurrent pericardial thickening or effusions. In our case, ESR and CRP levels were elevated. ESR elevation commonly occurs during a flare, but CRP is usually normal or slightly elevated. Serositis is an SLE manifestation that increases both CRP and ESR levels [6].

It is crucial to include SLE in the differential diagnosis of patients who present with cardiovascular symptoms because cardiac tamponade and pleural effusion are uncommon early presentations of SLE. For prompt intervention and improved patient outcomes, early detection and diagnosis are essential. This case report highlights the need for more investigation and knowledge of the underlying pathophysiology while educating medical practitioners about these unusual SLE presentations.

Conclusions
The rare incidence of cardiac tamponade and pleural effusion as initial manifestations of SLE is explained by this case report. It highlights the difficulties in obtaining an early diagnosis and the significance of taking SLE into account when making a differential diagnosis for patients with cardiovascular symptoms. Raising awareness of these unusual appearances has the potential to save lives and improve patient outcomes. To determine the best management approaches and to gain a deeper understanding of the mechanisms underlying various manifestations, more research is necessary.

Additional Information
Disclosures
Human subjects: All authors have confirmed that this study did not involve human participants or tissue. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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