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Case Report

A Case of Quadruple Trouble: SLE's Multi-System Onslaught

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Abstract: This case report presents a diagnostic challenge in a 27-year-old Sudanese female with no past medical history who presented to the emergency department with progressive shortness of breath and pleuritic chest pain. Investigations revealed a constellation of findings suggestive of systemic lupus erythematosus (SLE) with serositis, pleuritic chest pain, pericardial effusion, pleural effusion, and positive autoimmune markers. However, the case became further complicated by the development of acute kidney injury (AKI) and the discovery of a fixed inferior vena cava (IVC) thrombosis during her hospitalization. This report highlights the complexities of SLE presentation, particularly the potential for coexisting diagnoses and the importance of a comprehensive diagnostic approach.

Keywords: Shortness of breath; Pleural effusion; Pericardial effusion; Systemic lupus erythematosus; Systemic disease.

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1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by the production of autoantibodies that target multiple organs and tissues. This aberrant immune response leads to inflammation and tissue damage throughout the body. While the clinical spectrum of SLE is wide [1]. The initial presentation often involves nonspecific symptoms, making early diagnosis challenging. This case report presents a 27-year-old female patient with no past medical history, presented with atypical initial manifestations of SLE, including pleural effusion, cardiac tamponade, that complicated with acute kidney injury (AKI), and a fixed inferior vena cava (IVC) thrombosis. These unusual presentations and complications highlight the complex and unpredictable nature of SLE and emphasize the critical need for a comprehensive diagnostic approach.

2. Case Report

A 27-year-old Sudanese female unemployed, nonsmoker, presented to the emergency department with a five-day history of progressively worsening shortness of breath, and nonspecific chest pain radiating to shoulders not responding to over-the-counter medication such as paracetamol. Regarding cardiopulmonary symptoms, there is no cough, no orthopnea or paroxysmal nocturnal dyspnea and there is no lower limb edema. She denied fever, chills, sick contacts, or other symptoms. No past medical history or surgical history, apart from muscle cramps developed one year ago, which were related to exercise. She sought rheumatologic evaluation and did an autoimmune panel which was negative at that time. Family history was unremarkable.

On examination at the Emergency department, the patient was awake, alert, and oriented. Her vital signs showed tachycardia (HR:149 beats per min), and tachypnea (RR:30 cycle/ min). Blood pressure was (BP) 143/99mmHg, oral temp was 37.4 °C, and oxygen saturation was 97 on a 5-liter non-rebreather mask (Table 1). Chest examination revealed decreased air entry bilaterally at the bases and midzones, normal percussion tone, and basal crackles on auscultation. The remainder of the physical examination was unremarkable. Bedside ultrasound showed pericardial effusion.

Table 1. Vital signs of the patient at presentation, in MICU, and at discharge.

	At the first ED visit	**Rapid deterioration before MICU admission**	**Vital signs at time of discharge**
Oral Temperature	37.4 °C	36,7 °C	37°C
Respiratory rate	30 cycle/min	47 cycle/min	15 cycle /min
Oxygen saturation	97 on non-rebreather mask	100 on high flow nasal cannula	975 on room air
Heart rate	149 beats /min	85 beats /min	70 beat /min
blood pressure	143/99mmHg	131/49 mmHg	120/69 mmHg

The patient was admitted to the ICU for monitoring and blood pressure control. Lab tests, including CBC, TFT, and ECG, were done. A radiological workup includes a chest radiograph, abdominal ultrasound, echocardiography, CT pulmonary angiogram (CTPA), and CT aortogram. Routine investigations showed anemia with an HB level of 7 g/dl and an elevated creatinine level of 181 umol/l, besides CRP of 103.3 mg/l and a high troponin level of 30 ng/l. The chest x-ray showed a significant pleural effusion, primarily on the left side, obliterating the left costophrenic angle (Figure 1). Echocardiography showed bilateral pleural effusion mainly on the left side (Figure 2). CTPA showed no evidence of pulmonary embolism, and CT aortogram was unremarkable.

Figure 1. Chest X-ray showed bilateral pleural effusion with mainly obliteration of left side.

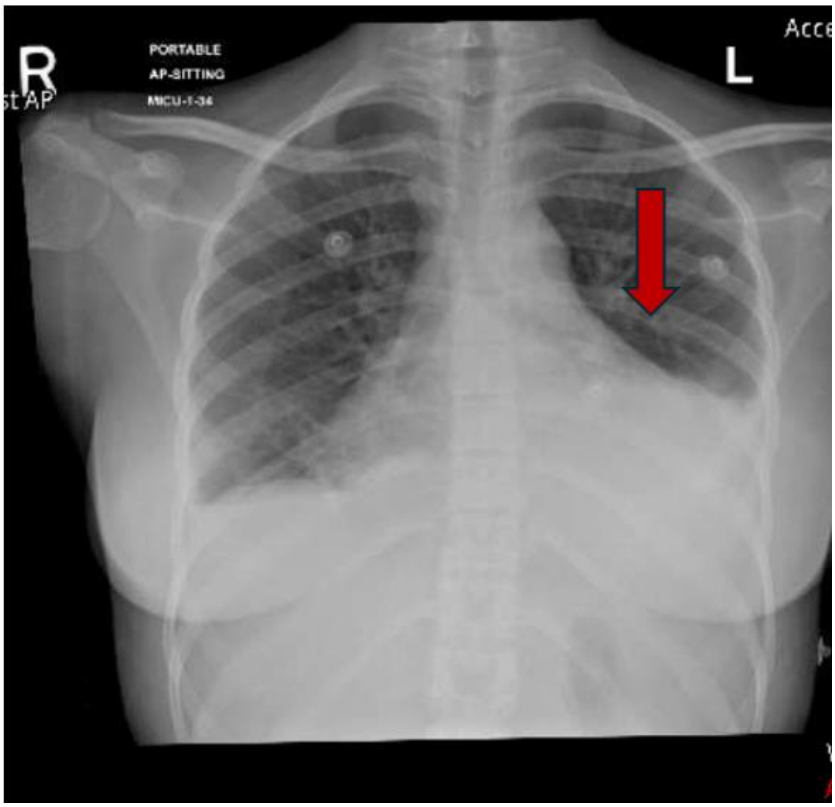
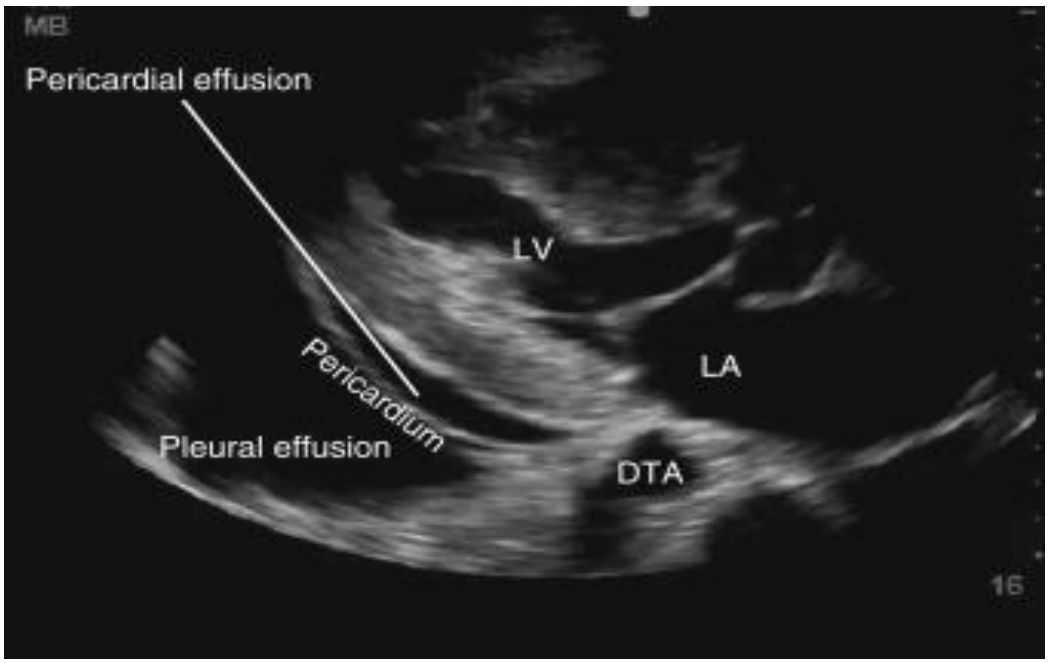


Figure 2. Echocardiography showed Pericardial and Pleural effusion.



Based on clinical manifestations, lab analysis, and radiological findings, a primary diagnosis of pericarditis due to a systemic disease such as tuberculosis or autoimmune disease was suspected. The patient started on antibiotics (ceftriaxone and azithromycin) and DVT prophylaxis (enoxaparin). Tuberculosis and autoimmune workups were ordered as suggested after rheumatology and nephrology consultations. At the end of the second day, the patient started to decline. Her SOB and chest pain increased rapidly; echocardiography showed rapidly expanding pericardial effusion with fibrin shred. The patient underwent a pleural tab and started on a steroid (prednisolone 40 mg) to guard against cardiac tamponade.

On the third day of admission, echocardiography was repeated. A fixed, non-mobile thrombus measuring 3cm by 0.9cm within the IVC was identified, and renal function deteriorated. The team weighed the risks of bleeding complications associated with anticoagulation against the potential for further thromboembolic events, ultimately, the patient commenced on heparin infusion, amlodipine 5 mg, and fractionated plasma. Serological workup showed positive ANA 1:1280, anti-dsDNA 299, RNP 240, anti-Smith 56, positive Coombs test, low C3 (Table 2). Diagnosis with SLE with seronegative antiphospholipid syndrome was made. The patients started to improve, and renal function started to get back to normal. She was transferred to the medical ward and started on rituximab 500mg, and hydroxychloroquine 200 mg, as well as continuing the same dose of steroids and starting warfarin 5 mg.

Table 2. Laboratory results during Hospitalization.

Group	Detail	Value w/ unit	Flags	Normal range
General hematology	Hb	7 g/dl	low	11,3 - 17
Blood chemistry	Creatinine	181 u mol/l	high	62 - 106
Blood chemistry	Troponin-T HS	30 ng/l	high	3 - 15
General hematology	CRP	103.3 mg/l	high	0.0 - 5
Immunology	C3	0.53 gm/l	low	0.90 – 1.80
Serology	ANA	1:1280 titer	positive	1:40 – 1:640

Serology	Anti-dsDNA	299 IU/mL	positive	0 – 25
Serology	Anti-RNP	240 U/mL	positive	Less than 20
Serology	Anti-Smith	56 U/mL	Positive	0 -7 U/mL
Serology	Coombs test		Positive	

Following a comprehensive rheumatologic workup, the patient was discharged after two weeks with instructions to follow up at a rheumatology clinic. At discharge, rituximab-mab therapy was continued, and prednisolone was gradually tapered. Follow-up and laboratory tests were scheduled one month later. Unfortunately, the patient lost follow-up and went back to her home country.

3. Discussion and conclusion

Systemic Lupus Erythematosus (SLE) presents a diagnostic challenge due to its clinical heterogeneity. Unlike diseases with characteristic presentations, SLE manifests with a diverse array of symptoms, often involving multiple organ systems. Patients frequently exhibit constitutional symptoms, musculoskeletal pain, and systemic involvement, with variations in symptom severity and combinations across different age groups. This broad spectrum of clinical manifestations documented in literature underscores the diagnostic dilemma [1]. While classification criteria such as the 2019 EULAR/ACR, 2012 SLICC, and 1997 ACR have been invaluable in standardizing the diagnostic process, their application often requires careful interpretation in conjunction with clinical judgment.

In contrast to diseases with well-defined diagnostic pathways, SLE necessitates a comprehensive evaluation, including a meticulous medical history, physical examination, and laboratory tests, to establish a definitive diagnosis. Compared to other autoimmune diseases with more specific diagnostic markers, SLE's diagnostic journey is often challenging. The overlapping symptoms with other connective tissue diseases can further complicate the diagnostic process. The nature of SLE is further exemplified by its rare and atypical presentations, which can involve any organ system. These unusual manifestations often confound diagnosis, as they mimic other diseases or present as isolated, seemingly unrelated symptoms. For instance, described a patient with SLE and antiphospholipid syndrome who developed isolated sterile tricuspid valve vegetations [6], while reporting a case of enteritis as the sole initial presentation in an adolescent girl [7].

Even within the realm of rare conditions, SLE can be an unexpected culprit, as evidenced by its association with acquired von Wille brand disease [8]. Moreover, the spectrum of SLE complications is vast, and sometimes, the complications themselves can serve as the initial presenting symptoms in previously undiagnosed patients. Serositis, for example, can be an early manifestation, as illustrated by a case of preeclampsia that ultimately unmasked lupus nephritis [5]. However, the rapid progression of SLE, emphasizes the critical importance of early recognition, as seen in a patient with late-onset SLE who developed effusive constrictive pericarditis, culminating in cardiac arrest within days of diagnosis [3]. This underscores the challenges in establishing a timely diagnosis, particularly when confronted with atypical presentations [4].

Unlike more predictable disease courses, SLE often defies conventional diagnostic pathways. In contrast to diseases with well-defined symptom clusters and specific biomarkers, SLE requires a high index of suspicion and a comprehensive diagnostic workup. While certain autoimmune diseases may have established diagnostic algorithms, SLE necessitates a more flexible and adaptable approach. The variable clinical course and the potential for rapid deterioration further distinguish SLE from other autoimmune conditions, demanding prompt intervention and close monitoring. In contrast to other cases, this previously healthy female presented with progressive shortness of breath and non-

specific chest pain, and she experienced rapid deterioration within the first hours of admission. This raised concerns about significant underlying pathologies, such as pulmonary embolism, which was ruled out by a CT aortogram.

The significant increase in pericardial effusion with fibrin strand formation, noted by transthoracic echo, prompted the team to start steroids to guard against cardiac tamponade. Escalation of challenges in this critical situation arose when the cardiologist identified a fixed, non-mobile thrombus within the IVC establishing the diagnosis of SLE with seronegative antiphospholipid syndrome. A decisive decision had to be made, weighing the benefits of starting anticoagulation to prevent thrombus progression against the risk of potential bleeding.

The choice to start anticoagulation in this quiet was basic, adjusting the dangers of thrombotic occasions against the potential for complications. The nearness of a settled thrombus inside the IVC showed a tall hazard of complications, such as pneumonic embolism or profound vein thrombosis. The main factors considered in the decision-making process for this patient began with assessing the severity of thrombosis. The measure and location of the thrombus can impact the likelihood of complications and the urgency of anticoagulation therapy. Another important factor is the patient's risk profile, which includes their general health status, age, comorbidities, and medication use, all of which can affect the risk-benefit analysis. Additionally, the patient's previous response to anticoagulation plays a role. If the patient has a history of bleeding or thrombotic events while on anticoagulation, this experience can inform the decision-making process.

There are notable benefits of anticoagulation, including the prevention of thrombosis. Anticoagulants can reduce the risk of recurrent thrombosis, preventing complications such as pulmonary embolism or deep vein thrombosis. Another benefit is improved outcomes early anticoagulation can improve long-term results in patients with systemic lupus erythematosus (SLE) and antiphospholipid syndrome [9]. However, there are also risks associated with anticoagulation. These include an increased risk of bleeding, especially in patients with underlying conditions or those taking other medications that affect clotting. Additionally, hemorrhagic stroke is a risk, particularly for patients with uncontrolled hypertension or other risk factors.

Ultimately, the patient was anticoagulated. Fortunately, she improved with treatment. Following discharge after two weeks of comprehensive rheumatologic workup and stabilization, the patient was scheduled for a one-month follow-up to monitor her condition, conduct lab tests, and adjust her management plan accordingly, unfortunately, she lost follow-up. This case report contributes to the developing body of writing on the complex interaction between Systemic Lupus Erythematosus (SLE) and antiphospholipid disorder, especially in the setting of thrombotic occasions. Whereas various case reports and observational thoughts have been distributed, there is still a need for more large-scale randomized controlled trials to build up authoritative rules for the administration of these patients.

SLE is a chronic autoimmune disease characterized by its diverse clinical manifestations. The management of SLE requires a comprehensive approach that addresses both the underlying disease and its associated symptoms. One of the primary goals of SLE treatment is to control disease activity and prevent organ damage. Corticosteroids are often used to suppress the immune system and reduce inflammation.

In addition to corticosteroids, immunosuppressants such as hydroxychloroquine, azathioprine, and cyclophosphamide are used to control disease activity and induce re-mission. These medications can help to prevent organ damage and improve long-term outcomes. For patients with antiphospholipid syndrome, anticoagulants are essential to prevent thrombotic events, such as blood clots. Studies have demonstrated the effectiveness of anticoagulation therapy in reducing the risk of recurrent thrombosis in patients with SLE and antiphospholipid syndrome [9]. Beyond medical interventions, supportive care plays a crucial role in managing SLE. Addressing symptoms such as fatigue, joint pain, and skin rashes can significantly improve the quality of life for patients.

The prognosis of SLE varies widely depending on factors such as disease severity, organ involvement, and response to treatment. While some patients may experience mild symptoms and a favorable prognosis, others may develop severe complications that can significantly impact their quality of life. Early diagnosis and effective treatment are crucial for improving outcomes in SLE patients [4]. This case report underscores the challenges associated with diagnosing and managing SLE and antiphospholipid syndrome (APS), particularly in acute settings with non-specific symptoms. The patient's rapid deterioration and the need for urgent intervention highlight the importance of a thorough diagnostic approach, timely intervention, and interdisciplinary collaboration.

The successful management of this patient required a multidisciplinary team, including rheumatologists, cardiologists, and other specialists. The use of corticosteroids and anticoagulation was essential in controlling disease activity and preventing complications. Moreover, this case emphasizes the importance of individualized long-term plans and follow-up care for patients with SLE and APS. Regular monitoring of disease activity, laboratory tests, and adjustments to the treatment plan can help improve outcomes and reduce the risk of complications.

In conclusion, this case report highlights the complexities of managing SLE and APS, emphasizing the need for a comprehensive approach that includes timely diagnosis, effective treatment, and ongoing monitoring. By addressing these factors, healthcare providers can improve outcomes for patients with these complex autoimmune diseases.

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