Primary Presentation of Systemic Lupus Erythematosus with Pulmonary Embolism: A Case Report

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KEYWORDS

Systemic Lupus Erythematosus. Pulmonary Embolism, Case Report

ABSTRACT

Pulmonary embolism (PE) as the first clinical manifestation of systemic lupus erythematosus (SLE) is an unusual and rare presentation. This case report discusses a 30-year-old male who presented with chest pain and hypoxemia at a hospital in Dubai. Initial workup revealed elevated D-dimer levels and a positive CT pulmonary angiogram confirming PE. The patient's medical history was unremarkable, except for a recent transient leg discomfort. Upon further investigation, hypoalbuminemia and proteinuria suggested nephrotic syndrome. A thorough autoimmune evaluation revealed elevated anti-dsDNA antibodies, leading to a diagnosis of SLE nephritis. This case emphasizes the necessity of considering SLE as a differential diagnosis for young patients with unexplained thrombotic events, even in the absence of traditional SLE symptoms. Treatment and early recognition are important in managing thrombotic complications in SLE.

1. Introduction

Systemic lupus erythematosus is a complicated systemic autoimmune disease; it may present itself in various ways, potentially involving many organs. The following is a case report of an interesting case where the disease was known for its myriad presentations for diagnosis when it was presenting with one alarming complication related to pulmonary embolism. The systemic autoimmune disease interplays in a complex manner with a probably lethal pulmonary complication, underlining the complexity of SLE and forming the hallmark of the thoroughness that is necessitated in diagnosis and management. In the present paper, an effort has been made to dissect the nuances of this case, detailing the interrelationship between SLE and pulmonary embolism and further complications while managing it clinically.

Case Presentation:

- Our case report is about 30 years old Emirati male patient, with an unremarkable medical history except for being a cigarette smoker of 20 packs/ year, our patient presented to the ER for another medical facility at first seeking medical help for chest pain where it was treated empirically using OTC pain killers and he was discharged after few hours, the pain did not improve, for this reason, the patient attended our ER because of chest pain in ER clinical evaluation revealed hypoxemia Spo2 93% on room air, oxygen supply started and diagnostic algorithm initiated, then the patient shifted to the ICU for further management and evaluation.
- When we asked the patient he denied any previous joint pain, swelling, or any other complaints except for mild left leg pain a week before and a feeling of leg swelling that was relieved spontaneously. The blood results came back as the following

Lab test	Value	reference range
HB	14.2 g/dl	13.00-17.00
WBC	10200 10^{3} /µL	4.00-10.00
CREATININE (BLOOD)	0.72 mg/dL	0.00-1.20 mg/dL
POTASSIUM(K)	4.03	3.50-5.10
SODIUM (Na)	135.00 mmol/L	136.00-145.00 mmol/L
CALCIUM	7.23 mg/dL	9.00-10.50 mg/dL

URIC ACID	4.43 mg/dL	3.40-7.00 mg/dL
BLOOD UREA	17.70 mg/dL	19.00-44.00 mg/dL
D-DIMER(QUANTITATIVE)	5.16ug/mL	0.00-0.50 ug/mL
ALBUMIN	2.03 g/dL	3.50-5.20g/dL
Blood(U)	Positive (2+)	Negative
Protein(U)	Positive (2+)	Negative

- The low albumin level and the urinary findings were surprising as they were unjustified clinically in the absence of signs and symptoms like peripheral pitting edema and urinary symptoms, for this reason, these results were double-checked.
- Based on the clinical and lab findings the diagnosis of pulmonary embolism was suspected and a detailed imaging study was done, including CXR, CTPE, lower limb Doppler, and echocardiogram.
- CTPE result: Positive for pulmonary embolism. Associated wedge-shaped pleural-based soft tissue and ground glass opacities seen in the right posterior basal segment, suggest infarct.
- The diagnosis of PE was done and treatment with enoxaparin anticoagulant dose was initiated.
- The chief complaint of the patient was chest pain with no other complaints, and the continuous clinical evaluation failed to find any other sign except for first-degree pitting edema in the lower limbs bilaterally.

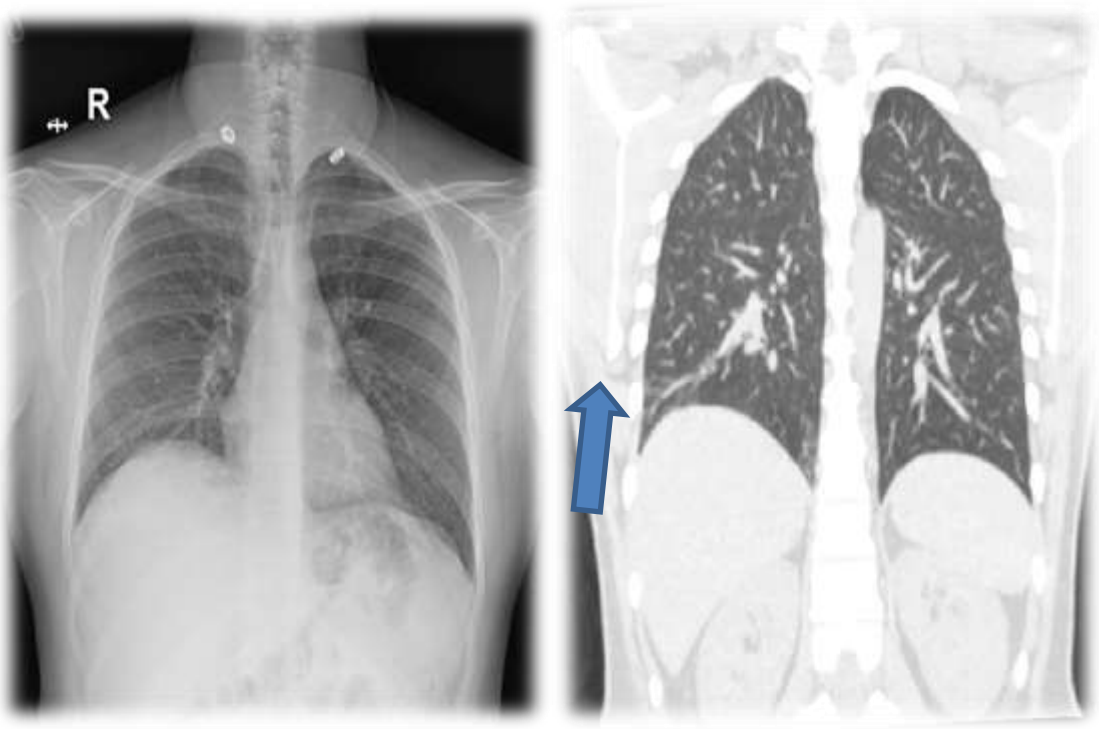
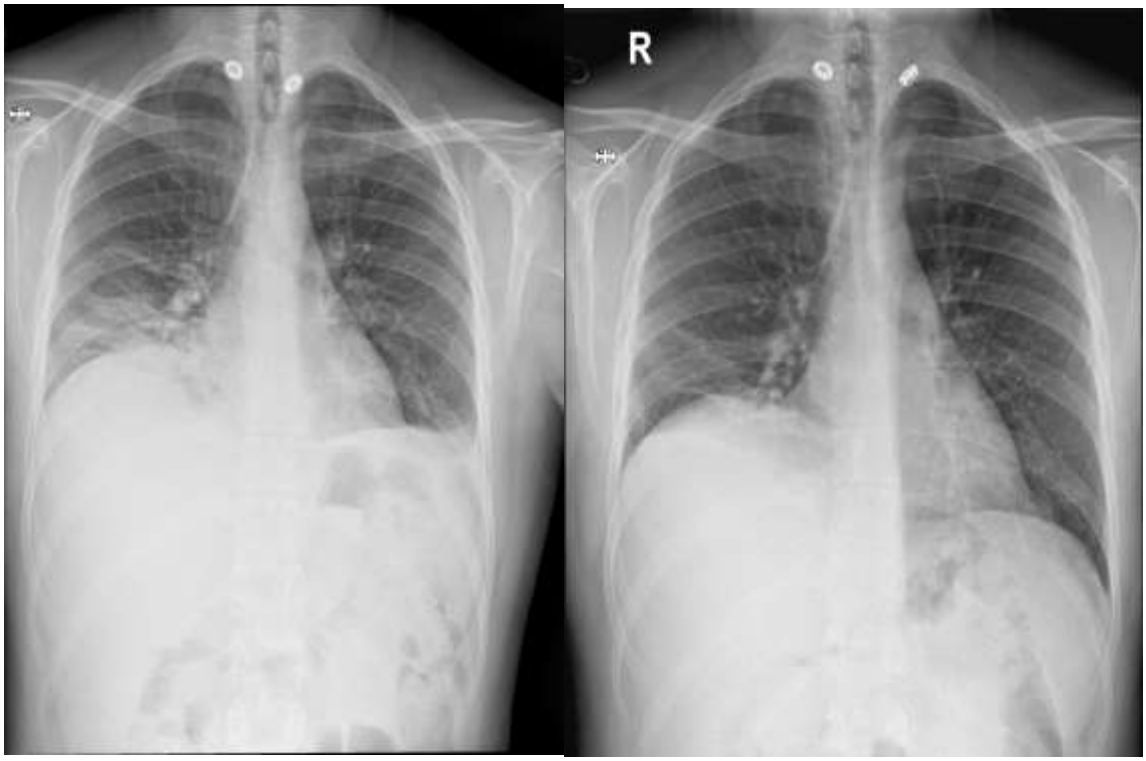


Figure 1-1 CXR that was done upon ER visit.



Diagnostic Assessment:

- As the lab tests revealed hypoalbuminemia in addition to high proteinuria, a diagnosis of nephrotic syndrome was formulated, then a comprehensive study for the nephrotic syndrome was done as follows.

Lab test	value	reference range
ANTI PHOSPHOLIPID ANTIBODIES Phospholipid IgG (EIA)	1.9U/ml	< 10.0 U/ml
ANTI PHOSPHOLIPID ANTIBODIES Phospholipid IgM (EIA)	1.9U/ml	< 10.0 U/ml
C3 COMPLEMENT	2.36 g/L	0.9-1.8 g/L
C4 COMPLEMENT	0.40 g/L	0.1-0.4 g/L
HIV - 1&2 Abs / P24Ag	NON-REACTIVE	NON-REACTIVE
MTHFR (1298 A>C):	Normal (Wild Type)	
ANCA-MPO (ANTI MYELOPEROXIDASE ANTIBODIES), SERUM	0.5 U/mL	Normal: < 5 U/mL
Anti-Nuclear Antibody (ANA) pattern nuclear	1: 100 (borderline)	< 1:100
HEPATITIS B SURFACE ANTIGEN (HBSAG)	NON-REACTIVE	NON-REACTIVE
ANCA-PR3 (SERIN PROTEINASE 3 ANTIBODIES), SERUM	2.3 U/mL	NORMAL: <10 U/mL

ANTI DOUBLE STRAND (DS DNA) IGG	24.4 IU/mL	Elevated:>=20.0 IU/mL
Phospholipase A2 rec.IgG Abs	<2.0 U/ml	<14 U/ml
Factor 2 Mutation (PCR)	negative	
Factor 5 Leiden Mutation (PCR)	negative	
ALBUMIN CREATININE RATIO (URINE)	>6523.48 H	0.00-20.00
MICROALBUMIN (URINE)	>8611.00 H mg/L	0.00-20.00 mg/L
24 HOUR URINE TOTAL PROTEIN	22125.0 H	0-140mg/24 hour

Based on the previous labs a diagnosis of SLE nephropathy was done.

Treatment, Outcome and follow up:

- Our main treatments were enoxaparin anticoagulant dose and prednisolone, in addition to painkillers to manage the chest pain and oxygen support.
- Gradually the case improved in terms of oxygenation and chest pain, for this reason, the patient was shifted to the medical ward, and with further improvement, the patient was discharged and followed as an outpatient.

The patient received a wide spectrum of immune suppressants which was escalated gradually overtime during the follow-ups in OPD to control his lupus, at the beginning it was started with prednisone then mycophenolate mofetil was added, after that tacrolimus, then rituximab was given, and 24 hrs. urine protein was monitored, the best response was achieved after using the cyclophosphamide as the 24 hrs. urine protein came to 3060 mg/24 hrs.

2. Result and Discussion

Patients with SLE have an increased risk of venous thromboembolic disease based on healthcare database analyses. Among hospitalized patients with SLE, the risk of venous thromboembolism (VTE) is 1.23 (95% CI 1.15-1.32) compared with hospitalized patients without autoimmune disease. SLE is associated with many risk factors for VTE, such as membranous nephropathy and the presence of antiphospholipid antibodies.

Antiphospholipid syndrome (APS) is a clinical and laboratory diagnosis characterized by both persistent laboratory evidence of antiphospholipid antibodies (aPL) **and** related complications, which may include venous thrombosis, arterial thrombosis, adverse pregnancy outcomes, and non-thrombotic manifestations (e.g., heart valve thickening, livedo reticularis/racemosa). APS occurs either as a primary condition or in the setting of an underlying disease, usually systemic lupus erythematosus (SLE).

Kidney damage (lupus nephritis) is one of the more common health problems caused by lupus. In adults who have lupus, as many as 5 out of 10 will have kidney disease. In children who have lupus, 8 of 10 will have kidney disease

The risk of thrombosis varies among the causes of nephrotic syndrome and appears to be highest in patients with membranous nephropathy This was illustrated in a cohort of 1313 patients with idiopathic glomerular disease due to membranous nephropathy, focal segmental glomerulosclerosis, or immunoglobulin A (IgA) nephropathy. The incidence of venous thromboembolic events was much higher in membranous nephropathy (7.9 percent) and focal segmental glomerulosclerosis (3.0 percent) than in IgA nephropathy (0.4 percent). The histologic diagnosis remained a predictive factor for thrombosis after adjustment for the degree of proteinuria (which was much higher at presentation in membranous nephropathy and focal segmental glomerulosclerosis [median 5.6 and 3.7 versus 1.6 g/day in IgA nephropathy]) and the serum albumin concentration.

Patients with membranous nephropathy associated with lupus also appear to be at high risk for thromboembolism. In a study of 66 such patients, venous thromboembolism occurred in 15 (23 percent) at a mean follow-up of 6.9 years. Factors other than nephrotic syndrome can contribute to an increased incidence of thromboembolism in patients with lupus, such as antiphospholipid antibodies.

Medical literature was reviewed seeking similar cases, just one case was found, and what makes this case unique is: that it was an SLE case in a male patient, with the first manifestation to be Pulmonary embolism based on nephrotic syndrome with no usual SLE presentations like joints pain, hair loss, photosensitivity in addition to the fact that clinical evaluation did not reveal any specific SLE cutaneous findings and (the only complaint the patient mentioned was just a mild transient discomfort feeling in the right leg with the sensation of this leg to be swollen which improved spontaneously one week before the current presentation), and finally the antiphospholipid study was negative.

Such a case raises the awareness of SLE as the cause behind thrombosis in young individuals even in the absence of traditional signs and symptoms of it.

3. Conclusion and future scope

It further suggests that systemic lupus erythematosus may be an important factor in the underlying cause of pulmonary embolism, even without symptoms typical for SLE. In this patient, SLE diagnosis was established based on the presence of nephrotic syndrome and high titer anti-dsDNA antibodies at the development of a PE as the first clinical manifestation. It is atypical of this presentation to highlight the necessity of further work-up of autoimmune disorders in young patients with thrombotic events of unknown causes.

SLE thus needs to be one of the differential diagnoses for any unexplained VTE, many a time presenting with features of nephrotic syndrome. Very early diagnosis and timely treatment, including anticoagulation and immunosuppression, will help in an improved prognosis of such patients. Further studies are indicated to understand the pathophysiology behind thrombotic events in SLE and strategize regarding early identification and management of high-risk patients.

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