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The Hidden Peril: Unraveling A Catastrophic Pulmonary Crisis in Systemic Lupus Erythematosus - Case Report

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Abstract

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease known for its diverse systemic manifestations. Diffuse Alveolar Haemorrhage (DAH) is a severe and life-threatening complication of SLE, marked by extensive alveolar bleeding and high mortality. We report a case of a young female who presented with DAH, acute psychosis, and multi-organ involvement due to SLE, underscoring the importance of early recognition and aggressive management.

Keywords: Mucocutaneous, Pulmonary, Renal, Dyspnoea.

Introduction

Systemic Lupus Erythematosus (SLE) can present with a wide spectrum of clinical features with a reported prevalence of in India as 3.2 per 100,000 populations. It affects multiple organs in the form of constitutional symptoms, musculoskeletal involvement, mucocutaneous features along with widespread renal, cardiac, haematological and neuropsychiatric involvement which makes it challenging to diagnose and manage. Therefore, a classification criteria have been developed for SLE, which labels the patients as definite, probable, possible SLE based on ACR/SLICC criteria.

Pulmonary complications, such as DAH, are rare but critical. DAH results from damage to the alveolar-capillary membrane and is associated with high morbidity and mortality if not rapidly identified and treated. This case highlights the clinical challenges in managing severe SLE presenting with nonspecific symptoms which posed a diagnostic challenge and eventually developing DAH during hospital stay and the importance of a multidisciplinary approach involved in managing the same successfully.

Case Report

We present here a case of 24-year-old female who presented to a tertiary care hospital with chief complaints of headache in bilateral frontal region mild in

intensity with no associated photophobia or focal neurological deficits for 2 weeks along with generalised body aches. Four days prior to admission she developed abnormal behaviour characterised by irrelevant speech, not associated with fever, seizure or neck stiffness. She also reported bleeding from gums for 3 days. On general physical and systemic examination pallor was noticed, rest of the examination was normal.

On the basis of history and examination and patient belonging to a scrub typhus endemic area, a presumptive diagnosis of scrub typhus with sepsis associated encephalopathy was made and patient was started empirically on doxycycline, meropenem and vancomycin. Simultaneously a detailed workup was sent for the patient. Initial lab investigations revealed anaemia, AKI, elevated transaminases, urine RBCs and thrombocytopenia.

Subsequently on 2nd day the patient developed haemoptysis and tachypnea, along with crepitation on lung examination with a fall in saturation. A malar rash was observed as well, prompting reconsideration of the diagnosis. Immediately a chest X-ray was ordered for the patient along with ANA panel, C3, C4 and an urgent HRCT was done. Meanwhile patient was shifted to critical care unit in view of worsening dyspnoea and managed with non-invasive ventilation. Emergency HRCT revealed diffuse infiltrates which was consistent with DAH. The complete picture of malar rash and DAH along with labs of AKI, thrombocytopenia prompted the diagnosis of SLE flare and after a rheumatology consultation a steroid pulse was planned and given. The patient responded well to immunosuppressive therapy in the form of mycophenolate mofetil along with tapering steroid doses. Her sensorium and headache improved. Her complaint of bleeding gums and haemoptysis also resolved along with improvement in platelet count, RFT, LFT and urine RBCs also became nil.

Meanwhile her ANA reflux was also collected which showed ANA positivity in the titre of 1:320, nucleolar pattern and the diagnosis of SLE was confirmed along with systemic involvement in the form of pulmonary(DAH), renal(AKI), mucocutaneous (malar rash), neurological (acute psychosis).

Table 1: Initial lab investigations

	DAY1		DAY1		DAY1	DAY 7	
HB	10.3	ELECTROLYTE	135/4/88	PT/INR	NO COAGULATION	ANA	POSITIVE 1:320
TLC	7	CALCIUM	9.2	PROCAL	6.63	DCT	POSITIVE
PLT	26	PHOSPHATE	3.36	VBG pH	7.25		
QCRP	-	BILIRUBIN TOTAL	1.9	PCO2	25		
ESR	15	SGOT/SGPT	88/161	HCO3	12		
BUN	100	ALP	309	LACTATE	3.01		
CREAT	4.1	PROTEIN T/A	5.5/2.04	NCCT HEAD	NORMAL		

Figure 1: Cutaneous manifestation in the form of malar rash

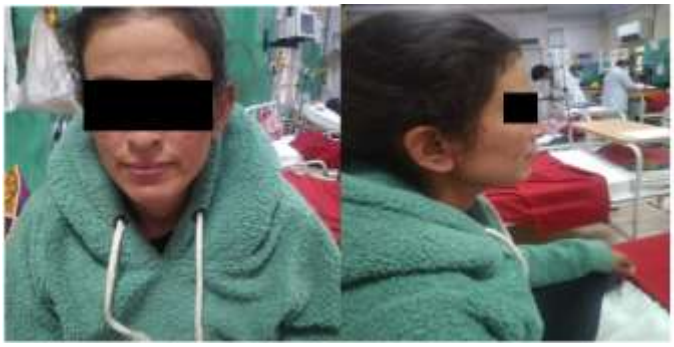

Figure 2: HRCT consistent with DAH (diffuse alveolar infiltrates)

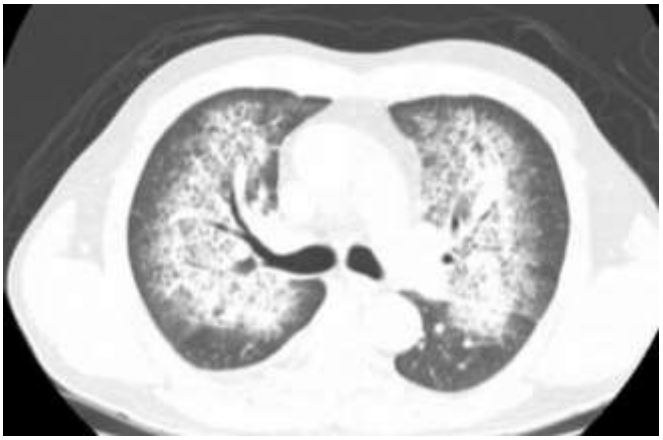

Table 2: Lab trends post administration of steroid pulse

	Day 15	Day 17	Day 18	Day 19
Hb	10.3	9.8	9.3	9.1
TLC	7000	6500	4900	6500
DLC	N94L4	N86L11	N72L15	N73L15
PC	$26X10^3$	$47X10^3$	$72X10^3$	$108X10^3$
PBF	-	NC/NC	-	-
QCRP	85	65		17.5

Discussion

Diffuse Alveolar Haemorrhage (DAH) is a life-threatening complication of Systemic Lupus Erythematosus (SLE), with a reported incidence of 2–4% among SLE patients and mortality rates reaching up to 50% despite advancements in medical care of SLE poses significant diagnostic and therapeutic challenges, as its initial presentation can be nonspecific and easily misinterpreted. This case highlights the complexity of diagnosing SLE with DAH, particularly in young patients with multi-organ involvement.

The initial symptoms in our patient included headache, abnormal behaviour, and bleeding from the gums, all of which were non-specific. The subsequent development of haemoptysis and respiratory distress prompted further investigation, revealing diffuse alveolar infiltrates on high-resolution CT, consistent with DAH. This underscores the importance of clinical vigilance and considering DAH in the differential diagnosis of SLE patients presenting with respiratory or bleeding symptoms.

Pathophysiologically, DAH in SLE is believed to result from immune complex deposition in the pulmonary capillaries, triggering inflammation, vasculitis, and eventual rupture of the alveolar-capillary barrier. Immune-mediated lead to rapid and severe respiratory failure, necessitating immediate intervention. The positive antinuclear antibody (ANA) in our patient was consistent with SLE, although the absence of anti-dsDNA antibodies highlights the variable serological profile often encountered in SLE-associated complications.

Our case also demonstrates the atypical manifestation of acute psychosis, which is part of the neuropsychiatric spectrum of SLE. Neuropsychiatric SLE (NPSLE) can present with a wide array of symptoms, including mood disorders, psychosis, and cognitive dysfunction, often complicating the clinical picture. The presence of DAH highlights the systemic and unpredictable nature of SLE, underscoring the need for a comprehensive and multidisciplinary approach to management.

The mainstay of treatment for DAH in SLE involves high-dose corticosteroids, which help to suppress the overwhelming inflammatory response. In our case, the patient received intravenous methylprednisolone, which led to significant clinical improvement. Immunosuppressive agents, such as cyclophosphamide and mycophenolate mofetil (MMF), are often required to achieve long-term disease control. Given the potential side effects, the addition of MMF was crucial in this patient to maintain remission while minimizing adverse effects.

Several studies have emphasized the importance of rapid initiation of immunosuppressive therapy. For instance, research by Kazzaz et al. showed that early and aggressive treatment with high-dose corticosteroids and adjunctive immunosuppressive agents reduces mortality in DAH patients. Our patient's favourable outcome highlights the approach, though it is essential to recognize that outcomes can vary based on the severity of disease and the presence of other organ involvement.

Despite the positive response in this case, the literature indicates that patients with SLE and DAH often experience relapses and require ongoing monitoring and treatment adjustments. Long-term follow-up, including regular pulmonary and rheumatological assessments, is critical for detecting and managing complications or recurrent disease activity. Moreover, collaboration among specialists, such as pulmonologists, rheumatologists, and intensivists, is key to optimizing patient outcomes.

Conclusion

This case of a young female patient with Systemic Lupus Erythematosus (SLE) complicated by Diffuse Alveolar Haemorrhage (DAH) emphasizes the importance of early recognition, a multidisciplinary approach, and aggressive immunosuppressive therapy. DAH in SLE remains a rare but severe manifestation with a high mortality risk, demanding prompt and targeted intervention. Despite the challenges of diagnosis and management, as demonstrated in this case, timely treatment with high-dose corticosteroids and appropriate long-term immunosuppressive agents like mycophenolate mofetil (MMF) can significantly improve outcomes and stabilize patients. Continued follow-up is essential to monitor disease activity and manage long-term complications.

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