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Coexistence of Pulmonary Embolism and Refractory Pulmonary Hemorrhage in Systemic Lupus Erythematosus: A Treatment Challenge

Calvin Diong Wai **Nga**[1], Zheng Hang **Yee**[1], Ping Seung **Ong**[1], Chiew Gek **Khor**[1]
[1]Department of Medicine, Hospital Raja Permaisuri Bainun Ipoh

INTRODUCTION

Diffuse alveolar hemorrhage (DAH) is a rare but severe pulmonary complication of systemic lupus erythematosus (SLE), affecting 2–5% of patients. It presents with hemoptysis, anemia, hypoxemia, and bilateral infiltrates, with mortality as high as 90%.

CASE REPORT

Miss NS, a 20-year-old woman with known SLE since 2018 (on azathioprine, hydroxychloroquine, and low-dose prednisolone)

- 1-month cough
- Loss of appetite and 10 kg weight loss
- 2 weeks of polyarthralgia, oral ulcers, alopecia
- 4 days of frothy urine

Initial findings:

- SLE flare involving musculoskeletal, hematologic (bicytopenia), and renal (24-hour urine protein: 823 mg/day) systems
- Started on IV hydrocortisone 100 mg TDS
- Planned for renal biopsy

Day 7 – Onset of DAH:

- Developed desaturation and blood-streaked sputum
- CTPA: No PE, but bilateral reticulonodular opacities and ground-glass changes
- dsDNA titer: 1530 IU/mL
- Hemoglobin dropped from 8.5 to 6.4 g/dL; new thrombocytopenia
- Sepsis markers were low (PCT 0.15 ng/mL, CRP 3.1 mg/L)
- DAH suspected clinically; renal biopsy postponed
- Initiated on plasma exchange (PLEX), IVIG, and pulsed IV methylprednisolone (0.5 g/day)

Day 11 – DVT Complication:

- Right lower limb swelling observed
- Doppler ultrasound: Long segment DVT from right common femoral to external iliac vein
- Femoral catheter removed; new right IJV catheter placed
- APLS workup was negative

Challenge in Anticoagulation:

- Despite confirmed DVT, initiation of anticoagulation (clexane) was **delayed** due to **ongoing hemoptysis and suspicion of active DAH**
- This posed a major therapeutic challenge: balancing thrombotic risk against the high risk of worsening alveolar bleeding
- To mitigate risk, an **inferior vena cava (IVC) filter** was inserted as a temporary measure

Day 14 – Clinical Worsening:

- Persistent hemoptysis, worsening respiratory distress
- Required non-invasive ventilation and ICU admission
- Repeat CTPA: Progressive bilateral consolidations, no PE
- Continued immunosuppressive therapy

Day 22 – Intubation and Escalation:

- Recurrent hemoptysis and respiratory failure → intubated
- Immunosuppressive regimen included:
  - PLEX ×10 cycles
  - IV methylprednisolone 0.5 g OD ×3 days
  - IVIG ×5 doses
  - IV cyclophosphamide 500 mg Q2 weekly
  - IV rituximab 500 mg weekly

Recovery Phase:

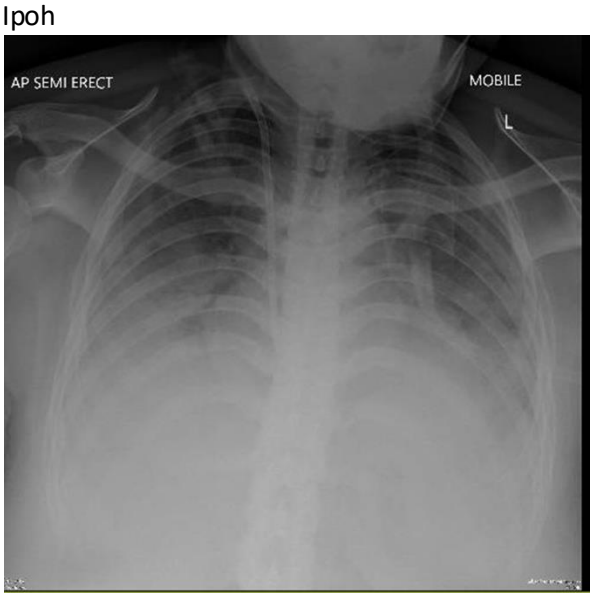
- Gradual clinical improvement after cyclophosphamide and rituximab
- Successfully extubated on Day 32

Complication:

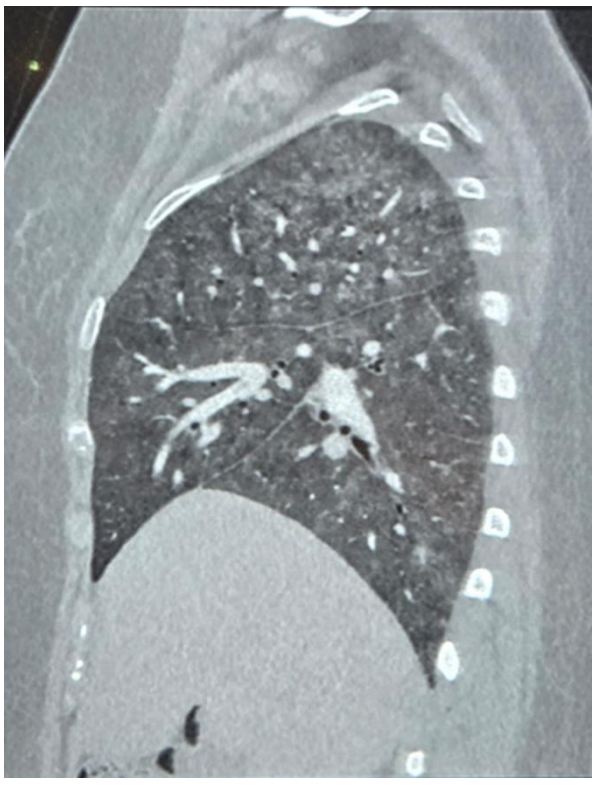
- Developed ventilator-associated pneumonia
  - CRP 58 mg/L
  - Endotracheal aspirate: *Achromobacter xylosoxidans*, sensitive to ceftazidime

Day 44 – New PE Identified:

- Repeat CTPA revealed new PE in a subsegmental branch of right middle lobe
- Likely due to **prolonged subtherapeutic anticoagulation**
- Anticoagulation cautiously resumed with LMWH (clexane)
- No recurrence of hemoptysis after reinitiation



1. Chest X-ray during hemoptysis: diffuse bilateral airspace opacities, predominantly in the mid-to-lower lung zones



2. CTPA: small filling defect at branch of right descending pulmonary artery, with background of diffuse ground glass opacities and patchy consolidations

DISCUSSION

DAH in SLE is driven by immune-mediated pulmonary capillaritis. Risk factors include:

- High SLE activity (SLEDAI >10)
- Elevated anti-dsDNA
- Hypocomplementemia
- Hematologic abnormalities (e.g. thrombocytopenia)

This case highlights:

- **Concurrent lupus nephritis and DAH**, suggesting severe systemic immune activation and endothelial injury.
- **Therapeutic dilemma**: managing confirmed thrombosis (DVT/PE) in the setting of active alveolar hemorrhage.
- **Bridging strategy**: An IVC filter was inserted while anticoagulation was withheld, reducing thromboembolic risk.
- **Aggressive immunosuppression** with corticosteroids, PLEX, IVIG, cyclophosphamide, and rituximab led to clinical improvement.

In refractory cases, **recombinant activated factor VII (rFVIIa)** has been reported to arrest alveolar bleeding by enhancing local hemostasis, though it was not used in this patient. It remains a potential adjunct in severe, non-responsive DAH.

CONCLUSION

This case illustrates the complex overlap of thrombosis and hemorrhage in SLE. A multidisciplinary, individualized approach was critical, balancing immunosuppression with thromboprophylaxis to achieve recovery.

REFERENCES

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