

Rare but deadly manifestation of systemic lupus erythematosus

Sebastian Alejandro Mikulic ¹, Michael Chahin ¹, Sukhraj Singh,² Myint Thway²

¹Internal Medicine, University of Florida College of Medicine, Jacksonville, Florida, USA

²Department of Medicine Division of Rheumatology, UF Health Jacksonville, Jacksonville, Florida, USA

Correspondence to

Dr Sebastian Alejandro Mikulic; sebastian.mikulic@jax.ufl.edu

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DESCRIPTION

A 26-year-old African-American woman with systemic lupus erythematosus (SLE) and end-stage renal disease due to lupus nephritis was sent to the hospital from the dialysis centre following oxygen saturation of 73% and fever of 101°F. The patient reported worsening dyspnoea and fatigue for 3 days, which led her to miss her routine dialysis session the day before hospitalisation. The patient has a complex history of SLE manifesting as diffuse cutaneous manifestations, lupus nephritis classes II and V, and autoimmune haemolytic anaemia. She was being treated with prednisone and hydroxychloroquine, but mycophenolate mofetil was discontinued 4 weeks prior due to methicillin-susceptible *Staphylococcus aureus* bacteraemia.

On examination, she was in moderate respiratory distress requiring supplemental oxygen. The patient's haemoglobin was noted to be 72 g/L (baseline 80 g/L) and platelet count 89 thou/cumm (baseline around 160 thou/cumm). Lupus activity markers showed low complement (C3) and elevated double-stranded DNA. Chest X-ray revealed multifocal patchy airspace opacities in bilateral lung fields (figure 1). Her hypoxia had minimal improvement with haemodialysis. Our suspicion for pneumocystis pneumonia was high, given her evolving clinical picture, elevated lactate dehydrogenase and increased arterial-alveolar gradient. We proceeded with bronchoscopy and bronchoalveolar lavage (BAL) revealed serial aliquots of serosanguinous saline indicative of diffuse alveolar haemorrhage (DAH). The patient was immediately started on pulse doses of intravenous methylprednisolone

500mg daily followed by rituximab 375 mg/m² induction therapy. Cyclophosphamide was not chosen given her recent infection, haemodialysis status, and young age. Her respiratory status improved dramatically, and follow-up chest X-ray revealed improvement in the diffuse patchy airspace opacities.

DAH is an uncommon but life-threatening complication of SLE. It is reported in the literature to occur in 1.0%–5.4% of cases with mortality ranging anywhere from 50% to 80%.^{1,2} The hallmarks of the disease include cough, dyspnoea and haemoptysis, with diffuse lung infiltrates on chest imaging and increasing red blood cells on serial aliquots on BAL. The most commonly associated organ involved in patients with SLE complicated by DAH is nephritis.³ In an effort to identify early predictors of DAH development, multivariate analyses found a history of thrombocytopenia and low C3 in one study¹ and coexisting neuropsychiatric lupus and high Systemic Lupus Erythematosus Disease Activity Index in another study⁴ as strong predictors for developing DAH. High doses of intravenous glucocorticoids is the cornerstone of therapy, along with additional immunosuppressive agents to sustain remission.³ Traditionally, cyclophosphamide has been the agent of choice.⁵ During the acute phase of DAH, immediate hemostasis can be achieved with bronchoscopic administration of intrapulmonary recombinant factor VIIa.⁶ Also in cases of refractory hypoxaemia, supportive care with extracorporeal membrane oxygenation has been described.⁷ Recent advances in our understanding of pathogenic mechanisms driving the development of DAH highlight the pivotal role played by B lymphocytes and humoral immunity.⁸ Thus, rituximab (anti-CD20 monoclonal antibody) has been applied successfully in multiple case reports⁹ and in our case as well. This case illustrates a textbook example of how DAH can present, the classic risk factors that serve to increase suspicion and use of emerging immunosuppressive options.

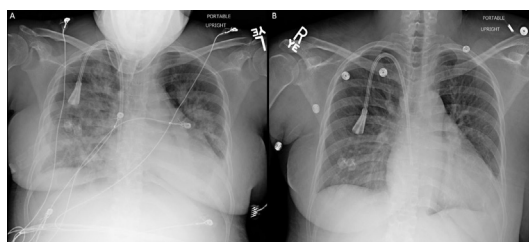


Figure 1 (A) Portable AP chest X-ray showing multifocal patchy airspace opacities through bilateral lung fields, a right lower lung opacity and a right tunneled catheter. This film was taken after the patient underwent urgent haemodialysis on presentation. (B) Portable AP chest X-ray showing marked improvement in the bilateral patchy opacities with only mild interstitial prominence remaining. This film was taken after receiving methylprednisolone and rituximab. AP, anteroposterior.

Patient's perspective

I was aware of my systemic lupus erythematosus diagnosis for multiple years; however, I was surprised to learn how deadly the disease could be once it started to affect my lungs. This has been a very frightening experience that has taught me a lot about my condition. I am very fortunate to have recovered, and I feel extremely relieved to have the opportunity to go back home to my family.



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Learning points

- Diffuse alveolar haemorrhage (DAH) is a life-threatening complication of systemic lupus erythematosus that classically presents with shortness of breath, hemoptysis and bilateral alveolar infiltrates.
- Early bronchoscopy is indicated in patients who are suspected of having DAH as it is used to aid in the diagnosis and rule out infection. A rising red blood cell count in sequential bronchoalveolar lavage aliquots from the same location is considered diagnostic of DAH.
- Corticosteroids and immunosuppressive agents remain the gold standard for the treatment of DAH.

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ORCID iDs

Sebastian Alejandro Mikulic <http://orcid.org/0000-0003-4518-2584>

Michael Chahin <http://orcid.org/0000-0001-9371-0252>

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