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Double trouble: Diagnostic and treatment challenges in overlap syndrome (systemic sclerosis and ANCA-associated vasculitis)

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INTRODUCTION

ANCA-associated vasculitis(AAV) can occur in systemic sclerosis (SSc) with a reported prevalence of 5.9-8%. Understanding the correlation between SSc and AAV is crucial for improving diagnosis, management and treatment strategies as both disorders can complicate clinical outcomes.

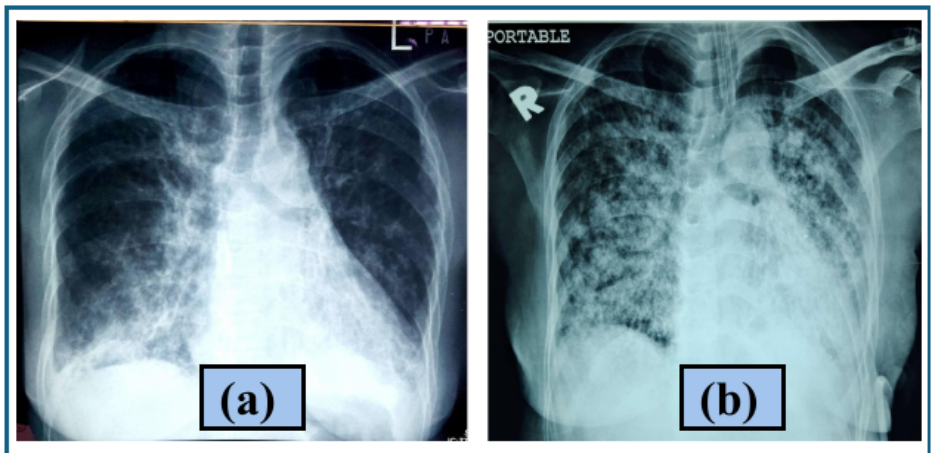
REPORT

A 43-year-old woman with systemic sclerosis sine scleroderma (ssSSc) met ACR/EULAR criteria based on the presence of Raynaud's phenomenon, telangiectasia, positive antinuclear antibody (ANA), Scl-70 autoantibodies and interstitial lung disease (ILD). She received six cycles of intravenous cyclophosphamide (CYC) for ILD, specifically the non-specific interstitial pneumonitis (NSIP) type in 2021, followed by maintenance with mycophenolate mofetil (MMF).

Fourteen months after completing IV CYC, she experienced a rapid decline in renal function evidenced by proteinuria, hematuria, and an increase in creatinine from 101 to 1437 mmol/l, noted during a routine clinic follow-up. She was diagnosed as having missed normotensive scleroderma renal crisis, and tablet captopril 12.5mg thrice daily was initiated. However, she required regular continuous ambulatory peritoneal dialysis (CAPD). Renal biopsy was not performed as the ultrasound showed bilateral renal parenchymal disease.

She remained stable until January 2025, when she developed sudden shortness of breath and hemoptysis, requiring mechanical ventilation. Tracheal aspirate cultures, multiplex test, influenza, COVID, and acid-fast bacilli tests were negative, while positive MPO-pANCA serology led to a diagnosis of pulmonary hemorrhage due to AAV.

She underwent five cycles of plasma exchange (PLEX), followed by five doses of intravenous immunoglobulin (IVIG) 0.4g/kg. We could wean down her fraction of inspired oxygen (FiO2) from 0.8 to 0.45. However, her condition deteriorated when she developed a hospital-acquired infection, which ultimately led to her death.



(a)Chest Xray upon admission
(b)Chest Xray on Day 4 of admission showing diffuse bilateral alveolar infiltrates

DISCUSSION

Presence of hemoptysis, new infiltrates on chest imaging, drop in hemoglobin and positive ANCA serologies (especially MPO-ANCA) should prompt evaluation for pulmonary hemorrhage and possible coexisting AAV. Immunosuppressive regimens used in AAV, such as corticosteroids, cyclophosphamide or rituximab may be warranted. However, caution is needed in SSc patients, who are at higher risk for complications such as scleroderma renal crisis and infections.

CONCLUSION

This case report underscores the coexistence of AAV and SSc. The overlap of these two autoimmune disorders can complicate diagnosis and management. Thus, a more effective therapeutic approach can be devised with early recognition of the coexistence of AAV and SSc, potentially mitigating severe complications like pulmonary hemorrhage and renal failure.

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