MSR MALAYSIAN SOCIETY OF RHEUMATOLOGY
23RD MSR - SSR 2025 Workshop in Rheumatology
Singapore Society of Rheumatology

A RARE COMPLICATION OF SSc-ILD: BILATERAL SPONTENOUS PNEUMOTHORAX

NF Mohd Nazri [1] I Ismail [1] M.Asrulshah [1] AF Md Salleh [1]
Mukhtar MA[2]
Medical Department, Hospital Sultan Abdul Halim[1]
Medical Department, Hospital Sik [2]

KAMI PRIHATIN
HOSPITAL SULTAN ABDUL HALIM

Introduction

Spontaneous bilateral pneumothorax is a rare complication in a patient with diffuse cutaneous systemic sclerosis and associated interstitial lung disease.

Case Report

A 54-year-old Malay lady with a 5-year history of dry cough and progressive skin tightening presented with worsening shortness of breath. Clinical evaluation revealed salt and pepper skin discoloration with significant weight loss. Initially denying skin changes. She later reported recent occurrence of Raynaud's phenomenonn. Examination confirmed diffuse cutaneous systemic sclerosis with Raynaud's, digital pitting, and sclerodactyly, with bilateral inspiratory fine crackles with reduced air entry, raising suspicion for interstitial lung disease (ILD).

The patient's serology was notable for a positive antinuclear antibody (ANA) and a negative anti-double-stranded DNA (dsDNA) antibody, along with a strongly positive anti-Scl-70 (DNA topoisomerase I) antibody. Imaging revealed significant pulmonary findings, where the chest radiograph showed bilateral pneumothorax. Further detailed assessment with HRCT thorax demonstrated bilateral pneumothorax, reduced lung volumes, widespread lung scarring characterized by fibrosis and bronchiectasis, and the presence of multiple air-filled sacs (bullae). Spirometry revealed restrictive pattern of lung function. Right video-assisted thoracoscopic surgery (VATS) was performed, and proceeded with bullectomy and pleurodesis.

Post-operatively patient was initiated with mycophenolate mofetil (MMF) and nifedipine, as well as plans for regular vaccinations and repeat imaging for follow up review and surveillance.

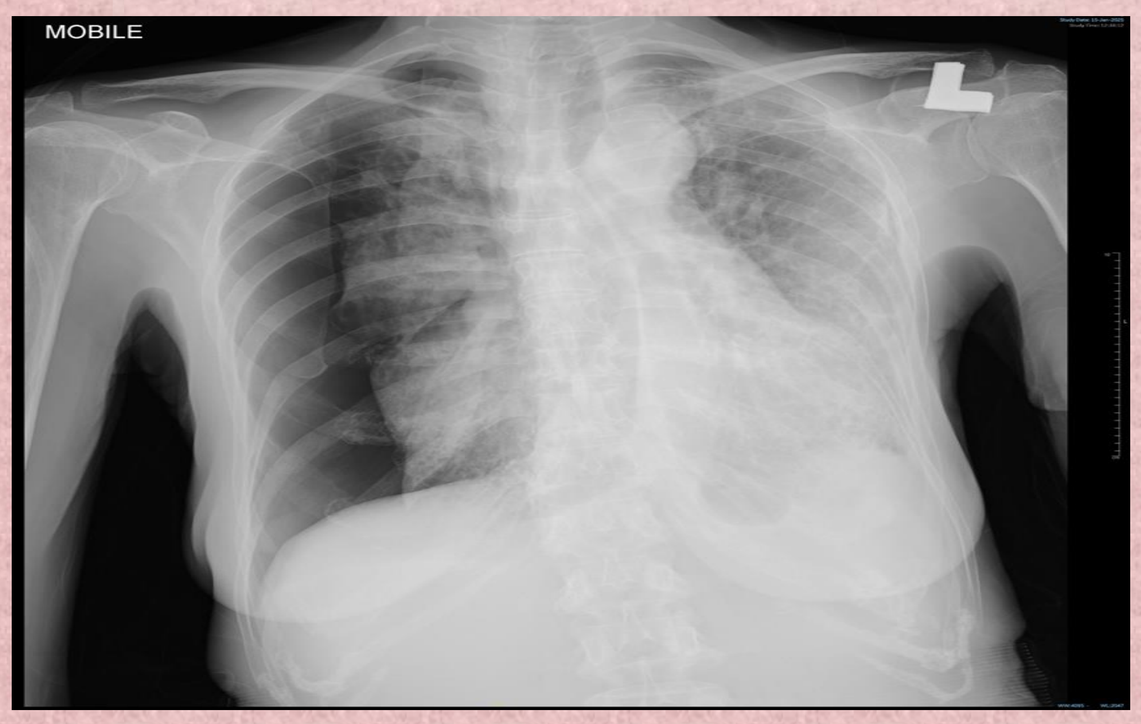

Figure1:Bilateral pneumothorax

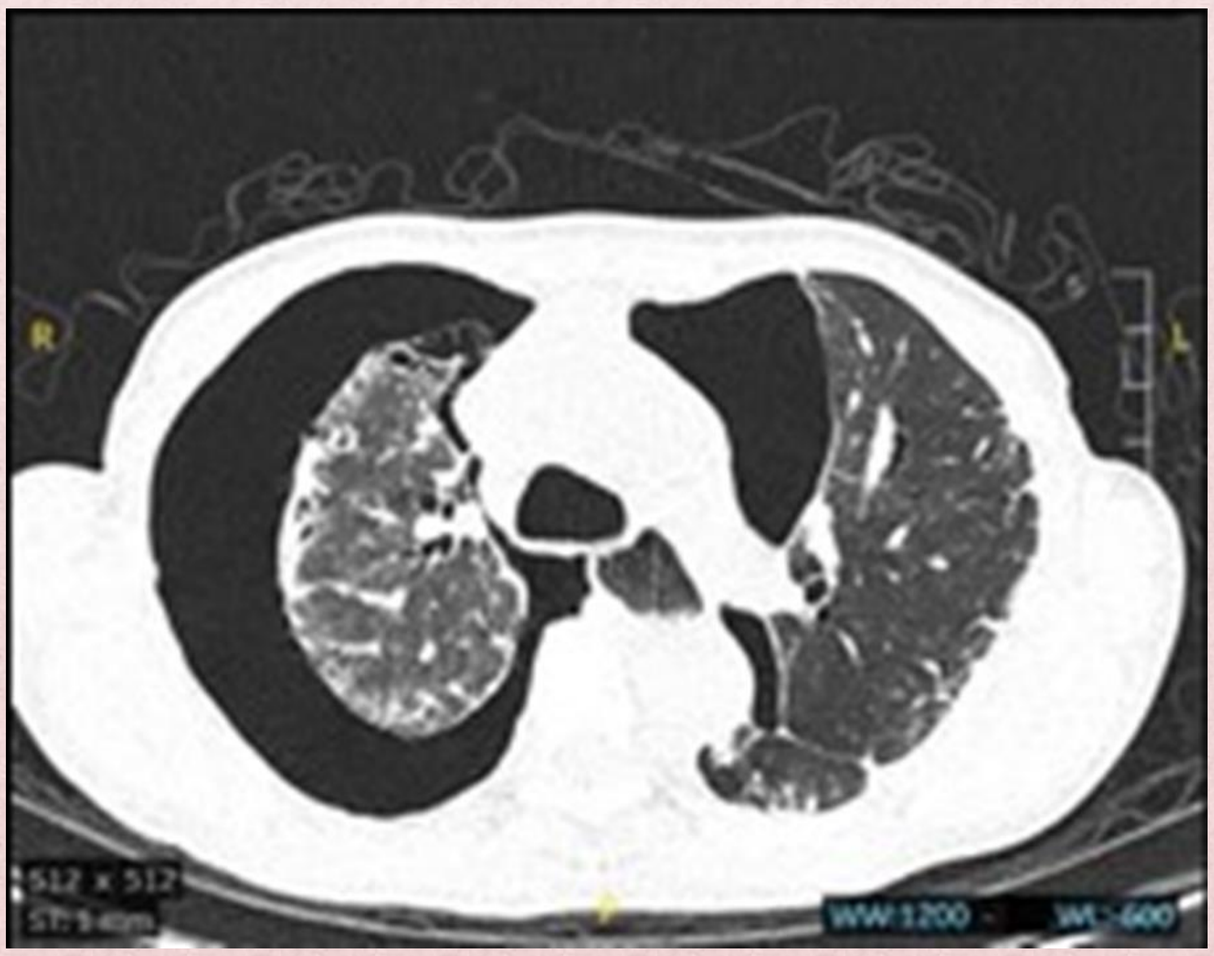

Figure 2:NSIP

Discussion

Spontaneous pneumothorax is an uncommon but serious lung issue in scleroderma, usually happening in cases of advanced lung scarring (pulmonary fibrosis) when cysts rupture. While a chest tube is the initial fix, it often comes back, needing more involved treatments like pleurodesis or partial lobectomy. For the underlying scleroderma lung disease (SSc-ILD), steroids aren't proven effective and can even cause a dangerous kidney problem called scleroderma renal crisis at high doses. Instead, mycophenolate mofetil (MMF) is a well-tolerated and better treatment. It helps slow down lung function decline (FVC) and improves survival compared to other medications.

Conclusion

Our case demonstrated a rare but important recognition of pneumothorax in scleroderma-associated ILD cases. We emphasize the necessity of clinicians being vigilant for pulmonary complications in this population, ensuring rapid diagnosis and tailored management strategies, which may involve surgical approaches for spontaneous pneumothorax. Ongoing follow-up is crucial to determine the long-term impact of treatment and the trajectory of the underlying disease processes.

Reference

1. *PLOS One (Nishimoto et al., 2020):* A study on connective tissue disease-associated ILD (CTD-ILD)
2. *Spandidos Publications (Kagohashi et al., 2014)*
3. *European Respiratory Society (ERS Publications):* Offers a review on scleroderma lung disease, mentioning pneumothorax in the context of ILD.