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A Recent Diagnosis Of Systemic Lupus Erythematosus In A Male With A History Of Recurrent Pneumothorax And Cystic Bronchiectasis: A Case Report

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ABSTRACT

Recently published case reports showed that Pneumothorax can be associated with active systemic lupus erythematosus (SLE). Herein, we present a smoker man with a positive history of connective tissue disease (CTD) in the family, who was admitted to the hospital many times during the previous year for recurrent chronic obstructive pulmonary disease (COPD) exacerbations and multiple chest infections. The patient had a history of recurrent spontaneous pneumothorax 20 years prior, which required treatment with pleurodesis. Furthermore, a year ago, he received a diagnosis of SLE. The association of pneumothorax with smoking and COPD is well known. Still, in our patient's case, he may have been predisposed to develop recurrent pneumothorax requiring pleurodesis because of his underlying undiagnosed and untreated active SLE disease.

INTRODUCTION

Pneumothorax is a serious condition that can potentially lead to severe complications such as respiratory failure, cardiovascular collapse, and death if not recognized and treated immediately. Primary spontaneous pneumothorax is primarily seen in individuals who are healthy and have no prior history of chest trauma or underlying lung conditions. Predominantly, it affects tall and thin males in the age range of 20 to 40 years. Secondary spontaneous pneumothorax affects individuals who already have lung disease, particularly emphysematous lung disorders; other causes include lung cancer, interstitial lung disease, pneumonia, asthma, cystic disorders, and pulmonary tuberculosis (Leys *et al.,* 2020; Nishimoto *et al.,* 2020). Spontaneous pneumothorax can be a complication that occurs in patients with connective tissue disease-associated interstitial lung disease (CTD-ILD) and predicts a poor prognosis. It is also reported in some studies to be a rare pleuropulmonary manifestation of active systemic lupus erythematosus (SLE). Studies associating SLE with pneumothorax are limited; therefore, the association between the two conditions remains unclear. However, all connective tissue disorders may theoretically be associated with a decrease in the integrity of the pleural membranes, which could lead to pneumothorax (Graves *et al.,* 2022a; Imad *et al.,* 2022; Sharma & Sharma, 2019). Bronchiectasis shares many clinical features with COPD, including inflamed and easily collapsible airways, airflow obstruction, and frequent office visits and hospitalizations. The diagnosis is established clinically based on cough on most days with tenacious sputum production, often one or more exacerbations/ year, and radiographically by the presence of bronchial airway dilatation on chest computed tomographic (CT) scans. Bronchiectasis has many causes including Cystic fibrosis, mycobacterial infections, Foreign bodies, airway compression, human immune deficiency virus (HIV) and hypogammaglobulinemia, Primary ciliary dyskinesia (PCD), Allergic bronchopulmonary aspergillosis (ABPA), Fibrosis of the lung tissue, Alpha-1 antitrypsin deficiency, Autoimmune or inflammatory disorders, like rheumatoid arthritis (RA), inflammatory bowel disease (IBD), lupus (SLE), and Sjögren's syndrome. This case report presents a Saudi male who developed a left-sided recurrent spontaneous pneumothorax which was treated with chest tube insertion twice then eventually pleurodesis. Subsequently, he was diagnosed with bilateral bronchiectasis and treated accordingly without significant improvement in clinical condition. However, 20 years later, the patient was diagnosed with SLE due to the presence of musculoskeletal symptoms and positive results of antinuclear antibody and anti-double-stranded DNA antibody tests.

Case Presentation

In March 2023, a 48-year-old male from Saudi Arabia, who works as a nurse, presented to the emergency department at King Faisal Hospital in Makkah City complaining of severe shortness of breath (SOB) for several hours of a one-day duration with no aggravating or relieving factors. His SOB was associated with cough and whitish sputum production with no fever, chest pain,

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or history of contact with sick patients. The patient, who was completely vaccinated against COVID-19, reported a history of repeated hospital admissions during the past year including intensive care unit admission due to community-acquired pneumonia with respiratory failure type 2. Previously he has been prescribed oral short courses of Prednisone (a steroid) in order to decrease the inflammation in the airways along with a combination of treatments including moxifloxacin (an antibiotic), inhalers such as salbutamol, fluticasone with salmeterol, tiotropium, and home BiPAP therapy for respiratory assistance but no significant improvement overall with the previously mentioned treatments. He reports a history of hookah smoking that had started 30 years ago; he had discontinued the activity over the past year. He also reported a history of CTD in the family, including a brother with Bechet disease and a history of left-sided recurrent spontaneous pneumothorax 20 years ago which was treated by chest tube insertion twice and eventually by pleurodesis. Examination revealed tachypnea with a respiratory rate of 28 bpm, O2 saturation of 90% on 1 liter of oxygen, and venous blood gases showing a pH (7.33), PCO_2 (74 mmHg), and HCO_3 (39 mmol/L). Upon chest auscultation, bilateral crepitations were noted, clearly accompanied by expiratory wheezes and decreased breath sounds specifically over the right lung. Also, the patient was noted to be slightly pale with apparent finger clubbing in both hands. The chest X-ray shown in (Figure 1) shows clear hyperinflation in both lungs along with consolidation and severe bronchiectasis on the right middle lung lobe. The impression of bronchiectasis exacerbation plus pneumonia was made and the patient was admitted to the medical ward under the pulmonology team for further treatment. Subsequently, a decision was made to refer the patient for Spiral computed tomography (CT) of the lungs to rule out pulmonary embolism, but the patient refused this procedure due to his fear of contrast administration.

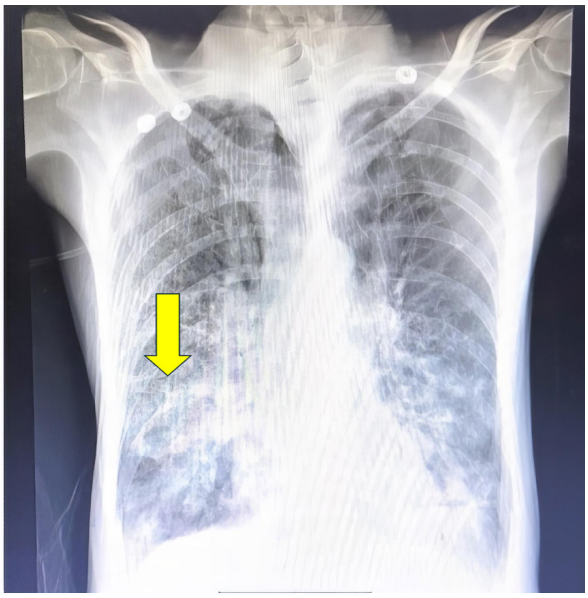

Figure 1: Chest X-ray showing hyperinflated chest with bilateral bronchiectasis

He agreed to have high-resolution computed tomography (HRCT) of the lungs shown in (Figure 2) which detected bilateral lung bronchiectasis predominantly in the right middle lobe and slightly less in the lower and upper lung lobes. Bronchiectasis is variable from varicoid to cystic. Air fluid levels are present within the cysts, which may represent mucus accumulation. Small areas of scarring are observed in the lung apices as well as some trees in bud nodularity. The mediastinal area exhibits multiple air cysts in the paratracheal and subcarinal areas around the main bronchi with no gross lymph node enlargement, no pleural effusion or pneumothorax, and no gross chest wall abnormality.

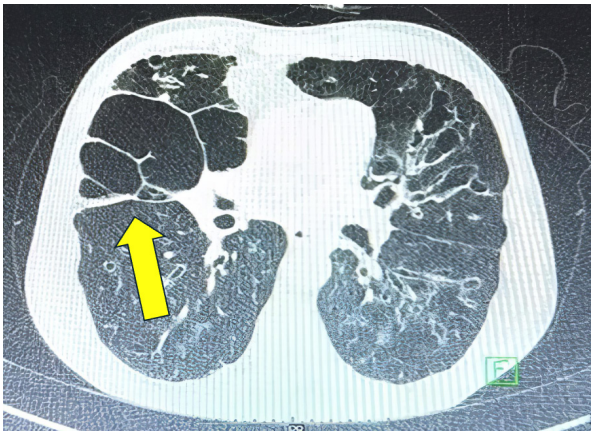

Figure 2: High-resolution computed tomography showing bilateral lung bronchiectasis predominantly in the right middle lobe.

The echocardiogram revealed that the left ventricular systolic function was within normal limits, with evidence of grade 1 diastolic dysfunction and the ejection fraction at 55%. The pulmonary artery systolic pressure (PASP) peaked at 50 mmHg. The left atrium was of normal size while the right atrium was a bit more enlarged compared to the other. No signs of atrial septal defect.

The blood test results were noted down at the time of admission at the hospital which is given in Table 1 in detail:

Table 1: Blood test results on admission

WBC: 6.11 x 109/L	(NORMAL)
Lymphocytes: 26.2 %	(NORMAL)
Monocytes: 15.5 %	(HIGH)
Neutrophil: 52.2 %	(NORMAL)
Eosinophils: 5.9 %	(UPPER LIMIT OF NORMAL)
Basophils: 0.2 %	(NORMAL)
Red blood cell count: 5.64 $X10^{12}/L$	(HIGH)
Hemoglobin: 10.4 g/dL	(LOW)
MCV: 72.9 fL	(LOW)
MCH: 19 pg	(LOW)
DAT: positive	

reticulocytes count: 0.1641	(HIGH)
Hematocrit: 39.8 %	(LOW)
RDW: 27.4%	(HIGH)
Platelets: 305 x 109/L	
Calcium: 2.43 mmol/L	
Phosphate: 1.04 mmol/L	
Na: 137 mEq/L	
Potassium: 5.4 mmol/L	
Total Protein: 81.2 g/L	
Albumin: 45 g/L	
Creatinine: 0.54 umol/L	
BUN: 9 mmol/L	
Vitamin D: 74 nmol/L	
CKI: 28.2 U/L	(LOW)
LDH: 417.8 U/L	(NORMAL)
ALT: 36 U/L	
AST: 27 U/L	
Total Bilirubin: 0.60 umol/L	
Direct Bilirubin: 0.2 umol/L	
INR: 1.1	
PT: 14.9 s	
PTT: 32.8 s	

WBC: White blood cell count; MCV: mean corpuscular volume; MCH: Mean corpuscular hemoglobin; RDW:red cell distribution width; CKI: Creatine Kinase with Isoenzymes; LDH: Lactate Dehydrogenase; AST: aspartate transaminase; ALT: alanine aminotransferase; PT: prothrombin time; PTT: partial thromboplastin time; BUN: blood urea nitrogen; INR: international normalized ratio.

Sputum culture revealed evidence of Pseudomonas aeruginosa growth and appropriate antibiotic treatment started. During his hospital stay, he was complaining of upper back pain and bilateral shoulder pain that had been bothering him over the last year but was responding to the previously described short courses of oral steroids (prednisone). Apart from a positive Coombs test and anemia, no other rheumatological disease manifestations were observed, so an impression of autoimmune hemolytic anemia was made. The rheumatology team consulted and a full autoimmune profile was requested (see Table 2).

The patient was diagnosed with SLE (based on positive ANA testing and anti-DS DNA) plus antiphospholipid syndrome with triple positivity of antiphospholipid antibodies. The management plan for the patient included oral prednisolone of 50 mg daily for 1 month, then tapered by 5 mg every 2 weeks, oral omeprazole of 40 mg daily, oral calcium of 600 mg thrice daily, oral Vitamin D of 50000 IU once weekly, oral bactrim of 960 mg thrice weekly, oral warfarin of 5 mg daily, oral hydroxychloroquine of 200 mg twice daily, oral mycophenolate mofetil of 1 gram twice daily, rituximab of one cycle every 6 months plus two IVIG doses of 1 g/kg, Sun-protection SPF of 50%, subcutaneous denosumab 60 of mg every 6 months, fluticasone/vilanterol of 200 micrograms inhalational once daily, tiotropium inhalational of 18 mcg once daily, Ventolin inhalation of 2 puff PRN, long term oxygen therapy, and daily home continuous positive airway pressure (C-PAP) from 12 midnight to 6 am. Additionally, he was referred to a transplant center for lung transplantation assessment. After initiation of the previously mentioned medications, the patient reports improvement in his overall symptoms

Table 2: The autoimmune profile of the patient, inflammatory markers, complement level, and immunoglobulins level

C4	13.8 mg/dL (low)
C3	103 mg/dL (normal)
IgG	1,000 mg/dL (normal)
IgA	318 mg/dL (normal)
IgM	102 mg/dL (normal)
ESR	62 mm/hour (high)
CRP	179.2 mg/L (high)
Anti-jo-1 antibody	4.28 negative
scleroderma Antibody	6.50 negative
Anti-beta 2 glycoprotein abs (IgG)	20.4 positive
Anti-beta 2 glycoprotein abs (IgM)	53.0 positive
Lupus anticoagulant	65.30 positive
Anti-cardiolipin IgM	3.58 negative
Anti-cardiolipin IgG	>100 positive
U1RNP antibodies	16.06 negative
SSB/La antibody	10.7 negative

ANA by IF	Titer = 1:80, with Nuclear Coarse Speckled pattern
anti-(DS)-DNA antibody	41.1 positive

ANA: antinuclear antibody testing; IF: immunofluorescence; C3 and C4: complement proteins; DAT: direct antiglobulin test; DS: double-stranded; CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; IgA: immunoglobulin A; IgG: immunoglobulin G; and IgM: immunoglobulin M; CRP: C-reactive protein.

and a reduction of repeated hospital admissions along with enhanced exertional dyspnea and an increased walking distance. The patient continued to follow up at the clinic with the pulmonology and rheumatology teams and currently staying in Riaydh city where he is undergoing assessments to establish his eligibility for the lung transplant.

DISCUSSION

Respiratory system involvement is common among SLE patients, it is estimated that up to 50% of patients will have lung involvement during the course of their disease which can involve the chest wall, pleura, airways, lung parenchyma, and pulmonary vasculature (Di Bartolomeo *et al.,* 2021). Sometimes it can mimic other aetiologies and be treated unproperly. In order of frequency, it includes pleuritis (40%–60%) pleural effusion (50%), acute pneumonitis (1%–12%), interstitial lung disease (3%–9%), shrinking lung syndrome (1%–6%), pulmonary hypertension (4%), diffuse alveolar hemorrhage, and thromboembolic disease (Amarnani *et al.,* 2021). Pneumothorax is rare to be encountered, yet some reports have mentioned a group of patients with synchronous pneumothorax and active SLE disease (Graves *et al.,* 2022b). Regarding bronchiectasis, it has been associated with rheumatoid arthritis (RA) and Sjögren's disease (SjD) more frequently than with SLE. Arthropathy and sicca features are usually advanced when bronchiectasis becomes apparent, yet in some cases, bronchiectasis occurs even before the rheumatic disease is diagnosed. The mechanism that lies behind the appearance of bronchiectasis in patients with autoimmune diseases is not well established but probable causes include the presence of abnormal cystic fibrosis gene (CFTR) allele in patients with RA, recurrent aspiration and traction in patients with interstitial lung disease, chronic inflammation with a weakened immune system, and multiple recurrent infections. All these mechanisms can contribute to the formation or worsening of a previously formed dilated airway. Bronchiectasis has also been noted in association with other systemic diseases, such as inflammatory bowel disease (IBD) and yellow nail syndrome (Jakharia *et al.,* 2022; Suarez-Cuartin *et al.,* 2016). A large Swedish population-based register study of lung disease among SLE patients showed that bronchiectasis occurs at a higher rate in patients with SLE compared to the general population (Forbess *et al.,* 2019). Another 10-year retrospective hospital-based study conducted at KingAbdulaziz University Hospital (KAUH) in Saudi Arabia by Prof. S. Attar and Prof. O. Alamoudi, involved 184 SLE patients (61 with pulmonary involvement and 52 with HRCT abnormalities), showed a significant association between abnormal HRCT and hypocomplementemia, high levels of anti-dsDNA, and disease activity while lupus nephritis and duration of SLE disease were not (Alamoudi & Attar, 2015). The patient in our case report had recurrent left-sided pneumothorax 20 years back secondary to underlying left lung apical lobe emphysematous changes; as written in his previously done Video-assisted thoracoscopic surgery (VATS) report; and bilateral cystic bronchiectasis, years following that event, he was diagnosed with active SLE. It is not clear whether remnant apical bullae or the presence of severe cystic bronchiectasis were solely responsible for the patient's recurrent pneumothorax. At that time, the possibility of an underlying CTD was not considered. Although smoking and bronchiectasis are all well-known causes of pneumothorax, we are suggesting that the presence of an undiagnosed SLE might have caused the patient to be more susceptible to recurrent pneumothorax and contributed to the formation of a more severe form of his cystic bronchiectasis with further worsening of the outcome for the affected organ, which is the lung tissue in this case.

CONCLUSION

Pulmonary system involvement in SLE is much wider than what is thought to be and can present in any shape with varying degrees of severity. The current 2019 European Alliance of Associations for Rheumatology (EULAR)/American College of Rheumatology (ACR) classification criteria for systemic lupus erythematosus include only pleural effusion, pericardial effusion, and acute pericarditis, therefore, a high level of suspicion is warranted when diagnosing unclassical SLE-related lung disease. Furthermore, it is crucial to screen for, diagnose, monitor, and treat all pulmonary symptoms in patients diagnosed with SLE, whether SLE is the cause or not, to prevent further deterioration and progression. We have to keep in mind that measures for SLE disease activity like SLEDAI and BILAG do not encompass the full spectrum of lung disorders encountered in SLE patients. Additional research is warranted to ascertain the optimal management strategies for pulmonary involvement in SLE. It is noteworthy that individuals with SLE who contract acute respiratory infections may experience a higher risk of developing structural lung problems such as pneumothorax and cystic changes. Physicians should be aware of this possibility and treat all patients with autoimmune diseases who present with active infections promptly to prevent further tissue damage.

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