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Case Report

A rare case report on rheumatoid arthritis with pleuro-parenchymal fibroelastosis

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ABSTRACT

Pleuroparenchymal fibroelastosis (PPFE) is a recently characterised type of interstitial lung disease that begins in the upper lung zones and extends to the entire lung. To date, the aetiology is unknown. However, the cause of PPFE in the current case report is rheumatoid arthritis. The symptoms are dyspnoea and dry cough. Pneumothorax is a common complication that occurs at the time of presentation or subsequently in the progression of the disease. It usually occurs in > 40% of ex-smokers with restrictive patterns in the pulmonary function test. Interstitial fibrosis appears pathologically as a thick consolidation in some preserved alveolar septal outlines and a markedly abrupt contact with the normal residual lung. Undiagnosed PPFE case scan be misdiagnosed as sarcoidosis, atypical idiopathic pulmonary fibrosis, or other unclassifiable interstitial pneumonia. The current case report will address the rare outline of Rheumatoid arthritis with PPFE.

Keywords: PPFE, Interstitial lung disease, Rheumatoid arthritis

INTRODUCTION

Rheumatoid arthritis (RA) is an autoimmune systemic inflammatory disease characterised by severe pain and inflammation in synovial joints. It leads to severe extra-articular symptoms that affect 1% of the Indian population. Extra-articular manifestations involve organs such as the skin, heart, lungs, and eyes.[1] The Lungs are primarily affected due to genetic mutation of protein citrullination. Though lung involvement in RA usually comes after articular signs, pulmonary symptoms could sometimes appear earlier than joint symptoms. In other terms, RA can affect any lung compartment, including the parenchyma, and induces interstitial lung disease (ILD) or rheumatoid nodules. The pleura can cause pleural inflammation or effusions and affects the small, large airways (cricoarytenoiditis, constrictive or follicular bronchiolitis, and bronchiectasis); and the pulmonary vasculature (vasculitis and pulmonary hypertension).[2] The most prominent and prevalent pulmonary manifestation of RA is ILD, a progressive fibrotic disease of the lung parenchyma that significantly increases morbidity and mortality.[3] Drug-related or viral precipitants can cause RA-ILD, or it can result from the chronic immunological stimulation and inflammatory processes that occur in RA, which promotes atypical fibroproliferation. Other extra-articular manifestations (EAMs), such as vasculitis, have declined over the past decade, although there has been an unexpected increase in the reported case of RA-ILD.

PPFE is a rare slow progressive disorder under the subtypes of ILD that causes pulmonary fibrosis, which is confined to the upper lobe of the lung. Dyspnoea or dry cough is the first symptom. However, chest discomfort occurs in some people due to pneumothorax. The typical histology suggests visceral pleural thickening with collagenous fibrosis, subpleural elastosis, and intra-alveolar collagenous fibrosis.[4] The patient in our case

study had no previous history or screening for RA but had a dry cough for a month with tremors and joint pain.

CASE REPORT

For ten days, a 65-year-old male retired teacher presented with dry cough, malaise, and anorexia. The patient had difficulty waking up in the morning and had pain in the small and large joints for 1.5 years and tremors for 20 years. He had lost around 10 kg weight within ten days. His vitals are stable, except SpO_2 was 94%. He was an ex-smoker who quit at the age of 30. The patient has been diagnosed with type 2 DM since January 2021. The patient had no history of COVID-19, hypertension, parkinsonism, or other comorbidities. He has been vaccinated with two doses of Covaxin. The patient was treated with tablet trihexyphenidyl 0.5 mg OD, tablet lornoxicam 8 mg BD, tablet serrato peptidase 10 mg BD, and tablet rasagiline 0.5 mg OD for one month. After a month, he still had the same complaints with anxiety-induced tachycardia. Then he was managed with tablet propranolol 10 mg TID, lornoxicam 8 mg and paracetamol 325 mg BD, and tablet oxazepam 10 mg HS. Basic laboratory investigations were normal. The patient was evaluated for further investigations such as ACE, anti-CCP, ANA, HRCT, RT-PCR, and ECG. ACE, anti-CCP showed positive results, and ANA showed negative values (Table 1).

Table 1: Observed values of ACE, Anti-CCP, ANA.

Parameters	Observed value	Normal range
Angiotensin-converting enzyme	76.32	8-68 U/l
Anti-CCP antibodies	>200	≤0.5
Anti-nuclear antibody	0.26	<0.8

High contrast computed tomogram showed multifocal confluent areas of interlobular septal thickening. The fibrotic bands were associated with tractional bronchiectasis with a honey-combing appearance and architectural distortion withvolume loss in both lungs. However,prominently in upper lobes and bilateral apical pleural thickening (Figure 1 and 2).

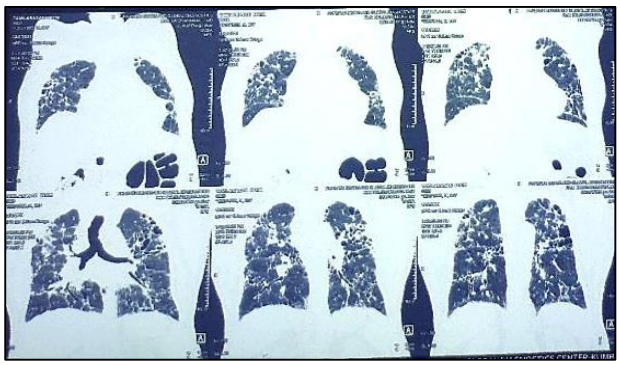

Figure 1: Coronal view of HRCT-bilateral apical thickening and traction bronchiectasis and PPFE.

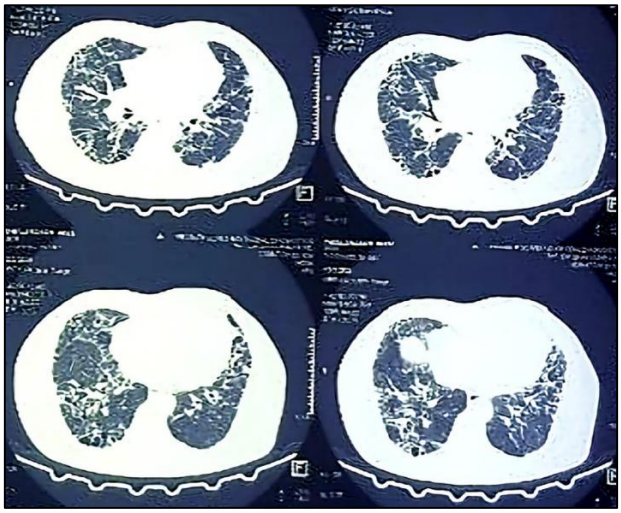

Figure 2: Axial view of HRCT-interlobular septal thickening and honey-combing appearance.

An electrocardiogram showed right ventricular hypertrophy. RT-PCR showed negative results for covid. On spirometry, forced vital capacity (FVC) was 40%, forced expiratory volume in one second (FEV_1) was 45%, and FEV_1/FVC ratio was 113% showing a restrictive pattern. The ultimate diagnosis was reactive rheumatoid arthritis with interstitial lung disease. It was associated with a usual interstitial pneumonitis atypical pattern with PPFE.

Additionally, a fraction of exhaled nitric oxide level of 6 implies that eosinophilic inflammation is unlikely, and therefore ICS treatment will be ineffective. The patient was then treated with tablet hydroxychloroquine 200 mg HS, tablet acetylcystine, acebrofylline ½ tablet BD, and neb. formoterol (6 µg+25 µg) glycopyrrolate 2 puffs BD.

DISCUSSION

Idiopathic PPFE is a rare clinicopathological condition with specific radiological and histological features and was first identified in 2004.The American thoracic society/ European respiratory society classification of idiopathic interstitial pneumonia was updated in 2013 that includes PPFE.

PPFE is now defined as a rare idiopathic interstitial pneumonia with a distinct clinical-pathological entity marked by extensive subpleural consolidation and traction bronchiectasis, architectural distortion, and upper lobe volume loss on chest CT, with a histological appearance of elastotic fibrosis and intra-alveolar fibrosis.[5] Symptoms include exertional dyspnoea, a dry cough, and recurrent respiratory tract infections.[6] Although the aetiology is uncertain, recurring pulmonary infections, autoimmune diseases, and genetic predisposition appear to be associated.[7] Cigarette smoking may influence PPFE, even though more than 40% of PPFE patients were ex-smokers. Patients had a restrictive pattern from the data exhibited,

with FVC% around 69 and FEV1% around 73.[8] Upper and middle lobe pleural thickening and subpleural fibrosis are seen on imaging, with no lower lobe involvement. There was honeycombing, traction bronchiectasis, and reticular anomalies. Histology revealed thickened visceral pleura and subpleural fibrosis composed of dense collagen and elastin (fibroelastosis). The transition between diseased and normal parenchyma is abrupt. Fibroblast foci and lymphocytic inflammation are seen in varying degrees.[9]

The characteristic hallmarks include radiographic Pleuroparenchymal anomalies, particularly in the upper lobe. The histological observations revealed alterations with a unique fibroelastosis, including the lung's peripheral/subpleural areas. In 30% of patients, pneumothorax, whether spontaneous or iatrogenic, worsens the condition of the disease. There have been reports of spontaneous pneumothorax and pneumomediastinum.[10] There is a substantial proportion of non-smokers, with fewer ex- and current smokers. The clinical result of PPFE is varied, with a considerable proportion of patients showing gradual deterioration and mortality. The stage of the disease determines the features of survival at the time of presentation. There is no proven effective therapy other than lung transplantation.

Interstitial lung disease is a group of chronic diseases characterised by inflammation and fibrosis, affecting gas exchange arduous. Pathogenesis may differ depending on the etiological variables, including connective tissue abnormalities, genetic disorders, occupational or environmental exposure, vasculitis, certain drugs, and idiopathic ILD.[11] Amiodarone, nonsteroidal anti-inflammatory medications, chemotherapeutic agents, colony-stimulating factors, interferon, anti-thymocyte globulin, intravenous immunoglobulin, and immunosuppressive treatments (methotrexate and cyclophosphamide) are all linked to ILD. This patient has no prior history of using any of these medications. Nevertheless, the patient had a rare case of RA with PPFE.

CONCLUSION

Pleuropulmonary fibroelastosis affects 11.8% of RA patients. The symptoms of PPFE were varied, but the most common ones were a non-productive cough and exertional dyspnoea. The CT scan confirms the diagnosis.

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