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**Clinical profile of pulmonary involvement in rheumatological disorders:
a cross-sectional study in a rheumatology-associated lung disease clinic**

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Abstract

Rheumatological diseases (RDs) are characterized by autoimmune-mediated organ damage, with the lungs being an important target organ. Almost all compartments of the pulmonary system may be involved, with considerable heterogeneity in the frequency, severity, and spectrum of pulmonary manifestations. This heterogeneity may result in delayed diagnosis and treatment, leading to adverse outcomes. The objective of this study was to characterize the clinical, demographic, serological, and radiological features of pulmonary manifestations across a spectrum of rheumatic diseases.

This cross-sectional study was conducted between 2016 and 2023 at a tertiary care center in New Delhi, India. Patients aged ≥ 18 years with a diagnosed or suspected RD and pulmonary manifestations were included. Patients who did not fulfill the American College of Rheumatology/European League against Rheumatism classification criteria for RD or the American Thoracic Society/European respiratory Society criteria for interstitial pneumonia with autoimmune features (IPAF), as well as those with chronic hypersensitivity pneumonitis or sequelae of previous pulmonary infections, were excluded. Of the 783 patients screened, 170 were diagnosed with rheumatology-associated lung disease (RALD) or IPAF. The mean age was 44.3 years, with a marked female predominance (85%). Rheumatoid arthritis (RA) and systemic sclerosis (SSc) accounted for 70% of cases, followed by IPAF, overlap syndromes, Sjögren syndrome (SjS), systemic lupus erythematosus (SLE), and idiopathic inflammatory myositis. Twenty-six patients with idiopathic interstitial pneumonia were reclassified as having IPAF, one of whom subsequently progressed to RA. Pulmonary symptoms preceded the diagnosis of RD in 13.6% of patients, occurred concurrently with the diagnosis in 17%, and developed during the course of RD in 69.4%. Antinuclear antibodies and rheumatoid factor were the most commonly detected autoantibodies, while anti-aminoacyl-tRNA synthetase antibodies were identified in 11 patients.

RA demonstrated the broadest spectrum of pulmonary manifestations. Interstitial lung disease was the predominant pulmonary manifestation across all RDs except SLE. Non-specific interstitial pneumonia and usual interstitial pneumonia patterns accounted for 87% of cases. Other pulmonary manifestations included infections in RA and SLE; vascular and pleural involvement in RA, SSc, and SLE; and airway involvement in RA and SjS. Pulmonary manifestations may precede, coincide with, or develop after the onset of systemic features in RDs. Comprehensive pulmonary evaluation and collaborative care involving rheumatologists and pulmonologists may facilitate earlier diagnosis and optimize the management of RALDs.

Key words: connective tissue diseases, interstitial lung disease, systemic autoimmune rheumatic diseases.

Introduction

Background and rationale

Rheumatological diseases (RDs) are characterised by autoimmune-mediated organ damage, and the lungs are an important target. Almost all components of the pulmonary system may be involved. There is a significant heterogeneity in the frequency, severity, and spectrum of pulmonary manifestations.

Patients may initially present to either a rheumatologist or a pulmonologist. Each specialist may have limited experience in the other's area of expertise. Consequently, rheumatology-associated lung diseases (RALDs) pose diagnostic and therapeutic challenges. Patients with RALDs often receive fragmented care, with rheumatologists and pulmonologists assessing them independently.

To address this gap, we developed a collaborative care model in which patients with suspected RALDs were jointly evaluated by a multidisciplinary team of pulmonologists and rheumatologists. In this cross-sectional study, we describe our experience with this collaborative clinic model.

Objective

The objective of this study was to provide insights into the clinical, demographic, serological, and radiological characteristics, including HRCT patterns, of pulmonary manifestations across a spectrum of rheumatic diseases, thereby helping to clarify their diagnostic relevance.

Methods

The Institutional Ethics Committee approved the study (TP (MD/MS) (52/2016) /IEC/PGIMER/RMLH 7786/16), and written informed consent was obtained from all participants.

The study was reported in accordance with the Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) guidelines and was conducted in accordance with the Declaration of Helsinki.

Study design and setting

This was a cross-sectional study conducted between 2016 and 2023. The study included patients referred to the RALD clinic in a tertiary care centre in New Delhi, India. Patients with a diagnoses or suspicion of RD attending the Rheumatology clinic were referred to the

RALD clinic if they had pulmonary symptoms. Patients attending the Respiratory clinic were referred to the RALD clinic if they had symptoms or signs of an RD as determined by an attending pulmonologist.

Participants

Patients aged 18 years or older with a diagnosed or suspected RD and pulmonary symptoms were referred to the RALD clinic. Patients who fulfilled the American College of Rheumatology/European League against Rheumatism (ACR/EULAR) classification criteria for RD or the European Respiratory society/American Thoracic Society (ERS/ATS) research criteria for interstitial pneumonia with autoimmune features (IPAF) were included in the study. Patients with chronic hypersensitivity pneumonitis or old infective sequelae, including tuberculosis, were excluded.

Data collection and variables

All patients underwent detailed history-taking, clinical examination, autoantibody profiling, high-resolution computed tomography (HRCT), spirometry, and echocardiography. The six-minute walk test and diffusion capacity for carbon monoxide (DLCO) were performed whenever feasible. Demographic, clinical, serological, and radiological data were collected. A myositis antibody profile was incorporated into the serological evaluation from 2019 onwards.

Definitions

Interstitial Pneumonia with Autoimmune Features (IPAF) comprises a subgroup of idiopathic interstitial pneumonias (IIPs) characterised by clinical and/or serological evidence suggestive of RDs. IPAF was diagnosed according to the 2015 ERS/ATS research criteria, based on the presence of features from the clinical, serological, and morphological domains in patients with radiological evidence of interstitial pneumonia, after exclusion of alternative causes of interstitial pneumonia and in the absence of a defined rheumatic disease.

Overlap syndrome included patients who fulfilled the classification criteria for more than one defined rheumatological condition.

Mixed Connective Tissue Disease (MCTD), a subset of OS, was defined by overlapping features of at least two connective tissue diseases, most commonly SLE, SSc, and

polymyositis/dermatomyositis, together with high titres of anti-U1 ribonucleoprotein (anti-U1RNP) antibodies.

Statistical analysis

Based on a study by Palmucci et al. [1], the reported prevalence of pulmonary involvement across various rheumatological diseases ranges from 9% to 80%. Assuming that 45% of patients with rheumatic diseases would have pulmonary involvement, a sample size of 170 participants was calculated to estimate the expected proportion with an absolute precision of 7.5% at a 95% confidence level. Sequential screening was undertaken until 170 eligible participants had been enrolled. Results are presented as the mean, number, or percentage, as appropriate.

Results

A total of 783 adult patients were screened. ACR/EULAR classification criteria for an RD were not met in 298 patients; 281 had sequelae of previous pulmonary infection on HRCT, and 34 had chronic hypersensitivity pneumonitis. All of these patients were excluded. Consequently, 170 patients fulfilled the inclusion criteria and were enrolled in the study (*Supplementary Figure 1*).

Demographics and disease spectrum

The mean age was 44.3 ± 16.0 years (range, 22–82 years), with a marked female predominance (84.7%). RA and SSc were the most common rheumatic diseases, accounting for 41.2% and 29.0% of cases, respectively. The remaining diagnoses included Overlap Syndrome (OS), Systemic Lupus Erythematosus (SLE), Sjögren Syndrome (SjS), and Idiopathic Inflammatory Myopathy (IIM). Baseline demographic and clinical characteristics are summarised in *Supplementary Table 1*.

Clinical and serological profile

Most patients (69.4%) developed respiratory symptoms after the onset of RD-related systemic manifestations. Respiratory symptoms preceded systemic manifestations in 13.6% of patients and occurred concurrently with systemic manifestations in 17.0%. In most patients with IPAF, respiratory symptoms heralded disease onset.

Exertional breathlessness and cough were the most common respiratory symptoms. Common extrapulmonary manifestations included polyarthritis/polyarthralgia, skin thickening, oral ulcers, Raynaud's phenomenon, gastro-oesophageal reflux, photosensitivity, digital ulcers, sicca symptoms, and muscle weakness. Certain findings, including mechanic's hands, acral osteolysis, reduced oral aperture, and malar rash, were disease-specific (*Supplementary Table 2*).

Among patients with MCTD, four had overlapping features of SLE, SSc, and SjS, whereas the remaining three had features of SLE, SSc, and RA.

Autoantibody profile

Antinuclear antibodies (ANA), rheumatoid factor (RF), anti-Ro, and anti-La antibodies were the most frequently detected autoantibodies. Anti-U1 ribonucleoprotein (U1 RNP) antibodies, considered a hallmark of MCTD, were also detected in several patients with SSc. Myositis-specific antibodies included anti-Jo-1, anti-PL-12, anti-PL-7, and anti-EJ.

The autoantibody profile was particularly diverse among patients with SSc, IPAF, and OS.

Amino-acyl tRNA synthetase (ARS) autoantibodies

ARS antibodies are a hallmark of Anti-synthetase syndrome (ASS). In patients with ILD, positive ARS autoantibodies fulfil the Connor et al. diagnostic criteria for ASS[2]. Eleven patients had positive ARS antibodies. The anti-ARS antibodies identified in this study included anti-Jo-1, anti-PL-7, anti-PL-12, and anti-EJ (*Supplementary Table 1*). These antibodies were detected in seven patients with IPAF, two with localised SSc, and one each with polymyositis and overlap syndrome. The clinical characteristics of these patients are summarised in *Supplementary Table 3*.

Pulmonary function evaluation

Pulmonary function tests were performed at baseline and during follow-up whenever feasible. However, many patients were unable to undergo pulmonary function testing, most of whom had severe clinical disease (*Supplementary Table 1*). Spirometry was successfully performed in 98 patients (56.5%), of whom 85 (86.7%) demonstrated a restrictive ventilatory pattern. Among these patients, 39 had mild restriction (FVC 70–80%), 24 had moderate restriction (FVC 60–70%), and 22 had severe restriction (FVC < 60%). An obstructive pattern was observed in 8 cases, and 7 of them had FEF 25-75% below the lower limit of normal. All

of these seven patients had RA and demonstrated HRCT features of centrilobular nodules and mosaic attenuation consistent with air trapping. These patients were therefore classified as having small airway disease. Five patients (5.1%) had normal spirometry. DLCO measurements were available for only 53 patients because adequate respiratory effort could not be achieved in the remainder. A DLCO of <70% predicted was observed in 21 patients (39.6%). The six-minute walk test (6MWT) was performed in 61 patients (35.9%). Severe pulmonary or musculoskeletal involvement precluded completion of the 6MWT in the remaining patients. An oxygen desaturation of $\geq 4\%$ during the 6MWT was observed in 23 patients (37.7%).

Pulmonary system involvement and radiological profile

All components of the respiratory system were involved across the spectrum of rheumatological diseases (*Supplementary Table 4*), with Rheumatoid Arthritis (RA) demonstrating involvement of all pulmonary compartments. Interstitial Lung Disease (ILD) was the most common pulmonary manifestation, occurring in 90.6% of the RDs. A broad spectrum of HRCT patterns was observed, with non-specific interstitial pneumonia (NSIP) being the most predominant pattern (72.7%), followed by usual interstitial pneumonia (UIP) (14.3%). Other HRCT patterns included organising pneumonia (OP), lymphocytic interstitial pneumonia (LIP) and the hypersensitivity pneumonitis pattern (HP) (*Supplementary Table 4*).

RA exhibited the widest spectrum of HRCT abnormalities, encompassing all the aforementioned ILD patterns. In patients with HP-pattern ILD, HRCT revealed centrilobular nodules, ground-glass opacities, and mosaic attenuation, however, no history of antigen exposure or positive serum precipitins was identified. Besides RA, LIP (Figure 1) was observed in Sjögren syndrome (SjS) and IPAF, whereas OP was documented in IPAF. A notable case involved a patient with RA receiving methotrexate who subsequently developed OP (Figure 2a), which resolved following discontinuation of methotrexate. Leflunomide was subsequently initiated for the management of RA in this patient. A patient with diffuse systemic sclerosis (dSSc) demonstrated an NSIP–OP overlap pattern (Figure 2b).

Specific CT signs including the anterior upper lobe sign (Figure 3a) and the exuberant Honeycombing sign (Figure 3b) were observed in one patient with established diffuse systemic sclerosis.

Bronchiectasis was observed in patients with RA and SjS whereas small airway disease was identified exclusively in RA. Pleural involvement occurred in RA, SSc, and SLE with pleural

effusion representing the most common thoracic manifestation in SLE. Vascular complications included pulmonary hypertension (PH), pulmonary thromboembolism and diffuse alveolar haemorrhage. PH was observed in SSc, RA, and OS, always in association with ILD, and was most prevalent in SSc (40.8%), occurring in both diffuse and limited cutaneous subtypes. Both PH and pulmonary thromboembolism was seen in patients with SLE, while diffuse alveolar haemorrhage was seen in RA.

Extra-pulmonary thoracic manifestations, including oesophageal dilatation, were observed in 15 cases of SSc (8 diffuse SSc and 7 limited SSc; 30.6%) and in four patients with OS, three of whom had features consistent with MCTD.

Discussion

Pulmonary involvement is a major contributor to morbidity and mortality in RDs. RA (42%), SSc (29%), IPAF (16%), and OS (6.5%) were the most common RDs identified in this study. Although the mean age of the study population was 44.3 years, patients with IPAF were older with a mean age of 61.4 years. The mean age was also higher among the patients with RA-ILD (54.6 years). Previous reports have also indicated that the incidence of ILD in RA increases with disease duration and is most commonly recognised during the fifth and sixth decades of life [3]. In contrast, patients with SLE were younger (mean age, 33.6 years), reflecting the higher prevalence of SLE among individuals aged 20–40 years. Most patients were non-smokers, which may reflect the marked female preponderance of the cohort as well as regional cultural factors [4].

Pulmonary involvement in various RDs may occur as a manifestation of the underlying disease itself, secondary to infections, drug toxicity, or vasculitis [5]. In the present study, pulmonary manifestations preceded, accompanied, or followed the onset of systemic disease. More than two-thirds of the study population (69.4%) had an established RD before the onset of respiratory symptoms. Respiratory and systemic manifestations occurred simultaneously in 17% of patients and were observed in RA, SSc, SjS, OS and IPAF. In both of these clinical scenarios, establishing the diagnosis is generally straightforward.

In 13.6% of patients, ILD was the initial manifestation of disease. This finding is consistent with previous reports showing that approximately 19% of patients initially diagnosed with idiopathic interstitial pneumonia (IIP) subsequently evolve into a defined rheumatic disease [6]. Previous studies have also suggested that 15–20% of patients presenting with chronic ILD either have an occult or latent rheumatic disease or subsequently develop clinically

overt disease [7]. In our cohort, 26 patients who initially presented solely with pulmonary manifestations and were managed as having IIP were subsequently reclassified as IPAF following multidisciplinary evaluation at our clinic.

IPAF lies at the interface between idiopathic interstitial pneumonias (IIPs) and rheumatic disease-associated interstitial lung diseases (RD-ILDs). According to the 2015 ERS/ATS research criteria, IPAF is defined as interstitial pneumonia in patients who do not have an established connective tissue disease or an alternative cause for the interstitial pneumonia. These patients should have at least one feature from two of the three domains of the proposed criteria viz. clinical, serological, and morphological to be diagnosed as IPAF [8]. Respiratory symptoms were the initial manifestation in 61.5% of patients with IPAF, while the remaining developed respiratory and systemic manifestations concurrently.

These findings highlight the diagnostic challenge encountered when pulmonary manifestations represent the initial or sole presentation of a rheumatic disease. This emphasises the importance of a comprehensive evaluation for missing rheumatic disease in all patients with IIP. In such cases, careful assessment of systemic features, serological findings, and HRCT abnormalities may facilitate an accurate diagnosis.

Clinical features

The most common extra pulmonary manifestations and red flags for Rheumatic disease observed in our cohort were consistent with those included in the ACR/EULAR classification criteria for the respective rheumatic diseases. When present, these findings strongly suggest an underlying rheumatic disease. Raynaud's phenomenon, inflammatory arthritis, myalgia, proximal muscle weakness, unexplained digital ulcers, sicca symptoms, and mechanic's hands prompted referral among patients with IIP. GERD is a recognised risk factors for both the development and progression of ILD and should therefore be actively evaluated and managed [9]. Additional clinical features, including mechanic's hands, heliotrope rash, the shawl sign, the V sign, Gottron's sign, and nailfold capillaroscopic abnormalities, may provide valuable diagnostic clues, particularly in patients with antisynthetase syndrome, amyopathic dermatomyositis, and scleroderma sine scleroderma presenting with ILD [10,11].

Autoantibody profile

Autoantibodies may be detected even in the absence of clinical features of a rheumatic disease. Antinuclear antibodies (ANA), rheumatoid factor (RF), anti-cyclic citrullinated

peptide (anti-CCP) antibodies, anti-Ro/La antibodies, anti-topoisomerase antibodies, and anti-aminoacyl-tRNA synthetase antibodies are included within the serological domain of the 2015 ERS/ATS IPAF classification criteria and, when present, should prompt a comprehensive evaluation for an underlying rheumatic disease [8]. Our findings highlight the value of comprehensive autoantibody testing, including a myositis antibody panel, in refining the diagnosis of both IPAF and established rheumatic diseases [12].

Radiological findings

ILD was the predominant pulmonary manifestation in our cohort. Many patients with RDs have subclinical interstitial abnormalities diagnosed incidentally on HRCT chest [13]. Any radiologic pattern of interstitial pneumonia can be seen in RD associated ILD and is similar to their idiopathic counterparts, as the response to injury has limited patterns [14]. Non-specific interstitial pneumonia (NSIP) was the most common HRCT pattern, followed by usual interstitial pneumonia (UIP), consistent with previous reports [10,11].

OP pattern can be due to a primary autoimmune process or secondary to drugs or infections and is common in RA with high titres of RF [15]. OP is rare in primary SjS, SLE and SSc [16]. LIP is a relatively rare pattern of ILD in RDs and is most commonly associated with SjS [17]. Certain HRCT signs such as straight-edge sign, anterior upper lobe fibrosis, exuberant honeycomb sign and four-corner sign in ILD may act as radiological clues to RD-ILD [18,19]. These signs, when present, may guide further workup in cases of IIPs or IPAF.

Airway involvement was seen in RA and Sjögren's syndrome. Tram-track sign, signet ring, thickened irregular airway walls, and lack of normal peripheral tapering on HRCT chest are characteristics of bronchiectasis [20]. Bronchiolitis is characterised by inflammation and fibrosis of bronchioles and is characterised by HRCT findings of mosaic attenuation due to air trapping, bronchial wall thickening and centrilobular nodules [21].

PH was defined radiologically as the main pulmonary artery to ascending aorta ratio greater than 1.0 and was noted in 17.1 % of cases. Pulmonary artery dilatation on CT has been shown to be better than the echocardiographic finding of an elevated right ventricular systolic pressure to detect PAH in a prior study [22].

Pleural effusions were common in SLE (4 out of 7 cases), RA and SSc. Serositis, rheumatoid nodules, infections, and pulmonary infarction can cause pleural effusion. Pleural pathology is seen in 5-20% and 17-60% cases of RA and SLE, respectively, in the literature [23].

Increased oesophageal diameter on HRCT in SSc patients is reported to be associated with severe radiologic ILD and lower lung volumes [24].

On pulmonary function testing, a restrictive pattern with DLCO reduction was typical in ILD. Obstructive pattern was seen in few RA patients. Radio-imaging evidence of small airway disease (SAD) in ILD act as radiological clues to RD-ILD. SAD on CT prompted us to look for mid expiratory flow rate (FEF25–75%) on PFT. These values, when less than the lower limit of normal, provided corroborative evidence of SAD [25].

Conclusions

Pulmonary involvement may precede systemic manifestations in patients with various rheumatic diseases. Patients presenting with idiopathic interstitial pneumonia, particularly younger women and those with suggestive clinical, serological, or radiological features such as NSIP, NSIP-OP overlap, OP or LIP, should undergo a comprehensive evaluation for an underlying rheumatic disease. IPAF may evolve into a defined rheumatic disease over time. Combined evaluation by rheumatologists and pulmonologists is essential for the diagnosis and management of RALD and IPAF.

Limitations

Resource limitations affected the availability of certain diagnostic modalities in this study. Further longitudinal studies are required to evaluate long-term outcomes and the evolution of IPAF into defined rheumatic diseases.

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Online supplementary material

Supplementary Table 1. Demographics and clinical characteristics of the study population.

Supplementary Table 2. Clinical profile of specific rheumatologic diseases.

Supplementary Table 3. Clinical profile of patients with amino-acyl tRNA synthetase autoantibodies in our study.

Supplementary Table 4. Pulmonary involvement and high-resolution computed tomography pattern of interstitial lung disease in the study population

Supplementary Figure 1. STROBE Diagram depicting the flow of participants through the study.

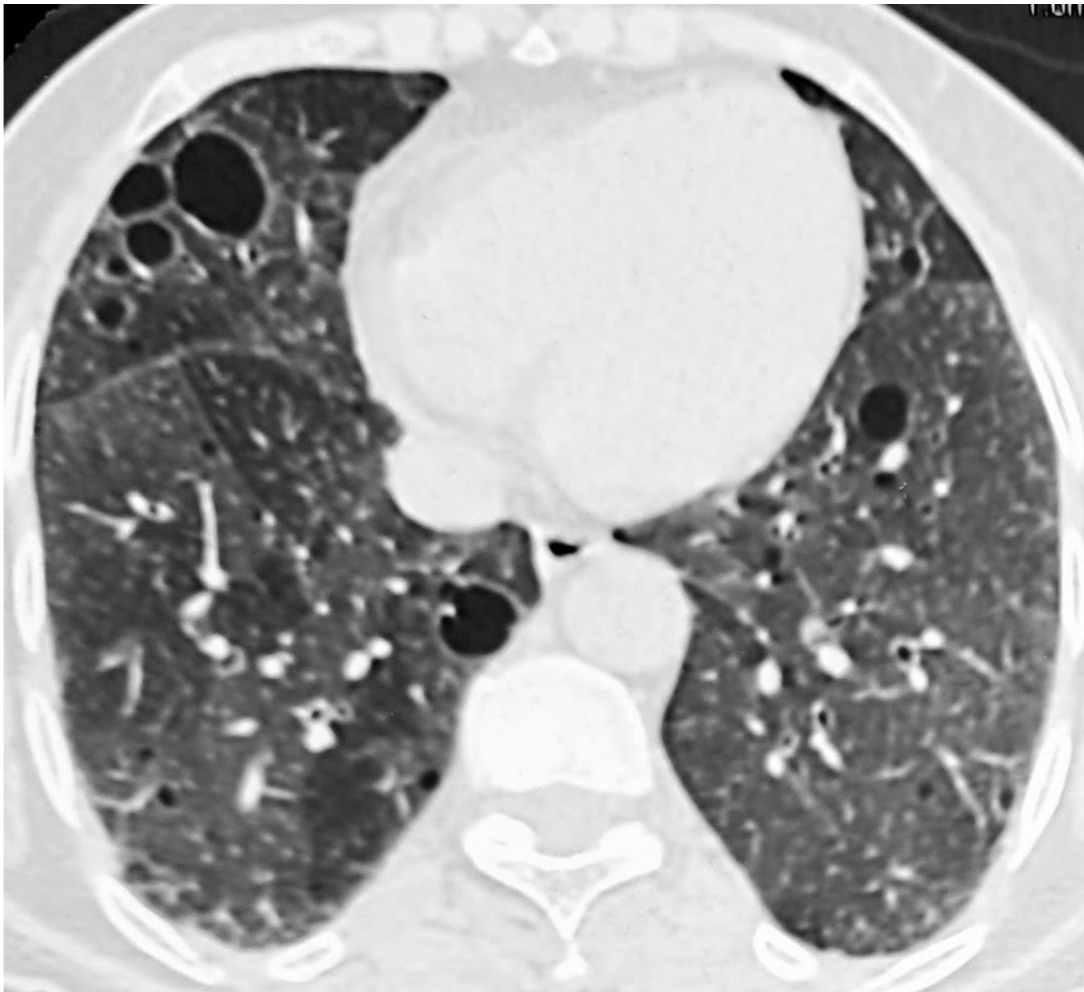


Figure 1. Axial section of High-Resolution CT scan of chest showing Lymphocytic Interstitial pneumonia in a 50-year-old lady with Primary Sjogren's Syndrome. The image shows prominent multiple perivascular cysts with diffuse ground glass opacities, and interlobular and intralobular septal thickening.

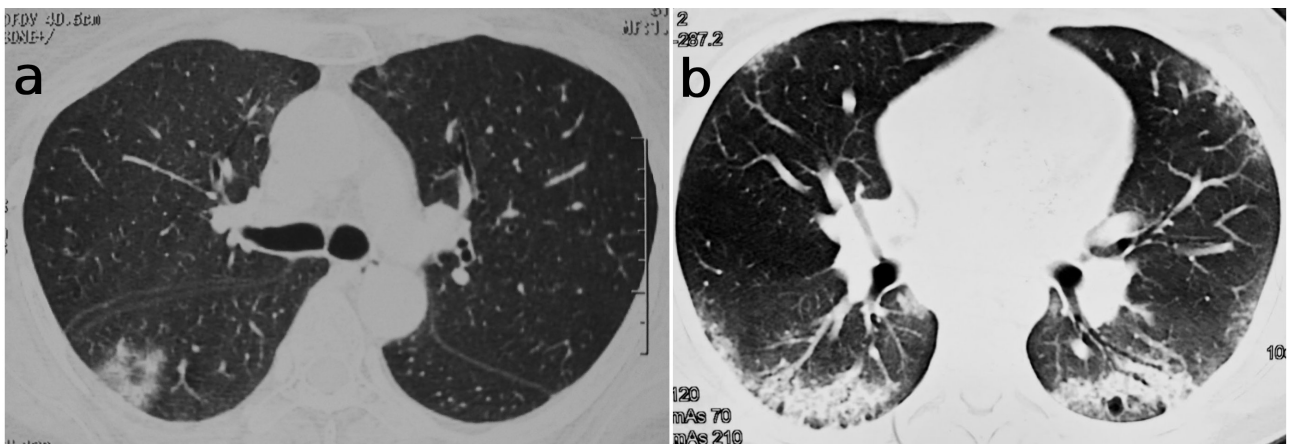


Figure 2. Axial sections of HRCT Chest showing Organising Pneumonia pattern. a) In a 52-years-old woman with Rheumatoid arthritis, the image shows organizing pneumonia as a patch of peripheral consolidation that appeared after introduction of Methotrexate and resolved with the discontinuation of the drug. b) NSIP-OP pattern in a 21-years-old lady with Systemic sclerosis. The image shows organizing pneumonia overlapping ground glass opacities with traction bronchiectasis in posterior regions of bilateral lower lobes and peripheral interstitial thickening. Vascular dilatation due to pulmonary hypertension is also noted.

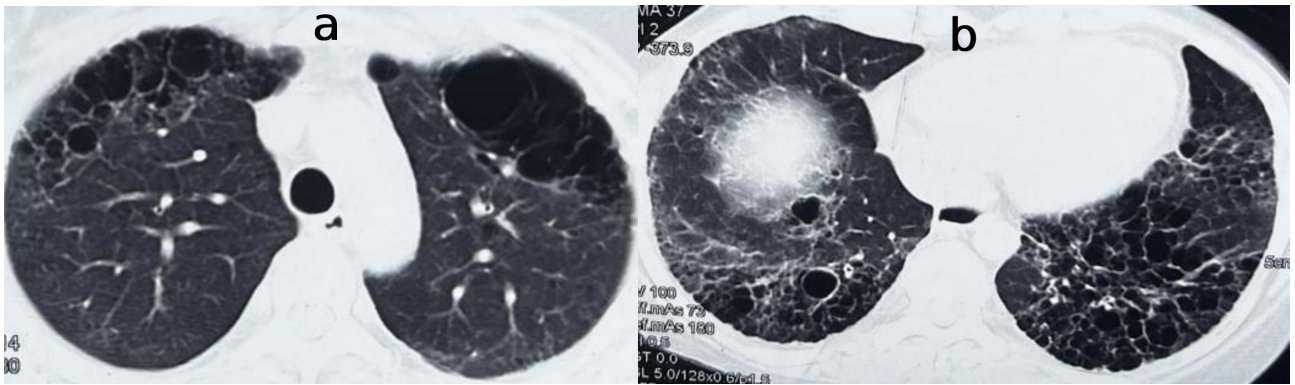


Figure 3. Axial sections of HRCT chest of a 31-year-old woman with diffuse systemic sclerosis showing signs suggestive of a rheumatology-associated interstitial lung disease. a) Anterior upper lobe sign - honeycombing in anterior regions of bilateral upper lobes with interlobular and intralobular septal thickening. b) Exuberant honeycombing and heterogenous destruction seen along with ground glass opacities, interstitial thickening, traction bronchiectasis and esophageal dilatation.