International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Spectrum of Interstitial Lung Disease as Seen on Computed Tomography in a Tertiary Centre

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Abstract: ***Interstitial lung disease is a broad spectrum of conditions, ranging from occasional self-limited inflammatory processes to severe debilitating fibrosis of the lungs. Imaging plays an essential role in characterizing this group of disorders and can often suggest the diagnosis, though the final interpretation requires a coordinated effort involving the radiologist, pathologist, and clinician. The challenge in diagnosing ILDs in India is constrained by lack of resources and standardized health care in India, lack of standardized approach in diagnosis and treatment of ILD. Empirical treatment is required for most patients diagnosed with ILD. The aim of this study is to find the prevalence of various interstitial lung disease in a developing country like INDIA at a tertiary centre.***

Keywords: Interstitial Lung Disease, Interstitial Pneumonia, Interstitial lung abnormality, honeycombing

1. Results

Among the total 100 subjects recruited, 46% were females and 54% were males with a mean age of 69 years.

Interlobar and intralobar septal thickening was the most prevalent HRCT chest findings. Wide spectrum of interstitial lung diseases were diagnosed, most common being Idiopathic pulmonary fibrosis / Usual interstitial pneumonia (IPF /UIP), Non specific interstitial pneumonia (NSIP) and chronic hypersensitivity pneumonitis along with less common ILDs like combined pulmonary fibrosis and emphysema (CPFE), lymphoid associated interstitial pneumonia (LIP), Langerhans cell histiocytosis (PLCH), Lymphangioleiomyomatosis (LAM) , as well as recently termed entity of interstitial lung abnomality (ILA).

Early diagnosis of IPF by using imaging is critically important due to the poor prognosis of the disease and progressive decline in pulmonary function.

2. Introduction

2.1 Materials and methods

We studied all patients who were diagnosed with any type of interstitial lung disease on HRCT CHEST at our institution, between 2022 and 2023. The Institute Ethics Committee approved the study protocol, and a written informed consent was obtained from all subjects.

2.2 Subjects and study procedures

100 patients with chest radiograph and HRCT thorax suggestive of interstitial lung disease were included in the study.

The relevant data included age, sex, occupation, presenting complaints, smoking history, environmental exposures, connective tissue diseases, family history and physical examination findings.

Contrast study was done in subjects suspected of mediastinal lymphadenopathy. Baseline 2D echocardiogram was done in all the patients.

HRCT CHEST was done on 128 slice SIEMENS SOMATOM go.Top Machine.

Breathlessness on exertion was the most common symptom (74%) followed by cough (65%), anorexia (34%), and fatigue (17%). Breathlessness at rest was seen in around 26% patients.The most common abnormalities on HRCT chest were interlobular septal thickening (74%), intralobular septal thickening (70%), ground glass opacities (22%), and honeycombing (36 %). Mediastinal lymphadenopathy was present in 23% subjects, whereas cardiomegaly was seen in 39% subjects. Traction bronchiectasis was seen in all the cases diagnosed with IPF and most of the cases of chronic hypersensitivity pneumonitis.

3. Discussion

Interstitial lung disease (ILD) is an umbrella term for over hundreds of different diseases that can be subdivided into those with an identifiable cause and those without; the latter being referred to as idiopathic interstitial pneumonias [1].Usual interstitial pneumonia / idiopathic pulmonary fibrosis (IPF) and Nonspecific interstitial pneumonia (NSIP) were the two most common diagnosis among the study. IPF was seen in 18 patients where as 16 patients were diagnosed with NSIP.

Reticular pattern, also sometimes referred to as reticulation, is the most common HRCT finding in IPF and is an indicator of fibrotic ILD in many cases along with fibrosis, including architectural distortion, traction bronchiectasis and bronchiolectasis [2] as seen in (figure 1a).

NSIP is interstitial pneumonia with the homogeneous appearance of interstitial fibrosis as seen in (figure 1b.)It is nonspecific as it lacks the histopathological features of the other subtypes of the ILD. NSIP typically has bilateral lung involvement and may have a predisposition for the lower lobes. [3]

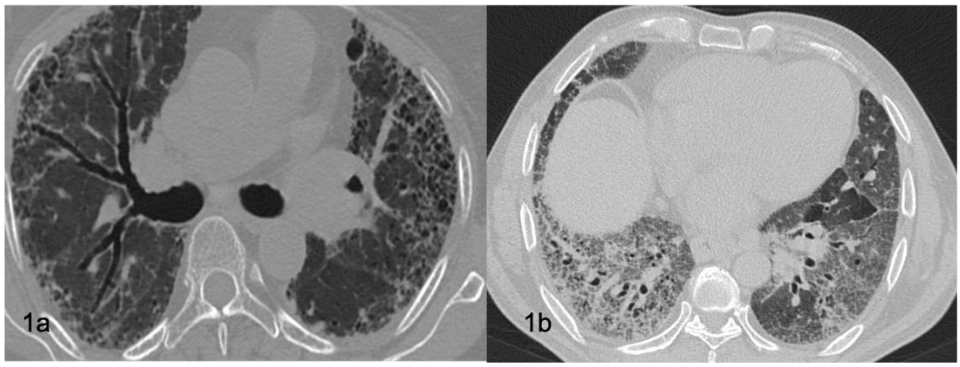

Figure 1(a): Shows traction bronchiectasis with honeycombing in IPF. **Figure 1(b):** shows changes of NSIP

UIP shows apicobasal gradient and is typically patchy and heterogenous, whereas NSIP is more homogeneous with no evidence of any apicobasal gradient.

Next common type of ILD encountered was chronic hypersensitivity pneumonitis (HSP), which was seen in 15 patients. It is a common interstitial lung disease which occurs due to from inhalation of antigens by susceptible individuals. Chronic HP is characterized by reticulation, traction bronchiectasis and honeycombing that can mimic UIP.

Compared with UIP or NSIP, a mid-to-upper lung predominant or random pattern of ground-glass opacification or reticulation is suggestive of HP [4] as seen in (figure 2a). HSP is also often characterized by mosaic perfusion and air-trapping, juxtaposed combination of lobular areas of high, normal and low attenuation (the so-called 'head-cheese' sign) that can help distinguish HSP from other ILDs [5], (figure 2b).

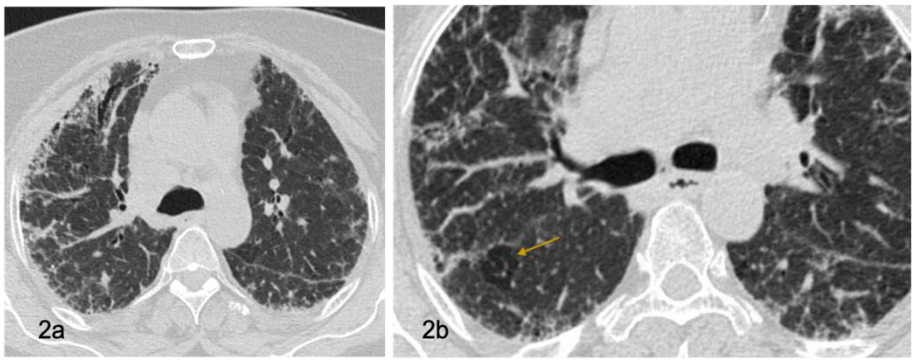

Figure 2(a): Upper lung reticulations bilaterally; 2b showing head cheese sign

14 patients presented with the newly coined entity of interstitial lung abnormality / ILA.

ILA is defined as incidental CT findings of nondependent abnormalities affecting more than 5% of any lung zone at complete or partial chest CT where interstitial disease was not previously suspected [6]. The findings include ground-glass or reticular abnormalities, lung distortion, traction bronchiectasis or bronchiolectasis, honeycombing, and nonemphysematous cysts. It is important to identify the changes related to ILA as it may eventually progress over 5 years in more than 50% of individuals, and are associated with worsened clinical outcomes, including respiratory symptoms, exercise capacity, lung function, and mortality [7].

Connective tissue related ILD were also prevalent and seen in 11 patients. We reported patients with scleroderma (fig 3a), rheumatoid arthritis, sarcoidosis and SLE related CT-ILDs. Extensive honeycombing was seen along upper lobes in RA-ILD patient (fig 3b), multiple small peri-fissural nodules and reticulations were seen in sarcoidosis related ILD (fig 3c).The spectrum of conditions included is broad, ranging from occasional self-limited inflammatory processes to severe debilitating fibrosis of the lungs. Rheumatologist opinion from the institute was considered and diagnosis of CTD was made using standard criteria.

Other diagnosed ILDs included cryptogenic organizing pneumonia (COOP), combined pulmonary fibrosis with emphysema (CPFE), smoking related ILDs like respiratory bronchiolitis related ILD and desquamative interstitial

disease (RB-ILD and DIP) and acute interstitial pneumonia which represented 8, 6, 3, 2 and 2 cases respectively.

One each case of Lymphangioleiomyomatosis (LAM), Langerhans cell histiocytosis (LCH), Lymphoid interstitial pneumonia (LIP), chronic eosinophilic pneumonia and airspace enlargement with fibrosis (AEF) were also noted.

COOP was seen as patchy consolidation with a predominantly subpleural and/or peribronchial distribution, (fig3d), and arcade-like sign of perilobular fibrosis describes an arch pattern in more than half of the patient with COOP [8]. The reverse halo sign (atoll sign) considered to be highly specific, although only seen in around 20% of patients with COOP [9].

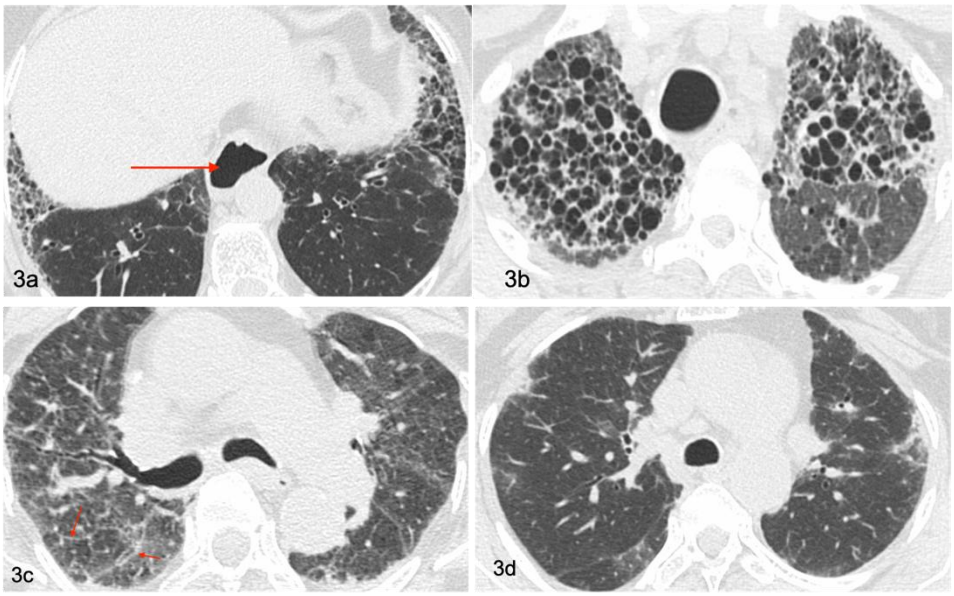

Figure 3: (a) Dilated esophagus with changes of ILD in case of scleroderma, (b) Profound honeycombing in RA related ILD, (c) Peribronchovascular nodularity in Sarcoidosis related ILD, (d) Peripheral consolidations as seen in COOP

Characteristic radiologic findings in the CPFE include emphysema as well as lower lobar interstitial fibrotic changes (fig 4a). The emphysema in CPFE is predominantly in the upper lobes. Honeycombing and reticular abnormalities are frequent, but areas of ground glass attenuation also are commonly present, as in 66% of subjects with CPFE in a series by Cottin et al.[10].

Smoking related ILDs like respiratory bronchiolitis related ILD and desquamative interstitial disease (RB-ILD and DIP) (fig 4b). The most frequent findings in RB ILD reported by Park et al. were central and peripheral bronchial wall thickening (in 90% and 86% of patients, respectively), patchy ground glass opacities and centrilobular nodules (71%) [11].

DIP was associated with parenchymal distortion, traction with bronchiolar dilatation. Honeycombing was seen in less than one-third of cases [12].

The overlap of findings is most significant between RB-ILD and DIP, with RB-ILD showing centrilobular patchy ground-glass opacities whereas DIP shows widespread ground-glass changes.

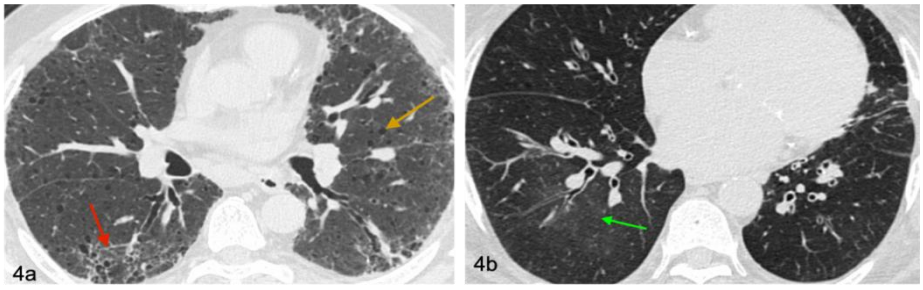

Figure 4: (a) CPFE: Orange arrow showing centriacinar emphysema and red arrow showing fibrosis, (b) RB-ILD : Patchy ground glass opacities in a chronic smoker

Lymphangioleiomyomatosis (LAM) and pulmonary Langerhans cell histiocytosis (LCH) are characterised by the presence of parenchymal cysts (fig5a) with LAM affecting predominantly women of childbearing age whereas LCH usually identified in young adults of 20-40 years age. A history of current or previous cigarette smoking is identified in up to 95% of cases [13].Distribution is the key in differentiating pulmonary Langerhans cell histiocytosis from other cystic lung diseases as it has predilection for the mid and upper zones [14]. In our study, LCH cases showed cavitatory nodules as well as non cavitatory nodules along with cysts. (fig5b).

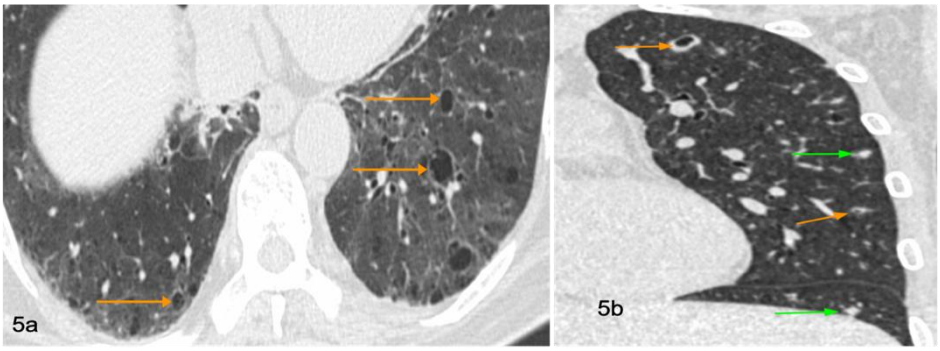

Figure 5: (a) shows multiple varying sized cysts in LAM; (b): Green arrows show non cavitatory nodules whereas orange arrows suggest cavitatory nodules in LCH patient.

The most frequent CT findings observed in patients with LIP were multiple cysts, small nodules, and ground-glass opacities (fig 6a). LIP is most commonly described in association with Sjögren syndrome, followed by infectious causes, such as HIV and Epstein-Barr virus [15].

Typical findings chronic eosinophilic pneumonia include non-segmental patchy areas of airspace fibrosis involving the lung periphery (fig 6b) and mostly the upper lobes. Less common findings are the presence of ground glass change, nodules and reticulation. Interval CT performed after more than 2 months from beginning of symptoms will show linear opacities parallel to the pleural surface. Only 9% of cases have a pleural effusion [16].

AEF showed peripheral subpleural cysts, which appeared lobulated, branched, and with bizarre shapes (fig 6c). Those cysts are termed as AEF and are categorized as a separate entity in smoking-related interstitial pneumonia according to a recent update of idiopathic interstitial pneumonias [17]. Watanabe et al. reported that multiple, thin-walled cysts are among high resolution CT features of AEF [18].

Acute interstitial pneumonia(fig 6d) shows combination of ground-glass opacities and air space consolidation [19]. It is rapidly progressive ILD of unknown aetiology and considered as acute process among the idiopathic interstitial pneumonias.

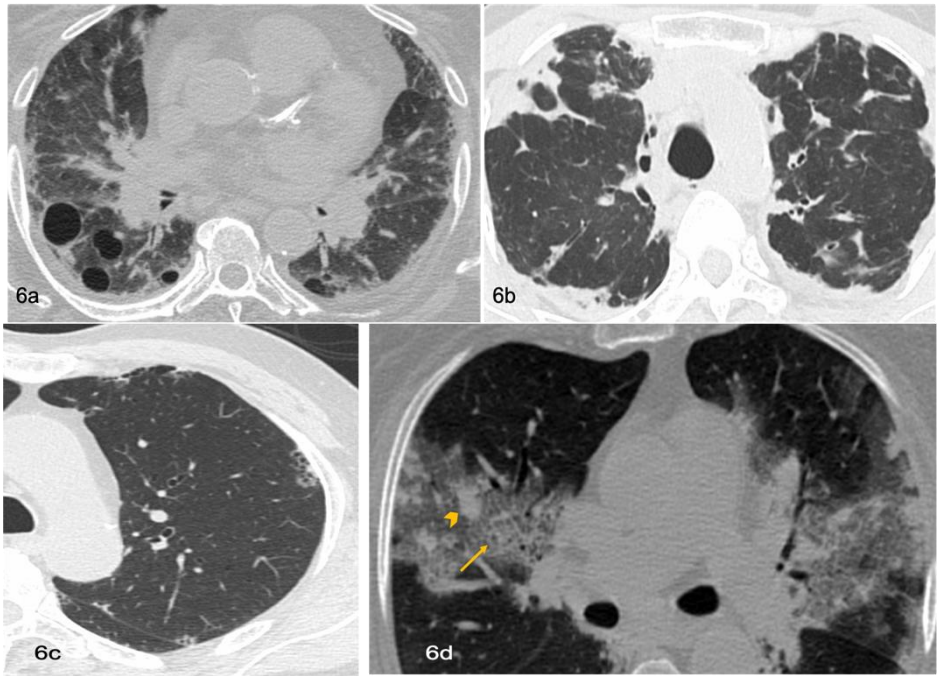

Figure 6: (a) Large subpleural cysts in LIP, (b) subpleural fibrosis in chronic eosniphilia, (c) subpleural lobulated bizzare shaped cysts in AEF and (d) AIP showing areas of ground glass opacities and consolidations.

4. Conclusion

Our study analyzed the spectrum of ILD encountered with radiological characteristics of the patients with interstitial lung disease in a tertiary care setting. Usual interstitial pneumonia / idiopathic pulmonary fibrosis and Nonspecific interstitial pneumonia were the two most common diagnosis among the study, followed by chronic hypersensitivity pneumonitis. A wide spectrum of ILDs were detected in our study, including the rare types, like LIP, DIP, LCH and LAM. 14% patients were found to have ILA / interstitial lung abnormality which is a relative recently assigned term.

To conclude, apart from commonly studied ILDs, a number of rare entities are also encountered at a tertiary centre.

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