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Idiopathic Interstitial Pneumonias (IIPs): Review of Clinical, Radiographic and High-Resolution Computed Tomography (HRCT)

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Abstract

Making a confident diagnosis is a complex task for a specific form of interstitial lung disease and providing appropriate management in an attempt to achieve normalization of the disease can put up an alarming process for the clinicians. A set of diffuse and restrictive lung diseases incorporate with idiopathic interstitial pneumonias, showing inflammation and fibrosis of the interstitium due to parenchymal damage. High-resolution computed tomography (HRCT) has magnified the diagnostic standpoint in stepwise identification and classified various patterns in the evaluation of interstitial lung disease. The aim of our review is to elaborate clinical, radiographic and typical and atypical HRCT findings of idiopathic interstitial pneumonias by correlating with its differential diagnosis. Idiopathic pulmonary fibrosis is the most predominant idiopathic interstitial pneumonias and its diagnosis needs to omit all other well-known causes of interstitial lung diseases. According to the 2011 evidence-based guidelines, usual interstitial pneumonia can be diagnosed by HRCT when all criteria are fulfilled. Non-specific interstitial pneumonia is distinguished by bilateral patchy ground-glass opacities and irregular linear/reticular opacities. Respiratory bronchiolitis associated-interstitial lung disease and desquamative interstitial pneumonia show centrolobular nodules and ground-glass opacities as imaging patterns. Cryptogenic organizing pneumonia consists of patchy peripheral or peribronchial consolidations, while ground-glass opacities with tendency for migration, which is evolving to fibrosis, in acute interstitial pneumonia. Lymphoid interstitial pneumonia and idiopathic pleuro-parenchymal fibroelastosis are classified under rare idiopathic interstitial pneumonias. HRCT images help radiologists in diagnosis and mapping specific patterns of idiopathic interstitial pneumonias. This article reviews the stages of evolution in HRCT features for idiopathic interstitial pneumonias.

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Keywords

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1. Introduction

According to The American Thoracic Society (ATS) and European Respiratory Society (ERS), ILD is a heterogeneous group of non-neoplastic disorders incidental from damage to the lung parenchyma by inflammation and fibrosis that vanishes the lungs capacity for alveolar gas diffusion [1]. ILD has peculiar category which can be differentiated from one another when clinical data, radiologic imaging, and pathologic findings (if lung biopsy is needed) are combined to reach a positive diagnosis [2] [3].

The ATS/ERS in 2002 publicized classifications of idiopathic interstitial pneumonias (IIPs) [1].This classification includes usual interstitial pneumonia (UIP), non-specific interstitial pneumonia (NSIP), desquamative interstitial pneumonia (DIP), respiratory bronchiolitis-associated interstitial lung disease (RB-ILD), cryptogenic organizing pneumonia (COP), acute interstitial pneumonia (AIP) and lymphoid interstitial pneumonia (LIP). In 2011 ATS/ERS revised IIPs by putting them in simpler way as possible into major IIPs and rare IIPs. Major IIPs includes chronic fibrosing as (IPF and NSIP), smoking related as (RB-ILD and DIP) and acute/subacute as (COP and AIP). Idiopathic LIP and idiopathic pleuro-parenchymal fibroelastosis (PPFE) are grouped under rare IIPs [4].

As for the physicians due to limited symptoms it becomes a nightmare in diagnosing patient with or without ILD. In many diseases, commonly occurring breathlessness and cough are seen in mildly aged and elder patients, mainly taken as COPD or heart failure. As soon as ruling out non-respiratory causes for breathlessness is done, the physicians should see the potential of IDL in mildly aged or elderly patients which shows unexplained chronic dyspnoea on exertion, particularly those who are more breathless than would be expected based on their lung function and other contributory factors such as obesity, or the once with the long duration of cough [5]. A hunch for IDL should be upraised in patients with nonspecific X-Ray, patients with probable obstructive lung disease or congestive heart failure which shows no hope from therapy.

Significant contribution of HRCT in finding and diagnosing of diseases has made the radiologist play an important role in the diagnosis of IIP. According to 2011, ATS/ERS have revised HRCT features for IDL (**Table 1**), hence HRCT is the most sensitive tool for non invasive of the lung parenchyma in patients with suspected IPF [6].

2. Literature Review and Discussion

2.1. Chronic Fibrosing IIPs Consists of UIP and NSIP

2.1.1. Usual Interstitial Pneumonia (UIP)

According to international guidelines idiopathic pulmonary fibrosis (IPF) is defined asa specific form of chronic, progressive, fibrosing interstitial pneumonia of unfamiliar influence, which limits to the lungs of elderly adults, with the histopathological and/or radiological pattern of UIP [6]. In some research the ratio of IPF in elderly males is more than 65 years, corresponding tendency has been seen in European studies, too [7]. In patients with more than 75 years much higher ratio is seen [7] [8]. According to the revised criteria of ATS/ERS in 2011 [6], the diagnosis of IPF involves following: 1) Exclusion of other known causes of ILD such as domestic and occupational environmental exposures, connective tissue disease and drug toxicity. 2) The presence of a UIP pattern on HRCT in patients not subjected to surgical lung biopsy (SLB), and 3) specific combinations of HRCT and SLB patterns in patients subjected to SLB.

On a chest X-Ray the image findings can be misdiagnosed in the early stages, lower lobes and costophrenic angles with reticular pattern and bronchiectasis are engaged preponderantly. Honeycombing is the most important picturing feature on HRCT for IPF [9], which is characterized as cluster of cystic air sacs with diameter varying from 3 - 10 mm, which can also hit to 2.5 cm in size [9] [10]. The mediastinal lymph nodes which are usually found in the paratracheal areas shows enlargement in 70% - 80% cases on HRCT [11]. According to the international guidelines, HRCT and histological appearances are used in diagnosing UIP, furthermore, UIP was classified in three main forms as UIP pattern, possible UIP, inconsistent with UIP which should be diagnosed on HRCT with their specific patterns [6] (**Table 2**). Even though HRCT is decisive component in diagnosing UIP,

Table 1. High-resolution computed tomography criteria for usual interstitial pneumonia pattern.

UIP pattern (all)	Possible UIP (all)	Inconsistent with UIP (any)
1. Basal and subpleural predomination 2. Reticular patterns with traction bronchiectasis association 3. Honey combing appearance 4. Deficits of features listed as inconsistent with UIP pattern	1. Reticular pattern with presence or absence 2. Typical gradients (lower lobes & subpleural) 3. Deficits of features listed as inconsistent with UIP pattern	1. Upper and middle lung predomination 2. Peribronchiovascular predomination 3. Extensive ground glass abnormality 4. Profuse micronodules (bilateral, upper lobes predominance) 5. Separate cysts(multiple, bilateral, off honeycombing areas) 6. Diffuse air trapping(bilateral 3 or more lobes) 7. Parenchymal consolidations

Criteria for usual interstitial pneumonia on high-resolution computed tomography.
UIP: Usual interstitial pneumonia.
UIP pattern and Possible UIP: all criteria ought to be fulfilled.
Inconsistent with UIP: among 7 any 1 of the criteria ought to be fulfilled.

Table 2. High-resolution computed tomography features of Idiopathic interstitial pneumonias.

Idiopathic interstitial pneumonias	High-resolution computed tomography features
UIP	Reticular pattern with presence or absence of transaction bronchiectasis, honeycombing appearance, basal and subpleural predomination, in UIP pattern deficits of features listed as inconsistent with UIP.
NSIP	Bilateral ground glass regions, reticular opacities.
RB-ILD	Badly defined centrilobular nodules, bronchial wall thickening or centrilobular emphysema.
DIP	Diffuse ground glass opacities, irregular linear opacities, Microcysts.
COP	Distinctive peripheral or peribronchial patchy consolidations, Ground-glass opacities with migrating tendency, rarely a mass or nodule that may cavitate the typical appearance of an “atoll sign”.
AIP	Ground glass attenuation regions with mosaic pattern, independent area with air space consolidation.
LIP	Perivascular cysts and ground glass opacities, centrilobular and subpleural nodules.
PPFE	Apical regions with subpleural thickening, small subpleural consolidations.

Features of idiopathic interstitial pneumonias on high-resolution computed tomography.
UIP: Usual interstitial pneumonia; NSIP: Non-specific interstitial pneumonia; RB-ILD: Respiratory bronchiolitis with associated interstitial lung disease; DIP: Desquamative interstitial pneumonia; COP: Cryptogenic organizing pneumonia; AIP: Acute interstitial Pneumonia; LIP: Lymphoid interstitial pneumonia; PPFE: Pleuroparenchymal fibroelastosis.

it should be correlated with histological appearances of IPF. The guidelines for matching histological results are, 1) Histology procedures should not be done for the confirmation of UIP pattern. After exclusion of identifiable causes of ILD, HRCT should be done in diagnosing of IPF, even if surgical lung biopsy is possible, probable or non-classifiable [6]. 2) Correlation with biopsy is needed for the possible UIP pattern. Diagnosing IPF in patients with a possible UIP pattern on HRCT and UIP or probable histological UIP pattern should be allowed. Possible UIP pattern is diagnosed by identifying any one feature from inconsistent with UIP and surgical lung biopsy which shows the pattern of UIP [6].

2.1.2. Non-Specific Interstitial Pneumonia (NSIP)

NSIP is one of the most common interstitial pneumonias after UIP. Respective studies based on histological features have shown some fact that some cases of interstitial pneumonia do not show resemblance with UIP, DIP or AIP [12]. Reference [13] reported these pneumonias as non specific interstitial pneumonias. The clinical features of NSIP are similar to those of UIP, except that patients with NSIP are more commonly female and generally have a younger mean age than do those with UIP. NSIIP morbidity is still a mystery, where there have been researches showing a range of 14% - 36% of all IIPs [3] [14]. Comprehensive variety of lung diseases shows analogous histopathological and radiological pattern of NSIP, hence this disease have been argued as independent clinical entity in some literatures by authors [15]. However, idiopathic NSIP has been classified as clinicopathological entity in revised 2002 ATS/ERS guidelines [4]. Reference [16] notified that clinical diagnosis of NSIP should be engaged with idiopathic and biopsy-proven cases when no contributory factors can be

identified. When disease shows no connection with any underlying disease, NSIP can be categorized as idiopathic NSIP [1]. However, cases which shows possibility of identifying a known aetiology, NSIP is considered as secondary [17].

With infiltrates preponderantly in lower lobes, showing reticular patterns and bronchiectasis, making image findings of chest X-Ray (CXR) not precise. Some important HRCT features shows [14] [18] [19]. 1) Bilateral ground-glass areas in middle and lower lung, sometimes with comprehensive distribution. 2) Reticular opacities, which can be overlying on ground glass patterns. 3) Traction bronchiectases, which can be parallel in their trend through the lungs or extremely irregular. NSIP shows rare sign of honeycombing [20] [21]. Mostly NSIP comes across with bilateral patchy areas of ground glass in middle and lower lung, with or without traction bronchiectasis or reticulation. Other HRCT features discovered are lower lobe volume loss and absence of ground glass opacifications in subpleural areas [22]. However studies in a series of 50 patients represented half majority of cases with non diagnostic imaging appearance or more corresponding with other chronic infiltrative lung diseases, pulmonary alterations compatible with a UIP pattern was seen in 32% of patients and only 22% of patients showed bilateral patch areas which is described as typical pattern for NSIP [20] [23]. Differential diagnosis for NSIP includes UIP, DIP, AIP and extrinsic allergic alveolitis.

2.2. Smoking Related IIPs Consists of RB-ILD and DIP

2.2.1. Respiratory Bronchiolitis with Associated Interstitial Lung Disease (RB-ILD)

RB-ILD is known as pathological lesion of RB related with clinical manifestation of interstitial lung disease [1]. Reference [24] reported first description of smoking related bronchiolitis, after demonstration on smokers he discovered presence of clusters of pigmented macrophages in respiratory bronchioles and neighboring alveoli. Similar alterations of respiratory bronchiolitis were found in patients with clinical and radiological features of chronic interstitial lung diseases [25]. Reference [26] demonstrated presence of RB in all smokers and about 50% of former. Mostly affects smokers of 30 - 40 years of age with a history of more than 30 pack-years of cigarette smoking [27] [28]. This disease is considered as slight male dominance [29]. Accumulation of alveolar macrophages within respiratory bronchioles is represented as histological hallmark of RB-ILD. Characterization of macrophages is done by eosinophilic cytoplasm, constituents of cigarette smoke portraying with brown granular pigmentation. The main difference between RB and RB-ILD is based on the stage of fibrosing and adjacent alveolar walls are also involved by inflammatory process that present in RB-ILD [25].

Radiographic findings are comparatively not obvious [25] [30]-[32]. In 20% to 28% of patients with histological proven RB-ILD were reported with normal chest radiographs [25] [30] [33], whereas both normal chest X-Ray and HRCT appearance were reported in one of the case [34]. In patients with RB-ILD the typical chest X-Ray findings shows diffuse fine reticulonodular interstitial opacities or are prevailing in basal lung areas [25] [30]. Some important HRCT features shows [32], 1) Ground glass opacities (related with accumulation of macrophages in alveolar spaces). 2) Poorly defined centrilobular nodules or also known as centrilobular ground-glass nodules. 3) Diffuse lung distribution. 4) Other important CT features: centrilobular emphysema and/or bronchial wall thickening due to cigarette smoking [18] [27] [34]. Due to absence of traction bronchiectasis and honeycombing some number of patients showed reticular pattern due to fibrosis [3] [28] [29] [31]. Differential diagnosis for RB-ILD includes NSIP, DIP and acute hypersensitivity pneumonitis [30].

2.2.2. Desquamative Interstitial Pneumonia (DIP)

Reference [35] coined the term desquamative interstitial pneumonia. DIP is considered as a form of interstitial pneumonia in which the alveolar spaces shows the presence of diffuse exudation of pigmented macrophages [4]. Reference [31] by comprehensive evaluation of HRCT findings, various degrees of severity of a reaction of small airways and lung parenchyma to cigarette smoke should be well advised in DIP and RB-ILD. Drug reactions and connective tissue diseases such as scleroderma, lupus and rheumatoid arthritis interrelate with DIP [36]-[38]. Reference [39] studies shows less than 3% of interstitial lung disease comes under DIP; patients in their third to fifth decade are usually affected, with value of males twice to that of females. Definite histological patterns to differentiate DIP from RB-IDL are given according to the guidelines [4], however making it difficult in histopathological diagnosis of DIP from RB-IDL. Lesions affecting in DIP are in uniform manner while RB-IDL shows bronchiolocentric distribution [31].

Chest X-Ray may show bilateral hazy opacities like interstitial infiltrates, but for detection of DIP this pattern is non-specific. HRCT scans for spotting pathognomonic radiological features of DIP include, 1) Diffuse ground

glass opacities 2) Irregular linear opacities. 3) Microcysts (seen in half majority of patients) [18]. Most important imaging feature of DIP is ground glass opacification of lung, 86% showed bilateral and symmetric, basal and peripheral showed 60% patch and diffuse with 20% each [34] [40] [41]. Differential diagnosis for DIP includes RB-ILD and chronic hypersensitivity pneumonia.

2.3. Acute/Subacute IIPs Consists of COP and AIP

2.3.1. Cryptogenic Organizing Pneumonia (COP)

Reference [42] notably characterized Cryptogenic organizing pneumonia (COP) in one of its lecture. COP at its origin was said to be as pulmonary condition which propels due to chronic persistence of a pneumonitis. Reference [43] coined another term for COP as idiopathic bronchiolitis obliterans organizing pneumonia (BOOP), which was then changed to avoid puzzlement in an airway disease known as constrictive bronchiolitis obliterans. COP remains unrelated to bronchiolar obliteration because of its clinical, physiological and imaging features; hence the term COP is preferred [44]. Although the organizing pneumonia process is primarily intraalveolar, it was included in the classification of the interstitial pneumonias because of its idiopathic nature [4]. The mean age of 55 years is seen in the patients affected with COP. COP shows no gender orientation or connection with cigarette smoking. Mostly patients are nonsmokers or former smokers. Patients with COP typically present with cough and dyspnea of relatively short duration with sparse crackles at auscultation [45].

Three main patterns are observed in the imaging findings of COP, 1) Typical form (occurrence of multiple alveolar opacities) 2) Focal consolidation associated form (focal COP) 3) Infiltrative form (occurrence of infiltrative opacities). COP is characterized radiographically by multiple bilateral areas of consolidation [46]. Presence of granulation tissue at the alveolar spaces leads to patchy areas of consolidations. With the tendency of migration, these alveolar infiltrates could be seen in different lung areas on different chest X-rays. Some important HRCT features include: 1) Distinctive peripheral or peribronchial patchy consolidations (occasionally with subpleural spread) likewise air bronchograms and mild cylindrical bronchial dilatation. 2) After antibiotic therapy for several weeks, rise of more consolidations observed. 3) Ground-glass opacities with migrating tendency and transformation of size. 4) Rarely, a mass or nodule that may cavitate the typical appearance of an “atoll sign”. Occurrence of ring consolidation surrounding normal lung or ground-glass opacification leads to “atoll sign”. By striking 20% patients of COP it is considered relatively specific for OP. Sarcoidosis and Wegener’s granulomatosis also been recently described of showing atoll sign [47] [48].

2.3.2. Acute Interstitial Pneumonia (AIP)

AIP is a form of interstitial pneumonia which is severe and sudden onset with rapid progression. Reference [49] [50] first reported this disease while describing the cases of rapidly progressive disease. Reference [51] introduced the term AIP for these rapid interstitial disease after observing similar cases which had priorly observed. AIP as an idiopathic disease is observed in patients with the mean age of 50 years with equal occurrence in male and women [18] [52] [53]. Most patients within 3 weeks of symptoms develop severe dysponea and seek treatment with signs of pneumonic consolidation with diffuse crackles [1] [18] [52]. Mechanical ventilation is required with oxygen therapy as the condition rapidly progress to acute respiratory failure. Most patients fulfill the clinical criteria for acute respiratory distress syndrome: acute onset, ratio of arterial partial pressure of oxygen to fraction of inspired oxygen of 200 mm Hg or lower, diffuse bilateral opacities on chest radiographs, and pulmonary capillary wedge pressure of less than 18 mm Hg [51] [53] [54]. The mortality rate is 50% or higher. Lung fibrosis is observed in the survivors of the acute phase [52]. For a definitive diagnosis, histopathological pattern is necessary [18].

Chest X-Ray shows bilateral airspace opacifications with air bronchograms in which costophrenic angles are spared [55]. Some important HRCT features include: 1) Diffuse ground-glass attenuation areas with a mosaic pattern (due to occurrence of alveolar septal oedema and hyaline membranes). 2) Airspace consolidation in dependent areas of the lung (due to reflection of intra-alveolar oedema and hemorrhage in the exudative phase) [18] [52] [55] [56]. These signs are mostly diffuse or involve upper lobes and usually bibasilar [52]. Intraalveolar fibrosis leads to consolidations, which are usually seen in organizing phase, distinguished by lung architectural distortion, traction bronchiectasis and cysts [52] [55]-[58]. Ground-glass attenuation on HRCT can be double faced disease. Various studies in AIP show ground-glass attenuation and consolidation are overt throughout all histological phase, constantly emulates different histological finding which exaggerates in diagnosing actual

histology [56] [59] [60]. Ground-glass attenuation, consolidation and traction bronchiectasis are usually associated with fibrotic phase [59]. Differential diagnosis for AIP includes DIP, pneumocystis carinii pneumonia, hydrostatic oedema, hemorrhage, alveolar proteinosis and bronchioloalveolar cell carcinoma.

2.4. Rare IIPs Consists of Idiopathic LIP and Idiopathic Pleuroparenchymal Fibroelastosis PPFE

2.4.1. Lymphoid Interstitial Pneumonia (LIP)

Reference [61]-[63] introduced the term LIP, to describe a diffuse lymphocytic interstitial infiltrate that was distinguishable from other patterns of interstitial pneumonias [62]. LIP appears in patients with lymphoma, so some authors debated LIPs position in interstitial pneumonias, rather proposed to be among pulmonary lympho-proliferative diseases [62] [64]-[66]. According to revised ATS/ERS guidelines LIP has been again classified in IIPs because its source remains undiagnosed [1] [62]. LIP is rare and usually associated with connective tissue disorder such as Sjögren's syndrome, AIDS, immunodeficiency syndromes such as Castleman's disease and autoimmune thyroid disease [44] [52] [62] [66]. Women's in their fifth decade of life are mostly diagnosed with LIP [62]. Patients for over 3 or more years show sluggish onset of symptoms with gradual progressive cough and dysponea; in some cases, symptoms like fever, weight loss, chest pain, night sweats and artralgia are also observed [62].

Diffuse or predominantly lower bilateral abnormalities usually show in HRCT images. In patients with LIP, presence of lymphatic nodes is often seen [62].Thickening of the bronchovascular bundles and interlobular septa is customary as LIP is most severe in inflamed perilymphatic interstitium [67]. Bilateral ground-glass opacity is a typical finding in LIP, which can be uniform or patchy [68] [69]. Perivascular, thin walled cysts are some other findings in LIP [52] [67]. These cysts appear within lung parenchyma mainly in the middle section of lungs, neighboring with blood vessels [70] [71]. The best indicators of LIP are considered to be, the association of perivascular cysts and ground-glass opacities. Centrilobular and subpleural nodules reflect the inflammatory infiltration of the peribronchiolar interstitium showing thickening of the interlobular septa and are also typical [68]. In 50% of patients, areas with previous airspace abnormalities shows perivascular honeycombing and reticular pattern. Differential diagnosis for LIP includes hypersensitivity pneumonitis, sarcoidosis and lymphangitic spread of tumor pneumocystis carinii and most important Hodgkin's lymphoma [72].

2.4.2. Pleuroparenchymal Fibroelastosis (PPFE)

According to recent ATS/ERS revised guidelines, Idiopathic PPFE has been categorized under IIPs classification as rare IIP [4]. Reference [72]-[74] in the Japanese literature represented PPFE as idiopathic pulmonary upper lobe fibrosis. Pleural surfaces and subpleural parenchymal lung shows development of fibrosis, especially in upper lobe. According to many recent studies, many authors claim PPFE as pulmonary reaction which is correlated with chronic graft-versus-host disease, which results to bone marrow transplantation [74]. According to literature occurrence of PPFE in male/female ratio is 2/1 [74]. However, other literature series the ratio is reported as 1/1.4 [73]. Histological pattern comprise of pleural thickening and subpleural fibrosis.

Chest radiographs shows lesser subpleural pulmonary consolidations with apical regions showing aspecific pleural thickening. Such imaging pattern shows similarity with many chronic pulmonary infections such as tuberculosis. Reference [75] reported five cases with superior hilar retraction. HRCT shows pleural thickening of apical zones, occasionally associated with bronchiectasis. Upper and middle regions and reticulations observed with small subpleural consolidatioin. Interlobar fissures shows infiltration due to pleural irregularities [75]. Pulmonary volume loss is the result from chronic fibrosis.

3. Conclusion

Knowledge of imaging features of IIP on HRCT images brings help for radiologists in its diagnosis. Furthermore, multidisciplinary approach is needed due to overlapping of imaging features. Particularized IIPs shows specific pattern on HRCT, hence making radiologist more capable in diagnosing ILDs on HRCT. Chronic respiratory conditions presents with similar symptoms in ILD, leading to regular misdiagnosis in their primary care, mostly COPD and heart failure. Among all IIPs, most prevalling is IPF and by excluding all other well-known causes of interstitial lung diseases helps in its diagnosis with presence of reticulations, bronchiectasis and honeycombing. According to 2011 revised guidelines [6], HRCT has an upper hand in diagnosing UIP when all

criteria are accomplished. Radiologist faces diagnostic challenge in early detection of its pattern, in order to provide precise disease management. IIPs restrict diagnosis as overlap of imaging features is commonly seen. Diverseness in radiological appearances makes diagnosis in NSIPs challenging, though most encountered features of NSIP remain bilateral patchy ground-glass regions. Smoking cessation in smoking related IIPs could improve their course. RB-ILD shows characteristic feature of poorly outlined centrilobular nodules, whereas DIP with typical ground-glass appearance. For radiologists, clinical presentation could help edge up with acute/subacute IIDs. COP consists of patchy peripheral or peribronchial consolidations, while ground-glass opacities with tendency for migration, which is evolving to fibrosis, in AIP. LIP and PPFE are classified under rare idiopathic interstitial pneumonias. The association of perivascular cysts and ground-glass opacities is typical in LIP, whereas an apical region with subpleural thickening is typical of PPFE.

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