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ORIGINAL ARTICLE

Pleuroparenchymal fibroelastosis-like lesions on chest computed tomography in routine clinical practice

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Abstract

Purpose To evaluate the incidence and changes in the pleuroparenchymal fibroelastosis (PPFE)-like lesions on chest CT in routine clinical practice.

Materials and methods This study included 1284 patients who underwent chest CT in 2011 at a hospital. The incidence of PPFE-like lesions and their correlation with age, body mass index, and concomitant pulmonary findings were assessed. Moreover, predictors of lesion progression were evaluated on follow-up. The ethical review board waived the requirement of informed consent for the retrospective review of patient records.

Results In total, 397 (30.9%) of the 1284 patients presented with PPFE-like lesions. The presence of PPFE-like lesions was correlated with older age (mean 72.2 vs. 69.9 years, $p = 0.002$); lower BMI (mean 21.0 vs. 23.2, $p < 0.001$); and interstitial pneumonia (64.8%, $p < 0.001$), emphysema (40.4%, $p < 0.001$), chronic airway disease (64.8%, $p < 0.001$), and old tuberculosis (56.5%, $p < 0.001$). Multivariate analysis of the follow-up CT findings revealed that interstitial pneumonia, nodular opacity, and lesion thickness affected progression (odds ratio: 3.81, 3.78 and 1.21), respectively.

Conclusion Pleuroparenchymal fibroelastosis-like lesions were not rare and correlated with interstitial pneumonia, emphysema, chronic airway disease, and old tuberculosis. On follow-up, PPFE-like lesions in some patients with concomitant interstitial pneumonia exhibited progression.

Keywords Computed tomography · Lung · Pleuroparenchymal fibroelastosis · Apical cap · Interstitial pneumonia

Introduction

Idiopathic pleuroparenchymal fibroelastosis (PPFE) is a rare condition characterized by fibrosis involving the pleura and subpleural lung parenchyma, predominantly in the upper lobes [1]. Idiopathic upper lobe fibrosis was first described by Amitani et al. [2] and the term PPFE was coined by Frankel et al. [3]. Imaging characteristics of PPFE include marked apical subpleural opacity on chest radiographs and intense subpleural fibrosis with pleural thickening on high-resolution computed tomography (HRCT) [3–5]. Some patients may develop progressive PPFE, which is associated with a poor prognosis [6]. The American Thoracic Society/European Respiratory Society has considered PPFE as a rare pattern of idiopathic interstitial pneumonia [1]. However, the findings of PPFE are indistinguishable from those of apical cap fibrosis, a finding often observed on lung autopsy and chest X-ray [7]. PPFE sometimes coexists with usual interstitial pneumonia, nonspecific interstitial pneumonia (NSIP), or other patterns of interstitial fibrosis in the lower lobe [8, 9]. Therefore, dense subpleural consolidation with fibrotic change, which is the characteristic radiological finding of PPFE, is not exclusive to idiopathic PPFE. However, the incidence of PPFE-like lesions on CT remains unclear and the features of PPFE-like lesions are also not well

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characterized. In addition, Yoshida et al. have reported two distinct patterns of the decline in forced vital capacity in patients with PPFE as follows: a rapid decline over a short period and a slow decline over a long period [10]. Therefore, changes in PPFE-like lesions may be heterogeneous. The objective of this study is threefold; the first objective is to determine the incidence of PPFE-like lesions in routine clinical practice; the objective is to evaluate the features of PPFE-like lesions; the third objective is to characterize changes in PPFE-like lesions on chest CT in routine clinical practice.

Materials and methods

The ethical review board at our institution waived the requirement of patients' approval or informed consent for the retrospective review of their records and images.

All patients who underwent chest CT in 2011 at one institution that was general city hospital were identified. 1366 patients were scanned with chest CT, and 82 patients for whom the pleura of upper lobe could not be evaluated (36 with resected right or left upper lobe; 15 undergoing radiation therapy at the upper lobe, 12 with atelectasis caused by pleural effusion at the upper lobe, 6 with atelectasis of the right or left upper lobe, 6 with pneumonia affecting the upper lobe, 4 with pneumothorax, and 3 with lung cancer of the upper lobe adjacent to the pleura) were excluded. Remaining 1284 patients [mean age 70.6 (range 17–100) years; 731 males and 553 females] were included. Height and weight data were not available for 89 patients; body mass index (BMI) was calculated for 1195 cases (Fig. 1).

CT images and review

CT images of all patients were obtained at end inspiration and in the supine position using several scanners. CT examinations were performed according to the routine protocols comprising 5-mm collimation sections and 5-mm intervals. Images were reviewed and findings were recorded by a chest

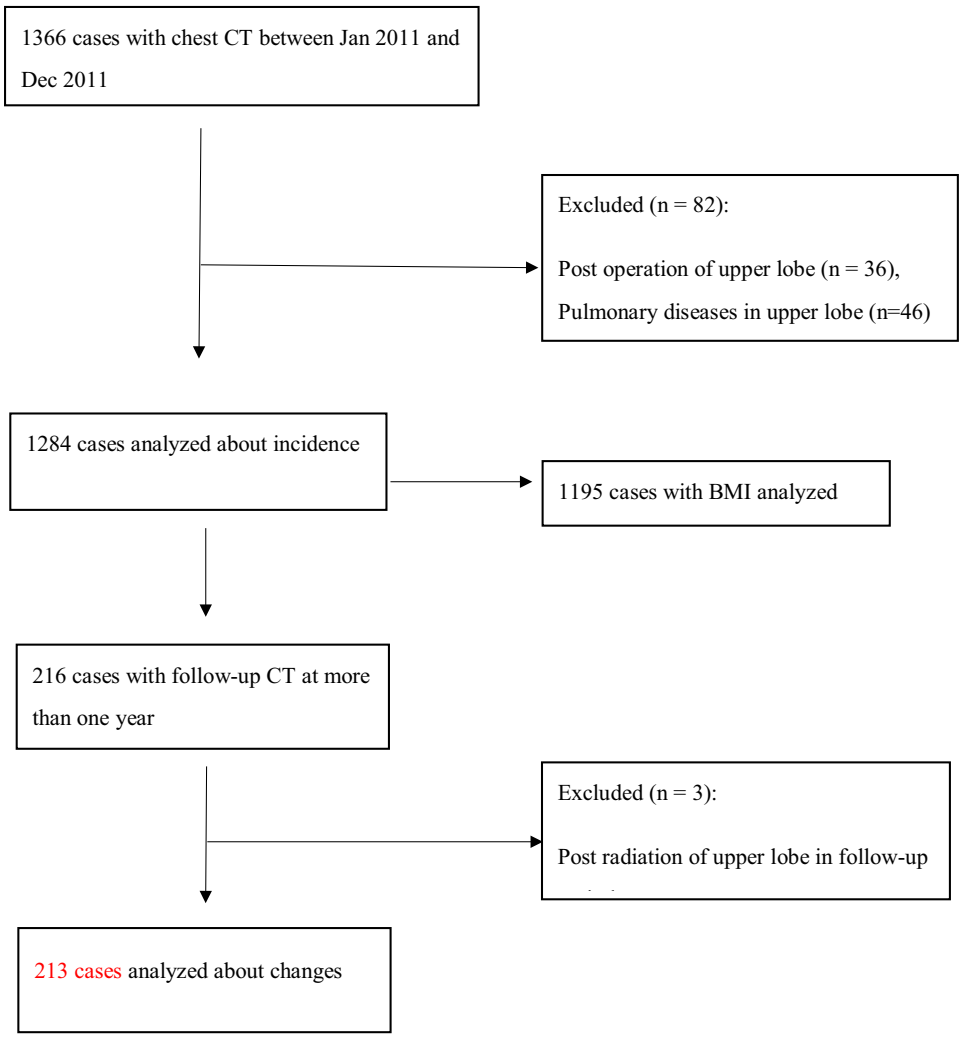

Fig. 1 Schematic illustration of the patient selection criteria

radiologist with 16 years of experience. Another radiologist with 29 years of experience reviewed 171 cases that are the cases for a period of 1 month to evaluate inter-observer variability.

The presence of PPFE-like lesion, which was defined as subpleural consolidation associated with the evidence of fibrosis in the upper lobe, was evaluated [5, 8]. The presence of other concomitant pulmonary findings, such as interstitial pneumonia (fibrotic changes in the bilateral lower lobes), emphysema, chronic airway disease (centrilobular nodules and/or bronchiectasis with segmental distribution), old tuberculosis (centrilobular nodules and/or segmental calcification in limited upper lung fields), silicosis (centrilobular nodules predominantly present in the upper lung), and pleural plaque, was also evaluated.

In cases that presented with PPFE-like lesions, the location, laterality, features, and thickness of the lesions as well as the surrounding features were evaluated. The caudal developments of lesions were evaluated by following four zones. The upper zone was defined as the area above the level of the sternoclavicular joint; the middle upper zone was defined as the area below the level of the sternoclavicular joint and above the level of the aortic arch; the middle lower zone was defined as the area below the level of the level of the aortic arch and above the level of the carina; the lower zone was defined as the area below the level of the carina. Four zones on axial images were divided according to their location. Medial zone was defined as the plane adjacent to the mediastinum. The other plane was divided into three equal zones, i.e., dorsal, lateral, and ventral zones. The laterality of the disease was evaluated according to the following five scales: 1, right only; 2, right predominant; 3, bilateral; 4, left predominant; and 5, left only. Features of PPFE-like lesions, including the presence of air bronchogram and calcification in the lesion, were evaluated. Lesion thickness was measured in the thickest part. Surrounding features of PPFE-like lesions, including the presence of nodular opacity, thickening of interlobular septa, consolidation along bronchus, bronchiectasis outside the PPFE-like lesion in the same cross-section, and the of volume upper lobe loss, were evaluated.

Follow-up

Cases with > 1-year follow-up CT were evaluated. Follow-up CT data were available for 216 patients; among these, three patients were excluded because they underwent radiation therapy during the follow-up. PPFE-like lesions were categorized according to their changes as follows: 1, resolved; 2, no change; 3, slight progression; and 4, distinct progression. Slight progression was defined as increase of PPFE-like lesions with no concomitant progression of volume loss. Distinct progression was defined as progression associated with volume loss. Another radiologist also reviewed 47 cases that that are the cases for a period of 1 month to evaluate inter-observer variability.

Statistical analysis

All statistical analyses were performed using statistical software (IBM SPSS Statistics, 2016; IBM, USA). Inter-observer variability with respect to the presence of, and the changes in, PPFE-like lesions was evaluated by kappa (κ) statistic. Inter-observer agreement was classified as follows: poor, $\kappa = 0$–0.20; fair, $\kappa = 0.21$–0.40; moderate, $\kappa = 0.41$–0.60; good, $\kappa = 0.61$–0.80; and excellent, $\kappa = 0.81$–1.00. The association between clinical findings and various CT characteristics of the lesions and their progression were assessed using univariate and multivariate analysis. A p value of < 0.05 was considered statistically significant. Correlations between age, BMI, and lesion thickness and progression were assessed using Mann–Whitney U test. Correlations between concomitant findings, features of PPFE-like lesions, and the location of axis and progression were analyzed using Fisher's exact test. Craniocaudal location was assessed by Chi-squared test.

Multivariate logistic regression analysis was used to assess the predictive value of various characteristics for the presence and progression of PPFE-like lesions. In multivariate analysis, variables were selected using a step-wise procedure. Findings that contributed to the power of regression equation ($p < 0.10$) were retained. BMI was not included in the multivariate analysis as relevant data was not available for all patients.

Result

The number of cases to evaluate the inter-observer agreement with respect to the presence of, and the changes in, PPFE-like lesions for 1 month was 171 cases and 47 cases. The result of inter-observer agreements with the presence of, and the changes in, PPFE-like lesions was fair and good ($\kappa = 0.26$ and 0.67, respectively). In addition, all cases of the broad extent PPFE-like lesion over middle zone were detected by both readers. PPFE-like lesions were observed in 397 of the 1284 patients (30.9%). The mean thickness of the PPFE lesions was 5.6 mm (range 2–22 mm). Correlations between the presences of lesions are presented in Table 1. PPFE-like lesions were observed in 221 of the 731 (30.2%) males and 176 of the 553 (31.8%) females; this difference was not significant ($p = 0.54$). Patients with PPFE-like lesions (mean age 72.2 ± 12.6 years) were significantly older than those without PPFE-like lesions (mean age 69.9 ± 13.2 years). In total, 32% (373 out of 1167) of the patients aged > 50 years showed PPFE-like

Table 1 Correlates of the presence of PPFE-like lesions

	All	Presence of PPFE-like lesion	percentage	*p* value
Sex (male/female)	731/553	221/176	30.2/31.8%	0.54†
Age (≤ 50/> 50 years)	117/1167	24/373	20.5/32.0%	0.006*†
Interstitial pneumonia	128	55	42.9%	0.002*‡
Emphysema	292	118	40.4%	< 0.0001*‡
Chronic airway disease	142	92	64.8%	< 0.0001*‡
Old tuberculosis	92	52	56.5%	< 0.0001*‡
Silicosis	3	2	66.7%	0.23‡
Pleural plaque	24	7	29.2%	1‡

PPFE pleuroparenchymal fibroelastosis

*There was a statistically significant difference ($p < 0.05$)

†p value with Mann–Whitney U test

‡p value with Fisher's exact test

lesions against 20% (24 of 117) of those aged < 50 years. BMI of patients with PPFE-like lesions was significantly lower than that of patients without PPFE-like lesions (mean BMI 21.0 vs. 23.2 kg/m^2, $p < 0.001$). Findings of concomitant interstitial pneumonia ($p = 0.002$), emphysema ($p < 0.001$), chronic airway disease ($p < 0.001$), and old tuberculosis ($p < 0.001$) were significantly correlated with the presence of PPFE-like lesions, whereas concomitant silicosis ($p = 0.23$) and pleural plaque ($p < 1$) did not significantly correlate with the presence of PPFE-like lesions. Among the 397 patients with PPFE-like lesions, 13.9, 29.7, 23.2, and 13.1% patients presented with concomitant interstitial pneumonia, emphysema, chronic airway disease, and old tuberculosis, respectively.

In the craniocaudal position of the 397 patients with PPFE-like lesions, 225 (56.7%) showed lesions in the upper zone, 110 (27.7%) in the upper middle zone, 62 (12.1%) in the lower middle zone, and 14 (3.5%) in the lower zone. Regarding the location of the axis, the most common location was dorsal ($n = 385$, 97.0%) and the least common location was ventral ($n = 44$, 11.1%). Lateral and medial positions were observed in 309 (77.8%) and 178 (44.8%) patients, respectively. In lateral position, 249 (62.7%) patients showed lesions in both lungs. In total, 18 (4.5%) patients showed lesions in the right lung only, 97 (24.4%) patients showed predominantly right-sided lesions, 29 (7.3%) patients showed predominantly left-sided lesions, and 4 (1%) patients showed lesions in the left lung only. In multivariate analysis, the presence of interstitial pneumonia [odds ratio (OR) 2.13; 95% confidence interval (CI) 1.45–3.14], emphysema (OR 1.83; 95% CI 1.37–2.44), chronic airway disease (OR 5.72; 95% CI 3.91–8.36), and old tuberculosis (OR 3.1; 95% CI 1.20–4.86) significantly correlated with the presence of PPFE-like lesions.

Follow-up

The mean follow-up period was 1276 (range 365–2182) days. Among 213 patients with follow-up CT, none of patients showed the resolution of lesions; 165 (77.5%) patients showed no change (Fig. 2); 33 (15.5%) patients showed slight progress; and 15 (7%) patients showed distinct

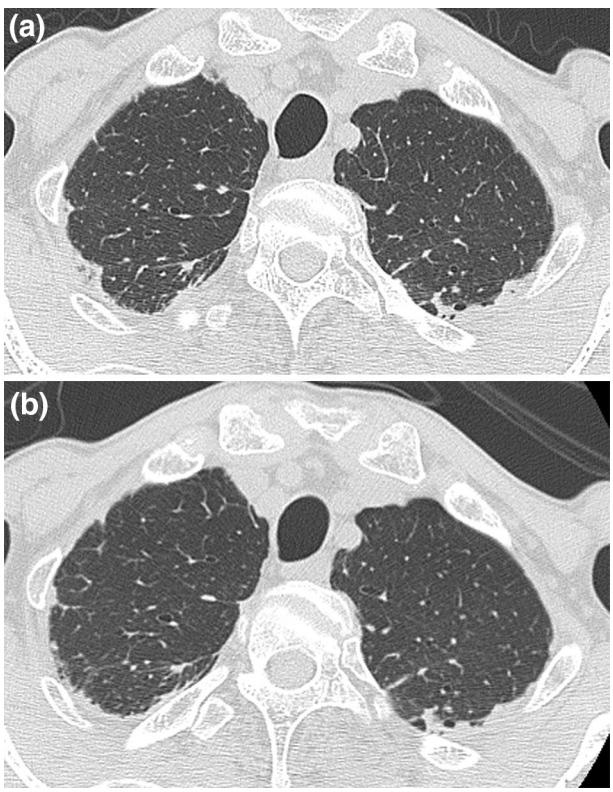

Fig. 2 CT radiographs of an 80-year-old man. **a** There are PPFE-like lesions in the bilateral dorsal portion and the right lateral portion. **b** Follow-up CT after 4 years showing no change in lesion

progress (Fig. 3). Patients who exhibited slight or distinct progress were classified as the progress group. Correlations between the features and progression of PPFE-like lesions are shown in Tables 2 and 3. Findings of interstitial pneumonia ($p = 0.001$), air bronchogram in PPFE-like lesions ($p = 0.001$), nodular opacity ($p < 0.001$), consolidation along the bronchus ($p = 0.025$), bronchiectasis outside PPFE-like lesions ($p = 0.001$), volume loss of the upper lobe ($p < 0.001$), the thickness of PPFE-like lesion ($p < 0.001$), caudal progression ($p = 0.001$), and ventral disease ($p < 0.001$) were significantly associated with lesion progression. In multivariate analysis, interstitial pneumonia (OR 3.81; 95% CI 1.62–8.94), nodular opacity (OR 3.78; 95% CI 1.17–12.20), and lesion thickness (OR 1.21; 95% CI 1.07–1.37) influenced progression.

Male sex, findings of interstitial pneumonia, air bronchogram in PPFE-like lesions, nodular opacity, consolidation along the bronchus, bronchiectasis outside the PPFE-like lesions, volume loss of upper lobe, thickness of PPFE-like lesions, caudal progression, and ventral disease were associated with distinct progress ($p = 0.03$, $p = 0.04$, $p = 0.021$, $p < 0.001$, $p < 0.001$, $p < 0.001$, $p < 0.001$, $p < 0.001$, and $p < 0.001$, respectively). In the distinct progress group, the number of male patients ($n = 12$) was more than that of female patients ($n = 3$). Seven male patients with distinct progression presented with interstitial pneumonia, while no female patients with distinct progression presented with interstitial pneumonia. In multivariate analysis, male sex (OR 7.14; 95% CI 1.13–45.1), interstitial pneumonia (OR 4.78; 95% CI 1.05–21.67), consolidation along the bronchus (OR 12.77; 95% CI 2.44–66.76), and ventral disease (OR 19.33; 95% CI 4.41–84.79) influenced distinct progression.

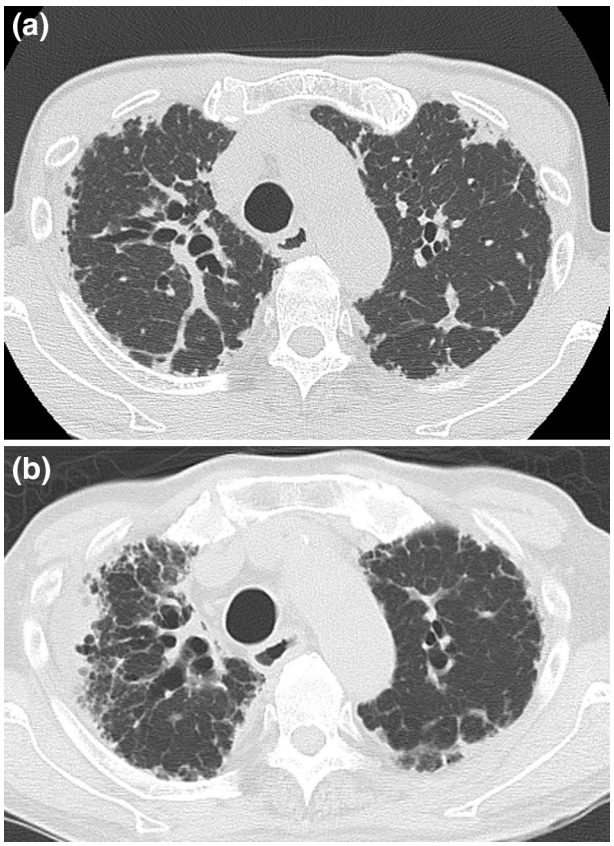

Fig. 3 CT radiograph of a 72-year-old man. **a** There are diffuse subpleural PPFE-like lesions with volume loss. **b** Follow-up CT after 3 years showing progression of PPFE-like lesions and volume loss

Discussion

On HRCT, PPFE shows dense subpleural consolidation with traction bronchiectasis, architectural distortion, and upper lobe volume loss [1]. These CT findings of PPFE are difficult to distinguish from those of an apical cap. PPFE lesions are broader and denser than apical cap; however, lesions borders between PPFE and apical cap remain unknown. Disease progression and associated clinical symptoms help differentiate PPFE from apical cap [11]. Therefore, we called the dense subpleural consolidation in upper lobe as PPFE-like lesion without differentiation between PPFE and apical cap.

In this study, a relatively high percentage (30.9%) of patients presented with PPFE-like lesions. Renner et al. have observed apical cap in 20.3% of cases on chest X-ray and in 40% of cases of lung autopsy [7]. Our results with CT findings were similar with the previous results of chest X-ray and lung autopsy. Although the exact cause of apical cap remains unclear, it is believed to be a consequence of intrinsic physiologic underperfusion compounded by recurrent inflammation and low-grade infection [12–14]. Both apical cap and PPFE are often accompanied by chronic infection. Im et al. have reported that apical cap is commonly observed in patients with tuberculosis [15]. Reddy et al. have reported that PPFE is associated with recurrent pulmonary infection [16]. In this study, old tuberculosis and chronic airway diseases strongly correlated with the presence of PPFE-like lesions. Repeated inflammatory damage might be the cause of PPFE-like lesions. In this study, emphysema correlated with PPFE-like lesions. In a previous study, a majority of patients with idiopathic PPFE have had a history of "never smoking" [17]; however, apical caps are common among smokers [18]. PPFE-like lesions associated with emphysema may cause apical caps.

On follow-up CT, interstitial pneumonia was found to be an important predictor of distinct progression. However, whether patients with interstitial pneumonia develop idiopathic PPFE remains unclear. Interstitial fibrosis in the lower lobe distant from PPFE has been described in patients with idiopathic PPFE [16, 19]. PPFE-like lesions have been

Table 2 Correlates of the progression of PPFE-like lesions

	No change ($n=165$)	Progress ($n=48$)	p value	Distinct progress ($n=15$)	p value
Sex (male/female)	82/83	30/18	0.14	12/3	0.032*†
Age (≤50/> 50 years)	9/156	1/47	0.46	0/15	1†
Thickness (mm)	5.3 ± 2.4	7.3 ± 4.1	< 0.001*	10.3 ± 5.0	< 0.001*†
Interstitial pneumonia	18	16	0.001*	7	0.004*‡
Emphysema	48	15	0.86	4	1‡
Chronic airway disease	38	13	0.57	7	0.054‡
Old tuberculosis	18	7	0.46	1	1‡
Silicosis	2	0	1	0	1‡
Pleural plaque	4	0	0.58	0	1‡
Air bronchogram	56	29	0.001*	10	0.021*‡
Calcification	12	7	0.14	0	0.37‡
Nodular opacity	6	12	< 0.001*	6	< 0.001*‡
Thickening of interlobular septa	77	25	0.52	10	0.18‡
Consolidation along the bronchus	37	19	0.025*	12	< 0.001*‡
Bronchiectasis outside PPFE-like lesion	6	9	0.001*	8	< 0.001*‡
Volume loss of the upper lobe	16	17	< 0.001*	12	< 0.001*‡
Ventral lesion	11	13	< 0.001*	11	< 0.001*‡

PPFE pleuroparenchymal fibroelastosis

*There was a statistically significant difference ($p<0.05$)

†p value with Mann–Whitney U test

‡p value with Fisher's exact test

Table 3 Correlates of the progression and the craniocaudal location of PPFE-like lesions

	Above the sternoclavicular joint	Above the aortic arch	Above the carina	Below the carina	p value
No change	100	49	12	4	
Progress	17	16	10	5	0.001*◆
Distinct progress	2	3	5	5	< 0.001*◆

PPFE pleuroparenchymal fibroelastosis

*There was a statistically significant difference ($p<0.05$)

◆p value with Chi-square test

reported in patients with idiopathic pulmonary fibrosis and connective tissue disease-related interstitial pneumonia [8, 20]. Enomoto et al. have reported the frequent progression of PPFE-like lesions (5/8, 63%) [20]. However, the classification of patients with concomitant interstitial fibrosis in the lower lobe and PPFE-like lesion in the upper lobe remains unclear. Moreover, interstitial fibrosis in the lower lobe is considered to be an important factor for PPFE-like lesion progression.

Male sex was a significant predictor of distinct progression; however, this association may have been confounded by the presence of interstitial pneumonia. A majority of males who showed distinct progression presented with interstitial pneumonia, but no female who showed distinct progression presented with interstitial pneumonia. Interstitial fibrosis in the lower lobe would be an important factor for PPFE-like lesion progression, particularly in males. Additional investigation with more cases is warranted.

The findings and the extent of PPFE-like lesion also influenced the progression. Thickness and perilesional characteristics, such as nodular opacity, consolidation along the bronchus, bronchiectasis outside PPFE-like lesion, and the volume loss of upper lobe, affected progression. Broader caudal and ventral distributions were important predictors of progression. However, few patients with narrow distribution only in the area above the level of the sternoclavicular joint showed distinct progression.

There are some limitations in our study. First, this was a retrospective study. Second, patients were diagnosed only by CT, and histological diagnosis was not performed in

any patient. Therefore, it was unknown whether idiopathic PPFE was included in this study. Whether patients with CT findings of subpleural consolidation showed the same histological features remains unknown. Third, clinical data were limited. Tuberculosis, small airway disease, silicosis, and other diseases were not clinically evaluated in detail. Fourth, weight and height data for all patients were not available; thus, BMI was not included in multivariate analysis. Fifth, although 1284 patients were included in this study, follow-up CT data were available only for 213 patients of which only 48 showed progression; this number is insufficient to discuss correlations with progression. Sixth, CT findings of most patients were evaluated by one reader; two readers could not evaluate data owing to the high number of cases. Therefore, the results in this study were not decided by consensus reading. Seventh, a fair level of inter-observer agreement was observed for the presence of PPFE-like lesions, likely because mild apical subpleural abnormalities such as old tuberculosis were common and were difficult to differentiate from PPFE-like lesions. On the other hand, all cases of broad extent PPFE-like lesion over middle zone were detected by both readers. The inter-observer agreement would be high in the cases with broad PPFE-like lesion.

In conclusion, PPFE-like lesions are not rare findings on chest CT and are correlated with older age; lower BMI; and the presence of concomitant interstitial pneumonia, emphysema, chronic airway disease, and old tuberculosis. On follow-up, PPFE-like lesions in some patients showed progression; interstitial pneumonia, lesion thickness, and findings surrounding PPFE-like lesions predicted progression.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical standards The ethical review board at our institution waived the requirement of patients' approval or informed consent for the retrospective review of their records and images.

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