SUNTEXT REVIEWS

SunText Review of Case Reports & Images
ISSN: 2766-4589

Open Access
Case Report
Volume 7:1

Combined Pulmonary Fibrosis and Emphysema (CPFE) Syndrome: A Case Report and Practical Approach

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Received date: 13 January 2026; **Accepted date:** 17 January 2026; **Published date:** 28 January 2026

Citation: Kumar P, Sinha S, Coombe MG (2026) Combined Pulmonary Fibrosis and Emphysema (CPFE) Syndrome: A Case Report and Practical Approach. SunText Rev Case Rep Image 7(1): 170.

DOI: https://doi.org/10.51737/2766-4589.2026.170

Abstract

Background: Combined pulmonary fibrosis and emphysema (CPFE) is a distinct clinico-radiological syndrome defined by the co-presence of emphysema and fibrotic interstitial lung disease (ILD). It is clinically important because it is frequently accompanied by disproportionate exertional hypoxemia, a high burden of pulmonary hypertension, vulnerability to acute respiratory deteriorations, and an increased risk of lung cancer.

Case Presentation: We report a 79-year-old male ex-smoker with severe smoking-related chronic obstructive pulmonary disease (COPD)/emphysema and bronchiectasis who presented with several days of worsening dyspnea, increased cough, and increased sputum production. He was diagnosed with an infective COPD exacerbation complicated by community-acquired pneumonia. A non-contrast computed tomography (CT) scan demonstrated severe centrilobular and paraseptal emphysema with basal-predominant subpleural reticular change, lingular consolidation, and scattered micronodules. Comprehensive pulmonary function testing showed severe airflow obstruction with marked hyperinflation and a disproportionately severe reduction in gas transfer (diffusing capacity for carbon monoxide [DLCO] 32.5% predicted). Transthoracic echocardiography performed during admission demonstrated normal right ventricular size and function with an estimated right ventricular systolic pressure (RVSP) of 28 mmHg, without evidence of significant pulmonary hypertension at that time.

Conclusion: This case illustrates a classic CPFE phenotype, integrating characteristic CT findings with the typical physiological signature of marked gas transfer impairment despite spirometric obstruction. It highlights a practical approach to identifying CPFE in patients labelled primarily as COPD, outlines a structured differential diagnosis (including asbestos-related disease in the appropriate exposure context), and supports a longitudinal management strategy focused on optimizing oxygenation, preventing exacerbations, surveilling for pulmonary hypertension, and maintaining vigilance for pulmonary malignancy.

Keywords: Combined pulmonary fibrosis and emphysema; CPFE; Emphysema; Interstitial lung disease; DLCO; Pulmonary hypertension; Pulmonary nodules; Bronchiectasis

Introduction

Combined pulmonary fibrosis and emphysema (CPFE) describes the coexistence of upper-lobe predominant emphysema and lower-lobe predominant fibrotic interstitial lung disease, most often seen in older individuals with a substantial cigarette smoking history. CPFE may be under-recognised because conventional spirometry can underestimate overall parenchymal disease severity; lung volumes may be relatively preserved or even increased due to hyperinflation, while gas exchange is often

markedly impaired. A key clinical clue is a disproportionate reduction in diffusing capacity for carbon monoxide (DLCO) relative to the degree of airflow obstruction. Recognition matters because CPFE carries a distinctive complication profile, particularly pulmonary hypertension and an increased incidence of lung cancer, and therefore benefits from deliberate diagnostic labelling and a structured follow-up approach that extends beyond COPD management alone [1-3].

Case Presentation

A 79-year-old man, an ex-smoker who ceased tobacco use approximately five years prior to presentation, was admitted with a several-day history of progressive worsening of baseline dyspnea, increased cough, and increased sputum production. His medical history included severe physician-diagnosed COPD/emphysema, bronchiectasis, hypertension, and hyperuricemia. He appeared cachectic with a body mass index (BMI) of approximately 17 kg/m². His resting oxygen saturation on room air was 94% at initial assessment. His remote surgical history included pleurodesis at approximately 18 years of age for recurrent pleural effusions of uncertain aetiology. Occupationally, he reported substantial historical exposure to inorganic dusts, including coal dust and asbestos. The clinical course is summarised chronologically. The patient initially presented on 27 December 2025, at which time venous blood gas sampling was performed. A non-contrast CT chest was completed on 28 December 2025. He was transferred to a tertiary centre for ongoing inpatient care on 29 December 2025. Further inpatient assessment included full pulmonary function testing on 7 January 2026, and transthoracic echocardiography with selected autoimmune serology on 8 January 2026. He was treated for an acute infective exacerbation of COPD complicated by community-acquired pneumonia. Examination findings documented in clinical notes included bilateral basal inspiratory crackles (more prominent on the right) and diffuse expiratory wheeze. The contemporaneous working diagnosis emphasised severe COPD/emphysema, consistent with his established history.

Diagnostic Investigations

Venous Blood Gas Analysis

A venous blood gas sample obtained on room air (FiO_2 0.21) showed pH 7.44, pCO_2 44 mmHg, and pO_2 41 mmHg, with bicarbonate 30 mmol/L, base excess +4.7 mmol/L, and lactate 2.7 mmol/L. This profile was interpreted as compatible with acute infective physiology and a metabolic component, without evidence of overt acute hypercapnic respiratory failure at that time.

Computed Tomography of the Chest

Non-contrast CT imaging demonstrated severe centrilobular and paraseptal emphysema throughout the upper and mid-lung zones. Basal-predominant subpleural reticular change and fine reticulation were present, most evident within the left lower lobe and lingula, consistent with fibrotic interstitial abnormality. Acute infective change was represented by consolidation in the superior segment of the lingula. A previously described nodular opacity had largely resolved; however, a new ill-defined 6 mm nodular opacity was noted in the left upper lobe/lingular region, with additional scattered micronodules. Bibasal pleural thickening was described, without pleural effusion or pneumothorax. Mediastinal lymph nodes were not enlarged by size criteria. Overall, the imaging pattern supported a CPFE phenotype (coexistent emphysema and basal fibrotic change) with superimposed infection.

Pulmonary Function Testing

Full pulmonary function testing (Table 1) demonstrated severe airflow obstruction post-bronchodilator (FEV_1 0.98 L, 43.5% predicted; FEV_1/FVC 52%) with marked hyperinflation and air trapping (TLC 152.9% predicted; RV 264.1% predicted). Gas transfer was severely impaired (DLCO 32.5% predicted; KCO 37.7% predicted). The combination of obstruction, profound hyperinflation, and a disproportionately severe reduction in DLCO is physiologically consistent with CPFE.

***Table 1**: Comprehensive Pulmonary Function Test Results (07 January 2026).*

Parameter	Measured Value	Percent of Predicted
Spirometry (post-bronchodilator)		
FEV_1	0.98 L	43.5%
FVC	1.89 L	63.3%
FEV_1/FVC	52%	—
Lung Volumes (Body Plethysmography)		
TLC	8.65 L	152.9%
RV	6.72 L	264.1%
RV/TLC	77.6%	—
Diffusing Capacity (Single-Breath)		

DLCO	6.64 mL/min/mmHg	32.5%
KCO	—	37.7%
VA	—	87.5%

Transthoracic Echocardiography

Transthoracic echocardiography was performed as screening for pulmonary hypertension. Left ventricular systolic function was preserved (ejection fraction 57%). Right ventricular size and systolic function were normal. Trace to mild valvular regurgitation was present. Estimated RVSP was 28 mmHg, within the normal range and not suggestive of significant pulmonary hypertension at the time of testing, although imaging windows were described as technically difficult.

Serological Investigations

Selected serology was performed to evaluate for connective tissue disease-associated ILD and other systemic drivers. Rheumatoid factor and anti-cyclic citrullinated peptide antibodies were negative. Serum angiotensin-converting enzyme level was within the reference range.

Diagnostic Reasoning and Integration

The diagnosis of CPFE was made by integrating radiological and physiological evidence within a compatible clinical phenotype. CT imaging demonstrated emphysema (severe centrilobular and paraseptal change) together with basal subpleural reticulation suggestive of fibrotic ILD. Pulmonary function testing quantified severe airflow obstruction and hyperinflation attributable to emphysema, alongside marked impairment in gas transfer consistent with significant alveolar-capillary involvement from fibrotic parenchymal disease. Clinically, the patient's demographic profile (older male ex-smoker), cachexia, and substantial dyspnea burden supported CPFE as the unifying chronic diagnosis, with the acute admission reflecting an infective exacerbation and pneumonia occurring on the background of this dual-pathology syndrome [1-3].

Differential Diagnosis Considerations

In patients with emphysema plus basal interstitial abnormality, a deliberate differential diagnosis helps avoid misclassification and supports appropriate surveillance and prognostication.

- Asbestos-related pleuroparenchymal disease: The exposure history and bibasal pleural thickening raise the possibility of asbestos-related disease. A specialist ILD radiology review is useful to evaluate for supportive features such as pleural plaques, parenchymal bands, rounded atelectasis, and a fibrosis pattern consistent with asbestosis [4].
- Smoking-related interstitial abnormalities: Heavy smoking may be associated with interstitial changes that range from subtle smoking-related fibrosis to smoking-related ILDs. Radiological overlap with other fibrotic entities is common, and multidisciplinary discussion is often required.
- Idiopathic pulmonary fibrosis (IPF) with coexistent emphysema: This is a clinically important CPFE subgroup. Determining whether the fibrotic component meets criteria for usual interstitial pneumonia (UIP) or probable UIP influences prognosis and the consideration of antifibrotic therapy [6].
- Chronic hypersensitivity pneumonitis: Fibrotic hypersensitivity pneumonitis can coexist with emphysema and air trapping. Features such as mosaic attenuation, mid-zone involvement, and poorly defined centrilobular nodules, combined with a careful exposure history, support this diagnosis [7].
- Connective tissue disease-associated ILD: Negative rheumatoid factor and anti-CCP reduce the likelihood of rheumatoid arthritis-associated ILD but do not exclude other connective tissue diseases. A systematic review for extrapulmonary features and broader serological evaluation may be appropriate depending on the clinical context [8].

Management Strategy

Acute Inpatient Management

Acute management followed standard approaches for an infective COPD exacerbation with community-acquired pneumonia. Treatment included intravenous antibiotics directed at common respiratory pathogens, systemic corticosteroids for airway inflammation, and frequent bronchodilator therapy. Because bronchiectasis was present, airway clearance strategies and chest physiotherapy were emphasised. Supportive measures included venous thromboembolism prophylaxis and careful oxygen titration to maintain adequate oxygenation while monitoring for carbon dioxide retention risk [9].

Longitudinal and Multidimensional CPFE Management Plan

Recognition of CPFE should trigger a structured plan that addresses emphysema, fibrosis, bronchiectasis, and the syndrome's major complications.

- Definitive ILD phenotyping and baseline assessment: Review of CT imaging within an ILD multidisciplinary team (MDT) is central to clarifying the fibrotic pattern (UIP/probable UIP/indeterminate/alternative diagnosis) and guiding prognosis and potential disease-modifying therapy. Baseline symptom quantification, resting and exertional oxygenation assessment, and pulmonary function measures establish a reference point for monitoring [6].

- Optimisation of inhaler therapy and exacerbation prevention: Long-acting bronchodilators should be continued for symptom control. The decision to include inhaled corticosteroids should be individualised, balancing exacerbation reduction against pneumonia risk, which may be particularly relevant in older patients with bronchiectasis. Vaccination against influenza, pneumococcus, and COVID-19 should be ensured, and a written action plan for exacerbations should be provided [9,10].
- Bronchiectasis-specific management: Airway clearance techniques should be reinforced with physiotherapy support. Sputum culture should be obtained during exacerbations to guide antibiotic selection. In patients with frequent exacerbations, consideration of long-term macrolide therapy or inhaled antibiotics should align with bronchiectasis guideline recommendations and specialist oversight [10].
- Oxygenation, exercise capacity, and nutrition: A six-minute walk test with continuous oximetry is recommended to document exertional desaturation and guide ambulatory oxygen prescription. Assessment for long-term oxygen therapy should follow standard criteria where indicated. Pulmonary rehabilitation should be offered to improve functional status and quality of life. Given marked cachexia, dietitian involvement is appropriate to address nutritional depletion and muscle wasting, which adversely affect outcomes in chronic lung disease [9].
- Surveillance for pulmonary hypertension: Pulmonary hypertension is a major determinant of morbidity and mortality in CPFE, and progression can be clinically subtle. Repeat echocardiography within 6–12 months (or earlier if symptoms worsen disproportionately, DLCO declines, or signs of right heart strain emerge) is reasonable. If echocardiography suggests significant pulmonary hypertension, right heart catheterisation remains the diagnostic gold standard and guides management decisions [11].
- Pulmonary nodule follow-up: The new 6 mm ill-defined nodule and scattered micronodules warrant structured, risk-based surveillance. Follow-up should align with established guidance such as the Fleischner Society recommendations, incorporating smoking history, age, and nodule characteristics to balance early cancer detection with avoidance of unnecessary imaging [12].
- Consideration of antifibrotic therapy: CPFE alone does not automatically indicate antifibrotic therapy. If the fibrotic component is classified as IPF or if criteria for progressive pulmonary fibrosis are met, then nintedanib or pirfenidone should be considered after discussion of risks and expected benefits [6].
- Safety-netting and advance care planning: Because acute deteriorations in CPFE may arise from infection, COPD exacerbation, acute ILD exacerbation, pulmonary embolism, or cardiac decompensation, clear safety-net advice should be provided. Values-based discussions regarding goals of care and escalation preferences are an important part of comprehensive, longitudinal management.

Outcome and Recommended Follow-up

The patient improved clinically with inpatient therapy including antibiotics, systemic corticosteroids, bronchodilator optimisation, and airway clearance measures, and he was stabilised for discharge. The broader diagnostic work-up clarified the underlying CPFE syndrome. Post-discharge review in 4–6 weeks was recommended to reassess symptoms and oxygenation following recovery from acute infection. Pulmonary function testing, with attention to DLCO trajectory, should be repeated at least annually. A follow-up non-contrast CT chest in 3–6 months was recommended to document resolution of consolidation and to reassess pulmonary nodules within a structured protocol. Repeat echocardiography for pulmonary hypertension surveillance should be incorporated into ongoing care (Table 2).

***Table 2**: Summary of Key Findings on Non-Contrast CT Chest.*

Region / Pattern	Findings
Emphysema	Severe, confluent centrilobular and paraseptal emphysema, predominantly in upper and middle lung zones.
Interstitial / fibrotic change	Basal-predominant, subpleural reticular opacities and fine reticulation, most pronounced in the left lower lobe and lingula.
Consolidation (acute)	Patchy consolidation in the superior segment of the lingula consistent with pneumonia; a smaller focus of ground-glass opacity/consolidation also described in the right upper lobe.
Pulmonary nodules	New 6 mm ill-defined, non-calcified nodular opacity in the left upper lobe/lingular region; additional scattered sub-centimetre micronodules.
Pleura	Symmetrical bibasal pleural thickening; no pleural effusion or pneumothorax.
Mediastinum / hila	No lymph nodes >1 cm short-axis; cardiac silhouette described as normal.

Discussion

This case highlights the clinical value of recognising CPFE when emphysema and fibrotic interstitial abnormality coexist. The severe DLCO reduction (32.5% predicted), occurring alongside pronounced hyperinflation and airflow obstruction, is a characteristic physiological pattern that should prompt clinicians to consider CPFE rather than attributing breathlessness solely to COPD [1-3]. Reliance on FEV_1 alone can obscure the true extent of parenchymal and vascular involvement, particularly when emphysema-related hyperinflation masks restriction. Management in CPFE is inherently anticipatory. Pulmonary hypertension is a central complication that may develop over time and can dominate prognosis, supporting a low threshold for repeat assessment when symptoms or physiological markers change [11]. Similarly, the increased risk of lung cancer necessitates disciplined follow-up of pulmonary nodules, particularly in older ex-smokers with emphysema and fibrotic change [12]. Acute deteriorations are often multifactorial, and distinguishing between infection, COPD exacerbation, cardiac failure, pulmonary embolism, and acute ILD exacerbation is crucial because treatment strategies differ substantially [6,9].

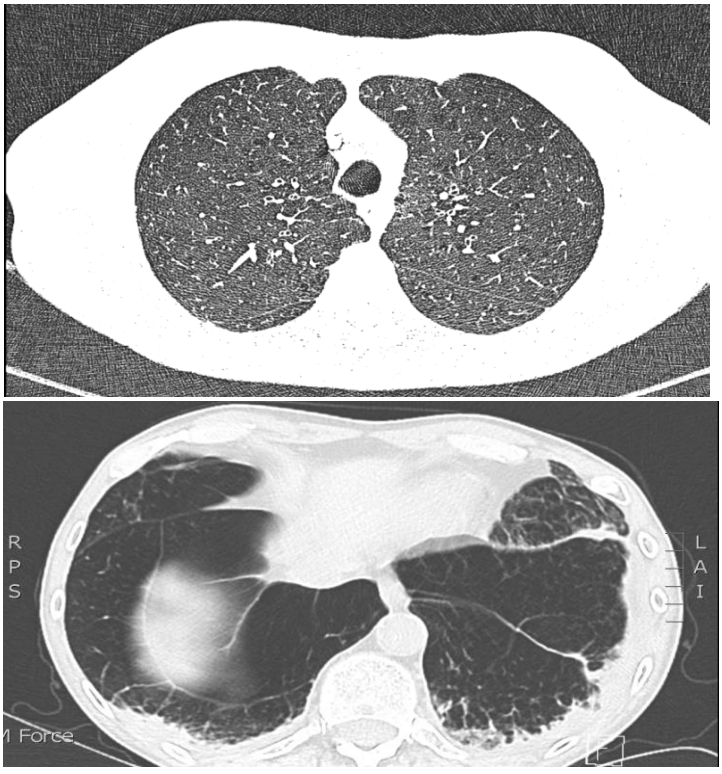
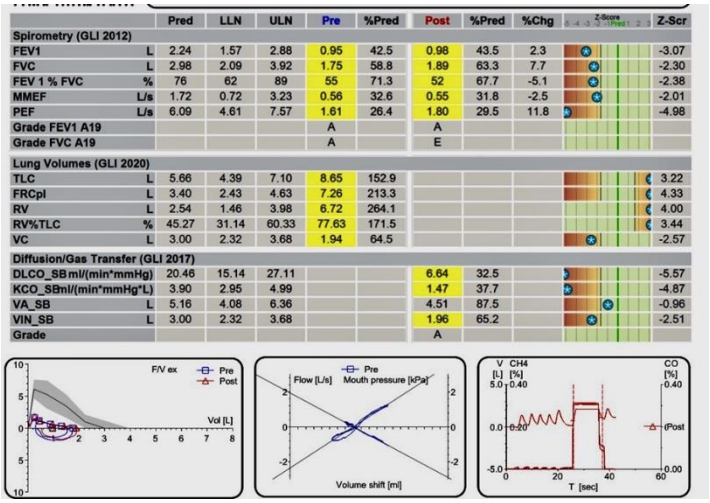


***Figure 1,2**: Representative axial non-contrast CT image. Areas of severe centrilobular and paraseptal emphysema are identified by focal regions of low attenuation without visible walls. Concurrent basal subpleural reticulation is present, seen as linear opacities parallel to the pleural surface. The juxtaposition of emphysema and basal fibrotic change demonstrates the characteristic imaging phenotype of CPFE.*



		Pred	LLN	ULN	Pre	%Pred	Post	%Pred	%Chg	Z-Scr
Spirometry (GLI 2012)										
FEV1	L	2.24	1.57	2.88	0.95	42.5	0.98	43.5	2.3	-3.07
FVC	L	2.98	2.09	3.92	1.75	58.8	1.89	63.3	7.7	-2.30
FEV 1 % FVC	%	76	62	89	55	71.3	52	67.7	-5.1	-2.38
MMEF	L/s	1.72	0.72	3.23	0.56	32.6	0.55	31.8	-2.5	-2.01
PEF	L/s	6.09	4.61	7.57	1.61	26.4	1.80	29.5	11.8	-4.98
Grade FEV1 A19					A		A			
Grade FVC A19					A		E			
Lung Volumes (GLI 2020)										
TLC	L	5.66	4.39	7.10	8.65	152.9				3.22
FRCpl	L	3.40	2.43	4.63	7.26	213.3				4.33
RV	L	2.54	1.46	3.98	6.72	264.1				4.00
RV%TLC	%	45.27	31.14	60.33	77.63	171.5				3.44
VC	L	3.00	2.32	3.68	1.94	64.5				-2.57
Diffusion/Gas Transfer (GLI 2017)										
DLCO_SB ml/(min*mmHg)		20.46	15.14	27.11			6.64	32.5		-5.57
KCO_SBml/(min*mmHg*L)		3.90	2.95	4.99			1.47	37.7		-4.87
VA_SB	L	5.16	4.08	6.36			4.51	87.5		-0.96
VIN_SB	L	3.00	2.32	3.68			1.96	65.2		-2.51
Grade							A			

***Figure 3**: Graphical representation of key pulmonary function abnormalities. The flow-volume loop demonstrates severe expiratory airflow obstruction with marked concavity. Lung volume indices illustrate pronounced hyperinflation with a markedly elevated residual volume. Gas transfer indices demonstrate severe DLCO reduction, reflecting disproportionate impairment in alveolar-capillary diffusion typical of CPFE.*

A notable nuance in this patient is the history of asbestos exposure and bibasal pleural thickening, which raises the possibility of asbestos-related pleuropulmonary disease. Accurate differentiation between asbestosis and other fibrotic ILDs (including IPF) can be challenging and is best resolved through ILD MDT review that integrates imaging pattern recognition with occupational history and clinical context [4]. In summary, CPFE should be approached as more than "COPD plus scarring." Diagnostic recognition provides a framework for surveillance, risk stratification, and holistic care planning that targets the syndrome's dominant complications and aims to preserve functional status and quality of life (Figures 1-3).

Learning Points for Clinicians

- CPFE should be considered in older smokers/ex-smokers with emphysema when CT imaging shows concomitant basal fibrotic change and pulmonary function testing demonstrates a markedly reduced DLCO that is disproportionate to spirometric obstruction.
- Management of CPFE extends beyond standard COPD pathways and should include structured surveillance for pulmonary hypertension, guideline-based pulmonary nodule follow-up, and careful phenotyping of the fibrotic ILD component.
- Optimal care is multidisciplinary and should address bronchodilator optimisation, airway clearance, oxygen assessment, pulmonary rehabilitation, nutrition, and advance care planning.

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Patient Consent Statement

Written informed consent for publication of this de-identified case report and any accompanying images will be obtained from the patient prior to submission. All personal identifiers have been removed to protect patient confidentiality

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