

## Combined Pulmonary Fibrosis and Emphysema: A Case Report and Literature Review

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### ABSTRACT

The syndrome of combined emphysema of the upper lobes and fibrosis of the lower lobes on chest computed tomography results in a characteristic functional profile, with preserved lung volumes, hypoxemia at exercise. Despite subnormal spirometry, combined pulmonary fibrosis and emphysema is a severe entity. The presence of pulmonary arterial hypertension at diagnosis is a critical determinant of prognosis.

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### Introduction:

Chronic obstructive pulmonary disease (COPD) and idiopathic interstitial pneumonias (IIP), with different radiological, pathological, functional and prognostic characteristics, have been regarded as separate entities for a long time. However, there is an increasing recognition of the coexistence of emphysema and pulmonary fibrosis in individuals (1). Combined pulmonary fibrosis and emphysema (CPFE) has been mentioned in

the past in series of patients with IPF or has been the subject of case reports or short series (2). Patients with this entity have relatively preserved lung volumes and spirometry but marked reductions in diffusing capacity on pulmonary function testing. Smoking appears to be the predominant risk factor for this disorder (3). Previous reported cases of CPFE describe upper lobe bullous disease and lower lobe fibrosis with honeycombing (4).

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### Case presentation

The patient was a 61-year-old man who presented with increased shortness of breath during activity and exacerbation of cough and sputum. The patient, a smoker, smoked 40 packs a year. He has been suffering from an increase of exertional dyspnea for about a year, which he considered to be related to smoking and didn't seek medical consultation. The patient's medical records includes a history of high blood pressure and a drug history of amlodipine 5 mg twice daily. On physical examination the following data were recorded: PR=100 beats/minute, RR=26 breaths/minute, BP=120/80 mmHg and his O<sub>2</sub> saturation in room air was 88%. On lung examination, Velcro-like crackles were heard at the base of the lungs and diffuse wheezing was heard during exhale. On limb examination, clubbing was observed and (+1) edema was present in the lower limb. Other examinations of the patient were normal. Routine tests such as CBC, ESR, BS, Renal and Hepatic function tests were performed and the results were in the normal range. The VBG test results were as follow: PH= 7.36, PCO<sub>2</sub>= 58, HCO<sub>3</sub>=33, indicating a chronic respiratory acidosis and metabolic alkalosis.

CXR imaging showed increased reticular pattern at the base of the lung and over inflation in the upper part of the lung therefore, due to lung involvement in the X-ray, a CT scan of the lung was done for the patient. The HRCT results were described as predominantly Para septal emphysema of the upper zone and lower zone fibrosis with intra lobular septa thickening and ground glass opacities and honeycombing Figure 1 (A-C). Based on the CT scan view, further decisions were made and serum immunologic profile including RF, P-ANCA, C-ANCA, HIV-antibody and ANA was checked, the results of which was all negative.

An echocardiography was performed for the patient according to which: EF=50%, pulmonary artery hypertension with pulmonary artery systolic pressure = 60 mmHg and dilated right atrium and ventricle.

Then a Pulmonary function testing (body box plethysmography) was done for the patient, the result of which was as follow: FVC: 70%, FEV<sub>1</sub>= 60%, TLC= 65%, RV=70%,  $\frac{FEV_1}{FVC}$  = 71%. It is worth mentioning that without a response to a bronchodilator agent FEV<sub>1</sub> increased approximately by 6%.

Based on the clinical and imaging findings, the patient was diagnosed with combined pulmonary fibrosis and emphysema (CPFE). COPD treatment was initiated for the patient and he was prescribed with triple spray therapy (LABA, ICS & LAMA), NAC tabs 600mg every 12 hours, pantoprazole 20mg daily and azithromycin 500mg every other day simultaneously. As for the patient's hypoxia, he was advised to supplementary oxygen use especially at nights. Following the start of drug treatment and the use of oxygen therapy, the patient's shortness of breath improved

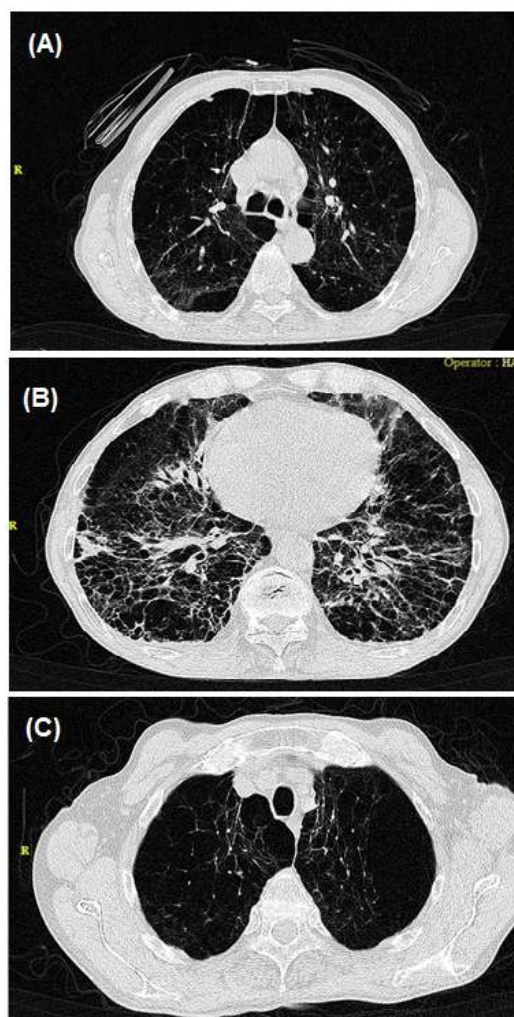


Figure 1 (A-C): lung HRCT

## Conclusion

CPFE is a separate entity that is considered less by doctors. In any patient presented with a clinical profile highly suspicious of obstructive pulmonary disease or emphysema, and in patients with a long history of smoking, if changes in lung volume (in spirometry or body box plethysmography) and alterations in CT scan - that is, despite emphysema, evidence Pulmonary fibrosis is also noticeable - is observed, then it's best to make sure of including this disease in the list of differential diagnosis held by the physician since these patients usually have a worse prognosis and higher mortality and suffer from complications of hypertension.

CPFE is still a rarely recognized clinical entity which may be overlooked due to subnormal lung volumes. Respiratory physicians should be aware of its existence and take more appropriate treatments while evaluating patients with severe impaired diffusing capacities out of proportion to their total lung volumes, or with pulmonary hypertension coexisting with a mixed obstructive/restrictive lung function abnormality, especially in a current or former smoker.

## References

1. Lin H, Jiang S. Combined pulmonary fibrosis and emphysema (CPFE): an entity different from emphysema or pulmonary fibrosis alone. *Journal of thoracic disease*. 2015;7(4):767.
2. Hasani, S., Mozdourian, M. Chronic obstructive pulmonary disease and bone health: A review on possible links and follow up studies. *Journal of Cardio-Thoracic Medicine*, 2020; 8(4): 684-692
3. Jankowich MD, Polsky M, Klein M, Rounds S. Heterogeneity in combined pulmonary fibrosis and emphysema. *Respiration*. 2008;75(4):411-7.
4. Cottin V, Brillet P-Y, Nunes H, Cordier J-F. Combined pulmonary fibrosis and emphysema. *Presse medicale (Paris, France: 1983)*. 2007;36(6 Pt 2):936-44.
5. American Thoracic S. Chronic bronchitis, asthma, and pulmonary emphysema: a statement by the Committee on Diagnostic Standards for Nontuberculous Respiratory Diseases. *Am Rev Respir Dis*. 1962;85:762-28.
6. Katzenstein A-LA, Myers JL. Idiopathic pulmonary fibrosis: clinical relevance of pathologic classification. *American journal of respiratory and critical care medicine*. 1998;157(4):1301-15.
7. Jankowich MD, Rounds SIS. Combined pulmonary fibrosis and emphysema syndrome: a review. *Chest*. 2012;141(1):222-31.
8. Marten K, Milne D, Antoniou KM, Nicholson AG, Tennant RC, Hansel TT, et al. Non-specific interstitial pneumonia in cigarette smokers: a CT study. *European radiology*. 2009;19(7):1679-85.
9. Cottin V, Cordier J-F. The syndrome of combined pulmonary fibrosis and emphysema. *Chest*. 2009;136(1):1-2.
10. Kitaguchi Y, Fujimoto K, Hanaoka M, Kawakami S, Honda T, Kubo K. Clinical characteristics of combined pulmonary fibrosis and emphysema. *Respirology*. 2010;15(2):265-71.
11. Odani K, Yoriko M, Shoji Y. Computed tomographic evaluation in the cases of coexistence of pulmonary emphysema with idiopathic pulmonary fibrosis. *Journal of Tomography (Japan)*. 2004;31(1):25-9.
12. Hiwatari N, Shimura S, Takishima T. Pulmonary emphysema followed by pulmonary fibrosis of undetermined cause. *Respiration*. 1993;60(6):354-8.
13. Ryerson CJ, Hartman T, Elicker BM, Ley B, Lee JS, Abbritti M, et al. Clinical features and outcomes in combined pulmonary fibrosis and emphysema in idiopathic pulmonary fibrosis. *Chest*. 2013;144(1):234-40.
14. Cottin V, Nunes H, Brillet P, Delaval P, Devouassoux G, Tillie-Leblond I, et al. Combined pulmonary fibrosis and emphysema: a distinct underrecognised entity. *European Respiratory Journal*. 2005;26(4):586-93.
15. Chamani J. Energetic domains analysis of bovine  $\alpha$ -lactalbumin upon interaction with copper and dodecyl trimethylammonium bromide. *Journal of Molecular Structure*. 2010;979(1-3):227-234.
16. Neghab M, Rahimi E, Emad A, Fard ARR. An epidemiological study of talc-related respiratory morbidity among employees of a rubber industry in Shiraz-Iran. *International archives of occupational and environmental health*. 2007;80(6):539-46.