Case Report

Combined pulmonary fibrosis and emphysema: A case report

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INTRODUCTION

Combined pulmonary fibrosis and emphysema (CPFE) syndrome is a rare and emerging combination of emphysema and fibrosis. In this syndrome two individual disease states, pulmonary fibrosis and emphysema coexist and presents into a distinct disease state that differs from either of the two components. Typical radiological finding in chest Computed Tomography (CT) images reveals emphysema in the upper lobe of the lungs, fibrosis in the lower lobe of the lung[1] and honeycombing.[2] These patients have a different natural history, complications, and mortality than those with pulmonary fibrosis or emphysema alone. There is a strong male predominance and the majority of patients are current or former heavy smokers. Major symptoms include dyspnea on exertion in almost all patients and a chronic productive or non-productive cough. Patients diagnosed with CPFE have predominantly bilateral crackles in the lower lung zones, while some patients can have wheezing and diminished breath sounds. Other possible signs and symptoms consist of finger clubbing, hypoxemia and chest pain.[3] The association was first described as a syndrome by Cottin in 2005, named "CPFE," which is characterized by exertional dyspnea, upper-lobe emphysema and lower-lobe fibrosis, preserved lung volume and severely diminished capacity of gas exchange.[4] Treatments for CPFE patients with severe pulmonary hypertension are less effective other than lung transplantation.[5]

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CASE REPORT

A 65 year old male with 35 pack year history of cigarette smoking and a diagnosed case of COPD on controller treatment (long acting beta2 agonist and long acting muscarinic antagonist) since 10 years presented with complaints of worsening dyspnea since 1 month, with symptoms even at rest and no relief even with increased use of SABA. On presentation the SpO_2 was 91% on room air, pulse rate of 82, blood pressure of 130/80 mmhg. Clinical examination revealed the presence of clubbing and B/L normal VBS with bibasilar fine and inspiratory crepts. On cardiac examination, heart sounds were normal with no murmur and JVP was not elevated. His abdomen was soft and nontender. ECG and ECHO were normal ruling out any cardiac involvement. Patient had history of prostate cancer treated with total prostatectomy and radiotherapy. He had strong family history of bowel cancer which led to increased suspicion of malignancy. CXR was done which showed hyperinflated lung fields with bibasilar interstitial markings raising concern of a superimposed pathology but Sputum examination came out to be sterile and routine blood analysis was within normal limits. 6MWT showed exertional desaturation to 79% after 2 min

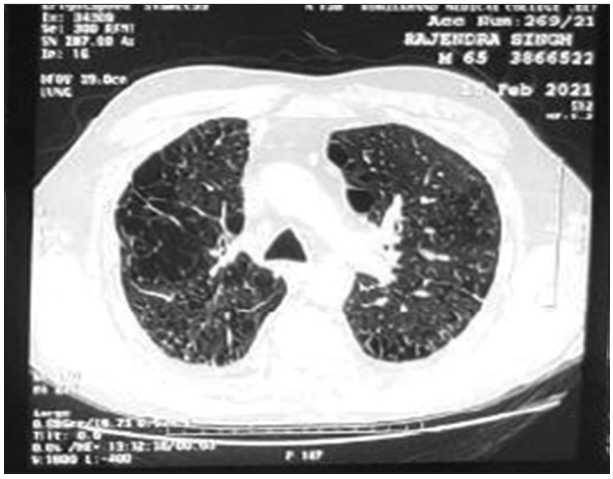

Figure 1: Depicts B/L upper lobe emphysematous changes

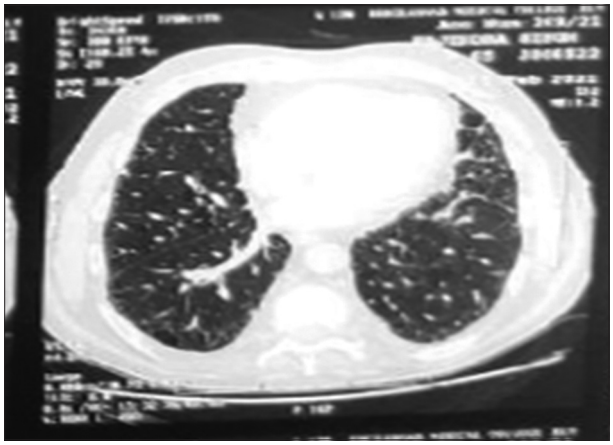

Figure 2: Depicts Postero-basal segment of the B/L lower lobes showing multiple cysts representing honey combing cysts

during 6-min walk testing. History and examination increased the suspicion of interstitial lung disease (ILD) and malignancy leading to increased need of HRCT for subsequent HRCT to establish the diagnosis. HRCT showed extensive areas of pan lobular and para septal emphysematous changes in B/L upper lobes. Postero-basal segment of the B/L lower lobes in the sub pleural location show multiple cysts representing honey combing cysts denoting superimposed ILD. Further to confirm the diagnosis of ILD, spirometry and DLCO were done. Spirometry showed mild obstruction with restriction (FEV1/FVC % = 66.1 and FVC=64%) and disproportionate reduction in DLCO 37% with TLC 63% describing the distinct diagnosis of CPFE.

DISCUSSION

CPFE is a rapidly progressing disease, be aggravated by pulmonary hypertension and usually leads to increased risk of lung cancer. Male sex and smoking are the most important risk factors. The pathogenesis of the disease is unknown. Pulmonary function tests show a normal or subnormal FEV1/FVC ratio but there is a markedly impaired DLCO. Reduced DLCO, that is, severe impairment of gas exchange in due to decrease in vascular surface area and pulmonary capillary blood volume plus alveolar membrane thickening resulting from the two coexistent disease processes. The both restrictive and obstructive disorder components in CPFE have counterbalancing effects in lung function measurements leading to a combined disorder pattern with a normal lung function. Finding of normal lung function is therefore misleading, underestimating the CPFE severity, or even lead to a missed diagnosis. Complications in CPFE include acute exacerbations, pulmonary hypertension and lung cancer, leading to a poor prognosis. Classical radiological findings of CPFE in CT images are emphysema in the upper parts of the lungs, fibrosis, and honey combing in the lower parts of the lung and Emphysema can be paraseptal, centrilobular, or most frequently mixed.

CONCLUSION

CPFE is a syndrome with clinical importance. This syndrome of combined emphysema of the upper lobe and fibrosis with honeycombing of the lower lobes on chest CT findings results in a typical characteristic functional profile, with normal lung volumes, strongly impaired carbon monoxide diffusing capacity of the lung, and decrease 6MWT, thus differentiating this entity apart from both idiopathic pulmonary fibrosis and pulmonary emphysema. Despite normal or subnormal spirometry, which is responsible for the underrecognition, CPFE is a severe entity. Early recognition with diagnosis with CT chest and DLCO with treatment is important for best patient care.

Parameter	PRED	LLN	Result	%PRED
DLCO (ml/min/mmhg)	27.6	20.7	10.1	37
VA	6.49	5.40	4.05	62
DLCO/VA (KCO)	4.03	-	2.49	62
TLC sb (L)	6.63	5.54	4.20	63
RV sb (L)	2.38	1.74	1.59	67
FRC	3.43	2.49	2.23	65

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