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A Rare Case of Combined Pulmonary Fibrosis and Emphysema in a Young Female Patient of Rheumatoid Arthritis and Systemic Lupus Erythematosis

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Abstract: ***Combined pulmonary fibrosis and emphysema is characterized by the co-existence of upper lobe emphysema and lower lobe fibrosis and is being increasingly detected as a new entity in smokers. Patients are usually men in their 6th to 7th decade[1].However smoking is not the only etiology and there are various other associations like occupational exposure and connective tissue disorders like rheumatoid arthritis and systemic sclerosis. In this case report we discuss the presentation and diagnosis of a 30-year-old female, known case of systemic lupus erythematosis and rheumatoid arthritis, diagnosed to have combined pulmonary fibrosis and emphysema on HRCT chest.***

1. Introduction

The presence of excess fibrosis has been historically excluded from the diagnosis of emphysema[2]. Emphysema and idiopathic pulmonary fibrosis were once recognized as two distinct entities. However due to the frequent coexistence of emphysema and pulmonary fibrosis, in 2005, Cottin et al. first time put forward a defined syndrome termed "combined pulmonary fibrosis and emphysema (CPFE)[3]. It is characterized by heavy smoking history, exercise hypoxemia, upper lobe emphysema and lower lobe fibrosis, relatively normal lung volumes and severe reduction of carbon monoxide transfer.

2. Case Presentation

We present the case of a 30-year-old female who is a known case of both systemic lupus erythematosis and rheumatoid arthritis and presented with complaints of dyspnoea on exertion and chest pain. Physical examination revealed basal rales. The patient had tachycardia with a heart rate of 130bpm. Her blood pressure was 127/78 mmHg and respiratory rate was34/ min. Frontal chest x-ray revealed radio-lucencies in the right lower zone. HRCT was performed which revealed diffuse interlobular septal thickening causing architectural distortion , mild traction bronchiectasis along with multiple thin walled cysts and areas of honey combing in bilateral lung parenchyma with an apico- basal gradient and predominant subpleural involvement, more marked changes being noted in the lateral segment of right middle lobe, lingula and basal segments of bilateral lower lobes. Paraseptal and few centriacinar emphysematous changes are seen predominantly in bilateral upper lobes. Few septated subpleuralmacrocysts were seen, best seen along the posterobasal segment of right lower lobe. The right and left branches of the pulmonary artery were prominent. There was no evidence of significant mediastinal lymphadenopathy. There was mild left ventricular dilatation with hypertrophy. A diagnosis of interstitial lung disease with emphysema pointing towards combined pulmonary fibrosis and emphysema (CPFE) was made on the CT findings.

3. Discussion

This syndrome of coexisting emphysematous changes and pulmonary fibrosis was first recognized by Cottin in 2005 and named it "*combined pulmonary fibrosis and emphysema* (*CPFE*)", which is characterized byexertional dyspnea, upper-lobe emphysema and lower-lobe fibrosis, relatively preserved lung volume and severe reduction in diffusion capacity. Even though cigarette smoking is the major risk factor, connective tissue disorders like rheumatoid arthritis and systemic sclerosis are also a causative factor. Patients with connective tissue disorder associated CPFE are more likely to be women of younger age group and tend to have less severe outcomes than their idiopathic CPFEcounterparts[4]. Cough and dyspnoea are the common symptoms. HRCT is a predominant diagnostic tool. Pulmonary function tests reveal mean values of forced vital capacity (FVC) and total lung capacity (TLC) in CPFE are usually within relatively normal range, whereas DLco is severely diminished. Blood gas analysis reveal changes of hypoxaemia. Surgical lung biopsy usually reveals a pattern of UIP, though the coexistent presence of pigmented alveolar macrophages in a pattern resembling RB-ILD or DIP may be found in areas. A histologic pattern of NSIP has also been recognized in some patients with CPFE with ground glass opacities in isolated cases[5]. The complications are pulmonary arterial hypertension, acute lung injury and lung cancer. There is a lower prevalence of pulmonary arterial hypertension in patients with connective tissue disorder associated CPFE[4].

4. Conclusion

CPFE is a distinct however under diagnosed entity and it is important for the radiologists to be aware of its imaging findings which are classical. It is also important to recognise that CPFE does not occur only in smokers but also in patients with connective tissue disorders in whom the presentation might be at younger age especially young females.

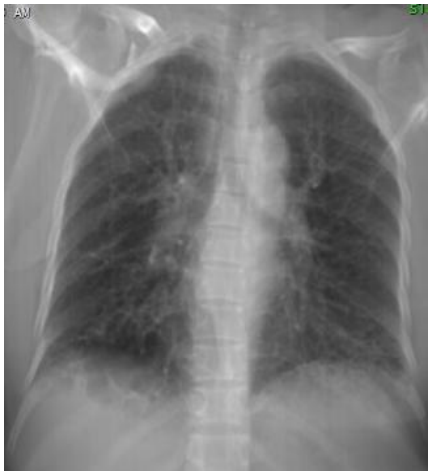

Figure 1

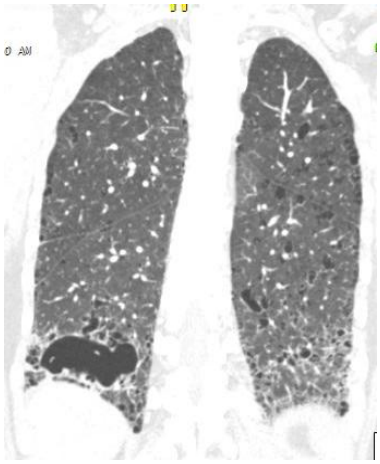

Figure 2

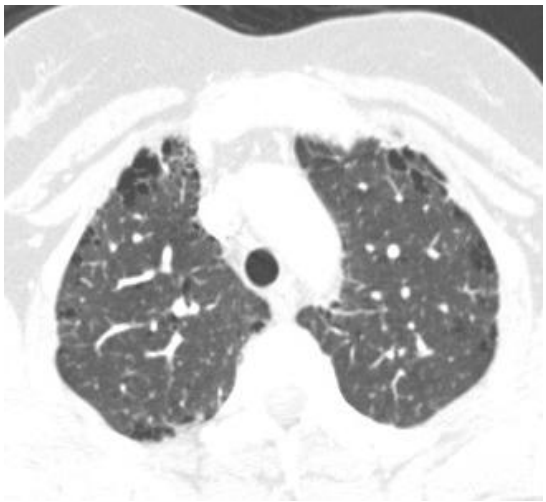

Figure 3

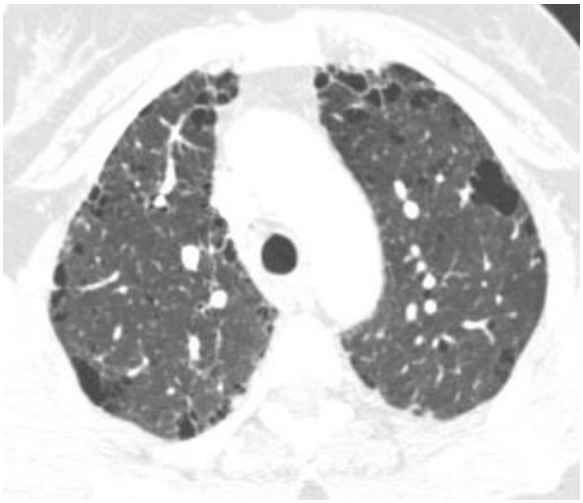

Figure 4

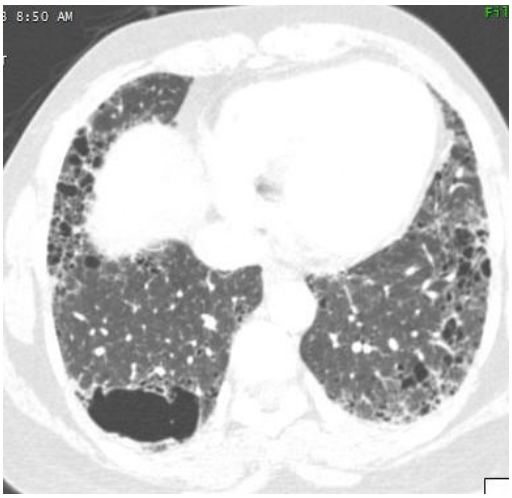

Figure 5

Figure 1: Frontal chest x-ray reveals multiple radio-lucencies in the right lower zone.

Figure 2: HRCT coronal images reveal multiple cysts in the basal segment of right lower lobe along with interlobular septal thickening.

Figure 3 and 4: HRCT axial images reveal centriacinar and paraseptal emphysematous changes in bilateral upper lobes.

Figure 5: HRCT axial images reveal interlobular septal thickening and honey combing in bilateral lung bases along with subpleural cysts in posterobasal segment of right lower lobe.

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