Case Report

COMBINED PULMONARY FIBROSIS AND EMPHYSEMA SYNDROME; AN UNCOMMON DISEASE, COMMONLY ASSOCIATED WITH PULMONARY HYPERTENSION

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CLINICAL DATA

A 54-year male presented with symptoms of progressive dyspnea on exertion and cough, predominantly dry for one year duration.There was no history of orthopnea. He was a cigarette smoker with a smoking index of 12 pack years. He had no history of fever, dust allergy or drug therapy. TB contact history was negative. There was no history of similar illness in the family members or any symptoms suggestive of connective tissue disease. His professional history was unremarkable. On physical examination, fingers clubbing was observed and on auscultation there were bibasilar fine end inspiratory crackles.

INVESTIGATIONS

X-Ray Chest (PA View) Figure 1 shows bilateral reticulation.

High resolution computerized tomography (HRCT) of thorax (Figure 2 A & B) showing multiple bullae and paraseptal emphysema involing both upper lobes. (Figure 3 A & B) show cystic honeycombing in sub plural region predominantly involving lower lobes. Main pulmonary trunk is enlarged suggesting pulmonary arterial hypertension.

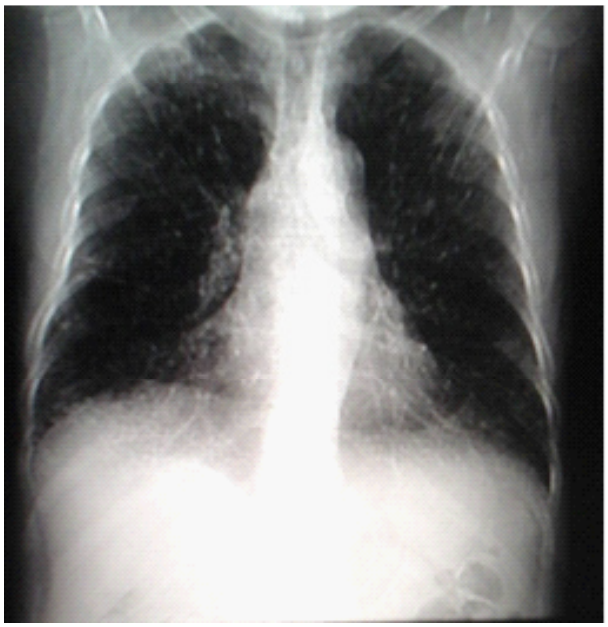

Figure 1: The chest radiograph (postero-anterior view) shows bilateral reticulation

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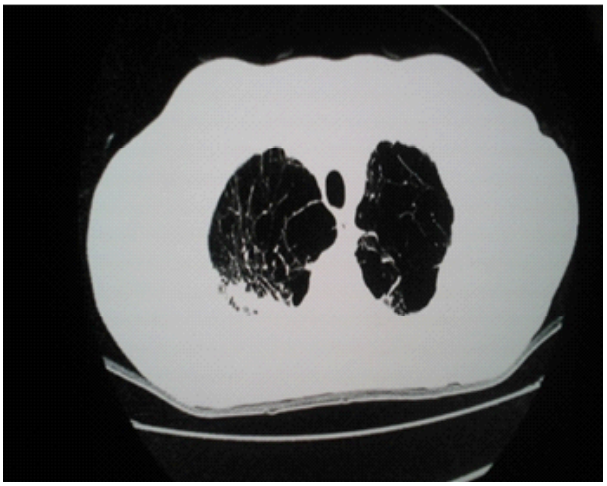

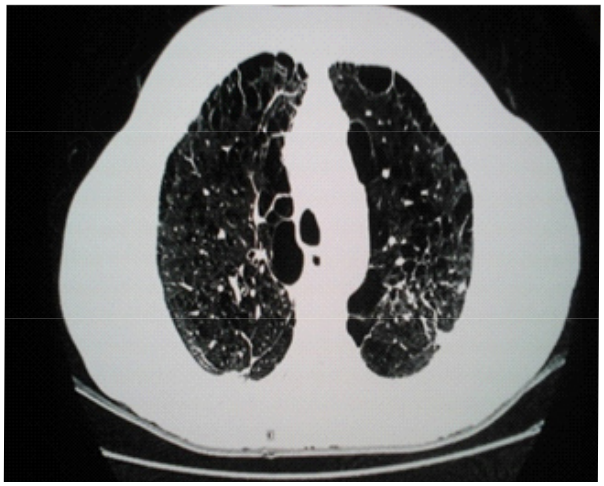

Figure 2: A & B HRCT of chest show multiple bullae and paraseptal emphysema involving both upper lobes

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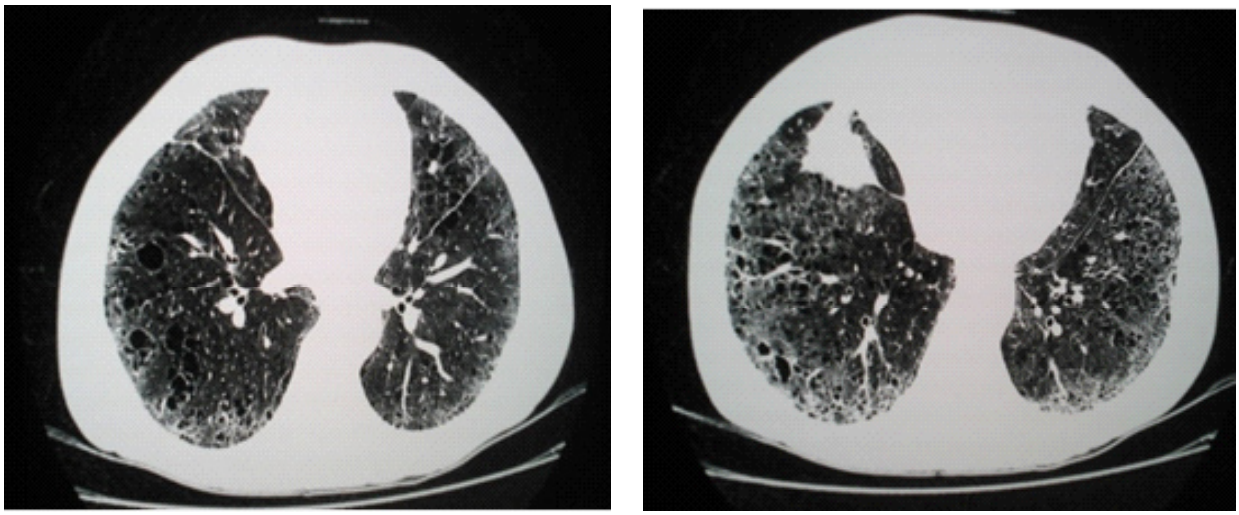

Figure 3 A&B show multiple micro cystic honeycombing involving lower lobes

DIAGNOSIS

Combined pulmonary fibrosis and emphysema (CPFE) syndrome.

DISCUSSION

Combined pulmonary fibrosis and emphysema (CPFE) Syndrome is defined as distinct clinical and radiological syndrome in which interstial pulmonary fibrosis (IPF) occur simultaneously with pulmonary emphysema. There is association of both disease entities with cigarette smoking. Emphsematous changes of this syndrome involves upper lobe while pulmonary fibrosis predominantly occur in lower lobe[1].

Auerbach et al provided initial his pathological proof of combination of both emphysema and pulmonary fibrosis in 1974[2] . Later on, in 1990 the Wiggins et al provided radiological evidence of this syndrome in his study which was based upon HRCT findings[3].

Apart from tobacco smoking which a major etiological factor in pathogenesis of CPFE, there are certain other risk factors such as exposure of agro compounds.

Mutation of ABCA3 gene is associated with this disease especially in non smoker and young patients[4]. Connective tissue disease associated with CPFE includes rheumatoid arthritis and systemic sclerosis[5]. The major complication of this syndrome includes pulmonary arterial hypertension and lung cancer. Incidence of pulmonary hypertension is increased in CPFE up to 50% which is higher as compared to emphysema or pulmonary fibrosis alone[6].

Clinical presentation of CPFE includes dyspnea on exertion with may or may not be associated with chest pain. Other symptom is dry cough. Physical findings include finger clubbing and fine end inspiratory crackles.

The diagnosis of CPFE based on classical HRCT findings which include emphysematous changes predominantly involving upper lobes and pulmonary fibrosis in lower lobes. Emphysema may be centrilobular variety or paraseptal with bullae formation predominantly in sub pleural region. The pattern of pulmonary fibrosis includes reticular opacities, honeycombing and traction bronchiectasis[7]. HRCT define the transitional zone between emphysema and pulmonary fibrosis.

In spite of sever clinical symptoms pulmonary lung function tests are usually preserved[8] because of co existence of emphysema with pulmonary fibrosis where increase in lung compliance due to emphysema counteract the loss of volume due to fibrosis however diffusion capacity for gases are markedly reduced due to synchronous effect of emphysema and pulmonary fibrosis. There is no specific curative therapy for CPFE[6].The therapeutic option includes smoking cessation and oxygen therapy for hypoxemia and lung transplantation.[8] Systemic corticosteroids and immunosuppressant therapy may be an option for patients with CTD-associated CPFE. certain cytokine antagonists , Pirfenidone, Nintedanib, and drugs used to treat pulmonary hypertension can be used in these patients[6].

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