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The Syndrome of Combined Pulmonary Fibrosis and Emphysema

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ABSTRACT

A syndrome associating an upper-lobe emphysema and pulmonary fibrosis of the lower lung was recently characterized. Few cases were identified in the literature. We report a case of a 68 year- old smoker man presented for exacerbation of a severe dyspnea. Physical examination revealed basal crackles and finger clubbing. Blood gas analysis showed hypoxemia. Chest radiography showed features compatible with emphysema of the upper lobes and fibrosis of the basis. Chest computed tomography confirmed chest radiography's findings and revealed fibrosis. The diagnosis of idiopathic pulmonary fibrosis was made. Pulmonary function tests showed obstructive pattern. Systolic pulmonary arterial pressure was elevated up to 87 mm Hg on heart ultrasonography.

The authors emphasize the importance of the diagnosis of this entity made through chest computed tomography and the fact that it is characterized by severe impairment of gas exchange, the high prevalence of pulmonary hypertension and poor survival. (*Tanaffos*2011; 10(3): 59-62)

Key words: Emphysema, Dyspnea, Gas exchange, Hypoxemia, Pulmonary fibrosis, Respiratory function tests

INTRODUCTION

Combined pulmonary fibrosis and emphysema (CPFE) is a recently individualized disease. The diagnosis is mainly based on high resolution chest tomography (HRCT) which reveals the association of emphysema in the pulmonary upper zones and fibrosis in the lower zones. This disease is rare. It has a particular profile on respiratory function tests and may become complicated by pulmonary hypertension

(PH) which worsens its prognosis. We report a patient suffering from this condition and describe its clinical, radiological and functional characteristics and its precise follow-up.

CASE SUMMARIES

Our patient was a 68-year-old man, smoking 63 packs/year, who presented to our department complaining of a 4-month history of dyspnea exacerbation (from stage I to stage III), associated with productive cough. On physical examination, the patient presented clubbing, tachypnea with respiratory rate of 30/min and bibasilar crackles.

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Blood gas analysis showed: pH: 7.43, PaO₂: 45 mm Hg, Pa CO₂: 23 mm Hg, HCO₃⁻: 16 mmol /l, and SaO₂: 85%. Chest x-ray revealed a thoracic distension, hyperlucency of the upper thoracic zones and honeycombing of the bases. The level of C-reactive protein was 110 mg/l. There were no acid fast bacilli in the sputum. The cyto-bacteriologic examination of the sputum isolated *Haemophilus influenzae* for which the patient was treated with amoxicillin 2 g per day associated with clavulanic acid for 7 days. Oxygenotherapy was prescribed for hypoxemia. Chest HRCT (Figures 1 and 2) showed centrilobular emphysema and bullae predominating in the upper lobes associated with honeycombing, a peribronchovascular and scissural distortion; and diffuse ground glass opacities predominating in the bases and the periphery compatible with idiopathic pulmonary fibrosis (IPF). Bronchial fibroscopy was normal. Bronchoalveolar lavage showed hypercellularity with 410×10³/ml cells including 65% macrophages, 22% neutrophil, 4% eosinophil, and 9% lymphocytes. Respiratory function tests revealed an obstructive syndrome with forced vital capacity (FVC): 3.14 L (67%), forced expiratory volume at the first second (FEV1): 1.97 L (53%), Tiffeneau ratio: 63% and total lung capacity: 5.57 L (80%). Immunological tests including antinuclear antibodies and ANCA were negative.

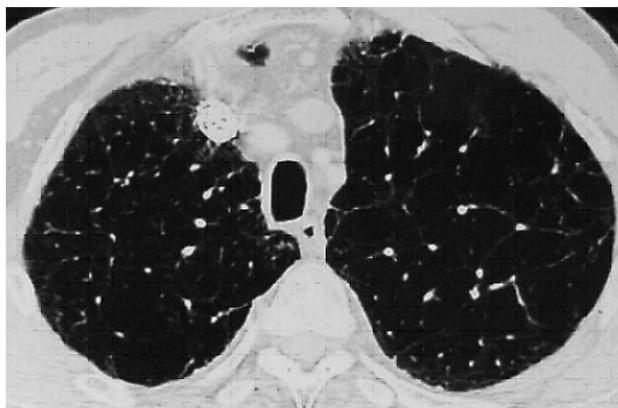


Figure 1. Chest HRCT showed centrilobular emphysema and bullae in the upper lobes.

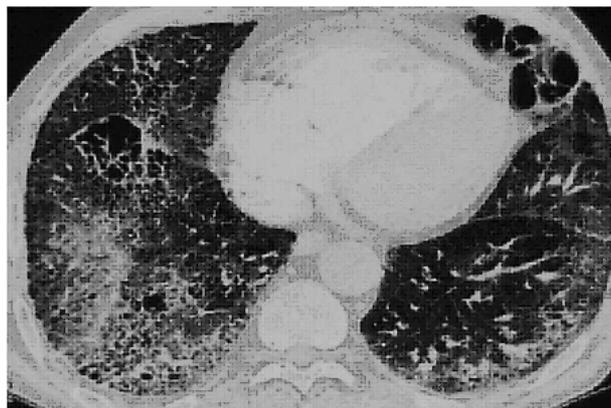


Figure 2. Chest HRCT showed honey combing, scissural distortion and ground glass opacities in the bases and the periphery of the lungs.

Cardiac ultrasonography showed dilated right chambers and pulmonary hypertension with systolic pulmonary arterial pressure of 87 mm Hg.

The patient clinically improved after 3 days. The current follow-up is 2 years in which the patient had a stable dyspnea (stage II of the NYHA) and slight hypoxemia but did not need oxygen at home.

DISCUSSION

The exact prevalence of CPFE is not known. However, Mejia et al, in a series of 110 patients with IPF found that 28% had CPFE (1). This syndrome typically affects heavy smoker males around 65 years of age or older as it was the case for our patient. The mean delay between the onset of symptoms and the diagnosis is 2 years. Exertional dyspnea is constant and most of the time significant. In half the cases it is associated with cough which is sometimes productive. As in IPF, bibasilar crackles and clubbing are the major findings (2). The disease occurs most of the time without any identified specific etiology (2). Neither the etiopathogeny nor the physiology of this syndrome is well known. A logical explanation is that the fibrosis of the lung in the bases exerts traction on the upper parts resulting in development of emphysema (2). However, emphysema most of the time precedes fibrosis (3).

However, studies found that tobacco smoking plays a major role in the development of the disease (2,4). Other factors such as agrochemical compounds are thought to be also possible risk factors (5). The diagnosis of CPFE lays on imaging findings. Chest HRCT reveals emphysema predominating in the upper zones and signs of IPF in the bases consisting of honeycombing, reticular opacities predominating on the basis and in the subpleural zones, distortion of the architecture of the lung and/or traction bronchiectasis. Ground glass opacity is more frequent in this syndrome than in IPF. There is almost always centrilobular and paraseptal emphysema. The latter represents a characteristic finding of the CPEF. In fact, despite it is related to tobacco smoking, it is rarely encountered in chronic obstructive pulmonary diseases (2,5). Bullae are also a common finding on HRCT. Our patient presented with dyspnea, clubbing, crackles and typical HRCT findings allowing the diagnosis of CPFE. However, in our patient FVC and FEV1 were low; whereas, this syndrome is characterised by the relative preservation of the pulmonary volumes contrasting with a major dyspnea (6). Another intriguing feature is that lung volumes are near normal contrasting with markedly impaired capacity of carbon monoxide transfer and arterial oxygen saturation (7,8). A proposed explanation is that hyperinflation and high compliance of the emphysematous areas of the lungs probably compensate the volume loss due to fibrosis of the lower lobes, while pulmonary emphysema and fibrosis may have additive or synergistic effects on carbon monoxide transfer and exercise hypoxemia (2). These findings indicate that the diagnosis may be under-recognized if the respiratory function tests are normal and carbon monoxide transfer not studied. Our patient, however, showed a mild hypoxemia during his follow-up. In CPFE bronchoalveolar fluid is compatible with that in IPF (9,10). A few series reported pathologic findings

but it seems that the aspect is that of a usual interstitial pneumonia (9). A major and frequent complication of the disease, also seen in our patient, is pulmonary hypertension occurring in 50% to 90% of the cases (1,2). This complication occurs more frequently in CPFE than in COPD or IPF. It significantly decreases the 5-year survival to 25% in those with PH versus 75% in those without it (2).

The authors emphasize the importance of the diagnosis of this entity made through chest computed tomography and the fact that it is characterized by severe impairment of gas exchange, the high prevalence of pulmonary hypertension and poor survival.

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