RESPIRATORY FAILURE FROM COMBINED EMPHYSEMA AND PULMONARY FIBROSIS

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ABSTRACT

An elderly man presented with hypoxemic respiratory failure and pulmonary hypertension. He was a heavy cigarette smoker and had been treated previously for pleuro-pulmonary tuberculosis. His lung volumes and airway function were normal. High resolution computer tomographic (HRCT) examination revealed combined upper lobe emphysema and lower lobe fibrosis. Routine pulmonary function tests may underestimate the degree of functional impairment in patients with combined emphysema and fibrosis. The HRCT examination however provided valuable information.

Keywords: Emphysema, pulmonary fibrosis, hypoxemia, high resolution computer tomography

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INTRODUCTION

Pulmonary function tests are essential investigations in the diagnostic workup of patients with hypoxemic respiratory failure[1]. In the presence of diseases of the airways, lung parenchyma or chest wall, pulmonary function tests will usually reveal abnormalities of ventilation. If the ventilatory function was normal, then shunts and pulmonary thromboembolism, should be considered as causes of hypoxemia[1,2].

This paper describes a patient with severe hypoxemic respiratory failure due to a combination of pulmonary emphysema and fibrosis. His ventilatory function however was normal and inappropriate for the degree of impaired gas exchange function. This was misleading information and resulted in some diagnostic uncertainty which was finally resolved by high resolution computerized tomographic (HRCT) examination of the lungs which demonstrated both the emphysema and fibrosis.

CASE REPORT

A 72-year-old man presented with progressive effort dyspnoea and mild non-productive cough. He was a heavy cigarette smoker, consuming about sixty cigarettes per day over 50 years, and had received treatment for pulmonary tuberculosis four years previously. The tuberculosis involved the left lower lobe and was associated with pleural effusions which required thoracenthesis. He was cyanosed, plethoric and had finger clubbing. Inspiratory crackles were heard over his lower chest.

The plain chest radiograph showed large pulmonary arteries consistent with long standing pulmonary hypertension (Fig 1). The lung fields were unremarkable except for mild hyperlucency in the left upper lobe. The left costo-phrenic angle was blunted consistent with healed pleuro-pulmonary tuberculosis. The ECGs showed non-specific clockwise rotation.

The arterial blood gas examined while the patients were breathing room air showed severe hypoxemia (PaO_2 of 36 mmHg), desaturation (arterial saturation of 74%) and an increased alveolar air-arterial blood gradient for oxygen tension (70 mmHg). There was no hypercapnia ($PaCO_2$ of 35 mmHg). The arterial desaturation was correctable by breathing 100% O_2, thus ruling out the presence of significant right-to-left sided cardiac or pulmonary vascular shunting.

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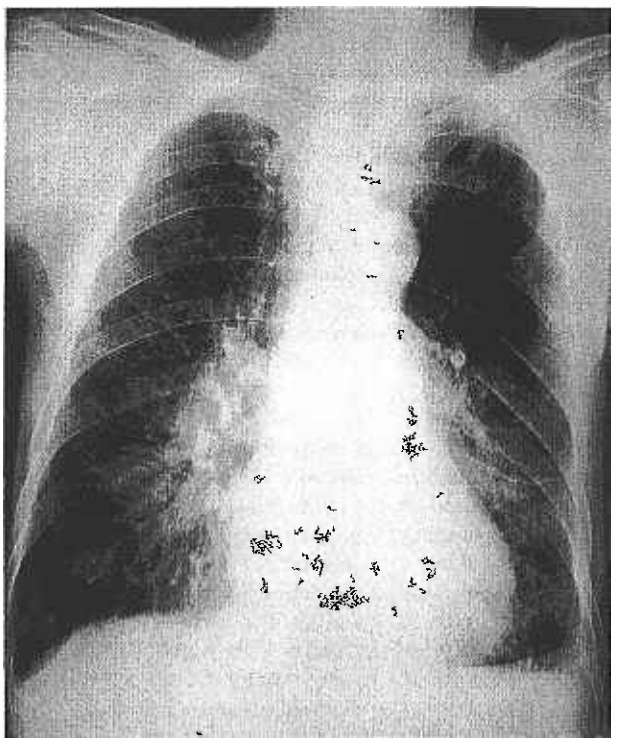
Fig 1 - Chest X-ray showing large pulmonary arteries, increased reticulation in both mid and lower zones, hyperlucency of the left upper lobe and blunted left costo-phrenic angle consistent with previous tuberculous effusion.

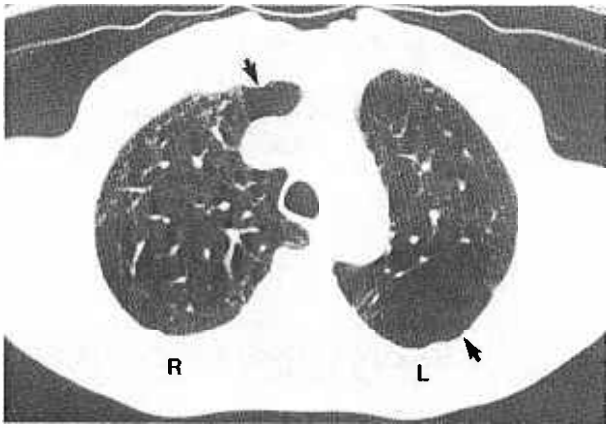


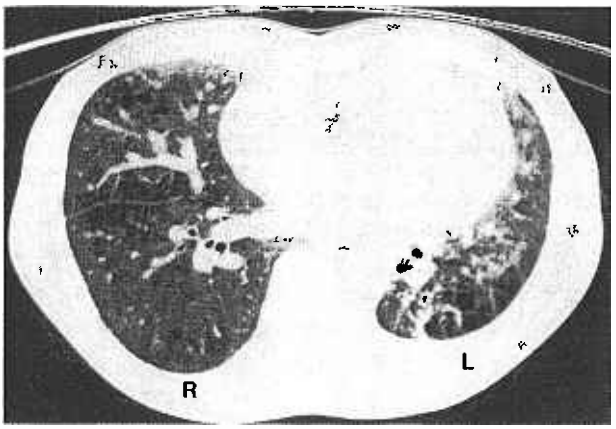
The pulmonary function tests showed normal lung volumes (VC of 78% predicted and TLC of 92% predicted) with only mildly reduced FEV_1 (62% predicted) and FEV_1/VC ratios (80% predicted). There was no increase in airway resistance (56% predicted). The carbon monoxide (CO) transfer capacity was however severely reduced (28% predicted).

Further investigations included 2D echocardiograms which confirmed enlargement of the right ventricles with normal left ventricular contraction and no valvular disease or septal defects.

High resolution computerized tomographic (HRCT) examination was undertaken to further characterize the underlying lung parenchymal disorder. The HRCT revealed bilateral apical emphysematous bullae with reticulo-nodular infiltrates forming microcysts in the left lower lobe (Fig 2). There was also loss of left lung volume and pleural thickening.

Fig 2 - The HRCT showing bi-apical emphysematous bullae (left panel - arrows) and loss of volume, pleural thickening and cystic reticulation in the left base (L) consistent with pleuro-pulmonary fibrosis (right panel).





DISCUSSION

The patient in this report had an unusual combination of emphysema and pulmonary fibrosis. The emphysema was associated with heavy cigarette smoking and advanced age while the lung fibrosis was the sequela of healed pleuro-pulmonary tuberculosis. Confirmation of the diagnosis would have required open lung biopsy which was declined by the patient. Nevertheless the lung lesions were clearly revealed in the HRCT examinations (Fig 2).

The initial diagnosis in the patient was chronic obstructive pulmonary disease (COPD). The relatively normal ventilatory function was unexpected and led to investigations to rule out intra-cardiac shunts and pulmonary thromboembolic disease. The normal TLC and VC combined with a relatively normal FEV_1/VC ratio and airway resistance would be incompatible with a clinical diagnosis of COPD. The pattern of pulmonary function abnormality usually associated with emphysema, that is - hyperinflation, reduced expiratory airflow and increased airway resistance - was obscured in this patient by the presence also of pulmonary fibrosis. Hunninghake and his colleagues, in a recent study on the influence of cigarette smoking on lung function in patients with idiopathic pulmonary fibrosis showed that measurement of lung volumes and FEV_1/VC ratio may not adequately define the severity of functional impairment[3]. Cherniack and his associates have shown that cigarette smoking and pulmonary fibrosis may have opposite effects on lung pressure-volume relation and airway function[4]. This would appear to be the case in this patient. The normal TLC and VC implies that the residual volume (RV) is also not elevated. This suggests that the tendency for emphysema to increase RV (from air trapping) is balanced by the tendency of pulmonary fibrosis to reduce it (loss of alveoli). The mildly reduced FEV_1 is both unexpected and unexplained since the FEV_1 is considered a polyvalent test which is sensitive to both restrictive and obstructive lung disorders[5].

The loss of functioning alveolar units due to a combination of emphysema and fibrosis in the patient would have accounted for the severe hypoxemia, pulmonary hypertension, and breathlessness. The reduced CO transfer capacity and widened alveolar-arterial O_2 gradient confirm the presence of severely deranged gas exchange function. There is a scatter of ventilation-perfusion (V_A/Q) ratios in normal lungs with a V_A/Q ratio of about 0.3 in the apex and 0.6 at the base[2]. In this patient, the spatial distribution of upper lobe emphysema and lower lobe fibrosis could have increased further the normally uneven distribution of regional ventilation and perfusion within the lungs. A predominance of apical alveolar units with high V_A/Q ratios associated with emphysema and basal units with low V_A/Q ratios associated with fibrosis would increase the usual V_A/Q inhomogeneity of normal lungs and produce hypoxemia[6,7].

The HRCT has been advocated as a useful technique for the imaging of lung parenchymal disease[8-13]. There are substantial improvements in image reconstruction, edge enhancement, scan times and spatial resolution when HRCT is compared with conventional CT[10,12,13]. The HRCT examination was the definitive test in this patient and demonstrated the presence, severity and spatial distribution of both emphysema and fibrosis in the lungs.

Turner-Warwick and her colleagues have described eight patients with combined cryptogenic fibrosing alveolitis and emphysema[14]. Their radiologic features, arterial blood gas and pulmonary function test results were atypical and very similar to the patient in this report. The patients had VC and FEV_1 in the normal range, unexpected for the degree of lung disease. This report, however, is the first description of combined emphysema and pulmonary fibrosis from healed tuberculosis giving rise to a similar clinical presentation and physiologic disturbance as the cases of combined emphysema and cryptogenic alveolitis described by Turner-Warwick and her colleagues.

In summary, this paper describes a patient with combined emphysema from cigarette smoking and fibrosis from treated tuberculosis and who presented with hypoxemic respiratory failure and pulmonary hypertension. Routine tests of ventilatory function were misleading and underestimated the degree of functional impairment. The most useful diagnostic information was obtained from the radiologic examination. This clinico-radiological-physiological complex is a result of the coincidence of two relatively common chronic respiratory disorders in the same patient.

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