A Functional Respiratory Imaging View on Idiopathic Pleuroparenchymal Fibroelastosis

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Introduction

Idiopathic pleuroparenchymal fibroelastosis (IPPEF) is a rather recently (Frankel et al. Chest. 2004; 126:2007-13) described rare entity, characterized by pleural and subpleural parenchymal fibrosis and elastosis mainly in the upper lobes. The etiology and pathophysiology are little documented and prognosis is poor, with no effective therapies other than lung transplantation.

This case report aims to describe some of the regional pathophysiology of IPPFE by using functional respiratory imaging (FRI).

Case subject

A 38 year old woman was diagnosed with IPPFE in December 2014 based on her clinical presentation. Diagnostic confirmation with biopsy was not possible at that time due to the advanced disease state, but IPPFE was confirmed was confirmed on the explanted lungs at the time of lung transplantation in September 2015. Lung function tests showed significantly reduced static volumes (TLC 1.82L(41%) and FRC 1.04L(41%) and even further decreased dynamic lung volumes (FEV1 0.76L(30%) and VC 0.88L(28%)p). Arterial blood gases (pCO2 43mmHg, pO2 94mmHg and SO2=97%) showed normal or borderline normal values.

In August 2015, as preparation for lung transplantation surgery, a two level (expiration-inspiration) CT scan was taken to allow for an FRI analysis of the IPPFE lung disease.

Functional respiratory imaging

FRI allows to get a regional insight on parameters linked to:

- Ventilation
- Lobe volumes
- Lobar airflow distribution
- Airway resistance
- Perfusion and tissue
- Blood vessel volume
- Emphysema
- Air trapping
- Fibrosis
- Particle deposition

Results

FRI results showed that the left lung (FRC 30%p, TLC 25%p, fibrosis 45%) was more affected by the disease as compared to the right lung (FRC 58%p, TLC 45%p, fibrosis 30%); and the upper lobes (FRC 20%p, TLC 16%p, fibrosis 50%) were more affected as compared to the lower lobes (FRC 58%p, TLC 44%p, fibrosis 31%). Airflow was greatly reduced towards the most affected zones (upper/lower ratio ~ 0.15). Particle deposition analysis demonstrated that less than 5% of inhaled particles (MMAD 2.6±1.5μm) reach these upper lobes. Furthermore, from the image it can be seen that the fibrosis causes emphysematous bullae in the periphery and increased airway volumes due to the traction on the lung tissue and changes in alveolar tissue stiffness. Also, infiltrations are seen at the oblique tissues.

Conclusions

FRI analysis was able to show and quantify the regional expression of a poorly documented disease such as IPPFE. The poor ventilation, and consequent quasiasbence of inhaled particle deposition, in the most affected lung regions explains why inhaled medication does not seem to be efficient in this small population, for which lung transplantation is still the only therapeutic option.