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Respiratory bronchiolitis-associated interstitial lung disease with obvious paraseptal emphysema

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Abstract

Respiratory bronchiolitis-associated interstitial lung disease (RBILD) is a distinct clinical–pathological disease described almost exclusively among cigarette smokers. Normally, the most common manifestations in computed tomography findings of RBILD are central and peripheral bronchial wall thickening, centrilobular nodules, ground glass opacity, and mild centrilobular emphysema in the upper lobes. In this study, we report a current smoker presenting with diffuse ground glass opacity in the entire lungs along with paraseptal emphysema. This case differs from past reports where obvious paraseptal emphysema in upper lung is seldom reported.

Introduction

Myers et al. [1] first described the concept of respiratory bronchiolitis-associated interstitial lung disease (RBILD) in 1987. RBILD is a mild inflammatory pulmonary disorder with good prognosis. The presence of smoker's macrophages is a fundamental requirement for diagnosing RBILD. Although some researchers have reported RBILD with paraseptal emphysema, the emphysema is mild. The chest computed tomography (CT) scan of our patient showed severe paraseptal emphysema that differed from past reports. Pathology also confirmed emphysema present at the same time.

Case Report

A 59-year-old farmer presented with non-productive cough and chest discomfort of 2 years. He had a 15 pack-year history of smoking with no exposure to dust or hay. He did not have any history of chronic disease or drugs. Clinical examination demonstrated fine 'Velcro'-like crackles of the lung bases and no finger clubbing. His haematological investigations were within normal limits, except that total cholesterol, triglycerides, and low density lipoprotein-C levels were elevated. Arterial blood gas analysis (ABG) showed pH7.359/$PaO_2$84.0/$PaCO_2$36.9/$HCO_3$21.0/Be-4.2. The pulmonary function test revealed mild obstructive abnormality with forced vital capacity (FVC) of 3.24 L (93.7% predicted), forced expiratory volume in 1 sec (FEV1) of 2.03 L (73.2%), and FEV_1/FVC of 62.48%, suggesting an obstructive abnormality. The diffusion capacity was normal. Echocardiography demonstrated mild pulmonary hypertension (PH; 33 mmHg). A chest CT scan (Fig. 1) revealed paraseptal emphysema and diffuse ground glass opacity. Pathological findings (Fig. 2) showed the accumulation of brownish pigmented macrophages within the respiratory bronchioles, with the infiltration extending into the neighbouring alveoli. An infiltration of chronic inflammatory cells was also present in the bronchiolar and alveolar wall regions. Prussian blue staining was positive. The lung biopsy was taken from the lower right lung. Bronchoalveolar lavage (BAL) contained large amounts of brown-pigmented macrophages.

The patient was started on inhalation therapy with budesonide–formoterol inhalation powder (160 μg/4.5 μg)

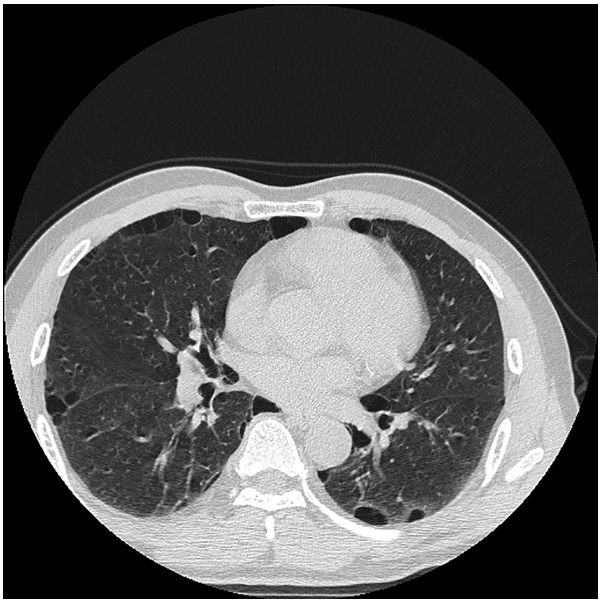

Figure 1. A chest computed tomography scan revealed paraseptal emphysema and pulmonary bullae mainly in the upper lungs and diffuse ground glass opacity was present in both lung fields. There is an absence of honeycombing.

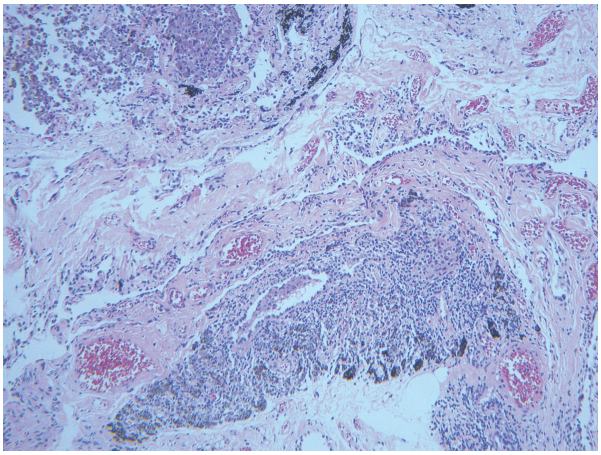

Figure 2. Pathology of medical thoracoscope biopsy samples. Presence of pigmented macrophages in the respiratory bronchiole and peribronchiolar inflammation along with fibrosis. Alveolar walls infiltrated with collagen and chronic inflammatory cell. Haematoxylin–eosin stain, 100×.

every 12 h, and was advised to quit smoking. He had mild improvement while on this therapy. Corticosteroid therapy was considered to be reasonable in some RBILD patients; as a result, the patient was treated with prednisone (20 mg/day) for 2 months. His cough symptoms disappeared, while pulmonary function test and chest CT image were never significantly changed.

Discussion

RBILD occurs almost exclusively in smokers [1,2]. As previously described, high-resolution CT (HRCT) scanning demonstrates centrilobular micronodules, ground glass opacities and peribronchiolar thickening [3,4], mild upper-lobe emphysema and air trapping are variably present, mostly centrilobular emphysema [5]. There were only two reports in which HRCT showed mild paraseptal emphysema. In the present case, apparent paraseptal emphysema both in upper and lower lobe was found, that differs from previous studies. Before a lung biopsy was performed, the alternative diagnosis may have been combined pulmonary fibrosis and emphysema (CPFE) because of the paraseptal emphysema and pulmonary fibrosis. CPFE and RBILD were both closely associated with smoking, but CPFE generally presents with severe dyspnoea, has a poor prognosis, frequently complicated by PH, preserved FEV 1.0, and severely impaired diffusion capacity. Characteristic radiological findings include upper-lobe emphysema and lower-lobe interstitial fibrotic changes. Honeycombing is frequent. Cottin et al. [6] noted that the interstitial pneumonia (IP) pattern may mimic usual interstitial pneumonia (UIP), non-specific interstitial pneumonia (NSIP), RBILD, and other such entities. Pathological characteristics included a predominance of UIP, NSIP, RBILD, airspace enlargement with fibrosis (AEF), and unclassifiable smoking-related interstitial fibrosis. Acute lung injury was one of the complications of CPFE, while RBILD hardly happened. There was no specific treatment for the CPFE syndrome. Smoking cessation is considered the most important factor in the management of RBILD. In conclusion, we considered that the patient was diagnosed with RBILD and obvious paraseptal emphysema.

Disclosure Statements

No conflict of interest declared

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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