

Case report

Respiratory bronchiolitis associated interstitial lung disease (RB-ILD) in a non-smoking female

Siribaddana A D¹

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Introduction

In the revised update of the International Multidisciplinary Classification of the Idiopathic Interstitial Pneumonias, respiratory bronchiolitis-interstitial lung disease (RBILD) is considered as a definite entity.¹ It is generally associated with smoking.^{1,2}

Histologic respiratory bronchiolitis (RB) is frequently present in current smokers and considered as a physiological response to smoking.² In a few individuals it becomes extensive enough to result in an interstitial lung disease. RBILD occurring in non-smokers is a rare entity³ and it has not been reported in Sri Lanka. We report a case of RBILD in a non-smoking female.

Case history

A sixty year old female presented with progressive shortness of breath of one year duration. She was a life time non-smoker. She had been exposed to firewood smoke for the past 40 years as she was engaged in cooking in a poorly ventilated kitchen.

On examination she was dyspnoeic at rest accounting to MRC dyspnoea scale of 4. She was centrally cyanosed and her oxygen saturation was 82%. There was no clubbing. Her pulse rate was 92 beats per minute and the blood pressure was 110/70 mmHg. Auscultation of the chest revealed vesicular breath sounds with end inspiratory fine crepitations predominantly of both lower zones of the chest.

Lung function tests showed a forced expiratory volume in first second (FEV1) of 1.12 l/s (36.5% of predicted) and a forced vital capacity (FVC) of 1.61 l (45.4% of predicted). The FEV1/FVC ratio of 70.2% (80.3% of predicted). Six minute walking distance (6MWD) was 100 m.

The total white cell count was $10.3 \times 10^9/l$ (neutrophils 67%, lymphocytes 30%, and eosinophils

3%). Erythrocyte sedimentation rate was 37 mm in first hour. HIV screening was negative. Rheumatoid factor and anti-nuclear antibody tests were negative.

The chest radiography (Figure 1) showed reticular and nodular shadowing with predominant nodular pattern of both lung fields. The changes were predominantly seen in the mid and lower zones of both lung fields. Some degree of air trapping was noted which signifies airway obstruction.



Figure 1. Chest radiograph showing reticular nodular shadowing of both lung fields with a predominant nodular pattern mainly seen in the mid and lower zones.

A high resolution computerized tomography (HRCT) of the chest (Figures 2a and 2b) showed ill-defined centrilobular nodules, reticulation, patchy ground-glass opacities with a mosaic perfusion pattern. The nodules were distributed predominantly in the mid and lower zones. Few dilated bronchi were present signifying traction bronchiectasis due to interstitial fibrosis. There was some degree of emphysema which is a common concomitant finding in people who are exposed to smoke.

¹ *Consultant Chest Physician, General Hospital, Kandy, Sri Lanka.*

Fibreoptic bronchoscopy was performed and transbronchial biopsy specimens and bronchoalveolar lavage (BAL) were obtained. The BAL showed a marked increase in cell number which was mainly pigment laden macrophages (Figures 3a and 3b). There were few bronchial epithelial cells and no lymphocytes. Histological examination showed prominent accumulation of pigment laden macrophages with mild chronic interstitial inflammation.

There were no fibroblastic foci. This was consistent with a diagnosis of RB-ILD.

The patient was commenced on oral steroids and she showed a remarkable clinical improvement. After 6 weeks of therapy her FEV1 improved to 1.32 l/s (46.5%) and FVC improved to 1.71 l/s (78.1%). The 6 MWD increased to 320 m.



Figure 2a.

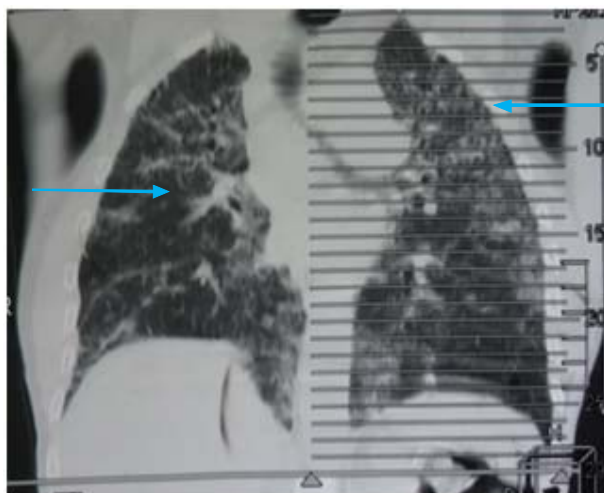


Figure 2b.

HRCT chest showing ill-defined centrilobular nodules (arrowed in the coronal section), centrilobular emphysema and patchy ground glass opacities with mosaic perfusion pattern suggestive of respiratory bronchiolitis. The axial section shows dilated bronchi signifying traction bronchiectasis. The thin walled cyst seen in the axial section is an incidental finding.

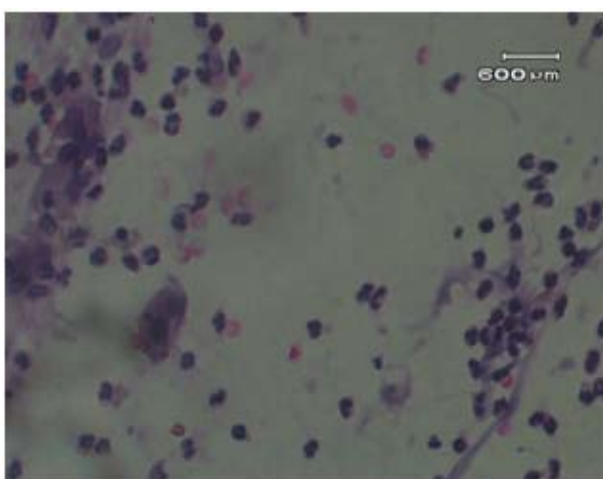


Figure 3a.

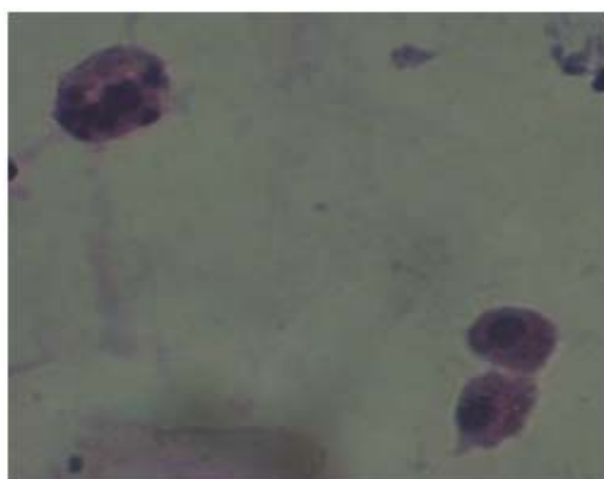


Figure 3b.

Bronchoalveolar lavage (BAL) cytology showing an increase in cell number, predominantly the characteristic pigment laden macrophages (figure 3a). Yellow brown pigment within the macrophages are seen in high power (figure 3b).

Discussion

RB-ILD is a rare clinical entity described in current heavy cigarette smokers. The typical pathological lesion of clusters of pigmented macrophages, which is observed in RBILD, is rarely seen in non-smokers.^{4,5}

The clinical presentation of this patient with predominant dyspnoea was compatible with a clinical diagnosis of interstitial lung disease.

The result of the lung function test was compatible with a mixed restrictive and obstructive ventilatory defect. The restrictive component signified the associated interstitial lung disease while the obstructive component was due to bronchiolitis. Markedly reduced 6MWD indicated a severe respiratory dysfunction.

The chest radiography showed a predominant nodular pattern with a mid and lower zone preponderance. The HRCT of this patient showed ill-defined centrilobular nodules, patchy ground glass opacities with a mosaic perfusion pattern which is compatible with RB. It lacked sub pleural honey-combing and apico-basal distribution pattern which is typical of idiopathic pulmonary fibrosis, an important differential diagnosis.¹

Although upper zone preponderance is commonly described in RBILD, our patient had a predominant mid and lower zone distribution. This pattern of distribution of nodules is also described in the literature.⁴

The characteristic pigment laden macrophages seen in the BAL and the transbronchial biopsy led to the diagnosis of RBILD in this patient. Hypersensitive pneumonitis is the main differential diagnosis here and it was excluded with the fact that having no notable exposure and absence of BAL lymphocytosis.

RBILD is rarely described in non-smokers. However, few inciting agents have been described other than cigarette smoke.³ In our patient, although a life

time non-smoker, exposure to smoke of biomass fuel may be the causative factor.

Due to severe respiratory dysfunction, the patient received inpatient treatment for 6 weeks and had no further exposure to smoke. Upon discharge the patient was advised to improve the ventilation of the kitchen which would reduce further exposure to fire-wood smoke.

Most of the published data on RBILD describes the heterogeneity of the disease. In some case series, smoking cessation alone had alleviated the symptoms while some had shown improvement with addition of a steroid. The response to steroid therapy is variable. However, our patient showed a significant clinical improvement with steroid therapy.

References

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