Pulmonary Alveolar Proteinosis Manifested by Patchy Peripheral Air-space Disease: A Case Report

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We describe the case of a 38-year-old woman with pulmonary alveolar proteinosis in whom a CT (computed tomography) scan of the chest showed patchy peripheral air-space disease, but in whom the central air-space in the lungs was relatively clear. The distribution of pulmonary alveolar proteinosis is usually predominantly central, and only rarely peripheral. (Kitakanto Med J 2003; 53 : 285−287)

Key words: pulmonary alveolar proteinosis, computed tomography, peripheral air-space disease

Introduction

Pulmonary alveolar proteinosis is a severe lung disease characterized by filling of the alveoli by a periodic acid-Schiff (PAS)-positive lipid-rich proteinaceous material.1 There have been many reports on pulmonary alveolar proteinosis since the original paper describing it, but its cause and pathogenesis are unclear. The distribution of pulmonary alveolar proteinosis is usually predominantly central, and only rarely peripheral.2

We report a case of pulmonary alveolar proteinosis associated with patchy peripheral air-space disease in both lungs.

Case Report

An asymptomatic 38-year-old Japanese woman was admitted to a hospital because of abnormal shadows on a routine chest x-ray. On physical examination, there were no crackles or rhonchi in either lung field, or any other abnormal findings. Laboratory tests were normal. A chest x-ray obtained on admission showed peripheral infiltrates in both lungs (Fig. 1), and a computed tomography (CT) scan of the chest showed patchy peripheral air-space disease (Fig. 2). The central air-space of the lungs was relatively clear.

Fig. 1. Chest x-ray on admission showing peripheral infiltrates in both lungs.

Fig. 2. Chest CT scan showing patchy peripheral air-space disease. The central air-space of the lungs is relatively clear. Patchy areas with sharply defined margins between normal and abnormal lung are seen.

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and the patchy areas were characterized by sharply defined margins with normal lung. No cavitations, pleural effusions, or lymphadenopathy was present. Bronchoalveolar lavage yielded a milky effluent, and transbronchial lung biopsy revealed alveolar filling with PAS-positive acellular material (Fig. 3). A diagnosis of pulmonary alveolar proteinosis was made, based on these findings, and a CT scan of the chest two years later showed no changes in the shadows.

**Discussion**

There have been many radiology studies of pulmonary alveolar proteinosis since it was first reported by Rosen. The radiographs of patients with typical pulmonary alveolar proteinosis show a bilateral, diffuse, perihilar or central, ill-defined nodular or confluent pattern, which is usually worse at the bases. The abnormal findings are often more pronounced in the perihilar or central regions and are suggestive of the "butterfly" or "bat wing" appearance of pulmonary edema, except that there are no radiographic signs of left cardiac decompensation, such as cardiomegaly, Kerley B lines, fissural thickening, and pleural effusions. An interstitial pattern, patchy disease, and peripheral infiltrates may also be present.

The chest CT scans of patients with pulmonary alveolar proteinosis show air-space filling with a variable and patchy distribution. Characteristic CT scans show a peripheral clear zone with the air-space opacification sharply demarcated from surrounding normal lung tissue, creating a "geographic" pattern. CT has been reported to be useful in the initial assessment or follow-up of patients with pulmonary alveolar proteinosis, but not in making the differential diagnosis. However, CT can be useful in making the differential diagnosis by enabling exclusion of hilar and mediastinal lymphadenopathy (expected in sarcoidosis) and pleural effusion (expected in congestive heart failure with pulmonary edema).

The distribution of disease is usually predominantly central, but sometimes peripheral, and cases with peripheral predominance have a small peripheral or central shadow. Our patient showed extremely unusual CT findings, i.e., patchy peripheral air-space disease without central shadows, the second case of pulmonary alveolar proteinosis manifested by peripheral shadows alone.

The clearing of the perihilar regions after bronchoalveolar lavage is known to be dramatic, but it is unclear why only peripheral shadows and no central shadows were observed in our patients. However, this finding may be attributable to enhancement of the clearing in the perihilar regions and delayed clearing in the peripheral regions.

There is a report of severer symptoms in patients with more peripheral shadows than in patients with shadows in more central regions. Although our patient had only peripheral shadows, she was asymptomatic. The amount of diseased lung present in each case is known to be correlated with patients' symptoms, and our patient may have had no symptoms because of the small amount of diseased lung present, not because of the distribution of the shadows.

There have been reports of prolonged spontaneous remission in patients with untreated pulmonary alveolar proteinosis and a report of a patient with more peripheral shadows who responded less well to therapy than patients with more central shadows. Thus, it is necessary to follow the symptoms and physical findings of the patient. Both our patient and the patient reported by Inui et al. had no symptoms or abnormal findings on physical examinations, and they were followed without treating their pulmonary alveolar proteinosis. It may be possible to follow asymptomatic pulmonary alveolar proteinosis patients with patchy peripheral air-space disease without treating them.

**References**