Air Leak Syndrome as One of the Manifestations of Bronchiolitis Obliterans Organizing Pneumonia

Tomoaki Iwanaga, Takako Hirota and Togo Ikeda

Abstract

A 46-year-old man developed respiratory distress with air leak syndrome (ALS), including pneumothorax, pneumomediastinum, and subcutaneous emphysema. Open lung biopsy was performed and revealed the histopathologic evidence of bronchiolitis obliterans organizing pneumonia (BOOP), which responded well to steroid treatment. As far as we know, this appears to be the first case of BOOP presenting with ALS as one of its major complications.

Key words: pneumothorax, subcutaneous emphysema, pneumomediastinum, open lung biopsy

Introduction

Idiopathic bronchiolitis obliterans with organizing pneumonia (BOOP) is a clinicopathologic syndrome first proposed by Epler et al (1) which is characterized by an indolent clinical course and a favorable prognosis, although a fulminating variant has been documented (2). BOOP features the ingrowth of polypoid fibroinflammatory granulation tissue from the bronchioles into the adjacent alveoli where organizing pneumonia forms. The typical patient presents with dyspnea, cough, fever, weight loss and single or multiple alveolar opacities on the chest radiograph. Air leak syndrome (ALS), comprising pneumothorax, pneumomediastinum, and subcutaneous emphysema, has never previously been reported as part of the presentation of BOOP. Here we report a case of BOOP in which ALS was the major presenting symptom.

Case Report

A 46-year-old man was hospitalized after presenting with dry cough, respiratory distress, and prominent subcutaneous emphysema. He was in his usual state of health until 4 months prior to hospital admission when he developed a persistent cough. He was treated with antitussives and bronchodilators without complete resolution of his symptoms. Four days prior to admission, he developed moderate respiratory distress associated with a severe dry cough and low-grade fever. He had no known allergies and no history of smoking.

Physical examination revealed a temperature of 37.7°C orally, a heart rate of 126 beats/min, a respiratory rate of 24/min, and massive subcutaneous emphysema extending over the chest and upper abdomen. Initial laboratory studies showed a WBC count of 13,490/µl with 10% eosinophils. Arterial blood gas analysis while breathing 5 l/min of oxygen showed a pH of 7.42, a PCO2 of 42, and a PO2 of 65. Blood and sputum cultures, and Gram staining and acid-fast preparations of the sputum were negative.

The chest radiograph (Fig. 1, upper) showed bibasilar infiltrates and prominent subcutaneous emphysema. Chest CT scanning (Fig. 1, lower) demonstrated bilateral airspace consolidation, limited pneumothorax, pneumomediastinum, and subcutaneous emphysema. Bronchoscopy was performed shortly after admission. Histologic examination of the transbronchial biopsy specimens revealed inflammatory infiltrates with focal fibrosis. Bronchoalveolar lavage fluid from the upper lobe of the right lung showed an increase in lymphocytes (54%) with a decrease in the CD4/CD8 ratio (0.29).

Open lung biopsy was performed on the left upper and lower lobes revealing multiple foci of myxomatous fibrous tissue obliterating the distal air spaces and associated with an interstitial inflammatory infiltrate (Fig. 2). These findings and the bronchoalveolar lavage (BAL) data were indicative of BOOP. None of the serologic parameters or tissue cultures for evaluation of infectious causes were positive. Therapy was started with 40 mg per day of prednisolone and there was gradual improvement of his symptoms. His ALS disappeared without any specific interventions including chest tube insertion. The prednisolone dose was progressively tapered and he showed a satisfactory course.
Figure 1. A chest radiograph (upper) and a CT scan of chest (lower) showing bilateral pulmonary infiltrates as well as massive subcutaneous emphysema, limited pneumothorax, and pneumomediastinum.

Discussion

ALS, comprising pneumothorax, pneumomediastinum, and subcutaneous emphysema, has been described as a complication of many lung diseases. The most common disorders underlying secondary ALS are chronic obstructive pulmonary disease and asthma, in which rupture of bullae occurs. For example, pneumomediastinum is a well-described complication of acute asthma, having been noted in 5.4% of 479 chest X-ray films of children admitted with asthma attacks (3). However, anything that produces alveolar overdistension, or a momentary discrepancy between the alveolar pressure and that in the adjacent tissues, will favor alveolar rupture (4). In parenchymal lung diseases, overexpansion of the distal air spaces beyond sites of small airway obstruction leads to alveolar rupture triggered by coughing or straining (5). Once it reaches the mediastinum, air tends to spread throughout the thorax into the subcutaneous tissues.

The hallmark pathologic change of BOOP is the presence of granulation tissue plugs within the lumens of the distal bronchioles extending into the alveolar ducts and alveoli (1). Although lung function studies show no airflow obstruction except in smokers (6), localized or regional peripheral obstruction can result in a ball-valve effect and distal overdistension leading to burst alveoli and entry of air into the bronchovascular sheath followed by manifestation of any form of aberrant air trapping, or ALS. The present patient had a severe cough which might have caused overpressurization of the alveoli. Though cough and dyspnea are common clinical features of BOOP (7), there has been no mention of severe cough causing ALS. To our knowledge, ALS has not previously been described as the cardinal presenting manifestation of this disease and this appears to be the first case of a BOOP patient with prominent ALS.

Since ALS, pulmonary infiltrates, and eosinophilia of the peripheral blood and sputum were the predominant presenting features in the present patient, eosinophilic pneumonia was a possible alternate diagnosis. However, lymphocytosis with a decreased CD4/CD8 ratio in the BAL fluid made this possibility unlikely and compatible with BOOP (8), which was confirmed by open lung biopsy. The possibility of asthma was not likely because of lack of airflow obstruction. Currently, no consensus exists regarding the optimum dose or duration of corticosteroid therapy for BOOP. Our patient responded gradually to intravenous prednisolone with an initial dose of 40 mg a day and slow tapering over several months. Since his pneu-
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Although the clinical and radiological features of BOOP are often distinctive, it may be masked by an unusual presentation such as ALS in the present case. We conclude that BOOP should be included in the differential diagnosis of patients presenting with ALS. Open lung biopsy for confirmation of the diagnosis is important, because BOOP usually shows a good response to corticosteroids, as was noted in our patient.

References


