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Case report

Successful unilateral partial lung lavage in a child with pulmonary alveolar proteinosis

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Abstract Pulmonary alveolar proteinosis (PAP) is a rare disorder in which lipoproteinaceous material acumulates within the alveoli. A 4-year-old child with autoimmune PAP, who was successfully treated with a series of unilateral partial bronchoalveolar lavages by selectively ventilating the other lung with a cuffed endotracheal tube, is presented.

1. Introduction

Pulmonary alveolar proteinosis (PAP) is a rare respiratory disease characterized by the accumulation of surfactant-derived material in the lungs. There have been important advances in the pathogenesis of the disease. Congenital PAP, secondary PAP, and autoimmune PAP previously were classified as idiopathic, acquired, and primary PAP [1].

Congenital PAP is characterized by acute onset immediately after birth, with respiratory distress and rapid progression [1]. Most cases of congenital PAP are transmitted autosomal recessively and caused by homozygosity for a frameshift mutation (121ins2) in the surfactant protein B gene (SP-B), which leads to secondary disturbances of surfactant protein C (SP-C) processing [2]. Despite glucocorticoids and exogenous surfactant substitutions, SP-B deficiency leads to death within the first year of life [3]. Mutations in the SP-C can lead to similar forms of neonatal respiratory distress [4]. Recently, mutations in the ATP binding casette transporter A (ABCA) gene, a gene implicated in surfactant metabolism, were shown in newborn babies with fatal respiratory distress [3]. The ABCA3 gene encodes a protein highly expressed in the lung, which has been localized to the limiting membrane of lamellar bodies, implicating ABCA3 as possibly important in the maturation surfactant production [5]. Four of 16 lung biopsies in one report [5] and two of 11 biopsies in another report [6] of children with ABCA3 mutations were found to be consistent with PAP; all died.

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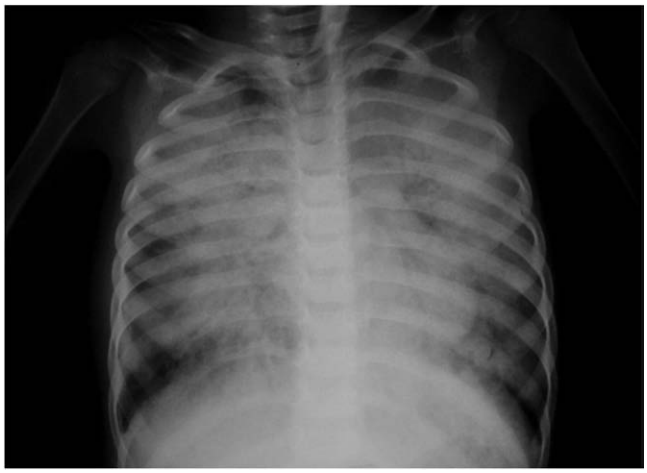

Fig. 1 Chest radiograph showing bilateral diffuse air space consolidation.

Autoimmune, or idiopathic, acquired, or primary, as it was once known, PAP is characterized clinically by cough, dyspnea, and progression to respiratory failure [1]. Animal studies have shown that genetically altered mice that were homozygous for a disrupted granulocyte-macrophage colony-stimulating factor (GM-CSF) gene (GM -/- mice) or its receptor (GM Rβc -/- mice), developed a lung lesion with histologic resemblance to PAP, which suggested that an abnormality in GM-CSF signaling may be involved. Clearance of surfactant lipids and surfactant proteins by alveolar macrophage in these mice is severely impaired, a finding that provides an explanation for the increase in surfactant accumulation [7]. These findings suggest that GM-CSF bioactivity in the lung is critical for surfactant homeostasis in humans. In addition, neutralizing autoantibody against GM-CSF was found in the bronchoalveolar lavage fluid and serum of patients with idiopathic PAP. These observations suggest that human PAP may be an autoimmune disease [8]. Recombinant human GM-CSF administered to adults with PAP, either subcutaneously or by inhalation, has resulted in significant benefit [9-13].

Secondary PAP is known to have a number of underlying causes, including lysinuric protein intolerance, acute silicosis and other inhalational syndromes, immunodeficiency syndromes, malignancies, and hematopoietic disorders [4].

Treatment of PAP depends on the underlying cause. The current therapy for the congenital PAP is supportive only, although the successful lung transplantation of two infants with SP-B deficiency has been reported [14]. Treatment for secondary PAP involves treatment of the underlying condition. Autoimmune PAP is treated with whole lung lavage, GM-CSF, and plasmapheresis [2]. Whole lung lavage is a method of physically removing the excess alveolar lipoproteinaceous material [2,15]. In the adult patient, it can be performed with a double-lumen endotracheal tube, by permitting ventilation of one lung during lavage of the other lung. However, in young children, there is always difficulty in performing this lavage because of the unavailability of small, double-lumen endotracheal tubes (ETTs).

A 4-year-old child with autoimmune PAP, who was successfully treated with series of unilateral partial bronchoalveolar lavages (BAL) while the other lung was selectively ventilated with a cuffed ETT, is presented.

2. Case report

A 4-year-old boy presented with cough, night sweats, and weight loss occurring for 5 months. In his past history, he was the second child of consanguineous parents and was diagnosed to have pneumonia at one and 1.5 years of age. Because of his dyspnea and cough, he was treated with antibiotics and antituberculous medications. As his complaints did not resolve, he was admitted to a university hospital. He was diagnosed with PAP based on alveolar-interstitial infiltrates on chest radiograph and high-resolution computed tomography (Figs. 1 and 2). The diagnosis was confirmed by open-lung biopsy, which showed periodic acid-Schiff (PAS) positive material within the alveoli (Fig. 3). He was referred to our hospital for treatment.

On admission, he still had cough and dyspnea. He weighed 14 kg and his height was 102 cm. His heart rate was 138 beats per minute and his respiratory rate (RR) was 60 breaths/min. He had cyanosis and intercostal and subcostal retractions; auscultation showed decreased aeration and tubular sounds on the left lung. His transcutaneous oxygen saturation while breathing room air was 76%. The minimum oxygen flow needed to increase his transcutaneous oxygen saturation to 92% was 5 L/min. His serum was tested at the Cytokine Biology Laboratory, at Cleveland Clinic Foundation, Cleveland, OH, where no detectable anti-GM-CSF antibody was noted.

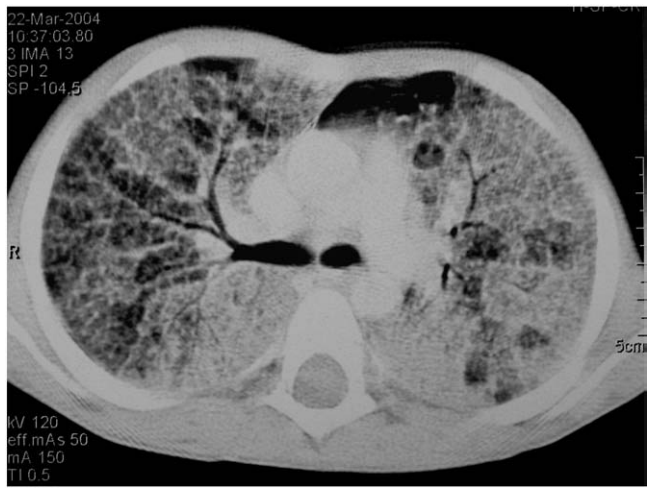

Fig. 2 High-resolution chest computed tomographic view at the level of the carina, showing patchy areas of ground-glass opacity and thickened interlobular septa bilaterally.