CHEST™

An Unusual Cause of Respiratory Failure in a 25-Year-Old Heart and Lung Transplant Recipient

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A 25-year-old woman, a never smoker with a history of heart-lung transplantation for World Health Organization group 1 pulmonary arterial hypertension performed 20 months prior to presentation, was evaluated for shortness of breath. Following transplantation, she was initiated on standard therapy of prednisone, tacrolimus, and azathioprine, along with routine antimicrobial prophylaxis. Her posttransplant course was complicated by persistent acute cellular rejection, as determined from a transbronchial biopsy specimen, without evidence of rejection in an endomyocardial biopsy specimen. The immunosuppressive medications were supplemented with pulse-dosed steroids, and the patient was transitioned from azathioprine to mycophenolate mofetil. Sirolimus was added 9 months prior to presentation. Three months prior to presentation, she was admitted for increasing oxygen requirements, shortness of breath, and bilateral infiltrates on the CT scans of the chest. CHEST 2015; 147(5):e185-e188

Physical Examination Findings

The patient was afebrile with a heart rate of 89 beats/min, BP of 124/82 mm Hg, respiratory rate of 20 breaths/min, and an oxygen saturation of 85% on room air and 94% on 2 L of oxygen. She was alert, oriented, and in mild respiratory distress. Examination revealed a clear oropharynx with moist mucosa. The neck examination showed no jugular venous distention, and lung auscultation revealed scattered, fine inspiratory crackles over the lung fields bilaterally. The heart rate and rhythm were regular with no murmur, gallop, or rub. There was no clubbing or cyanosis, but trace peripheral edema was present. The patient was noted to have mild wasting of muscles diffusely and appeared slightly Cushingoid.

Diagnostic Studies

The chest CT scan without IV contrast revealed diffuse ground-glass infiltrates with thickening of the alveolar septa in upper and mid-lung zone predominance (Fig 1). Pulmonary function testing revealed a restrictive pattern

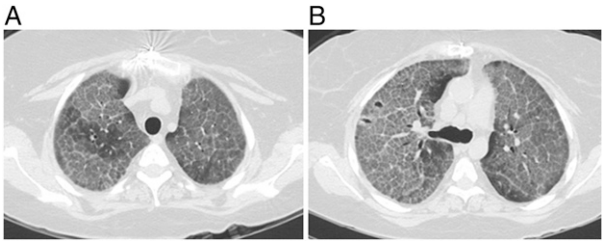

Figure 1 – *A, B, Different slices from a CT scan of the chest demonstrate diffuse ground-glass infiltrates with alveolar septal thickening.*

with a total lung capacity of 2.98 L (63% predicted) and a diffusion capacity of 33% predicted. Compared with prior pulmonary function testings, there had been a significant decline in total lung capacity and diffusion capacity of the lung for carbon monoxide.

Bronchoscopy was performed. Transbronchial biopsy specimens revealed scattered intraalveolar foamy histiocytes and edema fluid (Fig 2). The bronchoalveolar fluid was described as cloudy. A periodic acid-Schiff stain was positive.

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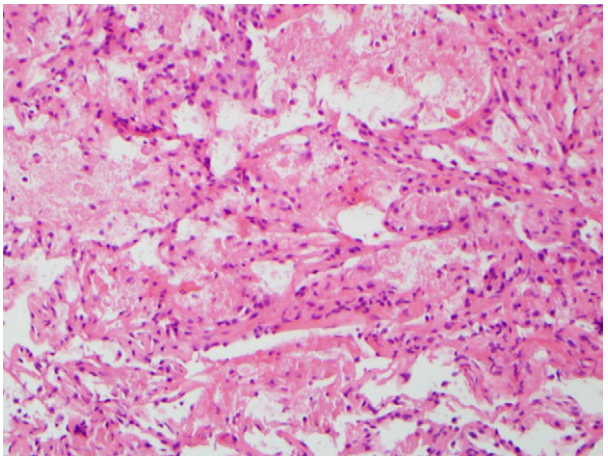

Figure 2 – *Hematoxylin and eosin stain showing scattered intraalveolar foamy histiocytes and edema fluid (original magnification × 400).*

What is the diagnosis?