Pulmonary Alveolar Proteinosis: Crazy Paving to Whole Lung Lavage

Shahnaz Ul Rehman, MD, and Jeffrey R. Hammersley, MD

Pulmonary alveolar proteinosis (PAP) is a rare disorder in which lipoproteinaceous material accumulates in alveoli. A 47-year-old white man with a 15–pack-year smoking history visited his primary care physician for progressive shortness of breath. A chest radiograph showed diffuse pulmonary opacification. A high-resolution chest computed tomographic scan showed the crazy-paving pattern of diffuse interstitial and ground-glass opacification throughout the lung fields (Figure 1). A wedge lung biopsy was performed; histologic examination showed periodic acid–Schiff (PAS)–positive granular material (Supplemental Figure, available online at http://www.mayoclinicproceedings.org) and alveolar macrophages with intracytoplasmic debris consistent with PAP.

Pulmonary alveolar proteinosis is a diffuse lung disease characterized by accumulation of PAS-positive lipoproteinaceous material in alveoli, with little lung inflammation and preserved architecture. Clinical presentation is usually the insidious onset of dyspnea and cough in a middle-aged male smoker. However, one-third of patients are asymptomatic. Physical examination findings are usually unremarkable; expectoration of white gelatinous material may occur. Laboratory abnormalities include polycythemia and increased lactate dehydrogenase levels. Typical findings on high-resolution chest computed tomographic scans are the crazy-paving pattern, and bronchoalveolar lavage or lung biopsy staining positive for PAS is diagnostic. The autoimmune form of PAP accounts for 90% of cases and is characterized by the presence of granulocyte-macrophage colony-stimulating factor autoantibodies that are believed to contribute to macrophage dysfunction and impaired surfactant processing.

Whole lung lavage is a widely accepted form of treatment (Figure 2). The median duration of clinical benefit from lavage is

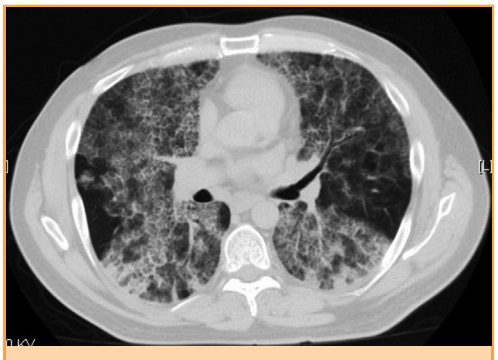

FIGURE 1. High-resolution chest computed tomographic scan shows the crazy-paving pattern of diffuse interstitial and ground-glass opacification throughout the lung fields.

FIGURE 2. The fluid retrieved from the whole lung lavage shows milky lipoproteinaceous material. The first bottle is more milky and turbid.

From the Department of Pulmonary, Critical Care, and Sleep Medicine, University of Toledo Medical Center, Toledo, OH.

Scan code to view additional images

approximately 15 months. Administration of subcutaneous or inhalational granulocyte-macrophage colony-stimulating factor for PAP remains experimental, but shows promise. Lung transplant has been performed in refractory patients.

ACKNOWLEDGMENTS

We thank Nada Siddique Khan, MBBS; Thomas Vacek, MD; Abdulmonam Ali, MD; Natalya Kozodoi, MD; Zaidi Ammari, MD; and Ragheb Assaly, MD, for their assistance with this submission.

SUPPLEMENTAL ONLINE MATERIAL

Supplemental material can be found online at: http://www.mayoclinicproceedings.org. Supplemental material attached to journal articles has not been edited, and the authors take responsibility for the accuracy of all data.

Abbreviations and Acronyms: **PAP** = pulmonary alveolar proteinosis; **PAS** = periodic acid—Schiff

Correspondence: Address to Shahnaz Ul Rehman, MD, Department of Pulmonary, Critical Care, and Sleep Medicine, University of Toledo Medical Center, 3000 Arlington Ave, Toledo, OH 43614 (drshahnazmd@gmail.com).