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...
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.

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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

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