

Silicoproteinosis Masquerading as Community-Acquired Pneumonia

CPT J. Esteban Palacio, MD, USA MC, and CPT Anne Champeaux, MD, USA MC

Pulmonary alveolar proteinosis is a rare disorder first described in the literature in 1958. It has been associated with numerous causes, including silica exposure,¹⁻⁴ infection,² malignancy,^{2,5} and inhalation of assorted irritant dusts or solvents.⁶ It is a disease process characterized by an accumulation of granular periodic acid-Schiff (PAS)-positive material in the alveoli. Disruption of type II pneumocytes might contribute to the disease process.⁴ The chest radiograph classically shows bilateral symmetrical air space disease, although variations have been reported. The clinical symptoms and signs of pulmonary alveolar proteinosis are varied, ranging from asymptomatic to acute respiratory failure.⁷

The case described below illustrates the symptoms of mild silicoproteinosis, a specific variant of pulmonary alveolar proteinosis associated with silica dust exposure and silicate particles in alveolar material. Silicoproteinosis can imitate other pulmonary disease processes, such as community-acquired pneumonia. Silicoproteinosis can be due to a unique pathophysiologic insult or a superinfection. Infectious agents include multiple possible organisms, such as *Nocardia*, staphylococcus, *Haemophilus influenzae*, mycobacterium, and *Cryptococcus*.^{2,8,9} In addition to the treatment of different infectious causes, recognition of pulmonary alveolar proteinosis is essential in its management. Bronchoalveolar lavage is the effective treatment in the acute setting, whereas longer term management includes removing the patient from the inciting exposure and treating residual symptoms supportively.

Case Report

A 25-year-old man complained of flu-like symptoms of diaphoresis, chills, headache, and mini-

mally productive cough. Penicillin was prescribed for treatment of a presumed upper respiratory tract infection. He returned the following day with increasing cough and dyspnea and crackles on auscultation. He developed blood-tinged sputum lasting only 1 day, but his cough, malaise, and other symptoms persisted. On the third day he reported pleuritic chest pain, increased malaise, and worsening dyspnea. When examined, he appeared fatigued and diaphoretic, and he had scattered fine crackles throughout both lung fields, most predominant at the right base. Pulse oximetry saturation on room air was 90% to 93%. Oxygen was provided through a nasal cannula. Findings on a chest radiograph were notable for diffuse, symmetrical, bilateral air space disease. He had leukocytosis, with a white blood cell count of 14,700/ μ L, and on sputum analysis there were 10 to 25 white blood cells per low-power field without predominant organisms.

He was admitted to the hospital for respiratory distress and was given erythromycin and ceftriaxone for presumed atypical community-acquired pneumonia. His temperature rose to 100.5°F on day 1 of hospitalization; otherwise, he was afebrile, and there was minimal improvement of his pleurisy despite unchanged auscultatory findings on examination. His leukocytosis resolved, but an eosinophilia up to 15.6% developed on his fifth hospital day. During a review of occupational exposures, he reported he had inhaled fine cement dust while wet- and dry-grinding of containers made of cement 8 to 10 hours a day for the previous 3 weeks without using proper protective equipment. Thus, pulmonary alveolar proteinosis was entertained as a diagnosis. Bronchoscopy with segmental lavage returned a copious amount of pink milky appearing effluent (red blood cell count 940/ μ L, white blood cell count 680/ μ L, neutrophils 18%, lymphocytes 28%, eosinophils 32%, basophils 1%, macrophage 21%). His pleuritic discomfort improved considerably with bronchoalveolar lavage. Likewise, substantial clearing of bibasilar diffuse infiltrates appeared on a chest radiograph.

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From the MEDAC Family Practice Center (JEP, AC), United States Army, Fort Leonard Wood, Mo. Address reprint requests to J. Esteban Palacio, MD, CPT, USA MC, 126 Missouri Ave, Ft. Leonard Wood, MO 65473.

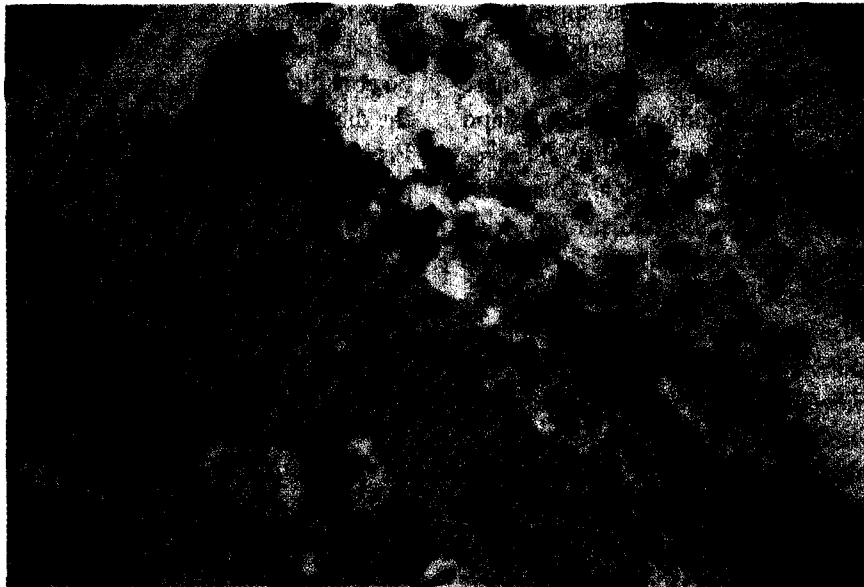


Figure 1. Cytologic findings from bronchoalveolar lavage: numerous eosinophils with scattered foam cells.

Cytologic examination of the bronchoalveolar lavage showed numerous eosinophils with scattered foam cells (Figure 1), PAS-positive amorphous granular material, and minute birefringent particles (Figure 2). Cold agglutinins, blood cultures, and sputum cultures all were negative except for an initial, single positive aerobic blood culture presumed to be contaminant. Soon after the bronchoalveolar lavage, the patient's condition returned to baseline function, oral antibiotics were pre-

scribed, and he was instructed to avoid any further exposure. During a follow-up examination 3 weeks later, he was asymptomatic, and there were normal findings on a chest radiograph. Efforts were made to change his job to avoid further exposures.

Discussion

Pulmonary alveolar proteinosis, silicoproteinosis, is a diagnosis based on finding silica particles with

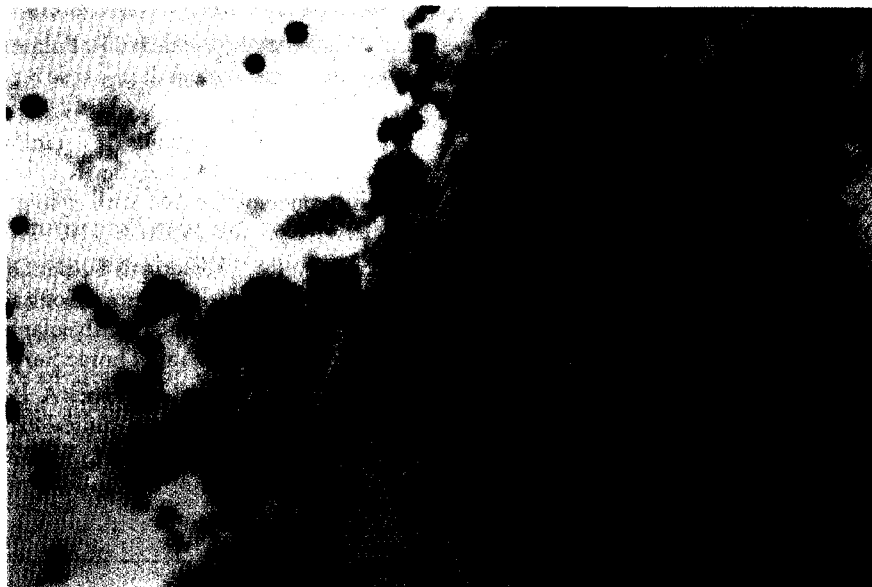


Figure 2. Cytologic findings from bronchoalveolar lavage: periodic acid-Schiff-positive amorphous, granular material and minute birefringent particles.

PAS-positive alveolar material in the appropriate clinical setting. Silicoproteinosis should not be confused with silicosis and acute silicosis, as these diseases are associated with pulmonary nodules and result in a high morbidity and mortality. The reactive lung process called pulmonary alveolar proteinosis is described as "one mechanism by which the lung responds to a variety of insults."⁷ It likely represents a clinical spectrum: the disease process and its severity resulting from an interplay among patient factors and the degree of exposure. A literature review retrieved multiple cases of sandblasters, miners, and individuals in quarries developing pulmonary alveolar proteinosis, but only one case report was found of pulmonary alveolar proteinosis related specifically to cement dust exposure.¹⁰

Patients with presumed pneumonia who do not respond as expected with appropriate antibiotics should have a careful evaluation of their occupational and recreational exposure history to assess whether they are potentially ill with pulmonary alveolar proteinosis. If there is a history of exposure, bronchoalveolar lavage should be considered with a PAS-stain of lavage effluent.

This patient improved considerably with a single bronchoalveolar lavage. Bronchoalveolar lavage has proved to be beneficial in the diagnosis and amelioration of symptoms in multiple reported cases of pulmonary alveolar proteinosis.^{4,9,11,12} In many of these cases of pulmonary alveolar proteinosis, there were multiple lavages performed within varying lengths of time, but several authors^{9,10,13} describe improvement of symptoms in patients with a single bronchoalveolar lavage.

The prognosis of pulmonary alveolar proteinosis is variable and depends on the severity of symptoms and the precipitating cause. Various causes include inhaled irritants, infectious sources, and malignancy. This range of causes emphasizes the importance of occupational history and illustrates how disease resulting from occupational exposures can mimic more common disease states. Mild disease can go unrecognized. Patients with presumed atypical community-acquired pneumonia who fail to show improvement or who have a history of expo-

sure to inhaled irritants could have pulmonary alveolar proteinosis. Bronchoalveolar lavage should be considered in the appropriate clinical setting for the diagnosis and acute management of this disease. Prevention of further hazardous environmental exposures constitutes long-term management.

References

1. Diseases associated with exposure to silica and non-fibrous silicate minerals. Silicosis and Silicate Disease Committee. *Arch Pathol Lab Med* 1988;112:673-720.
2. Rosen SH, Castleman B, Liebow AA. Pulmonary alveolar proteinosis. *N Engl J Med* 1958;258:1123-42.
3. Rosenman KD, Reilly MJ, Kalinowski DJ, Watt FC. Silicosis in the 1990s. *Chest* 1997;111:779-86.
4. Xipell JM, Ham KN, Price CG, Thomas DP. Acute silicoproteinosis. *Thorax* 1977;32:104-11.
5. Carnovale R, Zornoza J, Goldman AM, Luna M. Pulmonary alveolar proteinosis: its association with hematologic malignancy and lymphoma. *Radiology* 1977;122:303-6.
6. Davidson JM, Macleod WM. Pulmonary alveolar proteinosis. *Br J Dis Chest* 1969;63:13-28.
7. Rubin E, Weisbrod GL, Sanders DE. Pulmonary alveolar proteinosis: relationship to silicosis and pulmonary infection. *Radiology* 1980;135:35-41.
8. Claypool WD, Rogers RM, Matuschak GM. Update on the clinical diagnosis, management, and pathogenesis of pulmonary alveolar proteinosis (phospholipidosis). *Chest* 1984;85:550-8.
9. Wilson JW, Rubinfeld AR, White A, Mullerworth M. Alveolar proteinosis treated with a single bronchial lavage. *Med J Aust* 1986;145:158-60.
10. McCunney RJ, Godefroi R. Pulmonary alveolar proteinosis and cement dust: a case report. *J Occup Med* 1989;31:233-7.
11. Mason GR, Abraham JL, Hoffman L, Cole S, Lippmann M, Wasserman K. Treatment of mixed-dust pneumoconiosis with whole lung lavage. *Am Rev Respir Dis* 1982;126:1102-7.
12. Selecky PA, Wasserman K, Benfield JR, Lippmann M. The clinical and physiological effect of whole lung lavage in pulmonary alveolar proteinosis: a ten-year experience. *Ann Thorac Surg* 1977;24:451-61.
13. Kariman K, Kylstra JA, Spock A. Pulmonary alveolar proteinosis: prospective clinical experience in 23 patients for 15 years. *Lung* 1984;162:223-31.