Silicoproteinosis Masquerading as Community-Acquired Pneumonia

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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 minimally 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/μL, 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/μL, white blood cell count 680/μL, 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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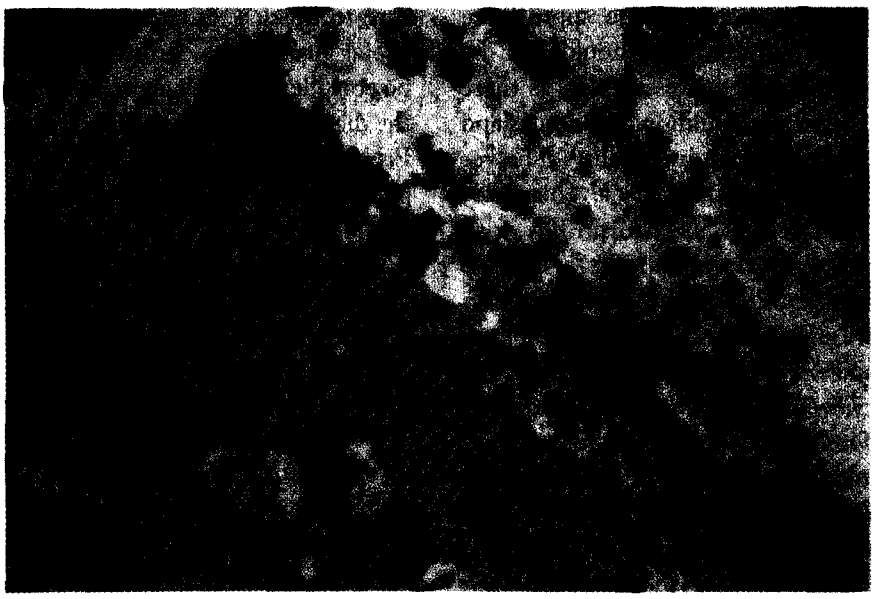

Figure 1. Cytologic findings from bronchoalveolar lavage: numerous eosinophils with scattered foam cells.

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 prescribed, 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 exposure 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.

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