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Pulmonary alveolar proteinosis

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A 32-year-old man presented with cough with white sputum, dyspnea, and intermittent dull pain in the right side of the chest for 3 weeks. He had also lost 6 kg of body weight over the previous month. He disclosed no relevant systemic disease, medication, or travel history. Chest radiography revealed a diffuse alveolar pattern over the bilateral lung fields, favoring a diagnosis of bronchiectasis with superimposed infection. The laboratory data were within normal limits. Empirical antibiotics were prescribed but his condition did not improve. Chest computed tomography showed bilateral ground-glass and crazy-paving patterns (Fig. 1), leading to suspicions of interstitial lung disease. A wedge resection of the lung was performed. Histopathologic examination revealed diffuse deposition of amorphous eosinophilic material in the alveolar spaces (Fig. 2A), with a positive periodic acid–Schiff stain (Fig. 2B).

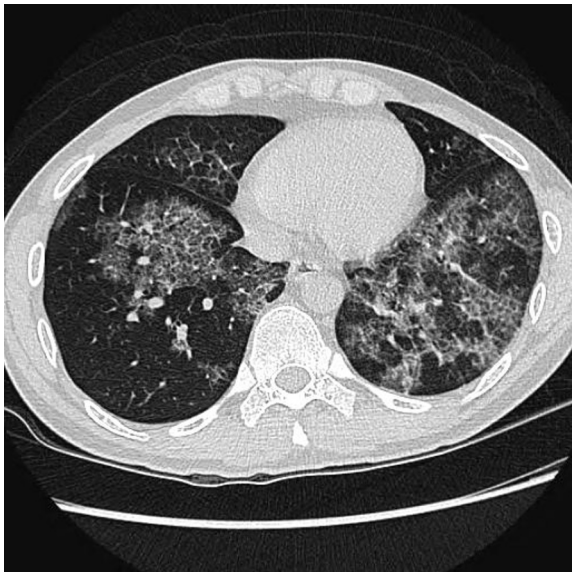

Fig. 1. Computed tomography showing bilateral ground-glass and crazy-paving patterns.

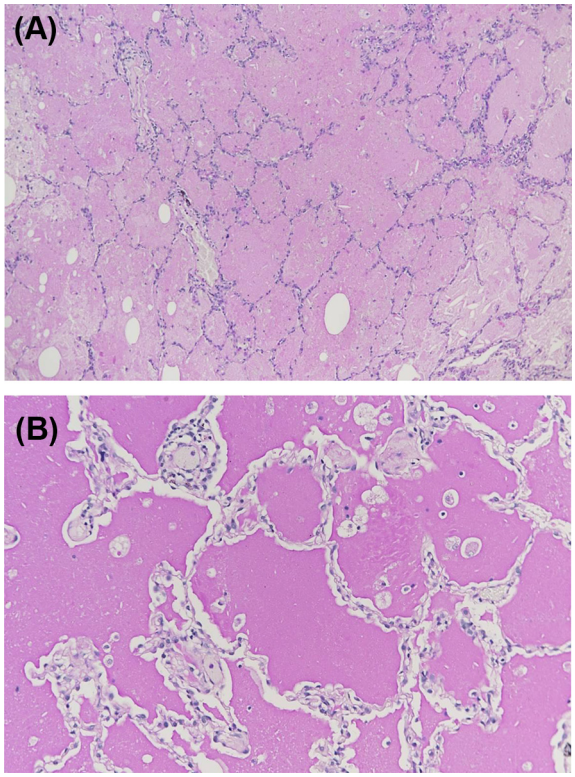

Fig. 2. Histopathology showing diffuse deposition of amorphous eosinophilic material in the alveolar spaces. (A) Hematoxylin–eosin stain, ×100; (B) periodic acid–Schiff stain.

Conflict of interest: none.

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Focal interstitial lymphocytic infiltrates and alveolar macrophages were present. No microorganisms were found on a Gomori methenamine silver stain. Pulmonary alveolar proteinosis (PAP) was diagnosed. There was no relevant history of exposure to occupational dust, infections, or hematologic malignancy. An acquired form of PAP was favored. The patient received supportive care with gradual improvement in his symptoms and follow-up images.

PAP is a diffuse lung disease with an accumulation of acellular surfactant in the distal alveolar spaces. Altered granulocyte macrophage-colony stimulating factor function, alveolar macrophage dysfunction, and impaired clearance play key roles in PAP. There are three forms of PAP: congenital, acquired, and secondary. The acquired form (also known as idiopathic PAP) is the most common. The secondary form is usually associated with hematologic malignancies, exposure to high levels of mineral particles and inhaled chemicals, and infections. Common symptoms include dyspnea on exertion and coughs. High-resolution computed tomography reveals ground-glass opacity and may show a crazy-paving pattern. The differential diagnosis on histology includes pulmonary edema, *Pneumocystis jirovecii* pneumonia and atypical infection. The fluid in pulmonary edema and *Pneumocystis jirovecii* is PAS negative. The choice of treatment depends on the severity of symptoms. Observation and supportive therapy are suggested for asymptomatic patients and those with mild symptoms. Whole lung lavage is recommended for patients with severe symptoms and hypoxemia.

Further reading

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