A case of pulmonary alveolar microlithiasis with Cor Pulmonale

Wen Chen, Tao Gu

Department of Radiology, Beijing Hospital, the Fifth Clinical Medicine College of Peking University, Beijing, China

ABSTRACT Pulmonary alveolar microlithiasis (PAM) is a rare disease characterized by the formation and deposition of microliths within the alveoli and a paucity of symptoms in contrast to the imaging findings. It has familial tendency and is thought to be an autosomal recessive disorder with the mutation in the SLC34A2 gene. We describe a case of PAM with Cor Pulmonale. Ultrasonic cardiogram showed pulmonary hypertension (82 mmHg). Chest radiography revealed diffuse, bilateral sandstorm-like micronodules with greater density in the lower lung fields. HRCT scans demonstrated diffuse ground-grass opacities, thickening and calcification of interlobular septa and confluent calcified nodules. A diagnosis of PAM was suggested and confirmed by transbronchial lung biopsy (TBLB).

KEY WORDS Pulmonary alveolar microlithiasis; Cor Pulmonale; HRCT

Quant Imaging Med Surg 2012;2:65-67. DOI: 10.3978/j.issn.2223-4292.2012.02.02

A 41-year-old man presented with intermittent cough, mild expectoration and progressive shortness of breath for 8 years. His younger brother had similar syndrome and died of unknown lung disease at the age of 25. Physical examination revealed inspiratory crackles, a loud P2 (pulmonic valve closure sound), cyanosis and clubbed fingers. Arterial blood gas demonstrated type I respiratory failure. Pulmonary function test revealed restrictive lung disease with impaired diffusing capacity. Ultrasonic cardiogram showed pulmonary hypertension (82 mmHg) and enlargement of right atrium and ventricle. Chest image examination was then performed. Radiography revealed diffuse, bilateral sandstorm-like micronodules with greater density in the lower lung fields. The heart borders and diaphragmatic surfaces were obliterated (Figure 1). High-resolution CT scan demonstrated diffuse ground-grass opacities, thickening and calcification of interlobular septa and confluent calcified nodules (Figure 2,3). A diagnosis of Pulmonary alveolar microlithiasis (PAM) was proposed. Later the patient underwent transbronchial lung biopsy (TBLB). The pathology showed laminated calcific concretions within the alveoli (Figure 4). The final diagnosis of PAM was established.

No potential conflict of interest.

Corresponding to: Tao Gu, MD. Department of Radiology, Beijing Hospital, Dongcheng, 100730, Beijing, China. Tel/Fax: +86-10-85133901. Email: gutao001@sina.com.

Submitted Jan 31, 2012. Accepted for publication Feb 06, 2012.

Available at http://www.amepc.org/qims

ISSN: 2223-4292

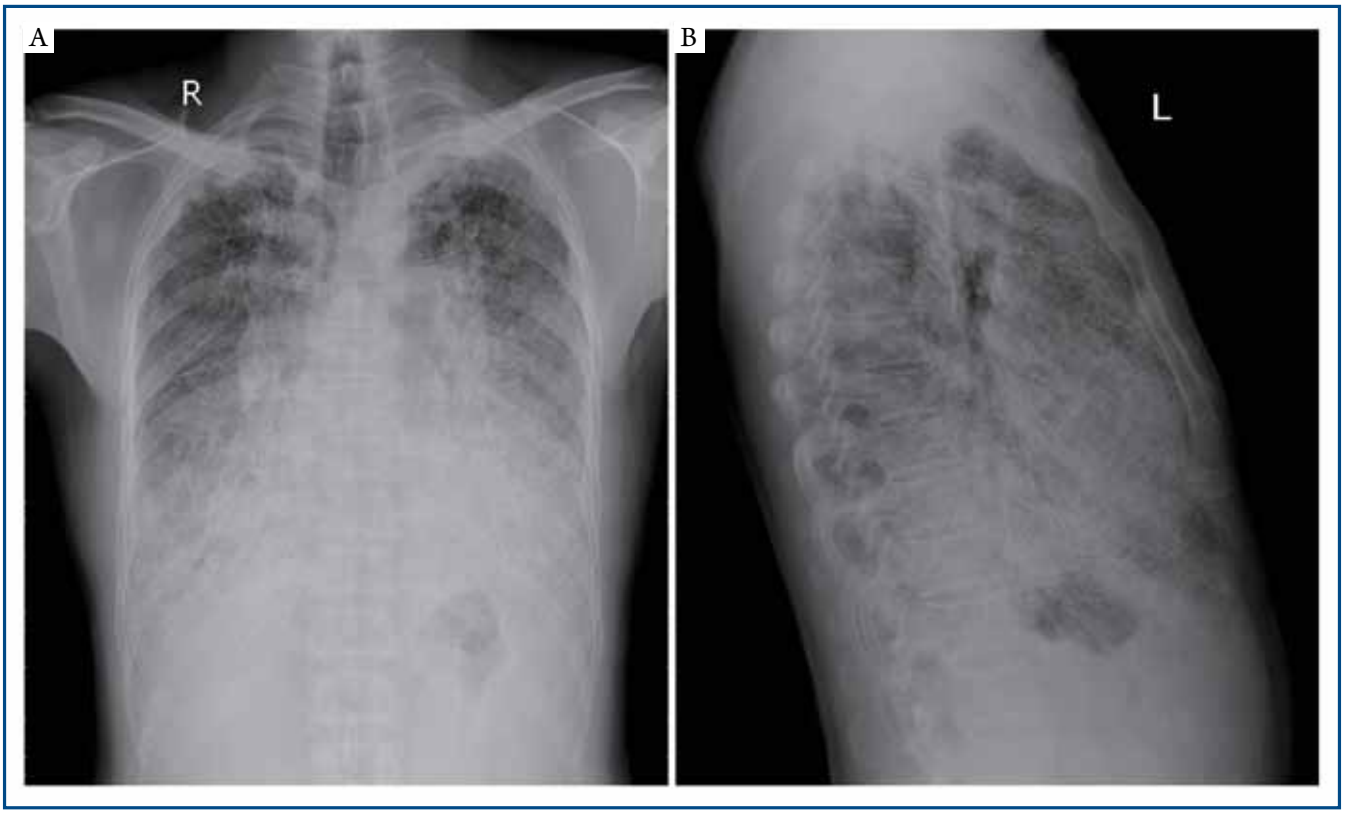

Figure 1. A-B:Chest radiographs reveal the classic sandstorm-like shadow with greater density in the lower lung fields. The heart borders and diaphragmatic surfaces are obliterated.

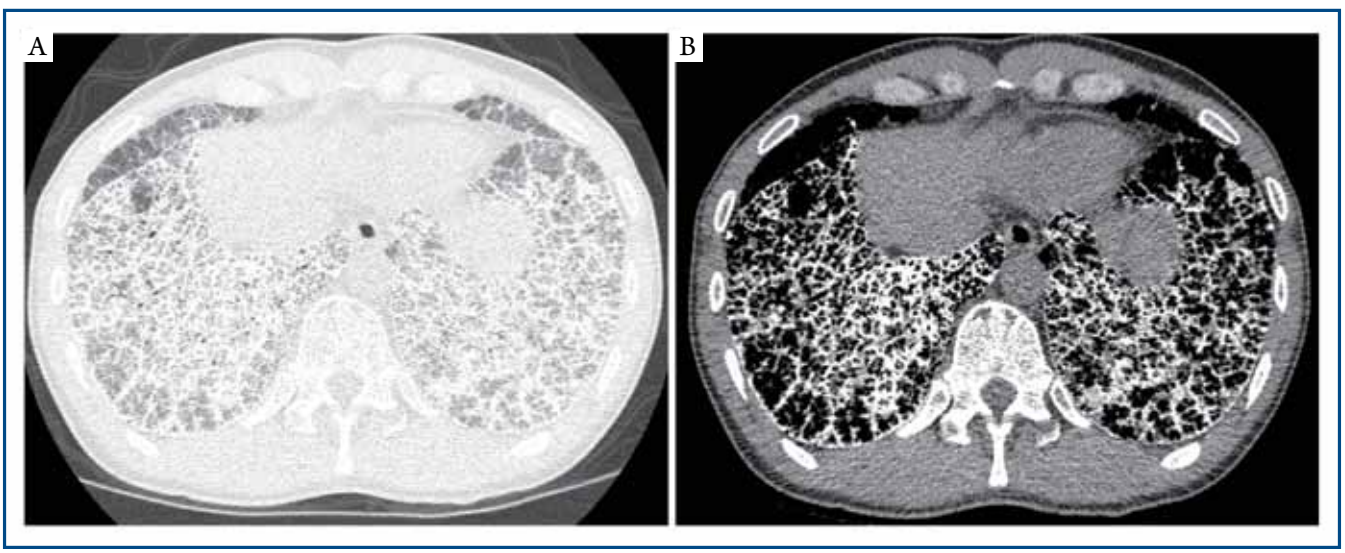

Figure 2. A: Axial CT scan (lung window) demonstrates diffuse ground-grass opacities and thickened interlobar septa; B: Axial CT scan (mediastinal window) shows diffuse calcifications along the interlobar septa, subpleural linear calcification, confluent and calcified nodules.

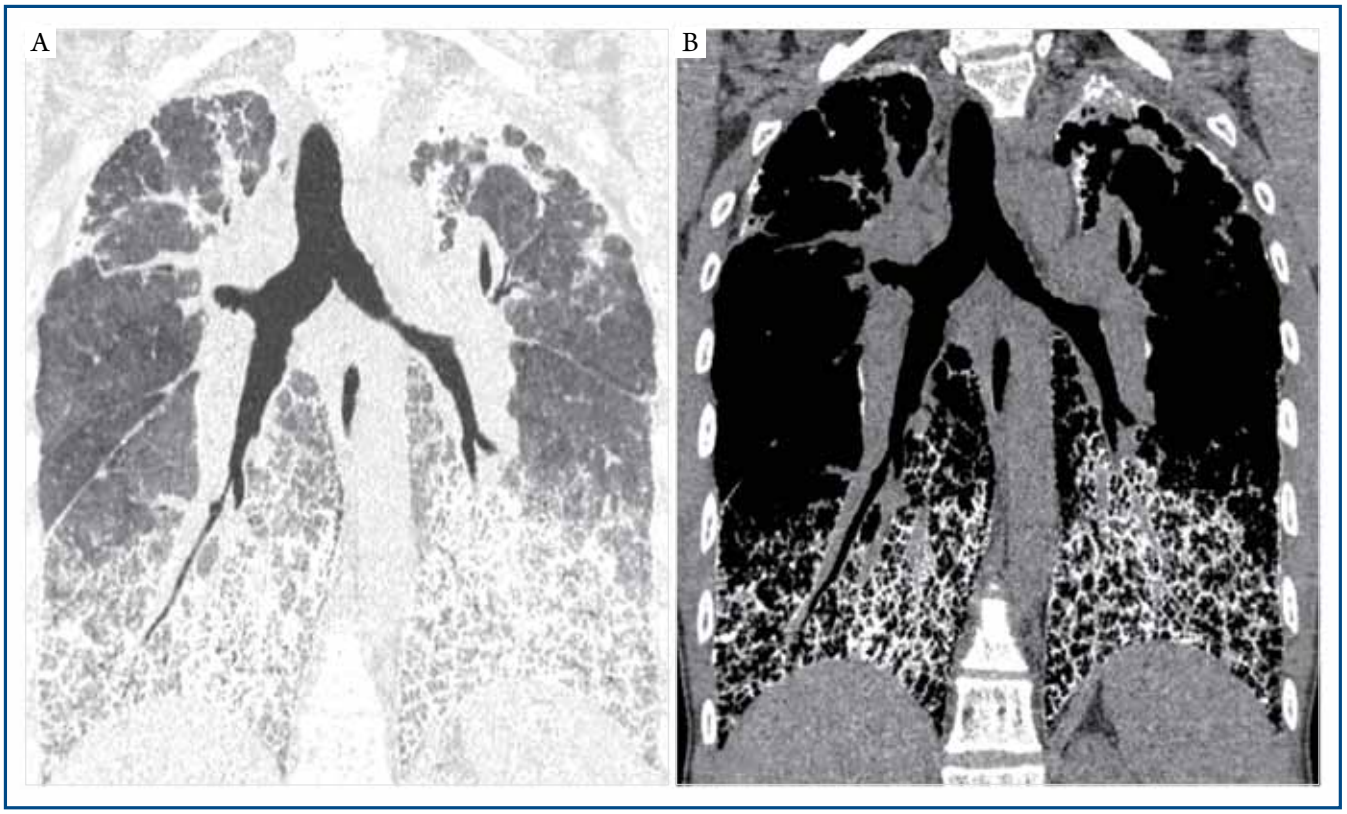

Figure 3. A-B: Coronal multi-planar reformation images show diffuse, bilateral involvement of both lungs a preponderance in the basal segment of lower lobes.

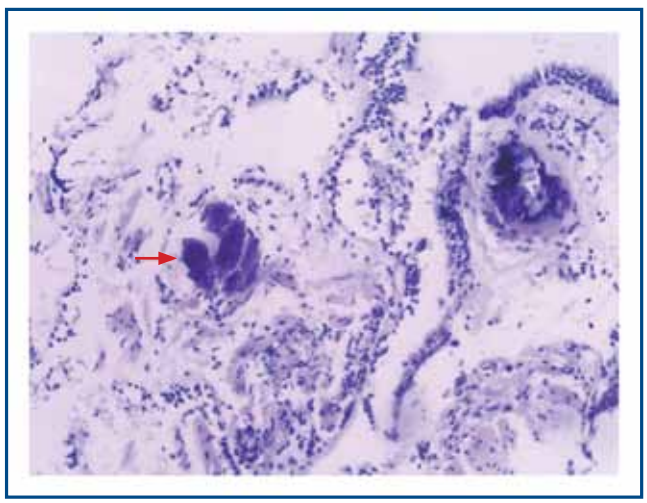

Figure 4. Photomicrograph (original magnification, ×100; hematoxylin-eosin stain) of lung tissue obtained by TBLB reveals laminated calcific concretions within the alveoli (arrow).

Cite this article as: Chen W, Gu T. A case of pulmonary alveolar microlithiasis with Cor Pulmonale. Quant Imaging Med Surg 2012;2:65-67. DOI: 10.3978/j.issn.2223-4292.2012.02.02