



Pulmonary Alveolar Microlithiasis

Nishant Gupta¹ and Francis X. McCormack¹

¹Division of Pulmonary, Critical Care, and Sleep Medicine, Department of Internal Medicine, University of Cincinnati, Cincinnati, Ohio



Figure 1.

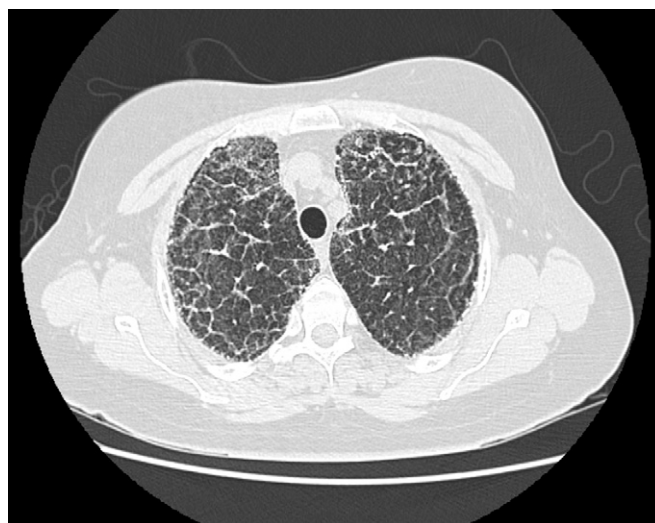


Figure 2.

A 59-year-old Middle Eastern female with a history of pulmonary alveolar microlithiasis (PAM) was referred to our clinic for second opinion. The patient had been diagnosed in her childhood but was asymptomatic until she developed exertional dyspnea at the age of 54 years. Chest examination revealed bibasilar crackles. Pulmonary function tests revealed mild restriction and reduced diffusing capacity. Six-minute-walk test revealed normal exercise capacity (distance walked, 1,800 ft) with mild exertional desaturation to 88%. Chest X-ray (Figure 1) showed remarkable, fine nodular lower lung zone opacities, and computed tomography of the chest (Figures 2–4) showed symmetrical lower lobe predominant micronodular calcifications along bronchovascular bundles and interlobular septal thickening, findings pathognomonic for PAM (1). PAM is caused by inactivating mutations in the SLC34A2 gene (2, 3). The mutation leads to reduced phosphate reuptake by type IIb sodium phosphate transporter in the apical membrane of type II alveolar cells, resulting in calcium phosphate chelation and microlith formation in alveolar air spaces. PAM is associated with consanguinity, and the incidence is higher in Turkey, Japan, India, and Italy. PAM can be diagnosed on the basis of characteristic radiological findings (4), and biopsy is not usually required. The clinical course of PAM is highly variable, with some patients developing pulmonary fibrosis and others remaining asymptomatic for decades. Etidronate, a bisphosphonate, reduces the formation of calcium hydroxyapatite crystals and has led to clinical and radiological improvement in few cases of PAM (5). The patient was started on treatment with etidronate.

Author disclosures are available with the text of this article at www.atsjournals.org.

Author Contributions: N.G. wrote the manuscript. F.X.M. edited the manuscript and provided the images.

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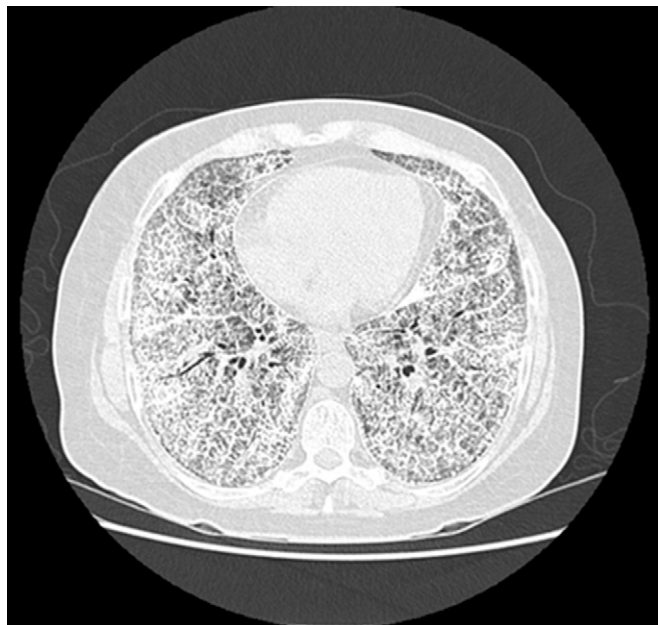


Figure 3.

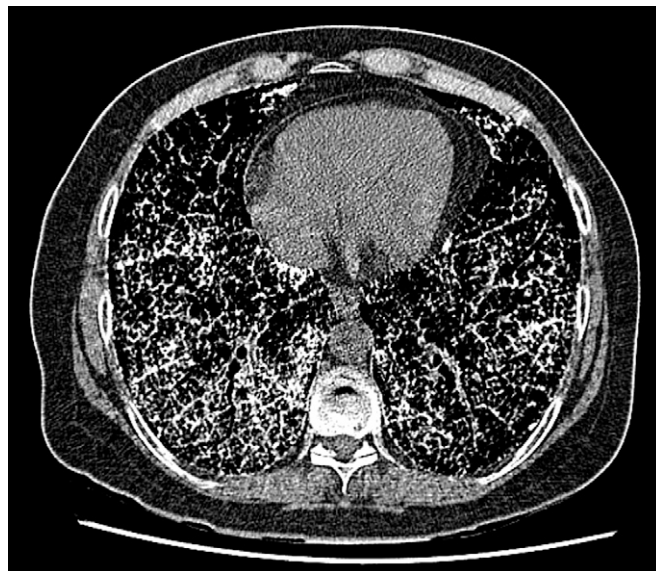


Figure 4.

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