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Emad Alkankan

Hasan Yamin

Hazim Bukamur

Fadi Alkhankan

Yousef Shweihat

See next page for additional authors

Authors
Emad Alkankan, Hasan Yamin, Hazim Bukamur, Fadi Alkhankan, Yousef Shweihat, and Fuad Zeid

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Case Report

Pulmonary alveolar microlithiasis diagnosed with radiography, CT, and bone scintigraphy

Emad Alkhankan, MD[a,*], *Hasan Yamin, MD*[a], *Hazim Bukamur, MD*[a], *Fadi Alkhankan, MD*[a,b], *Yousef Shweihat, MD*[a], *Fuad Zeid, MD*[a]

[a] *Pulmonary and Critical Care, Marshall University, 1249 15th St, Huntington, WV 25701, USA*
[b] *Pulmonary and Critical Care, Mclaren Oakland, Pontiac, MI, USA*

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ABSTRACT

Pulmonary alveolar microlithiasis is rare disease characterized by accumulation of calcium phosphate microlithis in the alveoli. The pathogenesis relates to mutation in the gene SLC34A2 (solute carrier family 34 member 2) located on chromosome 4p15.2, which produces a defective sodium-phosphate cotransporter in alveolar epithelial type-2 cells, making these cells unable to clear phosphorus released during recycling of surfactant [1].

In this article we report a case of pulmonary alveolar microlithiasis who presented with progressive dyspnea on exersion, and was diagnosed based on typical imaging findings, obviating the need for lung biopsy.

Case presentation

A 67-year-old female presented with complaints of slowly progressive shortness of breath on exertion and intermittent dry cough for 7 years. She was a nonsmoking teacher with no previously known pulmonary disease until 2012. She was suspected to have an interstitial lung disease because of persistent bilateral infiltrates on chest xray (CXR) (Fig. 1); however, no further workup was done at that time. She denied fever or hemoptysis. She was treated empirically multiple times in the past with steroids with no improvement in her symptoms.

On physical examination, she was found to be tachypnic RR 24, and hypoxemic with SpO_2 of 87% on room air improved to 92% on 2 L nasal O_2. Bilateral fine crackles were noted on auscultation with finger clubbing but no peripheral edema. Routine biochemistry including serum calcium and phosphorus were normal.

A chest radiograph showed bilateral diffuse opacities with sandstorm-like appearance suggestive of alveolar

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* Corresponding author.
E-mail address: alkhankane@live.marshall.edu (E. Alkhankan).

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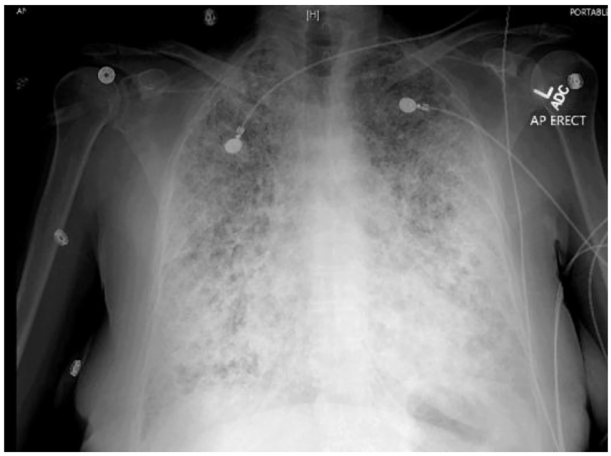

Fig. 1 – Chest X-ray showing diffuse bilateral infiltartes.

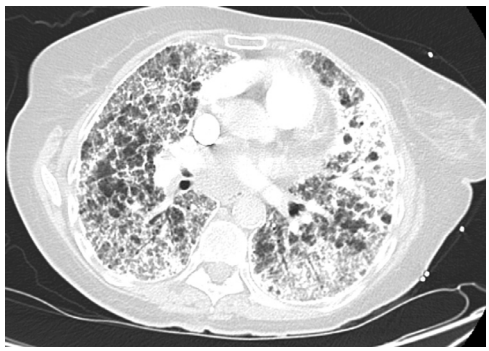

Fig. 2 – CT Scan of the chest without contrast showing diffuse calcification with septal thickening.

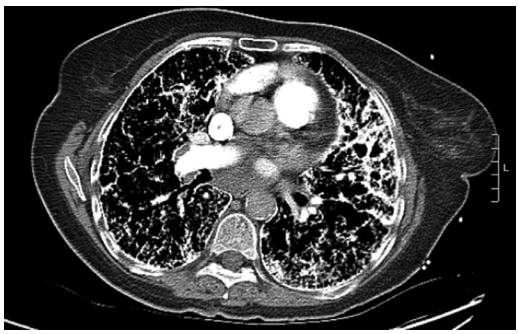

Fig. 3 – CT Scan of the chest without contrast showing diffuse ground glass attenuation with septal thickening.

microlithiasis. A chest CT showed diffuse calcifications in the form of intra-alveolar microliths, calcified interlobular septa, and diffuse ground-glass attenuation with septal thickening (Figs. 2,3). To further confirm the diagnosis Technetium 99^{m}-methylene diphosphonate bone scan was done, and showed diffuse radiotracer uptake (Fig. 4). These finding were deemed adequate to establish a diagnosis of pulmonary alveolar microlithiasis (PAM). Patient refused lung transplant referral

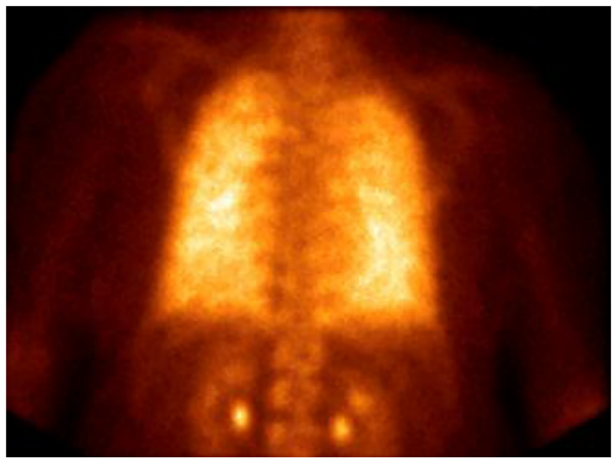

Fig. 4 – Technetium 99^{m}-methylene diphosphonate ($Tc99^{m}$-MDP) showing diffuse radiotracer uptake.

on discharge and she was provided with 3 L nasal O_2 after assessed for home oxygen requirement.

Discussion

PAM is a rare autosomal recessive disease with high penetrance, in which concretions composed of calcium phosphate fill alveolar spaces, despite normal serum calcium and phosphorus, and absence of any systemic disease of calcium metabolism [2]. Over 1000 cases were reported worldwide especially in Mediterranean countries [3]. Several mutations in the gene SLC34A2 on chromosome 4p15.2 are well known to cause the disease [1,4].

This disease is typically diagnosed accidently between 30 and 50 years of age with no significant gender differences found. The golden key in the diagnosis of this disease is clinical-radiological dissociation as patients have less severity of symptoms compared to radiologic finding. Symptomatic patients tend to have dyspnea, as well as a nonproductive cough, mainly on exertion. Blood chemistry tests usually produce results within the normal range. Chest radiographic findings in PAM show diffuse, sandstorm-like infiltration, particularly in lower bases. Chest radiographs may include black pleura sign seen as a strip of tangential peripheral lucency underlying the ribs as compared to the adjacent diffusely dense calcified lung. CT scan of the chest shows numerous sandstorm-like calcifications throughout the lungs with subpleural and peribronchial distribution. Bone scintigraphy using technetium-99m labeled diphosphonate compounds have affinity for calcification foci at soft tissue and can detect early pulmonary calcification [5]. Histopathology is not always required and it is done by open lung or transbronchial biopsy, showing intra-alveolar laminated calcium phosphate concretions. There is no treatment available for this disease. Thus, long-term oxygen therapy is required for hypoxemia and chronic respiratory failure. In our patient lung biopsy was not performed as she had typical findings on the bone scan.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2019.03.032.

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