Clinical Case Reports

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CLINICAL IMAGE

# Pneumothorax secondary to pulmonary alveolar microlithiasis

Oliver P. Devine[1] & Andrew C. Harborne[2]

[1]School of Medicine, University College London, Gower Street, London, WC1E 6BT, UK
[2]Department of Pathology, Hull Royal Infirmary, Anlaby Road, Kingston upon Hull, HU3 2JZ, UK

**Correspondence**
Oliver P. Devine, School of Medicine, University College London, Gower Street, London WC1E 6BT, UK. Tel: 0203 108 2158; E-mail: oliver.devine@gmail.com

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## Key Clinical Message

Pulmonary alveolar microlithiasis (PAM) is a rare impairment of pulmonary phosphate clearance that leads to gradual precipitation of intra-alveolar calcium phosphate microliths. There is often a striking difference between alarming clinical imaging and a relatively well patient. Pneumothorax in PAM often only respond to surgical intervention.

## Keywords

Chronic lung disease, interstitial lung disease, microliths, pneumothorax, pulmonary alveolar microlithiasis.

## Case

A 56-year-old Turkish man presented himself to the hospital with mild dyspnoea and a lifelong history of chronic lung disease. Despite an alarming bilateral whiteout with right apical pneumothorax, (Fig. 1A) he was comfortable at rest and appeared otherwise well. The pneumothorax remained refractory management with a chest drain. A computed tomographic scan revealed diffuse calcific micronodules throughout the lung parenchyma (Fig. 1B), consistent with a prior diagnosis of pulmonary alveolar microlithiasis. The patient underwent talc pleurodesis which resolved his pneumothorax. PAM is caused by mutation of the gene encoding the sodium-dependent phosphate cotransporter protein (SLC34A2) expressed by type II pneumocytes [1]. The transporter clears phosphate

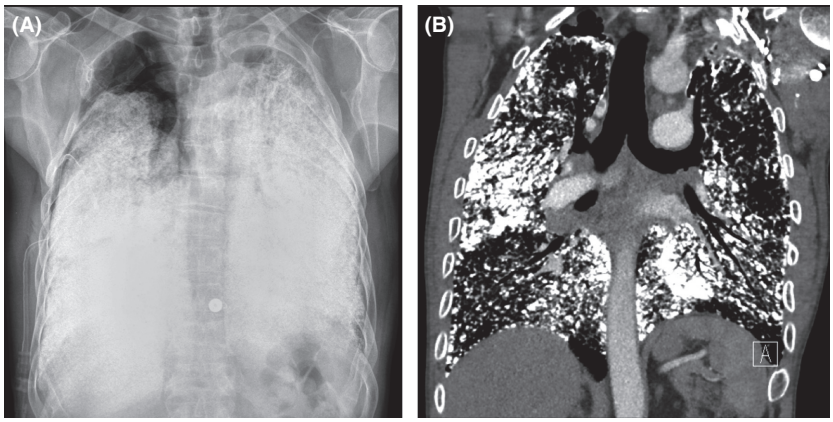


**Figure 1.** (A) Posterior–anterior plain chest radiograph showing bilateral whiteout of both lung fields and a right apical pneumothorax. (B) A coronal midthoracic computed tomography showing a multitude of small intra-alveolar microliths obstructing the lung parenchyma.

from phospholipoprotein-based pulmonary surfactant. Loss of function impairs phosphate clearance and precipitates calcium phosphate as intra-alveolar microliths. Approximately one thousand cases have been documented, one-quarter of which are in patients of Turkish ancestry [2]. The condition is notable for producing alarming radiographic findings in otherwise clinically well patients. Conservative management of pneumothorax in PAM patients typically fails. Here, we report an unusual presentation of a pneumothorax in a patient which – due to the stereotypic reduced parenchymal compliance of PAM – responded only to talc pleurodesis.

## Authorship

OPD: was responsible for researching and drafting the final manuscript. ACH: appraised the manuscript for critical content and provided clinical supervision and research oversight.

## Conflict of Interest

None declared.

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