Images in Thorax

Pulmonary alveolar microlithiasis

A 37-year-old patient was admitted to the hospital with severe dyspnoea and cough. Baseline investigations revealed haemoglobin of 16.2 g/dl, haematocrit of 48.8% and a total leucocyte count of 8800/mm³. Comprehensive metabolic profile and serum and urine protein electrophoresis findings were normal. Arterial blood gas analysis showed type 1 respiratory failure. Spirometry showed restrictive ventilatory disturbances (vital capacity 53%, forced expiratory volume in 1 s 91%). A posterior–anterior chest radiograph showed bilateral diffuse, nodular ('sandstorm-like') calcifications in both lungs (figure 1A). A high-resolution CT scan demonstrated multiple bilateral micronodulation of calcific densities throughout both lungs (figure 1B). On transbronchial biopsy, the diagnosis of massive parenchymal and alveolar calcification was confirmed. Echocardiography showed valve calcification with moderate mitral-aortic regurgitation and severe pulmonary hypertension. Abdominal ultrasonography revealed cholelithiasis and medullary nephrocalcinosis. The diagnosis of pulmonary alveolar microlithiasis (PAM) with concomitant valve calcification, cholelithiasis and idiopathic medullary nephrocalcinosis was established.

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Learning points

- PAM is a rare idiopathic slowly progressive disease; usually with familial association leading to alveolar deposition of calcium/phosphate microliths.¹ The underlying disturbances in calcium and phosphate homeostasis have not yet been identified. A discrepancy between the paucity of symptoms and the degree of pulmonary involvement is usually reported. PAM is found worldwide, but particularly predominates in Japan, India, Turkey and Italy.²
- PAM is prevalent among family units with a high rate of consanguinity among the parents of affected individuals. It has been recently found that mutations in the SLC34A2 gene (the type IIb sodium phosphate co-transporter gene) are associated with PAM.^{3 4}
- The characteristic picture of PAM on the chest radiograph shows the infiltrates as fine sand-like calcific micronodules ('sandstorm lung') diffusely involving both lungs, usually most marked in the middle and lower zones. CT reveals relatively symmetrical abnormalities in all patients and that calcifications were most prominent in the peripheral, mediastinal and fissural subpleural regions, and each lobe appeared surrounded by a fine dense outline, giving the overall appearance of a 'stony lung'. The use of high-resolution CT enables the identification of parenchymal abnormalities and provides additional information about the distribution of the disease at the level of the secondary pulmonary lobule.

REFERENCES

1. Hagiwara K, Johkoh T, Tachibana T. Pulmonary alveolar microlithiasis. In: Panos R, Trapnell B, McCormack F, eds. *Molecular Basis of Lung Disease, Insight from rare Lung Disorders*. New Jersey: Humana Press, 2010:325–38.
2. Castellana G, Lamorgese V. Pulmonary alveolar microlithiasis. World cases and review of the literature. *Respiration* 2003;**70**:549–55.
3. Huqun Izumi S, Miyazawa H, Ishii K, et al. Mutations in the SLC34A2 gene are associated with pulmonary alveolar microlithiasis. *Am J Respir Crit Care Med* 2007;**175**:263–8.
4. Corut A, Senyigit A, Ugur SA, et al. Mutations in SLC34A2 cause pulmonary alveolar microlithiasis and are possibly associated with testicular microlithiasis. *Am J Hum Genet* 2006;**79**:650–6.

Figure 1 Chest radiograph showing bilateral diffuse, nodular ('sandstorm-like') calcifications in both lungs (figure 1A). A high-resolution CT scan demonstrating multiple bilateral micronodulation of calcific densities throughout both lungs (figure 1B).