

## PULMONARY ALVEOLAR MICROLITHIASIS: HIGH-RESOLUTION CT SCAN

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Pulmonary alveolar microlithiasis (PAM) is a rare disease characterized by widespread calcific intra-alveolar concretions of both lungs.<sup>1</sup> The disease was first reported by Harbitz in 1918,<sup>2</sup> and since then over 200 cases have been reported worldwide in the medical literature. Although the cause of PAM is unknown, one possibility is that it is an inborn error of metabolism, and so may be regarded as an autosomal recessive hereditary lung disease.<sup>3</sup> The radiographic appearance of the disease is pathognomonic, and chest radiographs show apical bullae and diffuse micronodular shadow, which is sometimes described as sandstorm or snowstorm lung.<sup>3</sup>

### Case Report

A 60-year-old woman was admitted to our hospital in July 1998 for elective cholecystectomy, as she was suffering from acute calculus cholecystitis. As part of the routine preoperative investigations, chest radiograph was performed, which showed abnormally fine, sand-like micronodulations with calcific densities throughout both lung fields, predominantly in the lower zones (Figure 1). There was no evidence of apical bullae; the so-called black pleural line was not obvious, and heart borders and diaphragm were obscured. The appearance was highly suggestive of PAM.

Physical examination of the patient did not reveal any abnormal findings. Pulmonary function test was within normal limits. Transbronchial lung biopsy was performed, and it showed typical findings of pulmonary alveolar microlithiasis. High-resolution CT scan was performed on a Somatom DRH (Siemens, Erlangen, Germany) scanner. All images were obtained at maximal inspiration, using a 2-mm collimation, 125 KV, 28 mA, at 8 mm intervals in the supine position. Images were reconstructed by using bone algorithm and a 512x512 matrix. There were no apical bullae in this case. Small thin-walled subpleural cysts, ranging from 5 to 10 mm in diameter, were noted. The



FIGURE 1. Chest radiograph shows sandstorm lung. Heart borders and diaphragm are obscured; the microlithiasis is more marked in the middle and lower zones.

cysts were ribbonlike and arranged along the parietal pleura on both sides, and anteriorly from the apex to the costodiaphragmal sinus (Figure 2).

The pleura itself was thickened and irregularly delineated, containing punctate calcifications. Lung parenchyma showed high attenuation, increasing from the apex to the lung base. In the upper and middle lung zones, the high attenuation was distributed homogeneously throughout the lung parenchyma. In the lower lung zones, however, instead of diffuse involvement, higher attenuation was seen in the dorsal zones, especially in the prone position, causing us to rule out hypostasis as the reason for the increased density. In addition to high attenuation of the lung parenchyma, small (<5 mm) circumscribed intrapulmonary calcifications were visible, and irregularly arranged cysts smaller than 1 cm were seen throughout the lung parenchyma. The central bronchial system, the pulmonary vessels and the heart showed no major abnormalities.

### Discussion

Felson<sup>4-6</sup> first reported a linear radiolucency in the area of the lateral pleura on chest radiographs and called it a

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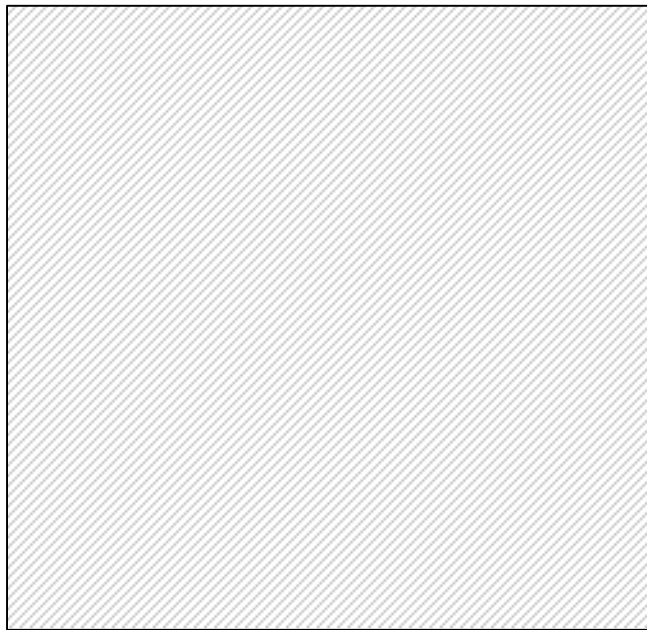


FIGURE 2. High-resolution CT scan (middle lung zone) shows subpleural cysts along the lateral and anterior aspects of the pleura. High attenuation of lung parenchyma is due to alveolar calcifications.

black pleural line. Later reports,<sup>7,8</sup> using high-resolution CT (HRCT) scan, suggested that this line was caused by subpleural cysts. Hoshino et al.<sup>1</sup> demonstrated that the black pleural line on the chest radiograph corresponded to the fat dense layer between the ribs and calcified parenchyma, and also reported for the first time MRI findings in this case as diffuse calcific micronodules, characterized by an increased signal intensity on T<sub>1</sub>-weighted images, predominantly in the posterior lower zones.

There are several reports about findings of PAM on conventional CT and HRCT.<sup>7-13</sup> To our knowledge, high-resolution CT findings have not been previously reported in the Arab region.

High-resolution CT shows that the so-called black pleural line on chest radiographs is caused by thin-walled subpleural cysts, ranging from 5 to 10 mm in diameter. The cysts are ribbon-like, and arranged along the parietal anterior and lateral pleura from the apex to the lung bases. These subpleural cysts can be shown on high-resolution CT scans and they correlate with the so-called black pleural line seen on chest radiographs. This zone of increased translucence in the location of the parietal pleura has been considered a visual illusion created by the contrast between extreme density of lung parenchyma on one side of the pleura and the ribs on the other side.<sup>4</sup> The high-resolution CT findings of small subpleural cysts not only explain the increased translucence seen on chest radiographs, but also the typical recurrent pneumothoraces that sometimes occur.

The advantages of high-resolution CT in detecting minimal structural changes in the lung parenchyma have been reported previously.<sup>12,13</sup> With thin-section and high

spatial frequency (bone) algorithms, structures approximately 1 mm in size can be detected which are not possible to detect with conventional CT techniques.<sup>11,12</sup> High-resolution CT scans show that there is a gradient of distribution of the calcifications in which lung bases, especially posteriad, are more involved than the middle and upper lung zones.<sup>10,11</sup> As well, high attenuation of lung parenchyma caused by calcispherites smaller than 1 mm (up to 5 mm) are visible on high-resolution CT scans.

Pracyk et al. proved that the composition of the calcifications is calcium and phosphate, in a ratio of 2:1.<sup>14</sup> Calcific micronodules and polygonal opacities are distributed throughout the lung parenchyma.<sup>1</sup>

This high-resolution CT scan technique can also reveal small cysts in the lung parenchyma, as well as pleural calcification<sup>10,11</sup> and small calcispherites within thickened pleura (irregular thickening of the pleura is due to fibrosis). In our patient, high-resolution CT scan also showed that both the bronchial system (including small bronchioles) and the size of pulmonary vessels were normal.

The differential diagnosis of this disease includes a variety of pulmonary conditions, such as tuberculosis, sarcoidosis, pneumoconiosis (desert lung syndrome), ornithosis or pulmonary adenomatosis.

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