Pulmonary Alveolar Microlithiasis with Pleural Calcification and Nephrolithiasis*

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A case of pulmonary alveolar microlithiasis (PAM) with pleural calcification and nephrolithiasis is reported. The significance of peripheral white lines, a rarely reported observation on chest roentgenogram in PAM, is discussed. Extrapulmonary calcification in multiple sites raises the possibility of PAM being a systemic disorder.

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Pulmonary alveolar microlithiasis (PAM), a rare disease of unknown etiology, is now beginning to emerge from obscurity. Its clinical profile, radiologic appearances, and pulmonary function abnormalities have been documented.1-3 Although the pathologic findings in this condition are said to be restricted to the lungs,4 cases of associated nephrolithiasis,5-8 milk-alkali syndrome,5 and pericardial calcification7 have been reported. In addition, a patient with calcification of lumbar sympathetic chain and probable testicular calcification has also been described.8 We present a unique case of a patient with PAM who had pleural calcification and nephrolithiasis.

Case Report

A 32-year-old woman with recurrent nonradiating colicky pain in the right loin area was found to have a right renal calculus for which surgery was advised. A chest x-ray film done as part of a preanaesthetic check was found to be abnormal for which she was referred to this Institute for evaluation. The patient did not have any respiratory symptoms. She had never worked outside her house, denied exposure to any kind of dust, and did not use snuff. There was no history suggestive of tuberculosis or any other chronic pleuropulmonary involvement. Results of physical examination were unremarkable. Hemogram was within normal limits. Chest roentgenogram showed bilateral reticulonodular opacities that were confluent and more dense at the bases. Dense, discrete miliary

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FIGURE 1. PA (penetrated) chest film shows bilateral reticulonodular opacities at both lung bases. Calcified pleura is visualized (arrowheads) as white lines along the costal surfaces, diaphragm, and pericardial reflection.

FIGURE 2A, B. Computed tomography of the thorax shows calcification of costal and mediastinal pleura (arrowheads).
opacities were also visualized. In addition, pencil-thin, sharp, dense white lines were seen bilaterally along the costal surfaces and over the hemidiaphragms. Similar lines could also be visualized along the left border of the heart and left paravertebral region (Fig 1).

Assessment of pulmonary function indicated a mildly restrictive ventilatory defect with normal Krog's constant and arterial blood gases. Serum urea nitrogen level was 28 mg/dl and serum creatinine level was 1.1 mg/dl. Intravenous pyelography demonstrated right ureteral dilatation with normally functioning kidneys. Levels of serum calcium, phosphorus, and alkaline phosphatase, and 24-hour urinary excretion of calcium and phosphorus (all investigations performed three times) were within normal limits. Antinuclear antibodies and rheumatoid factor were absent and lupus erythematosus (LE) cell test was negative.

Both children of the patient were found to be free of the disease. Siblings, however, were not available for examination.

A diagnosis of pulmonary alveolar microlithiasis was made and computed tomography (CT) of the thorax was done to investigate the line shadows, which revealed streaky as well as milary calcification in the lung parenchyma at both bases (Fig 2). There was extensive bilateral calcification of the diaphragmatic and parietal pleura, as well as its mediastinal and pericardial reflections. Both apices and the mediastinum were free of disease. A fine-needle aspiration biopsy was done under CT guidance. A gritty sensation was felt on rubbing the slides for preparation of the smear. Classic laminated microliths were observed on examination of the preparation stained with Giemsa stain. These microliths stained positive for calcium by the von Kossa's stain.

Subsequently, the patient was referred for surgery and the renal calculus was successfully retrieved; thereafter, she was unavailable for follow-up.

**DISCUSSION**

Standard textbooks do not make any mention of pleural involvement in PAM.\(^4\)\(^-\)\(^6\)\(^-\)\(^10\) Necropsy reports,\(^5\)\(^1\)\(^1\) too, have not revealed pleural calcification in this condition. In most reported cases, the disease-free pleura is highlighted as a black line around the lungs and under the ribs due to dense pulmonary opacities on one side and the ribs on the other.\(^1\)

However, review of literature disclosed that sharp, white lines along the periphery of the lungs, akin to those seen in our patient, have been observed earlier.\(^1\)\(^1\)\(^-\)\(^1\) The authors believed that the concentration of dense, continuous intra-alveolar microliths at the extreme periphery of the lungs may be responsible for these lines. Considering this explanation, these white lines should have been visible in most, if not all, reported cases of PAM. Computed tomography of the thorax, which documented pleural calcification in our patient, was not available then, however. Although Winzelberg et al.\(^1\) observed pleural calcification on CT, no mention was made of the white lines on chest roentgenograms. In retrospect, it appears that reports by Balkan et al.\(^1\)\(^1\) and Cole\(^1\)\(^1\) were also cases of PAM with pleural calcification. It is likely that peripheral, sharp white lines, when seen on chest roentgenogram in PAM, indicate associated pleural calcification.

Although no consistent abnormality of calcium metabolism has been recognized in this condition, cases with ectopic,\(^1\)\(^2\)\(^-\)\(^8\) bronchiolar, and interstitial\(^9\)\(^-\)\(^1\)\(^5\) calcification as well as hypercalcemia\(^1\)\(^6\)\(^-\)\(^1\)\(^7\) have been reported. Nephrolithiasis has been reported three times before in PAM.\(^1\)\(^5\)\(^-\)\(^6\) These reports, together with our case, suggest the possibility of multiorgan involvement in PAM. These findings do not support any theory of local precipitation of calcium in alveolar spaces.\(^4\)\(^-\)\(^1\) It is conceivable that PAM may be a systemic disorder with intermittent hypercalcemia during disease activity. Similar hypercalcemia has also been observed in sarcoidosis.\(^1\)

**REFERENCES**

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