Absence of pulmonary uptake of Tc-99m methylenediphosphonate in alveolar microlithiasis

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Pulmonary alveolar microlithiasis (PAM) is a rare disease of unknown etiology characterized by accumulation of calcific concretions in the alveolar spaces. The paper reports a case of PAM in a 56-year-old man. The patient had persistent dry cough, and gradually progressive dyspnea on exertion. The diagnosis was established on the basis of roentgenography and confirmed by the sputum and transbronchial biopsy findings. Scintigraphy revealed the absence of Tc-99m methylenediphosphonate uptake of lungs. Familial occurrence was not observed. Chest roentgenogram, pulmonary function, and clinical status of the patient have remained stable for 41 months. Radiological and clinical follow-up of the disease continues.

Key words: pulmonary alveolar microlithiasis, Tc-99m MDP bone scintigraphy, lung

INTRODUCTION

PULMONARY ALVEOLAR MICROLITHIASIS is a rare disease of undetermined cause characterized by the presence of microliths within the alveoli of the lungs. Although the disease shows no clear geographic distribution,2 most of the reported literatures were from Turkey.1 The disease affects both sexes equally, afflicts patients primarily between 4th and 6th decades.2 The diagnosis is usually made on the basis of the typical radiological pattern, namely a very fine, sand-like micronodulation of calcific density diffusely involving both lungs with basal predominance.3 After the disease is diagnosed in a given patient, other family members should be screened by chest roentgenography, and parents should be counseled that future children are also at risk of developing the disease.4 Although some patients present with progressive respiratory symptoms, the discovery is in most cases an incidental finding on a chest radiograph. Tc-99m diphosphonate scanning usually reveals diffuse intense uptake throughout both lungs.5

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normal limits. The chest radiogram revealed diffuse high-density sand-like micronodules and reticular lines throughout the lung fields (Fig. 1).

High-resolution CT of the chest (Fig. 2) showed a unique and characteristic calcified reticular pattern and thickening of the interlobular septa of the lung parenchyma, with predominant basal and peripheral lung distribution. There was no hilar or mediastinal lymphadenopathy.

Whole body bone scintigraphy was performed 3 hours following i.v. injection of 740 MBq (20 mCi) of technetium-99m methylene diphosphonate using a gamma camera with a low energy, high resolution collimator (Toshiba GCA-901/SA). The photopake was centered at a 20% window in the camera. In obtained images, the radiotracer was distributed normally in systemic bones, and symmetric uptakes with increased activities were seen in joints and junctions. Also bone scintigraphy showed the absence of Tc-99m methylenediphosphonate uptake of lungs (Fig. 3).

Histopathological examination of the transbronchial lung biopsy specimen showed the characteristic calcispherites in the alveolar space but not in the interstitium. In microscopic examination some of the alveoli contained laminated eosinophilic calcispherites with Von-Kossa (Fig. 4).

His three brothers, two sisters, two sons, two daughters, and four grandchildren underwent chest radiography. There were no abnormalities suggesting alveolar microlithiasis. The last follow-up radiographs and pulmonary function studies were available on 15 March 2004 to document the clinical progression of the disease. His clinical status has remained stable for 41 months. No differences were seen on chest radiographs, lung function tests or arterial blood gas measurements. There was no progression in symptom severity. Vital signs and physical examination were also normal. Follow-up regarding the radiological and clinical course of the disease continues.

**DISCUSSION**

Pulmonary alveolar microlithiasis is a rare disease entity of unknown etiology that affects individuals of all ages, from the very young to elderly. A review in 2002 described 300 cases of PAM reported in the literature. To our knowledge, only two PAM cases with absence of