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A Speckled Chest Radiograph

To the Editor:

We describe a patient who was diagnosed as having pulmonary alveolar microlithiasis. We report the case and discuss this condition.

CASE PRESENTATION

A 45-year-old woman presented with history of mild exertional dyspnea of 10 years duration associated with cough and chest pain unrelated to exertion. She did not give a history of wheezing or ankle swelling, and there was no diurnal variation in her symptoms. She was a current cigarette smoker with a 15-pack-year history of smoking. The general physical and systemic examination was unremarkable. Pulse oximetry saturations were normal. An electrocardiogram, spirometry, full blood count, renal function, and bone profile were within normal limits. A plain chest radiograph (Figure 1A) showed multiple small nodules scattered across both lung fields. A contrast-enhanced computed tomography scan demonstrated these nodules to represent small calcific foci (Figure 1B). No fibrotic or inflammatory changes were seen within the lung parenchyma. The diagnoses considered included previous varicella infection, inhalational pneumopathy, and pulmonary alveolar microlithiasis. She did not remember having chicken pox, and her occupational history was not contributory.

A fiberoptic bronchoscopic examination subsequently performed was morphologically normal. A transbronchial lung biopsy showed characteristic calcified concentric lamellae in the form of "microliths" within the alveolar space (Figure 1C), diagnostic of pulmonary alveolar microlithiasis. Microliths consist of concentric lamellas that surround a central nucleus (Figure 1D) with a diameter between 0.25 and 1.0 mm. They contain dense calcium and phosphate mixed with a small amount of magnesium and aluminum. The microliths are Periodic acid-Schiff-positive on staining.

DISCUSSION

Pulmonary alveolar microlithiasis is a rare idiopathic condition with a little over 500 cases reported worldwide.[1,2] This condition was first characterized in 1918 by Harbitz,[3] who described at postmortem the presence of the minute calcific deposits in the lung. Puhr, in 1933,[4] described the condition as "Microlithiasis Alveolaris Pulmonum," from which the current nomenclature is derived. Usually, pulmonary alveolar microlithiasis is diagnosed in patients under the age of 40 years; however, it has been reported in premature twins and patients in their 8th decade as well. In sporadic cases a male predominance, and in familial clustering a female predominance has been reported. Around a third of the cases have a family history of the disease. Symptoms are noted in less than half and include cough, dyspnea, and chest pain. Often the diagnosis is suspected due to an incidental abnormality detected on the chest radiograph, the characteristic picture being that of diffuse, bilateral fine sand-like calcific nodules that may appear coalesced. The typical florid manifestation also is referred to as a "sandstorm" appearance; however, appearances can be subtle, as in our case. In most cases, the condition takes a protracted and relatively indolent course. In a relatively few cases, this condition might have an aggressive course with progressive pulmonary fibrosis ultimately leading to respiratory failure. A recent study has suggested a possible genetic association with this condition.[5]

The chest radiograph and computed tomography findings are often characteristic enough to suggest the diagnosis, which can be confirmed using a combination of bronchial lavage, transbronchial biopsy, or formal lung biopsy. Occasionally, microliths may even be demonstrated in sputum samples. Treatment options for pulmonary alveolar microlithiasis are limited. Systemic corticosteroids, diphosphonates, and large-volume bronchoalveolar lavage have been used; however, these have not been validated across a series. Lung transplantation remains the only treatment option if the disease is progressive.

Our patient underwent large-volume bronchoalveolar lavage with no benefit. Her symptoms remain indolent

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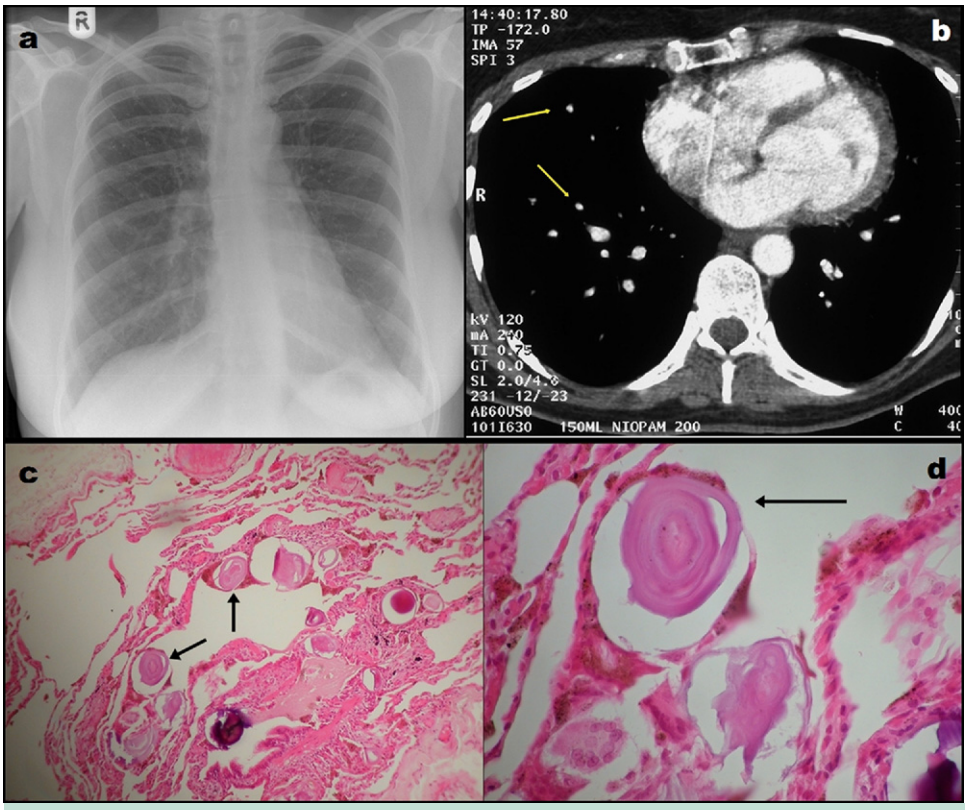

Figure (**a**) Very small calcific nodules are seen on the plain chest radiograph, prominent in both the upper zones. These nodules do not appear coalescent. Hilar, mediastinal, and bony structures are normal. (**b**) Chest computed tomography on mediastinal windows showing multiple (arrows) dense white small calcific nodules with have the same density as the thoracic spine. This is a contrast-enhanced computed tomography scan and hence the pulmonary vessels also have a similar density, but are larger. (**c**) Hematoxylin and eosin photomicrograph of the transbronchial lung biopsy. Calcified "microliths" are seen in the alveolar space (arrows). (**d**) A "microlith" in more detail (arrow) demonstrating the characteristic concentric lamellar appearance.

and static on follow-up, and the chest radiograph is unchanged.

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