Asymptomatic clinical course of pulmonary alveolar microlithiasis during a 12-year follow-up

Pulmoner alveoler mikrolitiazisin 12 yıllık takip süresince semptomsuz klinik seyri

Akif Turna, Ali Kılıçgün, Muzaffer Metin, Adnan Sayar, Atilla Gürses

Department of 1st Thoracic Surgery, Yedikule Teaching Hospital for Chest Diseases and Thoracic Surgery, İstanbul

Pulmonary alveolar microlithiasis is a rare disease of unknown cause, characterized by the presence of microliths within the alveoli of the lungs. An eleven-year-old girl presented with growth retardation. She had a history of myoplasty operation for the sternocleidomastoid muscle for torticollis. Findings of physical examination and laboratory investigations were normal. Chest radiography showed multiple nodular infiltrates and open lung biopsy confirmed the diagnosis of pulmonary alveolar microlithiasis. Treatment consisted of only observation. During a 12-year follow-up, her clinical status and pulmonary function tests remained unchanged. Her brother also had bilateral alveolar microlithiasis that remained asymptomatic. Pulmonary alveolar microlithiasis is a slowly progressing pulmonary disease with familial occurrence. No specific treatment is available and the need for treatment is yet to be justified.

Key words: Calculi/diagnosis/pathology; lung diseases/diagnosis/radiography; respiratory function tests.

Pulmoner alveolar mikrolitiazis, akciğerlerde alveoller içinde mikro taşların varlığı ile karakterize nadir bir hastalıktır. Nedeni henüz bilinmemektedir. On bir yaşında bir kız büyüme-gelişme geriliği yakınmasıyla başvurdu. Öyküsünde, tortikolis nedeniyle sternokleidomastoid kasa yapılan miyoplasti ameliyatı vardı. Fizik muayene ve laboratuvar incelemelerinde anormallik saptanmadı. Düz göğüs radyografisinde akciğerde çok sayıda nodüler infiltrasyon görüldü; açık akciğer biyopsisi sonucu pulmoner alveolar mikrolitiazis ile uyumlu bulundu. Sadece gözlemi içeren bir takip planlandı. On iki yıllık izleme sırasında hastanın genel durumu ve solunum fonksiyon testlerinde değişiklik olmadı. Hastanın erkek kardeşinde de semptomsuz seyreden iki taraflı alveolar mikrolitiazis saptandı. Pulmoner alveolar mikrolitiazis, ailesel bağlantısı olabilen yavaş ilerleyen bir akciğer hastalığıdır. Hastalığın spesifik bir tedavisi bulunmamaktadır ve tedavi gereksinimi olup olmadığı da tam kesinleşmemiştir.

Anahtar sözcükler: Kalkulus/tanı/patoloji; akciğer hastalığı/tanı/radyografi; solunum fonksiyonu testi.

Pulmonary alveolar microlithiasis is a rare disease of unknown cause characterized by the presence of microliths (calcispherites) within the alveoli of the lungs. Typically plain chest radiographs show a white lung or sandstorm appearance consisting of fine sand-like microcalcifications diffusely scattered throughout the lungs, with higher density at the base.[1] The heart borders and the diaphragm are usually obliterated.

The disease is so uncommon that a review in 1983 included approximately 120 published cases.[1] So far, no definitive treatment has been defined.[1-3]

CASE REPORT

An eleven-year-old girl was brought to our clinic for growth retardation. Physical examination and laboratory investigation showed no abnormality. A chest roentgenogram showed multiple small nodular infiltrates in both lungs (Fig. 1a). She had undergone a myoplasty operation involving the sternocleidomastoid muscle for torticollis which developed due to trauma during labor. A biopsy of the scalene lymph nodes yielded no pathological abnormality. Pulmonary function tests could not be performed because of lack of cooperation on the part of the patient. For a definite diagnosis, an open lung biopsy was performed via a mini-thoracotomy through the fifth intercostal space, which yielded a pathological diagnosis of microlithiasis of the lung. The postoperative course was uneventful and she was discharged on the eight postoperative day.

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Correspondence: Dr. Akif Turna. Yedikule Göğüs Hastalıkları ve Göğüs Cerrahisi Eğitim ve Araştırma Hastanesi, 1. Göğüs Cerrahisi Kliniği, 34020 Zeytinburnu, İstanbul. Tel: 0212 - 664 17 00 / 1318 e-mail: aturna@turk.net

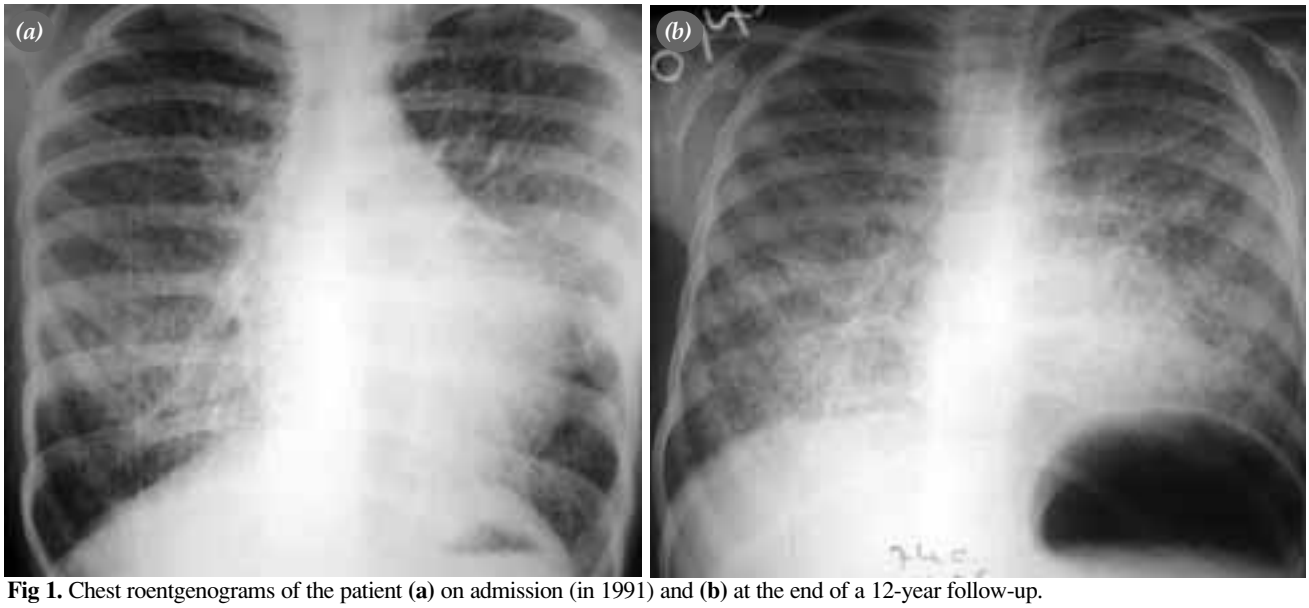

Fig 1. Chest roentgenograms of the patient **(a)** on admission (in 1991) and **(b)** at the end of a 12-year follow-up.

During a follow-up of 12 years, radiologic and clinical findings did not change. Spirometry performed at the last follow-up showed FEV_1 as 2.71 (89% of predicted) and FEV_1/FVC as 90.2% (103% of predicted). The roentgenologic appearance of the chest remained unchanged (Fig. 1b). Computed tomography of the chest showed bilateral calcific densities. She did not have pulmonary complaints and pulmonary function tests were in normal limits. A chest radiograph of her brother showed typical appearance of bilateral alveolar microlithiasis. He also did not have any pulmonary symptoms or signs.

DISCUSSION

Alveolar microlithiasis is a rare disorder that usually presents as an abnormal chest radiograph in an asymptomatic patient. The chest radiograph is diagnostic, showing a sandlike micronodulation throughout the lung fields.[1] This is caused by the presence of innumerable minute calcified spherules filling the alveolar space.

Our patient had typical radiologic features and no pulmonary symptoms. During a 12-year follow-up, no clinical and radiologic progression was shown. Alveolar microlithiasis exhibits familial occurrence.[1] The roentgenogram of brother of our patient also showed bilateral sandlike nodulation with no pulmonary symptoms. Despite an apparent roentgenologic abnormality, there is little evidence indicating a need for therapeutic intervention. Pathologic studies have shown that, in most cases, the alveolar structures are remarkably well preserved. Some evidence suggests, however, that microliths form in the alveolar walls and subsequently extrude into the alveolar spaces.[1,2] Long-term follow-up of patients with alveolar microlithiasis show that aging process per se is associated with a restrictive type of deterioration rather than diminution in lung function,[1] allowing most of the patients to remain symptom-free.

Caffrey and Altman[2] identified alveolar microlithiasis in premature twins, and suggested that it might have originated in utero resulting from an enzyme defect. It was also postulated that the disease was due to an inborn error of the respiratory metabolism at the alveolar interface, but thus far this theory has not been substantiated.[3]

No known therapy exists for pulmonary alveolar microlithiasis. The disease is reported to be unresponsive to corticosteroids and chelating agents. Treatment has been purely supportive and alleviative in a subset of patients. Bronchopulmonary lavage has been tried, but has had no effect on the course of the disease.[4] Moreover, the need for treatment has not been justified since the disease remains unchanged in a majority of patients. Similarly, our patient, together with her brother, remained symptom-free for 12 years without the need for any treatment.

In conclusion, alveolar microlithiasis is a pulmonary disease with no known therapy and cause. In some patients, the disease does not show progression in long-term follow-up. It may be speculated that no treatment is necessary in patients showing no development. Further studies with larger series are needed to justify the need for treatment.

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