CASE REPORT

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First Presentation of a Case of Pulmonary Alveolar Microlithiasis with Spontaneous Pneumothorax

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ABSTRACT

Pulmonary Alveolar Microlithiasis (PAM) is a rare disease of unknown origin. It is characterized by the presence of small calculi within the alveolar spaces, and has various clinical manifestations. Some patients may be asymptomatic for a long period of time with subsequent occurrence of dyspnea, dry cough, chest pain, and eventually, respiratory failure. Recurrent spontaneous pneumothorax episodes are a late complication of the disease. We report the case of an alveolar microlithiasis episode in a 42-year-old male, admitted to Tohid Hospital, Iran, whose first clinical presentation was symptoms of pneumothorax. He was admitted with sudden onset dyspnea and right-sided pleuretic chest pain. Following treatment of pneumothorax after chest tube placement, the pulmonary function revealed normal indices, and chest radiograph demonstrated diffuse confluence of dense micronodular infiltrate. High-resolution computerized tomography scan showed diffuse ground glass attenuation and calcifications along the interlobular septa and subpleural regions. Transbronchial lung biopsy confirmed the diagnosis of PAM.

Pulmonary Alveolar Microlithiasis (PAM) is a rare disease of unknown etiology, characterized by the formation and accumulation of laminated calcispherites in the alveoli.[1-3] However, its exact pathogenesis is still unknown. The clinical features may vary and patients may be asymptomatic for a long period of time with subsequent occurrence of dyspnea, dry cough, and chest pain, leading to cor pulmonale.[4,5] Spontaneous pneumothorax may occur years after the diagnosis.[6]

In this article, the authors present a case of PAM in a 43-year-old man with spontaneous pneumothorax without previous respiratory symptoms.

CASE REPORT

A 43-year-old male, non-smoker, presented to Tohid Hospital, Sanandaj, Iran, with sudden onset shortness of breath and right-sided chest pain, which was sharp and aggravated by deep inspiration. The symptoms started two days after a common cold. The patient was entirely asymptomatic and denied dyspnea, cough, hemoptysis, and weight loss before recent presentation. The patient had no family history of pulmonary disease, took no medications, and had regular daily exercises without difficulty.

On physical examination, the patient was anxious and tachypneic. Pulmonary auscultation revealed reduced breath sounds in the upper region of the right hemithorax and inspiratory crackles in lower third of both lung fields. Cardiac auscultation revealed normal heart sounds and the patient had no cyanosis, or observable peripheral edema.

Blood tests showed normal values for complete blood counts and serum chemistries and the patient's arterial oxygen saturation, without the use of supplemental oxygen, was 81%.

Plain chest films showed right-sided pneumothorax and extensive bilateral dense opacities most marked in the middle and lower zones [Figure 1]. In addition, computerized tomography (CT) examination on the admission day revealed right-sided pneumothorax and high attenuation of lung parenchyma.

After insertion of a chest tube in the right side of the patient's chest, on the second day of admission, the dyspnea subsided and repeated chest radiography revealed almost complete re-expansion of the right lung. Following chest tube removal, the patient was discharged on day four. Two weeks later, high resolution CT (HRCT) of lungs and pulmonary

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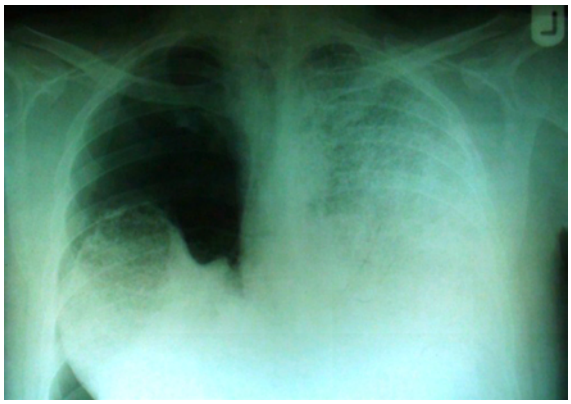

Figure 1: The chest plain film taken on admission revealed right side pneumothorax, extensive bilateral dense opacities obliterated the cardiac and mediastinal borders.

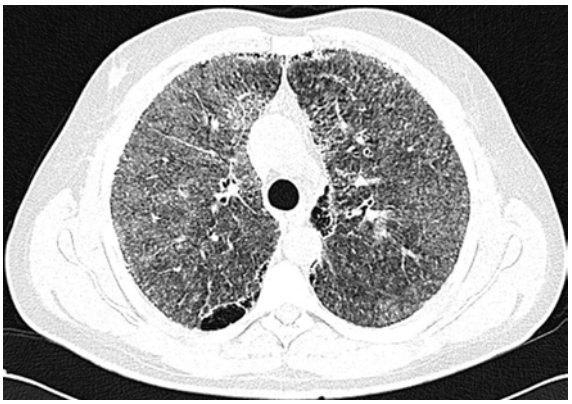

Figure 2: High-resolution computerized tomography of lungs reveals ground glass attenuation, septal thickening, and sub-pleural cysts.

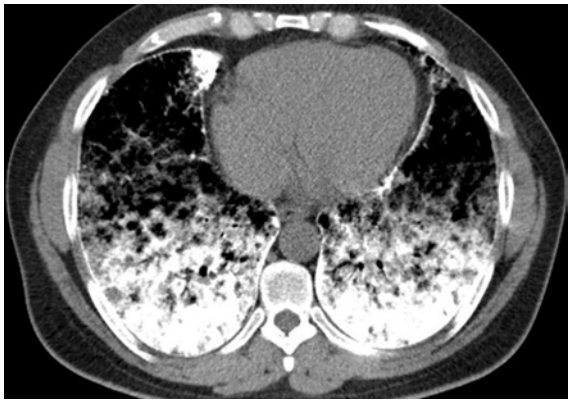

Figure 3: Chest computerized tomography scan of lower zone shows greater involvement: dens calcification of interlobular septa, sub-pleural zone, and pericardial layer.

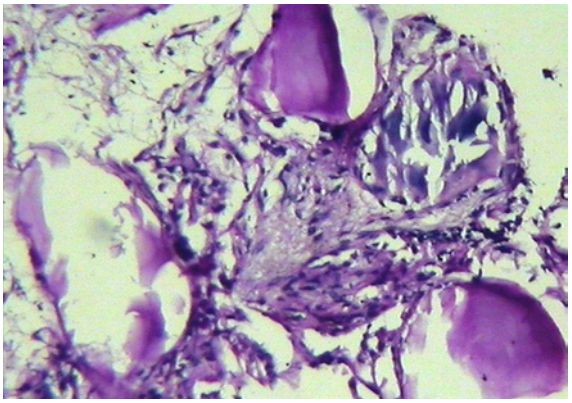

Figure 4: Histopathologic examination of lung shows interstitial infiltration by inflammatory cells, areas of fibrosis and laminated calcispherites within the alveoli.

function tests were performed. HRCT of the lungs showed high attenuation, increasing from the apex to the lung base, ground glass opacities, and sub-pleural cysts [Figure 2]. Calcification along the interlobular septa and sub-pleural calcifications were also noticed [Figure 3]. Spirometry results were normal with forced vital capacity (FVC) and forced expiratory volume in 1 second (FEV1) more than 80% of the predicted values.

The patient underwent transbronchial lung biopsy during the course of diagnostic fiber-optic bronchoscopy. Histopathologic examination revealed interstitial inflammatory cell infiltration in conjunction with areas of fibrosis. Multiple laminated calcispherites were found in the alveoli [Figure 4]. These findings were suggestive of PAM.

DISCUSSION

PAM was first described by Malpighi in 1686, who provideda concise and precise macroscopic description of the disease.[7] In 1918, Harbitz provided an accurate autopsy of a second case but it was Puhr in 1933 who named the disease mikrolithiasis alveolaris pulmonum.[8,9]

A review of 576 published cases showed that there was no gender predominance and the ages of affected patients ranged from newborn to those in their ninth decade of life .[10]

The most accepted etiology for PAM is that it is an inherited metabolic abnormality, involving the enzyme carbonic anhydrase, which promotes alkalinity of the alveolar surface and consequent precipitation of calcareous salt.[11] Furthermore, familial incidence is

common, particularly in siblings. A family history for the disease was found in one-third of patients.[1] Inactivating mutations in the Scl-34A2 gene have been reported in familial cases of pulmonary alveolar microlithiasis. The Scl-34A2 gene encodes a type IIb sodium phosphate cotransporter that is expressed in type II alveolar cells.[12]

There is a striking discordance between the extent of radiologic involvement and the severity of clinical presentation in PAM patients. Patients may remain asymptomatic for many years, and the condition may progress slowly leading to progressive dyspnea and ultimately result in cor pulmonale. Other reported symptoms are cough, fatigue, chest pain, hemoptysis, palpitations, and headache.[11-13] Recurrent pneumothoraces may occur over the course of the disease. Shishido et al[14] reported a case of PAM in a patient who developed pneumothorax 34 years after the diagnosis was established. Timothy et al,[15] described the surgical management of recurrent pneumothoraces in a case of PAM, decades after initial diagnosis. It is believed that progression of emphysematous bullae resulting in spontaneous pneumothorax occurs in the chronic phase of PAM but pneumothorax symptoms were the first presentation of the disease in our patient, which has not been reported in previous studies.

Although pulmonary function tests may initially yield normal results, restrictive pattern is a common finding during the course of the disease.[7] Our case, however, showed a normal pattern of pulmonary function studies after re-expansion of the lung.

The characteristic feature of PAM on the chest radiograph revealed a picture of infiltrates as fine sand-like calcific micronodules ("sandstorm lung") diffusely in both lungs.[16] This feature resembled the radiographic finding of calcified miliary tuberculosis,[17] small thin-walled subpleural cysts are described. In addition, which are responsible for "black pleura" sign seen in chest X-rays.[3,5,6]

As a diagnostic method, HRCT of the chest has greater sensitivity than a chest X-ray. HRCT findings in patients with PAM vary considerably, nevertheless, the most common findings are ground glass opacities and diffuse calcified nodules.[5,6,10,16] Others include linear calcifications, calcification of interlobular septa, mosaic pattern of attenuation, small calcipherites within the thickened pleura, crazy paving pattern, and calcifications along the heart borders.[3-7] The latter was seen in our patient. In addition, there were visible apical cysts on both sides. These cysts were ribbon like and arranged along the mediastinal and parietal pleura, and could be the source of spontaneous pneumothorax in our case. Whole body ^{99m}Techenetium methylen diphosphate scintigraphy may show diffusely increased radiotracer uptake over the lungs.[18]

There is no known medical treatment to reduce or halt the progression of PAM. Lung transplantation remains the only possible treatment for end-stage cases.[4,19] Recurrent pneumothorax in PAM should be managed with thoracotomy and excision of affected areas of lung, and subsequent lung defects should be oversewn and buttressed.[15]

CONCLUSION

In conclusion, PAM is a rare, chronic lung disease characterized by progressive clinical course. Spontaneous recurrent pneumothorax may occur in the later phase of the disease but it should be highlighted that the first presentation of a PAM case could be the occurrence of pneumothorax.

Disclosure

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Statistics

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