http://dx.doi.org/10.15218/zjms.2015.0005

Pulmonary alveolar microlithiasis: A case report

Received: 17/3/2014 Accepted: 28/5/2014

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Abstract

Background and objective: Pulmonary alveolar microlithiasis is rare disease characterized by the presence of numerous small calculi within the alveolar spaces.
Case report: 39-year-old female with pulmonary alveolar microlithiasis is described, in which the diagnosis was suspected clinically and radiologically and confirmed histopathologically. The clinical, pathogenesis and pathology are briefly reviewed.
Keywords: Diffuse lung disease, Microlithiasis.

Introduction

Pulmonary alveolar microlithiasis is uncommon chronic lung disease characterized by the presence of calcific concretions within the alveolar spaces.[1] Pulmonary alveolar microlithiasis was first described by Friedrich[2] in 1856 and then by Harbitz[3] in 1918. O'Neill reported a family with pulmonary alveolar microlithiasis in 1967 and noted that the total number of recorded cases at that time was seventy.[4] It occurs sporadically and the symptoms of lung disorder with restrictive pattern start in most cases in the third and fourth decades of life.[5,6] Pulmonary alveolar microlithiasis is regarded as an autosomal recessive lung disease with apparently no important gender predominance.[5,7] As of 2004, 576 cases have been studied, and most have originated from Europe (42.7%) and Asia (40.6%).[7] The major finding on chest radiograph is sand like diffuse symmetric micronodular calcification which is predominant in the middle and lower lobes with sparing of apices.[5,8] We report this case because it is very rare. We aim to emphasize a high index of suspicion for this uncommon disorder to avoid misdiagnosis of miliary tuberculosis and sarcoidosis and as far as we are aware this is the first case to be reported in Erbil, Kurdistan, Iraq.

Case presentation:

39-year-old housewife, presented with progressive shortness of breath and cough for the last 10 years. She had no history of fever or weight loss, nor history of dust inhalation or smoking. No history of similar condition in the family was noted. For the last two years, the case has been treated elsewhere as miliary tuberculosis based on chest x-ray findings, but with no clinical or radiological response. Physical examination revealed crackles at auscultation of the chest. Cardiac auscultation was normal and no cyanosis or peripheral edema was observed. Sputum examination for acid-fast bacilli was not performed and the routine laboratory findings were unremarkable including serum calcium and phosphorus. Pulmonary function test showed mild restrictive ventilator defect (mildly increased FEV1/FVC ratio). Arterial blood gas analysis showed reduction in O_2-saturation of 85%, and echocardiography showed no significant abnormal findings. Serial chest x-ray demonstrated diffuse bilateral dense micro nodular infiltrate predominantly at mid and lower zones with obliteration of cardiac and diaphragmatic borders (Figure 1). High-resolution computed tomography revealed diffuse minute pulmonary calcification, distributed predominantly over basal and posterior

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regions of the lung fields bilaterally as well as along parietal and mediastinal pleura with small apical bullae, black pleural line and thickened interlobar septa (Figure 2). Lung biopsy was performed and sent to histopathological analysis (Figure 3). On Pathological study, gross examination showed the specimen consisting of a piece of lung tissue measuring 2.5 cm in maximum diameter, brown in color, hard and gritty, while microscopic examination showed numerous rounded concentrically laminated calcific bodies within the alveolar spaces. The alveolar septa were irregularly slightly thickened. Decalcified section was made and showed numerous intraalveolar basophilic bodies (Figures 4-6).

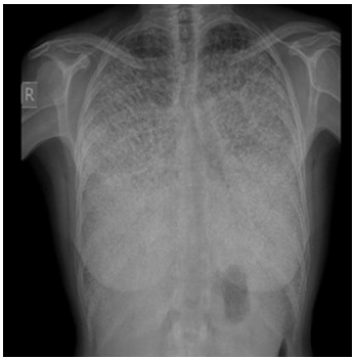

Figure 1: Chest X-Ray showing sandstorm lung, obscuring diaphragmatic and cardiac border, with predominant middle and lower zones distribution.

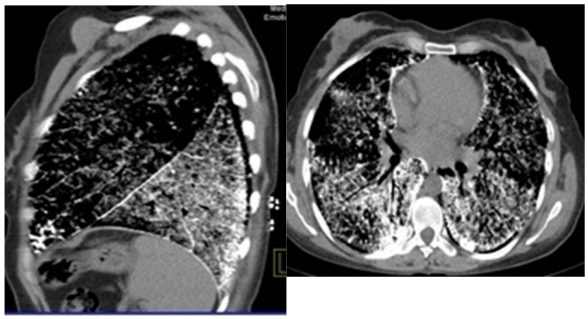

Figure 2: HRCT showing miliary calcification distributed all over both lungs with predominant basal and posterior distribution as well as along parietal and mediastinal pleura with black pleural line.

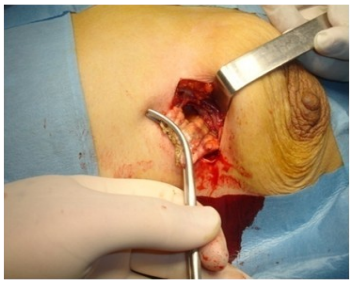

Figure 3: Open Lung biopsy; the lung tissue is studded with hard small nodules.

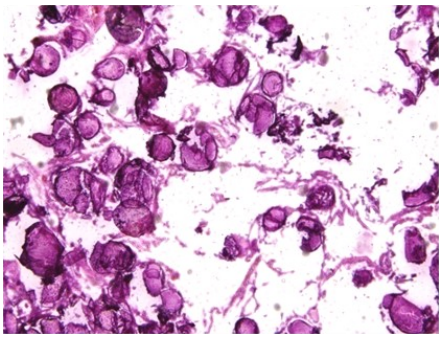

Figure 4: Pulmonary alveolar microlithiasis showing numerous rounded, laminated calcific bodies within the alveolar spaces.

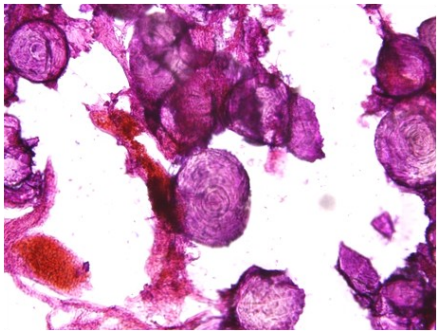

Figure 5: Pulmonary alveolar microlithiasis: High magnification

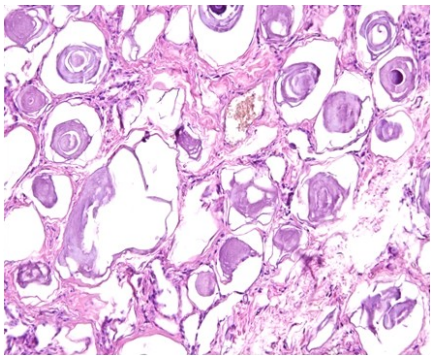

Figure 6: Pulmonary alveolar microlithiasis: Decalcified section showing numerous intraalveolar basophilic laminated rounded bodies.

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Discussion

Pulmonary alveolar microlithiasis is a rare disease characterized by progressive deposition of calcium within the alveolar spaces without alteration of calcium and phosphate metabolism.[1] Patients with pulmonary alveolar microlithiasis may remain asymptomatic for many years and present with restrictive lung disorder between the third and fourth decades.[8,9] The mean age at the time of diagnosis is 35 year[5] and the age of our patient is 39 year. Regarding the sex there is no significant predominance of gender[5,7] and our patient is female. Pulmonary alveolar microlithiasis has high incidence of familial occurrence suggesting an auto-somasl recessive inheritance pattern.[5,6] A few reports have described the role of mutation in the type II b sodium-phosphate co transporter gene (SCL34 A2 gene) which is involved in phosphate homeostasis in the lungs.[10,11] SCLC34 A2 expression is observed in type II alveolar cells. These cells use phospholipids to produce surfactant and are also responsible for recycling and degrading the out dated surfactant. It is believed that the dysfunction in SCLC34 A2 may reduce the clearance of the phosphate released in this process leading to formation of microliths.[11] The chest radiograph of our patient showed a diffuse symmetric lung lesion with dense micronodules which is consistent with many other studies.[5,8] Mosaic pattern on HRCT scan in which the interlobular septa are of calcium density due to the deposition of microliths within the peripheral lobular parenchyma adjacent to the septa may be considered pathognomonic of pulmonary alveolar microlithiasis.[5] Clinicians should be aware that some findings in pulmonary alveolar microlithiasis such as nodular calcification can be found in other diseases such as tuberculosis, silicosis, metastatic pulmonary calcification, sarcoidosis and amiodarone lung toxicity. In this way CT finding and clinical features should always be correlated[12] and biopsy will confirm the diagnosis. The patients show progressive deterioration of the lung function and death usually occurs in mid-life because of respiratory failure associated with cor pulmonale.[8,13,14]

Conclusion

Pulmonary alveolar microlithiasis is a rare chronic progressive lung disease that can affect young patients. Clinicians and radiologists should be aware of its existence and need to be considered in the differential diagnosis of diffuse lung lesions on chest radiography and high resolution computed tomography can reveal characteristic pattern and biopsy will confirm the diagnosis.

Conflicts of interest

The author reports no conflicts of interest.

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