

# Familial Pulmonary Alveolar Microlithiasis: A Rare Case Report with Emphasis on Imaging Findings

Dr. Qays Ahmed Hassan AL-Timimy M.B.CH.B., D.M.R.D., C.A.B.M.S.(RAD). Radiologist specialist, Instructor/ Al-Kindy College of medicine - University of Baghdad,

Dr. Abdullateef Aliasghar M.B.CH.B., C.A.B.M.S.(RAD) Radiologist specialist, Instructor/ Iraqi national cancer research center - university of Baghdad

Dr Muthanna D.R. Alassal, M.B.CH.B., F.I.C.M.S. Consultant thoracic and vascular surgeon, Instructor / Al-kindy college of medicine - university of Baghdad

**ABSTRACT** Pulmonary alveolar microlithiasis is rare infiltrative pulmonary disease characterized by intra-alveoli deposition of microliths. We present a familial case of an adult female with complaint of progressive shortness of breath on exertion. Chest radiograph showed innumerable tiny dense nodules, diffusely involving both lungs mainly the lower zones. High-resolution CT scan illustrated widespread intra-alveolar microliths, diffuse ground-glass attenuation areas and septal thickening predominantly in the basal regions. Chest radiograph is all that is needed for the diagnosis

of this case but CT scan was done to demonstrate the extent and severity of this disease.

**Key Words:** Pulmonary alveolar microlithiasis, Calcification, Computed tomography  
*Al-Kindy College Medical Journal Vol. 11 No.2. Page: 108-110*

*\*Received at 13<sup>th</sup> Dec. 2015, accepted in final 15<sup>th</sup> Dec. 2015.*

*Corresponding author to Dr. Dr. Qays Ahmed Hassan AL-Timimy, M.B.CH.B., D.M.R.D., C.A.B.M.S.(RAD). Radiologist specialist, Instructor/ Al-Kindy College of medicine - University of Baghdad,*

**P**ulmonary alveolar microlithiasis (PAM) is a rare chronic disease characterized by widespread intra-alveolar deposition of micro-calcifications called "calcspherites" range from 0.01 to 3 mm in size and are composed of calcium phosphate<sup>[1]</sup>. Up to December 2014 a total of 1022 PAM cases were described in the literature in 65 nations, the majority being in Asia<sup>[2]</sup>. Etiology is unknown, but there is high incidence of familial occurrence (approximately one-third of the cases) suggesting an autosomal recessive inheritance pattern<sup>[3,4]</sup>. Most cases are diagnosed in the third to the fifth decade and there is no predilection for one sex over the other, although there is slight female predilection in familial cases<sup>[5]</sup>. Herein, we report a familial case of pulmonary alveolar microlithiasis because of its rarity and to emphasize on its radiological features.

## Case Report:

A 35 year old female was admitted in January, 2016 in Al-Kindy teaching hospital, Baghdad, with a history of cough with expectoration and progressive exertional dyspnea for the last eight months. The expectorate contained sand like particles and there was no history of fever, hemoptysis or weight loss. She was non-smoker. She reported that two of her sisters had experienced similar breathing problems at their third-fourth decade and been dead at 40 years age. On physical examination of the respiratory system, harsh breath sounds with bilateral scattered creptations were found, mainly in the lung base. Clubbing was also present. Sputum for AFB was negative and serum calcium was normal. Pulmonary function studies revealed severe restrictive lung disease with diminished diffusing capacity.

The chest radiograph [Figure - 1] shows diffuse bilateral calcific infiltrates, which are seen predominantly in the mid and lower lung zones with obscuration of the mediastinal, cardiac, and diaphragmatic borders. These infiltrates are alveolar in nature and produce an air bronchogram. A few fibrotic strands were also seen. CT scan [Figure - 2] revealed the presence of widespread nodular intra-alveolar opacities of calcific density with diffuse ground-glass attenuation, mainly in the lower pulmonary regions. Calcifications were seen along the interlobar septa and subpleural regions. There was also evidence of septal thickening. Few fibrotic changes were also noticed.

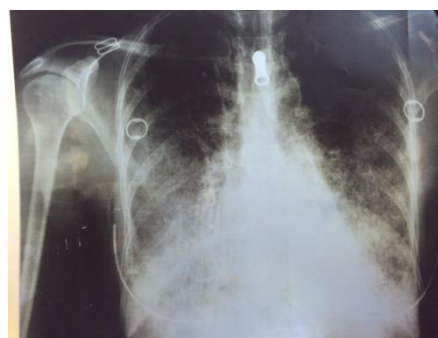
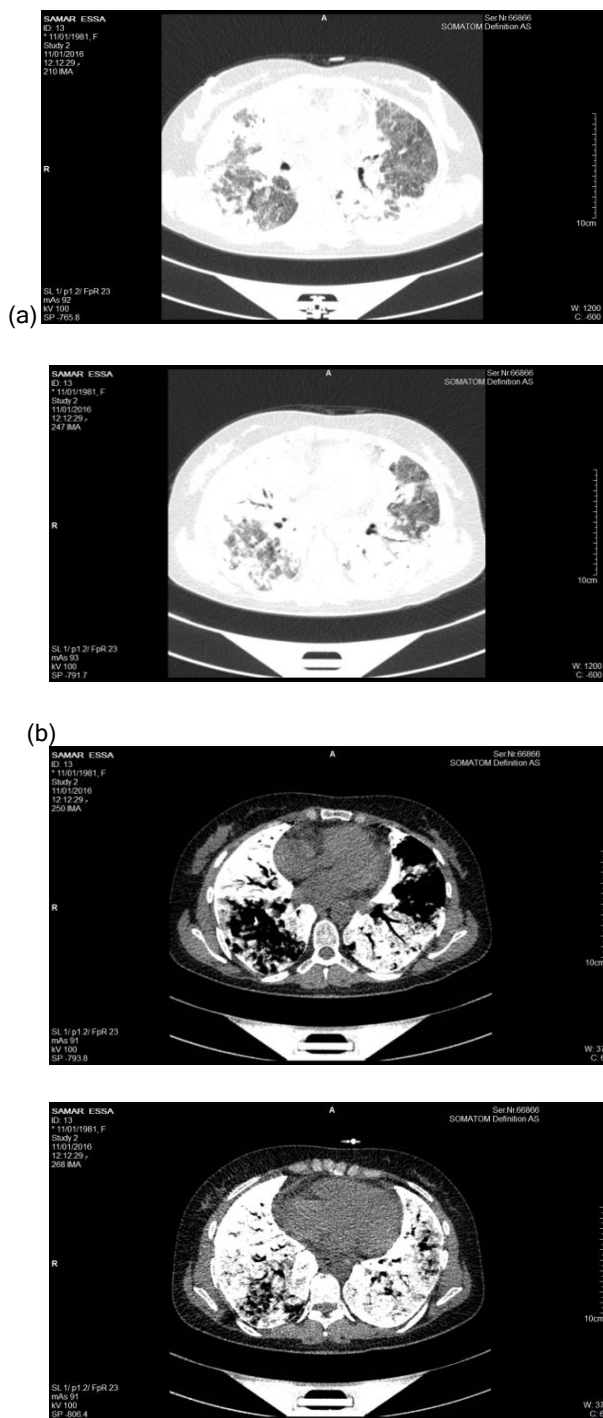


Figure 1. chest x ray showing bilateral diffuse high density micronodular infiltrates more toward the mid and lower zones. The heart borders and the diaphragm are obscured.



**Figure 2:** (a) Axial CT images (lung window) shows a diffuse intra-alveolar opacities of calcific density in bilateral lung parenchyma with subpleural and peribronchial distribution (b) Axial CT images (mediastinal window) shows diffuse bilateral dense alveolar calcification with air bronchogram, septal thickening, and calcification along the interlobar septa and subpleural regions.

**Discussion:** The best diagnostic work-up of sporadic PAM consists of chest radiography followed by the combination of chest HRCT with transbronchial biopsy, but a chest radiography along with information obtained

from the family history are sufficient for diagnosis in familial PAM as in our case. Moreover, HRCT make it possible to classify the evolutionary stage of the disease and its severity [1,2,5]. A striking feature of this disease is lack of significant symptoms despite extensive radiographic changes. Typically, as seen in our case, patients may remain asymptomatic for many years and usually become symptomatic between third and fourth decades, the condition may progress slowly leading to progressive dyspnoea with or without cough and ultimately end up with respiratory insufficiency, lung fibrosis and corpulmonale.

In the radiological diagnosis of PAM, chest radiographs usually reveal diffuse, bilateral areas of micronodular calcifications (“sand storm”) that predominate in the middle and lower lung areas [3,6]. The heart borders and the diaphragm are usually obliterated. The chest radiographs of our patient showed a diffuse symmetric lung lesion with dense micronodular aspect, corroborating the pattern described in the literature.

The HRCT findings in patients with alveolar microlithiasis vary considerably according to its severity. Ground-glass opacities, subpleural linear calcification, confluent and diffuse calcified nodules and dense consolidations are common findings described in literature [3,6]. The CT scan in our case show advanced changes manifested by diffuse dense alveolar calcification with few elements of fibrosis.

There is no known medical treatment to reduce the progression of the disease. Palliative treatments with systemic corticosteroids, calcium-chelating agents and serial bronchopulmonary lavage have been shown to be ineffective. Attempts to reduce calcium phosphate precipitation in pulmonary alveoli has been tried with diphosphonate [7]. Lung transplantation remains the only possible treatment for end-stage cases [2,8]

In conclusion, PAM is a rare disease that can affect young patients, with chronic and deteriorating evolution. Clinicians should be aware of it existence and the radiological features associated. The radiographic picture of PAM is highly characteristic and thus constitutes an extremely important diagnostic element. Superficial assessment of the radiological findings in patients with PAM can cause confusion with other diseases associated with miliary dissemination such as tuberculosis, mycosis, sarcoidosis, haemosiderosis, pneumoconiosis and amyloidosis, which can all present with diffuse opacifications, although the symptoms are more severe in these diseases. In this way, HRCT should always be performed since it can reveal characteristic patterns of alveolar microlithiasis, reserving lung biopsy for atypical and inconclusive cases.

**References:**

1. Castellana G, Lamorgese V. Pulmonary alveolar microlithiasis World cases and review of the literature. *Respiration* 2003;70:549-55.
2. Castellana G, Gentile M, Castellana R, and Resta O. Pulmonary alveolar microlithiasis: review of the 1022 cases reported worldwide. *EurRespir Rev* 2015; 24: 607-620.
3. Marchiori E, Gonçalves CM, Escuissato DL, Teixeira KI, Rodrigues R, Barreto MM, and Esteves M, "Pulmonary alveolar microlithiasis: high-resolution computed tomography findings in 10 patients," *Jornal Brasileiro de Pneumologia* 2007;33: 552-557.
4. Mariotta S, Ricci A, Papale M, Clementi F, Sposato B, Guidi L, and Mannino F, "Pulmonary alveolar microlithiasis: report on 576 cases published in the literature," *SarcoidosisVasculitis and Diffuse Lung Diseases* 2008; 21:173-181.
5. Tachibana T, Hagiwara K, Johkoh T. Pulmonary alveolar microlithiasis: review and management. *CurrOpinPulm Med* 2009;15(5):486-490.
6. Gasparetto EL, Tazoniero P, Escuissato DL, Marchiori E, Frare Silva RL, and Sakamoto D, "Pulmonary alveolar microlithiasis presenting with crazy-paving pattern on high resolution CT," *British Journal of Radiology* 2004;77:974-976.
7. Göcmen A, Toppare MF, Kiper N, Büyükpamukcu N. Treatment of pulmonary alveolar microlithiasis with diphosphonate: Preliminary results of a case. *Respiration*1992;59:250-52.
8. Raffa H, El-Dakhakny M, Al-Ibrahim K, Mansour MS. Single lung transplantation for alveolar micro-lithiasis: The first clinical report. *Saudi J Kidney Dis Transpl.* 1996;7:189-93.