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Pulmonary Alveolar Microlithiasis in Moroccan Child: A Case Report

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Abstract **Case Report**

Background: Pulmonary alveolar microlithiasis (PAM) is a rare but not uncommon disease; it showed worldwide distribution either familial or sporadic, the most reported cases in Europe, especially in Turkey. The presence of round shaped little bodies containing concentric calcareous lamellas in pulmonary alveolus is the hallmark of the disease. With this study, we report a case of PAM in moroccan child, who complained of class 2 exertional dyspnoea with no significant personal or family history. On physical examination, there was no evidence of clubbing or cyanosis of the extremities. On auscultation, fine crackles were present bilaterally. Chest X-ray showed innumerable small dense nodules diffusely involving both lungs. High resolution CT scan showed extensive micro nodules, areas of diffuse ground glass attenuation, septal thickening and in some cases black pleural lines. The diagnosis confirmed by opened right lung biopsy and Broncho alveolar lavage, biopsy showed numerous lamellar (Calculi).

Keywords: Pulmonary alveolar microlithiasis. PAM, Pediatrics.

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is a rare infiltrative lung disease characterized by deposition of spherical calcium phosphate concretions (microliths) in the alveoli and a paucity of symptoms in contrast to imaging findings [1, 2]. Although the aetiology remains unclear, a few reports have described the role of a mutation in the sodium phosphate cotransporter type IIb gene (SCL34A2 gene) in the pathogenesis of the disease [3].

CASE PRESENTATION

We present the case of a 14-year-old girl who complained of class 2 exertional dyspnoea with no significant personal or family history. On physical examination, there was no evidence of clubbing or cyanosis of the extremities and no hepatosplenomegaly. On auscultation, fine crackles were present bilaterally. Chest X-ray showed innumerable small dense nodules diffusely involving both lungs (Figure 1). High resolution CT scan showed extensive micro nodules, areas of diffuse ground glass attenuation, septal thickening and in some cases black pleural lines (Figure 2). Diagnosis confirmed by opened right lung biopsy and Broncho alveolar lavage, biopsy showed numerous lamellar (Calculi).

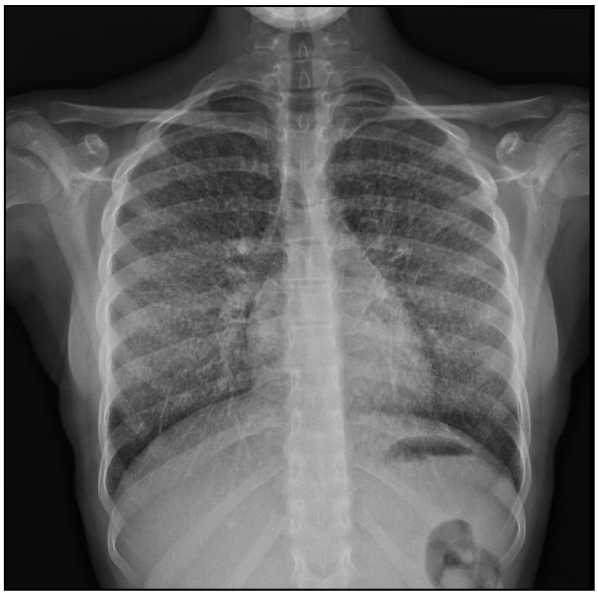

Figure 1: Chest X-ray showing bilateral diffuse high density micro-nodular opacities involving both lungs

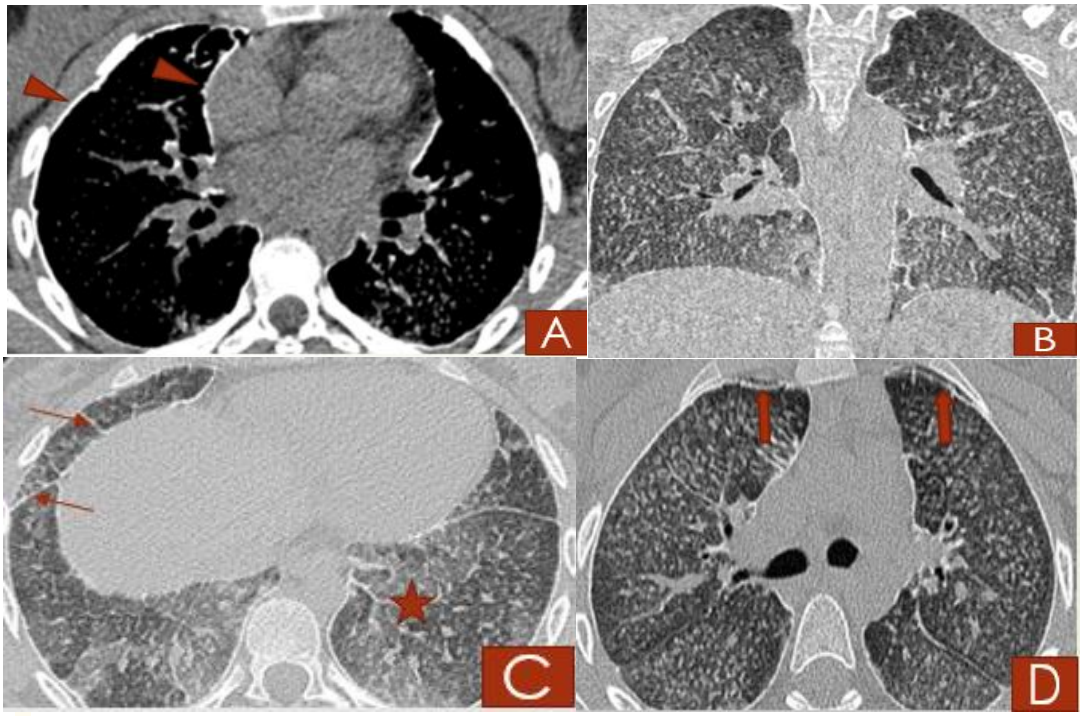

Figure 2: (a-b-c and D) High-resolution CT scan with a mediastinal axial window A, B Coronal view, C and D axial view of the chest, reveals: (A) Calcifications along the sub pleural regions (arrow head). (B and C) bilateral diffuse micro-nodular opacities and ground glass opacities (Asterix) in upper with dense reticulation (arrow) and attenuation and septal thickening, more pronounced in lower pulmonary regions (B), (D) subpleural cysts (arrow)

DISCUSSION

Pulmonary Alveolar Microlithiasis (PAM) is a rare but not uncommon disease, it showed a worldwide distribution [1]. It affects both genders equally; Although it is common between 20-30 years of age but pediatric and neonatal cases were reported [3, 4]. PAM considered idiopathic in spite of the various etiological theories that were reported in the literature. Genetic analysis revealed that there is heterogeneous mutation of SLC34A2 gene, which is responsible for the familial PAM [5].

Many patients have no clinical symptoms, and most of the cases either have normal pulmonary function or a mild restrictive pattern [6]. In symptomatic patients, typical complaints are dyspnea as in our case, nonproductive cough, chest pain and asthenia [7]. Although the clinical course varies, the disease is usually slowly progressive. In some patients, the disease remains static, while it in others worsens over time, progressing into pulmonary fibrosis, respiratory failure and cor pulmonale [6, 7].

The characteristic chest radiograph reveals a picture of fine sand-like micronodulation of calcific densities bilaterally, described as a sandstorm, involving mainly the middle and lower zones as like our case without any known disorder of calcium metabolism [8]. The distribution of the calcified nodules can also be explained by the relative higher blood supply to this area. Other typical findings include small apical bullae and a black pleural line, which is demonstrated as an area of increased translucence between the lung parenchyma and the ribs The chest radiographs of our patient showed a diffuse symmetric lung lesion with dense micronodular aspect, corroborating the pattern previously described in the literature [4, 6, 8].

CT usually shows diffuse ground-glass opacities in both lungs, associated with confluent and diffuse calcified nodules. It confirms the predominance of symmetrical abnormalities in the middle and lower zones. Calcifications along the bronchovascular bundles and at the central region of the bronchovascular tree can also be seen. A predominance of calcifications in the medial areas when compared with the lateral portions of the lungs is also evidenced in the CT scan [1, 2]. CT scans may reveal small cysts in the subpleural lung parenchyma, pleural calcification and small calcispherites within the thickened pleura [2, 3]. These findings were observed in our case.

CONCLUSION

Pulmonary alveolar microlithiasis (PAM) is a rare parenchymal lung disease characterized by deposition of spherical calcium phosphate concretions (microliths) in the alveoli and a paucity of symptoms in contrast to imaging findings. High resolution CT scan remains the imaging technique of choice to diagnose PAM.

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