PULMONARY ALVEOLAR MICROLITHIASIS: A CASE REPORT WITH EMPHASIS ON CLINICORADIOLOGICAL DISSOCIATION

Raghuram U1, Md. Badusha2, B. Anusha Bhargavi3, C. Ramanjula Reddy4, S. Satya Sri5

1Junior Resident, Department of Pulmonology, ASRAMS, Eluru.
2Junior Resident, Department of Pulmonology, ASRAMS, Eluru.
3Junior Resident, Department of Pulmonology, RMC, Kakinada.
4Senior Resident, Department of Pulmonology, ASRAMS, Eluru.
5Professor & HOD, Department of Pulmonology, ASRAMS, Eluru.

ABSTRACT

Pulmonary alveolar microlithiasis (PAM) is a rare disease characterised by the presence of small calculi in the alveolar space. The authors report a case of a 59-year-old woman with a 2-year history of dry cough. Physical examination was not significant except for fine crackles at auscultation. Pulmonary function revealed a mild restrictive ventilatory defect and the chest radiograph demonstrated bilateral dense micronodular infiltrates giving diffuse sandstorm appearance. High-resolution CT scan revealed diffuse ground glass attenuation and septal thickening, more pronounced in mid and lower pulmonary regions, with calcifications along the interlobar septa and subpleural regions. A transbronchial lung biopsy confirmed the diagnosis of PAM.

KEYWORDS

PAM, Pulmonary Alveolar Microlithiasis, SLC34A2 Gene, Micronodular Infiltrates, Sandstorm Appearance.


INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is a rare chronic disease characterised by multiple microscopic calculi within the alveoli and a paucity of symptoms in contrast to the imaging findings.[1-7] The disease is usually discovered incidentally during radiography of the chest for other reasons. Many patients are asymptomatic and the majority of patients either have normal or restrictive pulmonary function. The clinical course of the disease varies. While it remains static in some patients, it progresses into pulmonary fibrosis, respiratory failure and cor pulmonale in others. With the exception of lung transplantation, there is no known effective treatment for the disease. It is mostly accepted now that the disease has an autosomal recessive inheritance.[2,4,8] Recently, a few reports have described the role of mutation in the type IIb sodium-phosphate co-transporter gene (SLC34A2 gene) in the disease pathogenesis.[9,10] The authors present a case of symptomatic PAM in a 59-year-old woman, emphasising the role of high-resolution lung CT (HRCT) in the diagnosis of this unusual disease.

CASE REPORT

A 59-year-old female presented with a 2-year history of dry cough with no other significant symptomology. At physical examination, auscultation of the lungs revealed fine inspiratory crackles. Cardiac auscultation was normal, and no cyanosis or peripheral oedema was observed. There was no history of smoking or previous known pulmonary disease.

On routine blood examination, blood counts and serum chemistries were found to be normal. Arterial blood gas analysis and echocardiography showed no important abnormalities. USG abdomen was also normal.

Pulmonary function tests (PFT) showed a mild restrictive ventilatory defect. The sputum was negative for acid-alcohol resistant bacillus and human immunodeficiency virus testing was negative as well. The chest plain films revealed a diffuse symmetric dense bilateral micronodular pattern with middle to lower zone predilection giving a sandstorm appearance and black pleura sign could be made out (Figure 1). Based on this finding, HRCT scan was obtained, revealing diffuse ground glass attenuation with reticulations more pronounced in mid and lower pulmonary regions with thickening of the interlobar septa and calcifications of subpleural regions. Small subpleural cysts were also noticed (Figure 2). The patient underwent a fibroptic bronchoscopy with bronchoalveolar lavage and transbronchial lung biopsy. The lavage fluid was positive for tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi. Microliths were not found. Histology revealed round, concentrical tubercle bacilli or fungi.

Fig. 1: Chest Radiograph in Anteroposterior view showing Diffuse Symmetric Lung Lesion with Confluence of Dense Micronodular Infiltrate. Note the, Predominance of the Lesions in the Paracardiac Regions of the Lungs
PAM, causes an increase in permeability, leading to an increase in the levels of these two proteins in the blood. Therefore, SP-A and SP-D measurement may be an alternative to monitor the progression and activity of the disease. These exams were not performed in our patient. Nevertheless, no effective treatment for end-stage PAM currently exists, with the exception of lung transplantation.

Transplantation should be considered in cases where either severe respiratory failure or right heart failure are present. Patients who have undergone lung transplantation have shown an increase in right ventricular ejection fraction and regression of right ventricular hypertrophy.[14]

In the radiological diagnosis of PAM, chest radiographs usually reveal diffuse, bilateral areas of micronodular calcifications ("sandstorm") that predominate in the middle and lower lung areas.[2–7] The heart borders and the diaphragm are usually obliterated. Other typical finding includes a black pleural line, demonstrated as an area of increased transluence between the lung parenchyma and the ribs.[2,4,5,7] The chest radiographs of our patient showed a diffuse symmetric lung lesion with dense micronodular aspect, similar to the pattern described in the literature.

The HRCT findings in patients with alveolar microlithiasis vary considerably.[2–7] Ground-glass opacities are the most common finding described in literature.[2,4,5,7] This pattern occurs probably due to small calculi in the air space. Subpleural linear calcification, confluent and diffuse calcified nodules and dense consolidations are also a common finding. Calcifications along the bronchovascular bundles and at the central region of the bronchovascular tree can also be seen.[2–7] The CT scan confirms a predominance of symmetric calcifications along the heart borders and in the lower posterior portions of the lung.[2,4,7] Small thin-walled subpleural cysts are described as well, and they are responsible for the “black pleura” sign seen in the chest X-rays.[2,4,5,11]

Recently, it was described the unique characteristics of the mosaic pattern seen in alveolar microlithiasis, in which the interlobular septa are of calcium density is due to the deposition of calcospheres within the peripheral lobular parenchyma, adjacent to the septa. Some authors consider this pattern very specific and even pathognomonic of PAM on the HRCT scan, since this aspect has not been described in any other disease.[2,7] Clinicians should have in mind that some findings seen in PAM, such as nodular calcifications, can be found in other diseases like tuberculosis, metastatic osteosarcoma, amyloidosis and silicoproteinosis. Besides that, dense consolidations can also be found in metastatic pulmonary calcification, taklossis and amiodarone lung toxicity. In this way, associated CT findings and clinical features should always be correlated, since these diseases have different kinds of presentation and evolution.[10]

In conclusion, PAM is a rare disease that can affect young patients, with chronic and deteriorating evolution. Clinicians should be aware of its existence and the radiological features associated. The striking feature is lack of clinical findings despite significant radiological changes. The micronodular pattern seen in chest radiography can sometimes be misdiagnosed as milary tuberculosis or other diseases that present with this pattern. 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