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Research Article

PULMONARY ALVEOLAR MICROLITHIASIS WITH BRONCHIAL ASTHMA

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A B S T R A C T

Pulmonary alveolar microlithiasis (PAM) is a rare autosomal recessive disorder characterized by intra-alveolar accumulation of spherical microliths in the absence of any known calcium metabolism disorder. The hallmark of this disorder is clinical-radiological dissociation, meaning that there is a paucity of symptoms in contrast to characteristic imaging findings. At diagnosis, most patients are asymptomatic and changes in the lung parenchyma are found incidentally. As the disease advances patient may develop dyspnea, dry cough, chest pain. There are less than 1050 cases of PAM reported worldwide. We report a rare case of PAM with bronchial asthma.

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is a rare autosomal recessive disorder characterized by intra-alveolar accumulation of spherical microliths in the absence of any known calcium metabolism disorder. (1) it's caused due to the mutations of the *SLC34A2* gene, which encodes a sodium phosphate co-transporter. The hallmark of this disorder is clinical-radiological dissociation, meaning that there is a paucity of symptoms in contrast to characteristic imaging findings. (2) At diagnosis, most patients are asymptomatic and changes in the lung parenchyma are found incidentally. As the disease advances patient may develop dyspnea, dry cough, chest pain. There are less than 1050 cases of PAM reported worldwide. (3)We report a rare case of PAM with bronchial asthma.

CASE REPORT

A 60 year old lady, presented with complaints of exertional dyspnea, cough with scanty mucoid expectoration, wheeze on and off since 5 years, atopy symptoms since childhood. Her vitals were normal at the time of presentation. On examination she had bilateral diffuse rhonchi. A clinical diagnosis of bronchial asthma was made and a chest X ray and PFT were ordered. Chest X ray revealed diffuse sandstorm-like calcifications predominantly in the bilateral middle and lower lung zones. PFT showed restrictive pattern with good reversibility.

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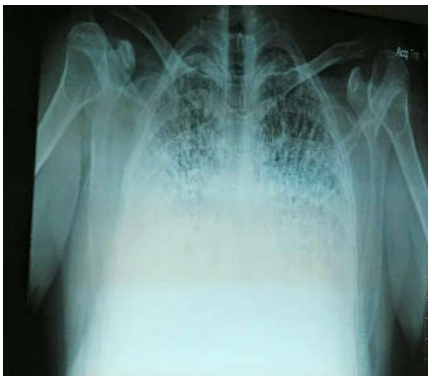
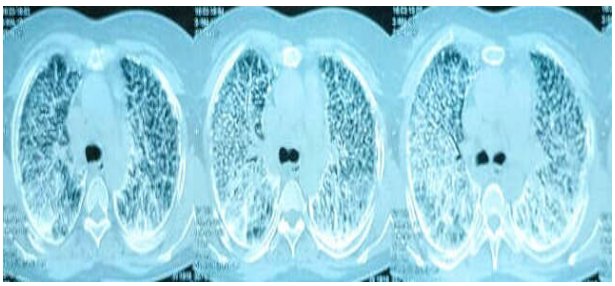
HRCT lungs then showed bilateral calcification predominantly in the middle and lower zone with inter and intralobular septal thickening and ground glass opacities with positive black pleura sign. With the characteristic X ray and CT findings a diagnosis of PAM with bronchial asthma was made. Lung biopsy showed intra alveolar microliths confirming the diagnosis of PAM.





DISCUSSION

Pulmonary alveolar microlithiasis is a rare disease entity that is characterized by innumerable diffuse calcospherites (microliths composed of calcium and phosphorus) within the alveolar space [(1)]. It is reported worldwide and 25% cases are from turkey. A slight female predilection may be present in familial form. The disease is usually discovered from birth up to 40 years of age and is often diagnosed incidentally during radiography of the chest for other reasons. Although the etiology remains unclear, PAM is thought to be an autosomal recessive disease caused by mutations of the *SLC34A2* gene, which encodes a sodium phosphate co-transporter.[(3)] PAM has been described in detail by Soaman in 1957.[1-10]

The patients are usually asymptomatic in mild form or may present as dyspnea, dry cough, chest pain and asthenia when the diseases advances. Our patient presented with asthma like symptoms. Respiratory insufficiency eventually progresses to pulmonary fibrosis, end-stage lung disease, and chronic pulmonary heart disease. Extrapulmonary involvement is uncommon [(5)]. The hallmark of this disorder is clinical-radiological dissociation meaning that clinicians will find a paucity of symptoms in contrast to imaging findings. [(6)]

On radiographs PAM is characterized by diffuse fine calcific micronodules that involve both lungs in a pattern that is classically described as sandstorm-like involving the mid and lower zones [(6-7)]. Calcified micronodules, ground-glass opacities, interlobular septal thickening, pleural and subpleural calcification and cysts are typical radiological findings seen in HRCT(8) As the disease progresses, pulmonary function tests reveal typical features of a restrictive defect with reduced forced vital capacity (FVC) and elevated FEV1/FVC. Reduced total lung capacity and tidal volume (V_T) have also been described.

There is no known effective treatment for PAM, with the exception of lung transplantation [(9)] 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 [(10)] The long term prognosis of this disease is poor.

CONCLUSION

The disease may progress with chronic alveolar calcification causing interstitial inflammation, and fibrosis leading to decreased lung volumes and eventually right heart failure. No treatment has been proven to effectively prevent the progression of PAM. Currently, the only effective therapy is lung transplantation, especially when it is performed before the disease progresses to an advanced stage.

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