

Pulmonary alveolar microlithiasis

Anita Flynn, Anuradha D Agastyaraju

ABSTRACT

Introduction: Pulmonary alveolar microlithiasis is an important cause of diffuse parenchymal lung disease and is characterised by calculospherites in the alveolar spaces. Usually occurring in the 4th to 6th decade. Sex distribution is roughly equal. Clinical course is progressive and fatal. About 300 cases of pulmonary alveolar microlithiasis have been reported in literature. **Case Report:** Here we report a case of a 56-year-old male who presented with progressively increasing breathlessness. Imaging revealed characteristic “sandstorm” appearance on X-ray. A transbronchial lung biopsy confirmed the diagnosis of pulmonary alveolar microlithiasis. **Conclusion:** The etiology of pulmonary alveolar microlithiasis is unknown and there is no definite medical treatment. Hence, therapy is mostly empirical.

Keywords: Breathlessness, Alveoli, Calculospherites

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INTRODUCTION

Pulmonary alveolar microlithiasis is a rare disease of unknown etiology. It is characterized by intra-alveolar deposition of calcific concretions in the absence of abnormal calcium metabolism [1]. Pulmonary alveolar microlithiasis is a slowly progressive disease leading to respiratory insufficiency associated with cyanosis, clubbing and pulmonary hypertension [1]. There is no satisfactory therapy for this condition [1]. This disease was first described by Malpighi et al. in 1686 [2]. Puhr et al. gave this disease an appropriate name in 1932 [2]. In 1947, Mariani et al. described the clinical, functional and radiological features of this entity [2].

The diagnosis of pulmonary alveolar microlithiasis depends on the characteristic “sandstorm” picture in chest X-ray and the finding of alveolar calculospherites on histology.

CASE REPORT

A 56-year-old male presented in the pulmonology out patient department with the chief complaint of progressively increasing breathlessness for the past one year accompanied by dry cough which was not relieved by medication. There was no history of fever, hemoptysis or chest pain. Patient was a non-smoker. There was no history of tuberculosis within the family members or any known history of pulmonary disease. On physical examination, the patient had no clubbing, cyanosis, lymphadenopathy or peripheral edema.

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Cardiac auscultation was normal. Auscultation of lungs revealed coarse crepitations in all the lung fields. Pulmonary function test (PFT) revealed mild restrictive ventilatory defect with reduced total lung capacity. HIV was non-reactive. Routine lab investigations were non-contributory. Chest X-ray showed bilateral finely granular calcified densities predominantly in the middle and lower lobes (Figure 1).

Fiberoptic bronchoscopy, was performed bronchoalveolar lavage and transbronchial biopsy. The bronchoalveolar lavage fluid was negative for acid fast bacilli and fungi. Calcospherites were not found. The transbronchial biopsy was sent for histopathology. Microscopy showed alveolar spaces filled with deeply eosinophilic lamellated calcospherites (Figure 2). The alveolar walls showed a scattered chronic inflammatory infiltrate and mild fibrosis. A histologic diagnosis of pulmonary alveolar microlithiasis was made. The patient was started on steroids and came for subsequent follow up visits for a period of six months. Minimal improvement in the repeat X-ray was documented. The patient was subsequently lost to follow-up.



Figure 1: Chest X-Ray showing typical “sandstorm” appearance.

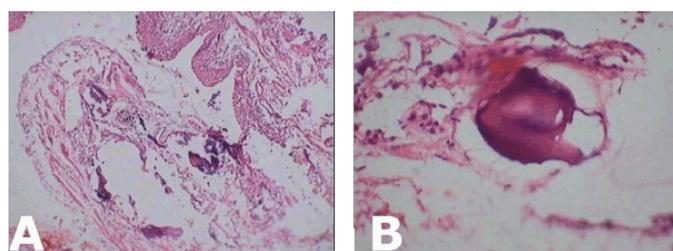


Figure 2: (A) Lamellated calculospherites scattered in the lung parenchyma, (H&E, x100), (B) Intra-alveolar calculospherites (H&E, x400).

DISCUSSION

Pulmonary alveolar microlithiasis is an autosomal recessive lung disease characterized by filling of the lung alveoli by calcospherites [3]. It is caused by mutations in the SLC34A2 sodium dependant phosphate transporter

that is normally expressed in the type II alveolar macrophages [4]. Half of the cases, however, are familial [5]. In the remaining, the cause is obscure [5]. The average age of presentation is between the 4th and 6th decade. The M:F sex ratio is roughly equal [3]. Patients are usually asymptomatic or present late, usually with cough or breathlessness [5]. In advanced cases, hemoptysis, pneumonia, end-stage interstitial pneumonia and cor pulmonale may develop [3]. Pulmonary function tests mirror the stage of the disease [6]. The progress of these patients is variable. Most patients suffering from a protracted disease which is usually progressive and fatal [2, 5]. There is no effective treatment except lung transplant [5].

Imaging findings are described as a typical “sandstorm” appearance with fine granular opacities throughout the lungs, worse at the bases [2]. The airways appear unremarkable on bronchoscopy; however, concretions may be aspirated in the BAL fluid [2].

At autopsy, the lungs are extremely difficult to cut. Cut surfaces are hard to gritty with numerous 0.2–0.3 mm, yellowish brown, sand-like particles [5]. Histologically, the alveoli are filled with concentric lamellated basophilic calcospherites measuring 50–200 microns in size [3]. The calcospherites are composed of calcium, phosphate or calcium hydroxyapatite with traces of magnesium, silicon, iron and aluminium [3]. The central cores of the microliths are PAS-positive while the surrounding laminations are PAS-negative and von Kossa positive [3, 6]. In our case, no attempt was made to do any chemical analysis of the calcareous material. The calcospherites are sometimes birefringent, giving a Maltese cross pattern with polarization [6]. The differential diagnosis of calcific bodies include pulmonary blue bodies, corpora amylacea, metastatic calcification and heterotopic ossification [3].

CONCLUSION

The etiology and pathogenesis of pulmonary alveolar microlithiasis is still largely unknown. Hence, there is no effective and definitive medical therapy and treatment is mostly empirical. Radiology may be confusing, keeping in mind the numerous differential diagnoses. It will only be possible to develop effective treatment of the disease after its etiology has been fully established.

Author Contributions

Anita Flynn – Conception and design, Acquisition of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Agastyaraju D Anuradha – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

1. Tahlan Anita, Bal Amanjit, Mohan Harsh, Saini Varinda, Deb Anirban. Pulmonary alveolar microlithiasis: A case report. *Clin Pulm Med* 2003;10:353–4.

2. Mariotta S, Guidi L, Papale M, Ricci A, Bisetti A. Pulmonary alveolar microlithiasis: Review of Italian reports. *Eur J Epidemiol* 1997;13(5):587–90.
3. Churg A, Myers JL, Tazelaar HD, Wright JL. Thurlbeck's Pathology of the Lung, 3rd edition. New York: Thieme 2005:1157.
4. Castellana G, Gentile M, Castellana R, Fiorente P, Lamorgese V. Pulmonary alveolar microlithiasis: clinical features, evolution of the phenotype, and review of the literature. *Am J Med Genet* 2002;111(2):220–4.
5. Warriar AR, Gowrinath K. Pulmonary alveolar microlithiasis. *Lung India* 2006;23:42–4.
6. Moran CA, Hochholzer L, Hasleton PS, Johnson FB, Koss MN. Pulmonary alveolar microlithiasis. A clinicopathologic and chemical analysis of seven cases. *Arch Pathol Lab Med* 1997;12(6):607–11.

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