

Case Report

Pulmonary alveolar microlithiasis

Gayathri Devi H. J., Mohan Rao K. N., Prathima K. M.¹, Jayanth K. Das²

Departments of Chest Diseases, MS Ramaiah Medical College, ¹Pathology, MS Ramaiah Memorial Hospital, and ²Cardiothoracic Surgery, MS Ramaiah Narayana Hrudayalaya, Bangalore, India

ABSTRACT

Pulmonary alveolar microlithiasis is a rare disease of unknown cause. We report a case in a young boy who presented with history of failure to thrive and chest X-ray finding suggestive of miliary mottling. Open lung biopsy revealed pulmonary alveolar microlithiasis.

KEY WORDS: Pulmonary alveolar microlithiasis, misdiagnosis, miliary tuberculosis, biopsy

Address for correspondence: Dr. Gayathri Devi H. J., Department of Chest Diseases, MS Ramaiah Medical College, Bangalore - 560 054, India.
E-mail: joshyev@gmail.com

INTRODUCTION

Pulmonary alveolar microlithiasis (PAM) is an uncommon idiopathic disease characterized by microliths in the alveoli, and was first described by Friedrich in 1856 and then by Harbitz in 1918.^[1,2] In 1957, Sosman emphasized that 50% of the cases were familial.^[3] It is regarded as an autosomal recessive lung disease. As of 2004, 576 PAM cases have been studied, and most have originated from Europe (42.7%) and Asia (40.6%). Pulmonary tuberculosis or sarcoidosis was misdiagnosed in 88 cases of the 576.^[4]

In this report, our aim is to emphasize a high index of suspicion for this uncommon disorder to avoid misdiagnosis of miliary tuberculosis.

CASE REPORT

A fifteen-year-old boy was investigated for failure to thrive. He was totally asymptomatic and no respiratory complaint at the time of evaluation. He had received two courses of anti tuberculosis treatment (2007 and 2009) elsewhere with the diagnosis of Miliary tuberculosis based

on radiological findings. Clinical examination did not reveal any abnormality.

Investigations

His CBC, electrolytes and calcium were normal. Ultrasound of abdomen was normal. Flow volume loop showed mild restriction. Serial chest X-rays from 2007 showed bilateral diffuse micronodular calcific shadows more toward the mid and lower zones [Figures 1 and 2]. HRCT scan showed bilateral diffuse micronodular opacities distributed predominantly over basal and posterior regions with thickening of interlobar septa [Figure 3]. Wedge biopsy of the Lingula was done. Histopathology examination revealed multiple calcispherites within the alveoli of lung parenchyma suggestive of pulmonary alveolar microlithiasis of lung [Figure 4]. Chest X-ray of his only sibling did not reveal any abnormality.

DISCUSSION

PAM is an uncommon disease characterized by widespread localization of calcispherites (salts of calcium and phosphate) in the alveolar spaces in the absence of any known disorder of calcium metabolism. Alveolar microlithiasis is reported worldwide and one-quarter of patients are from Turkey.^[5] PAM can be seen in any age group. Sex distribution is equal. A familial incidence particularly in siblings is common. Case reports of PAM have been published in Indian literature.^[6]

PAM has been described in detail by Sosman in 1957. Recently, the gene SLC34A2 has been identified and is considered to be responsible for PAM. This gene codes for

Access this article online	
Quick Response Code: 	Website: www.lungindia.com
	DOI: 10.4103/0970-2113.80334

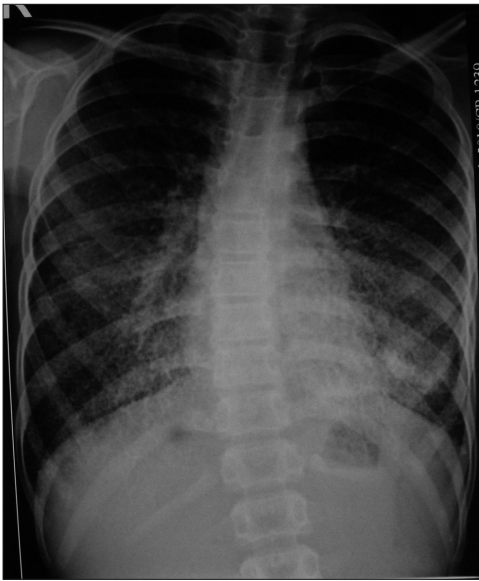


Figure 1: Chest X-ray showing bilateral diffuse high density micronodular opacities more toward the mid and lower zones

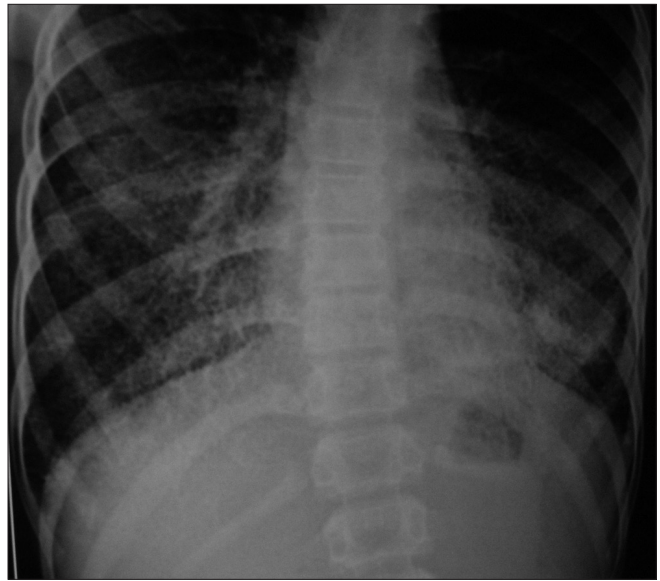


Figure 2: Chest X-ray showing bilateral diffuse high density micronodular opacities more toward the mid and lower zones

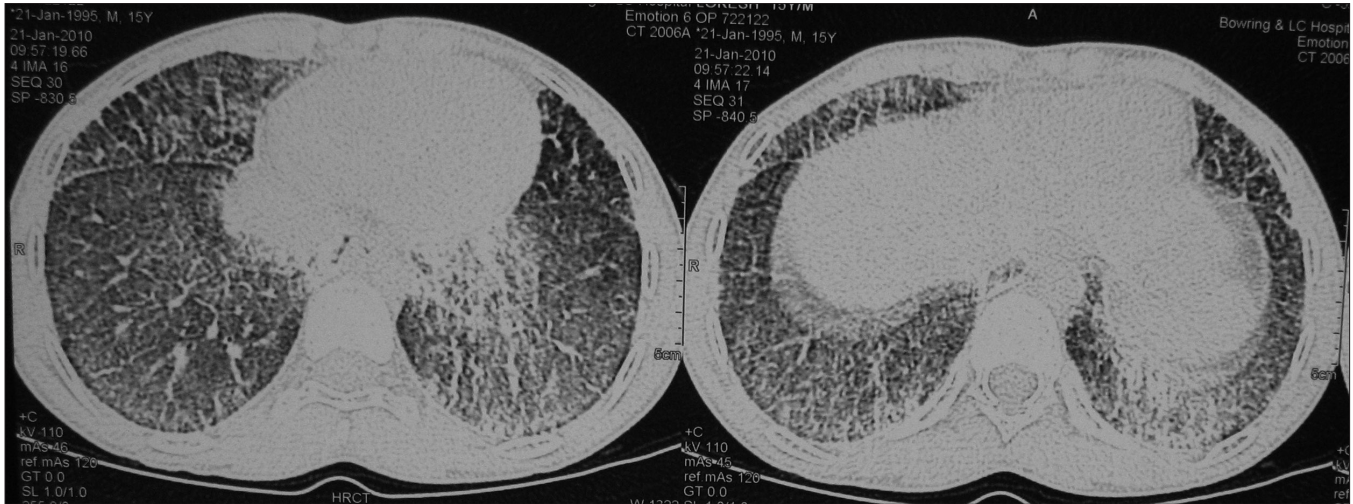


Figure 3: CT of Thorax showing bilateral diffuse micronodular opacities distributed predominantly over basal and posterior regions with reticular pattern

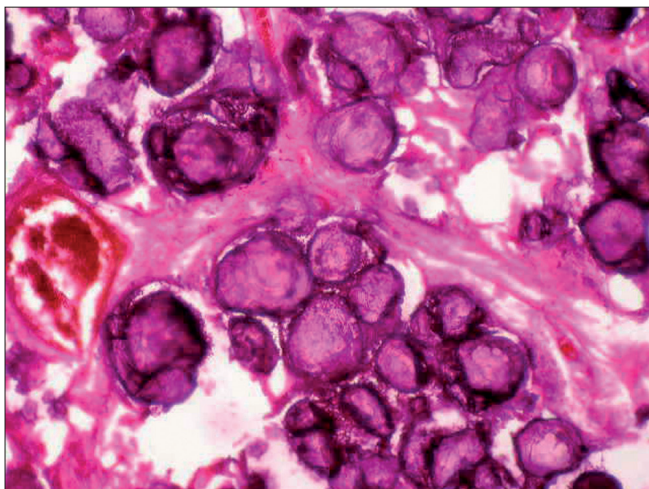


Figure 4: Histopathology section showing multiple calcispherites within the alveoli of lung parenchyma. H and E 100x

type IIb sodium-dependent phosphate transporter and its function may provide insight into the pathogenesis of this disorder. Confirmation of diagnosis of PAM can be done by demonstrating the mutation in the SLC34A2 gene.^[7]

A striking feature of this disease is lack of significant symptoms despite extensive radiographic changes. 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 and cor pulmonale. Plain chest radiographs usually reveal diffuse bilateral areas of micronodular calcifications that predominate in the middle and lower lung areas.

There is no known medical treatment to reduce or halt 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.^[8] Lung transplantation remains the only possible treatment for end-stage cases.^[9]

REFERENCES

1. Friedrich N. Corpora amylacea in den Lungen. Arch Pathol Anat 1856;9:613-8.
2. Harbitz F. Extensive calcifications of the lungs as distinct disease. Arch Intern Med 1918;21:139-46.
3. Sosman MC, Dodd GD, Jones WD, Pillmore GU. The familial occurrence of pulmonary alveolar microlithiasis. Am J Roentgenol Radium Ther Nucl Med 1957;77:947-1012.
4. Mariotta S, Ricci A, Papale M, De Clementi F, Sposato B, Guidi L, *et al.* Pulmonary alveolar microlithiasis: report on 576 cases published in the literature. Sarcoidosis Vasc Diffuse Lung Dis 2004;21:173-81.
5. Shah MS, Nanavati KI, Airon N, Shah RR, Joshi BD. Case report: Pulmonary alveolar microlithiasis. Indian J Radiol Imaging 2003;13:277-9.
6. Gowrinath K, Warriar AR. Pulmonary alveolar microlithiasis. Lung India 2006;23:42-4.
7. Tachibana T, Hagiwara K, Johkoh T. Pulmonary alveolar microlithiasis: Review and management. Curr Opin Pulm Med 2009;15:486-90.
8. Göcmen A, Toppare MF, Kiper N, Büyükpamukcu N. Treatment of pulmonary alveolar microlithiasis with diphosphonate: Preliminary results of a case. Respiration 1992;59:250-2.
9. 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.

How to cite this article: Gayathri Devi HJ, Mohan Rao KN, Prathima KM, Das JK. Pulmonary alveolar microlithiasis. Lung India 2011;28:139-41.

Source of Support: Nil, **Conflict of Interest:** None declared.

Announcement

"Quick Response Code" link for full text articles

The journal issue has a unique new feature for reaching to the journal's website without typing a single letter. Each article on its first page has a "Quick Response Code". Using any mobile or other hand-held device with camera and GPRS/other internet source, one can reach to the full text of that particular article on the journal's website. Start a QR-code reading software (see list of free applications from <http://tinyurl.com/yzlh2tc>) and point the camera to the QR-code printed in the journal. It will automatically take you to the HTML full text of that article. One can also use a desktop or laptop with web camera for similar functionality. See <http://tinyurl.com/2bw7fn3> or <http://tinyurl.com/3ysr3me> for the free applications.