

## Pulmonary Alveolar Microlithiasis

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### ABSTRACT:

*Pulmonary alveolar microlithiasis is a rare autosomal recessive ,diffuse lung disease characterised by bilateral filling of the pulmonary alveoli by numerous calcific concentrations known as calcispherites.*

*The pathogenesis involved in its mechanism is due to mutations in the gene encoding the solute carrier family 34 member 2 gene.*

*Although PAM can develop in children, the majority of patients are diagnosed in adulthood due to the slow progressive nature of this disease.*

*In children, majority of patients are asymptomatic and changes in the lung parenchyma are usually identified incidentally. Symptoms typically appear in the third or fourth decade of life. 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 may progress into pulmonary fibrosis, respiratory failure and cor pulmonale.*

*The radiological lesions in PAM closely resemble miliary tuberculosis. PAM should be suspected in cases when the radiological lesions do not correlate clinically.*

*A positive diagnosis of PAM is reached by the combination of a positive chest radiograph and histological examination.*

*This condition was first described in 1856 by Friederich as "Corpora-Amylacea in den lungen". The term "Micralithasis alveolaris pulmonum" was given by Puhr in 1933,*

*We report here a case of 45 year old Indian male who presented with complaints of shortness of breath on exertion and dry cough since 10 months*

### Key-words:

1. Pulmonary alveolar microlithiasis
2. Calcispherites
3. Solute carrier family 34 member 2
4. Microliths

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### I. Introduction

Pulmonary alveolar microlithiasis (PAM) is a rare hereditary lung disease in which calcium phosphate microliths, termed calcospherites, accumulate in the alveolar spaces. This was first brought into notice by Friedrich in 1856 and then by Harbitz in 1918

Most of the patients are asymptomatic at the time of diagnosis and it is usually incidental finding on radiographic examination of the chest for other purposes. The typical presentation on a chest X-ray is by a 'sandstorm' appearance. In contrast to the radiological findings, which are often severe, the clinical presentation of PAM is often relatively mild or absent. In symptomatic patients most common symptom is shortness of breath, followed by a dry cough, chest pain, hemoptysis.

There is no definitive treatment available to prevent the progression of PAM, although lung transplantation has been used to treat patients with end-stage and disodium etidronate has been administered due to its alleged calcium phosphate precipitation-reducing effect in PAM; however, its use is controversial.

## **II. Case History**

We report here a case of 45 year old Indian male who presented to OPD with complaints of shortness of breath on exertion and dry cough since 10 months. There was no history of fever, chest pain, hemoptysis, or weight loss. He was a non-smoker and had no pulmonary disease or significant family history.

Patient was then referred to Radiology department for Radiograph of chest, which revealed diffuse fine nodules in the lungs, mainly in the middle and lower zones.

Owing to symptoms of X- Ray , patient was then advised HRCT chest for further evaluation which revealed extensive areas of ground glass opacities with interspersed interlobular septal thickening involving bilateral lung parenchyma predominantly in lower lobes with sparing of thin peripheral subpleural rim of parenchyma. Multiple calcified nodules were noted in bilateral lower lobes and right middle lobe.

Extensive pleural calcifications were noted along the diaphragmatic and pericardial surface and also along the fissures.

The patient was then advised to undergo bronchoalveolar lavage(BAL) and lung biopsy, in which microliths were observed. The lavage fluid was not suggestive of tuberculosis or fungal infection. Transbronchial biopsy revealed concentric laminated microliths in the alveoli thus, confirming our final diagnosis of PAM.

## **III. Discussion**

PAM is a rare disease of unknown pathogenesis, characterized by the formation of microliths or calcispherites in the alveolar spaces with no underlying disorder of calcium metabolism. It is seen in both sexes with a slight predominance among males. It is seen in all age groups. It is present globally and >1,000 cases have been reported till date. The majority of patients are from Asia, followed by Europe . The country with the highest number of reported cases relative to the population is Turkey .The first case in India was reported by Viswanathan .

Familial incidence of PAM is 30–50%. The typical features of PAM on a plain chest radiographs are a fine, scattered micronodular pattern, producing a ‘sandstorm’ appearance or bilateral diffuse nodular lesions with translucent band in bilateral upper zone referred as (black pleura sign).

HRCT scan shows widespread intra-alveolar microliths, diffuse ground-glass attenuation areas, septal thickening, and black pleural lines - predominantly in the basal regions.

Diagnosis is confirmed by transbronchial biopsy.

There is no specific treatment available till date. Unlike pulmonary alveolar proteinosis broncho alveolar lavage of the whole lung is not effective for PAM. Disodium etidronate, which inhibits the micro-crystal growth of hydroxyapatite has been tried in some patients. Lung transplant may be needed for endstage disease.

### **DIFFERENTIAL DIGNOSIS-**

- Miliary tuberculosis

## **IV. Conclusion**

PAM is a rare disorder with an autosomal recessive trait with mutation of the SLC34A2 gene and should be considered as one of the possible differential diagnosis of nodular opacities on CXR. In a tuberculosis endemic country like India, there are chances for wrong treatment with ATT. Whenever possible a HRCT thorax should be performed.

A positive diagnosis should be considered in a patient presenting with the typical radiological features of the disease without clinical symptoms, particularly in the presence of a family history or consanguinity .



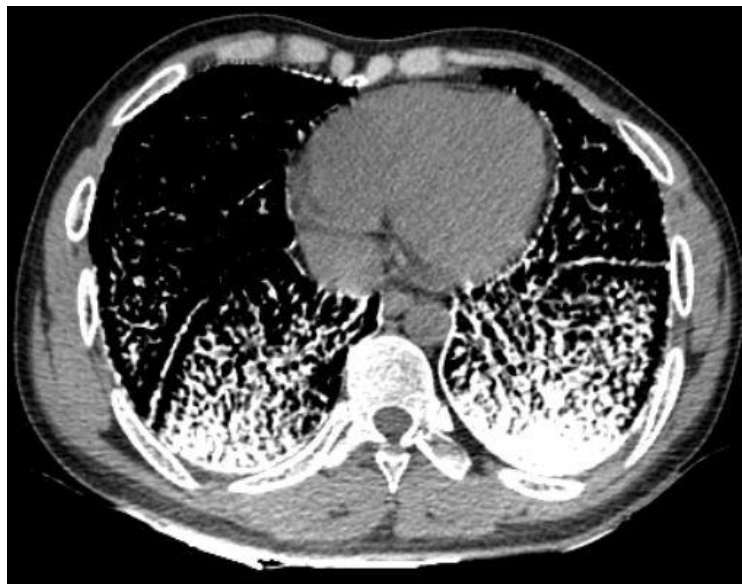
**Chest radiograph (posteroanterior view) shows innumerable widespread, small, dense nodules, diffusely involving both lungs-predominantly in the basal region.**



**HRCT chest showing extensive areas of ground glass opacities with interspersed interlobular septal thickening involving bilateral lung parenchyma predominantly in lower lobes with sparing of thin peripheral subpleural rim of parenchyma.**



**HRCT chest showing extensive areas of ground glass opacities with interspersed interlobular septal thickening involving bilateral lung parenchyma**



**Mediastinal window of the chest showing multiple calcified nodules in bilateral lower lobes and right middle lobe. Extensive pleural calcifications along the diaphragmatic and pericardial surface and along the fissures.**

### References

- [1]. <https://err.ersjournals.com/content/21/125/249>
- [2]. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5772952/>
- [3]. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3808952/>
- [4]. <https://www.hindawi.com/journals/crim/2010/819242/>
- [5]. [https://www.ijss-sn.com/uploads/2/0/1/5/20153321/ijss\\_apr\\_cr08.pdf](https://www.ijss-sn.com/uploads/2/0/1/5/20153321/ijss_apr_cr08.pdf)
- [6]. <http://www.jpgmonline.com/article.asp?issn=0022-3859;year=1982;volume=28;issue=4;spage=235;epage=7;aulast=Saigal>
- [7]. <http://www.lungindia.com/article.asp?issn=0970-2113;year=2013;volume=30;issue=2;spage=143;epage=147;aulast=Kashyap>

Dr. Nilesh Ingale. "Pulmonary Alveolar Microlithiasis." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 18, no. 7, 2019, pp 68-71.