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Pulmonary Alveolar Microlithiasis

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CLINICAL HISTORY:

34-year old male patient who is a paper mill worker came with complaints of:

- Cough with expectoration for 1 month
- Shortness of breath (Grade IV – MMRC) since 1 month
- Symptomatic since 2 years, increased since 1 month
- No h/o orthopnea, PND
- No h/o fever, chest pain, loss of appetite, weight loss, weakness

EXAMINATION AND INVESTIGATIONS:

Clubbing +

Cyanosis +

Lymphadenopathy absent

RR: 35cpm

SpO2: 66% RA, 88@5L of O2

RS: B/L Air Entry +

B/L NVBS +

B/L Basal crepts

CVS: S1, S2 +. No murmurs heard

PA & CNS: Normal.

CBC: Hb% – 20.2

RBC – 7.2

PCV – 60

Mantoux test – Negative

Sputum for AFB – Negative

ABG:

pH – 7.35;

pCO2 – 35 mmHg;

pO2 – 58 mmHg;

HCO3 – 23.7

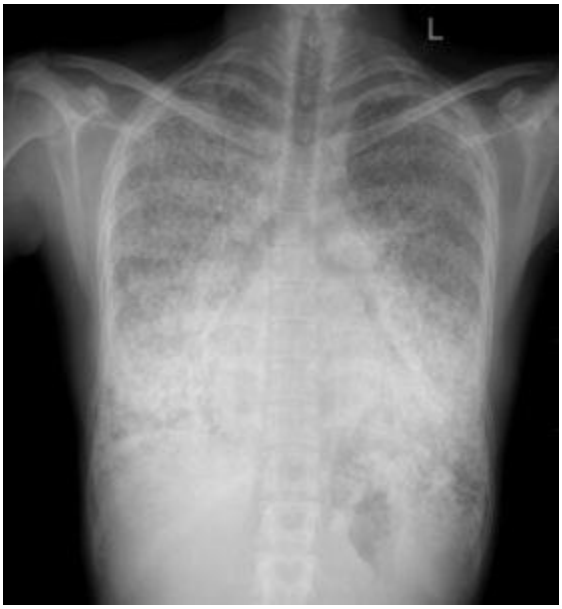
2D Echo –

RA, RV Dilated;

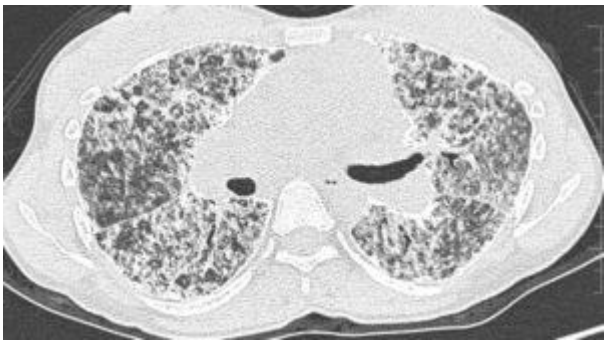
RVSP – 67mmHg indicative of pulmonary arterial hypertension.

PFT: Suggestive of Restrictive Defect.

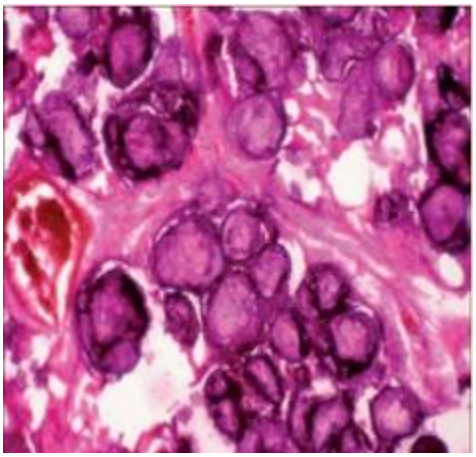
Bronchoscopic Lavage: Microliths +



CXR: Small nodules involving both the lungs.



HRCT Thorax: Widespread nodular intra-alveolar opacities of calcific densities + diffuse ground glass attenuation



Transbronchial biopsy: Concentric laminated microliths + thickened interstitial septa

FINAL DIAGNOSIS:

Pulmonary Alveolar Microlithiasis

Treatment Given:

Duolin Nebulization 8 hourly

O2 Supplementation – 2 lts / min

Advice on Discharge:

Home O2 2 Lts/min for 16 hrs -18 hrs / day

Deep Breathing exercises

DISCUSSION:

Pulmonary alveolar microlithiasis is a rare, chronic lung disease with bilateral intra-alveolar spherical calcium phosphate microliths (calcospherites).

Etiopathogenesis of PAM

- mutation in SLC34A2 gene that encodes a sodium-phosphate co-transporter in alveolar type II cells
- NPT2B is primarily on the surface of alveolar type II cells, absence of NPT2B results in increase in phosphate levels in the alveolar lining fluid leading to formation of complexes with calcium, resulting in formation of lamellated microliths. [1].
- Symptoms set in with advancement of the disease. – Non productive cough and dyspnea on exertion are the common symptoms.
- In the later course of the condition, respiratory insufficiency, cor pulmonale may occur or the condition may turn to be fatal. [1]

Radiology:

Classical example of clinical and radiological dissociation

Chest Radiograph:
Fine sand-like calcific micronodules also called ‘sandstorm lung’, diffusely involving both lungs, usually most marked in middle and lower zones

CT: Bilateral and symmetrical lung parenchymal abnormalities, usually as marked calcifications. The calcifications are most prominent in peripheral, mediastinal and fissural subpleural regions and each lobe is surrounded by a fine dense outline, giving the overall appearance of a stony lung. The black pleura sign is a feature described in PAM. A strip of peripheral lucency which is tangential is seen underlying the ribs while the adjacent lung shows diffuse dense calcifications. [2]

- PAM can be easily diagnosed by Bronchoalveolar lavage and sputum examination for the production of microliths but they are non-specific for the diagnosis of PAM.
- Confirmatory diagnosis can be established by open lung or transbronchial biopsy, showing intra- alveolar calcospherites, which in turn represent laminated calcium phosphate concentrations.

Apart from supportive care, there is no specific medical therapy for PAM, the only option is lung transplantation which can provide improvement in respiratory insufficiency. [1]

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