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Pulmonary alveolar microlithiasis at female patient

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Abstract

Pulmonary alveolar microlithiasis is a rare, autosomal recessive disease characterized by intra alveolar accumulation of spherical calcified calcium-phosphate crystals in the absence of calcium metabolism disorder. Most patients are asymptomatic, and the disease is discovered incidentally during a preventive examination. Symptoms appear most often in the 3rd-4th decade of life, but in general it is found in almost any age group. The youngest patient is an 8-month-old baby. Intra alveolar lamellar microlithiasis calcium deposits are found, most in the bases of the lung, and subsequently upwards towards the middle lobes. In the initial stage, deposits are in the middle of the alveolar space and do not interfere with air exchange. But in the last stages, the entire alveolar space is covered. The calcification itself begins to press the alveolar wall and damages it, leading to the formation of fibrous tissue.

Keywords: Pulmonary, microlithiasis, disease, asymptomatic, pulmonology

Introduction:

Pulmonary Alveolar Microlithiasis (PAM) represents an autosomal recessive genetic condition caused by a loss-of-function of the *SLC34A2* gene, which in turn causes deposition of miniscule calcium crystals, the so-called microliths. The disorder is rare with around 1100 cases having been reported worldwide. PAM was first described in 1686. [1]

At diagnosis, most patients are asymptomatic and changes in the lung are discovered incidentally. The most common symptoms are dyspnea, dry cough, chest pain, hemoptysis, asthenia. In some cases, pneumothorax may also be present.

Alveolar microlithiasis most likely begins to form from the beginning of childhood, but without any clinical symptoms, because a small part of the lung is involved. Later, when the disease progresses and a large part of the lung is involved, both mechanical ventilation and perfusion begin to be impaired. Then hypoxemia, increased amount of pCO2 in the blood, pulmonary hypertension and cor pulmonale are induced.

Other organs can also be affected - for example: the kidneys (medullary nephrocalcinosis, nephrolithiasis), the gall bladder (cholelithiasis), the cardiovascular system (aortic and/or mitral valve), etc. [2]

Pulmonary alveolar microlithiasis usually is diagnosed before age 40, there are rare cases like the one that will be presented after 60 years. Often the disorder is discovered when medical imaging is done for some other condition. The condition typically worsens slowly over many years, although some patients with this disease have signs and symptoms that remain stable for long periods of time.

Material:

For the purposes of the research, data were taken on patient diagnosed with Pulmonary alveolar microlithiasis from Public Hospital in Sofia, Bulgaria.

Case report:

A 67-year-old woman was admitted to the ER in the hospital with the following complaints: tightness in the chest, weakness and fatigue during physical exertion, dry irritating cough, and shortness of breath for 3-4 days.

From the status of the patient, it is seen that she is an adult woman in poor general condition, who needed medical help because of the cough and the shortness of breath from the previous days. She was oriented to place, time, and person. On her visible were her cyanotic skin and mucous membranes. Her tongue was dry and coated which can be seen on a lot of patients with dry irritating cough. Her chest was emphysematous and on percussion, sonorous tone was heard. The breathing of the patient was vesicular but weakened, exhalations were prolonged and on the base of the lungs there were small moist rales.

Her belly was soft, and there was not any swelling on her limbs.

Spirometry was also made: The FVC was decreased as it is on most of the patients with pulmonary alveolar microlithiasis whose disease is not in the initial phase.

Laboratory blood test was done and out of normal ranges were the: Leucocytes: (H), Lymphocytes: (L), Granulocytes: (H), Lymphocytes %: (L), Granulocytes %: (H), Serum Creatinine:(L)

Investigations for confirming the diagnosis:

Blood test results:
Leucocytes: 11.4 (H),

Lymphocytes: 1.2 (L),

Monocytes: 0.8,

Granulocytes: 9.4 (H),

Lymphocytes %: 10.6% (L),

Monocytes %: 7.1%,

Granulocytes %: 82.3% (H),

Hemoglobin: 132.0,

Erythrocytes: 4.75,

Hematocrit: 40.6,

Thrombocytes: 252.0,

Serum Glucose: 5.6,

Potassium: 5.0

Serum Creatinine: 34.3(L)

Sputum – Normal

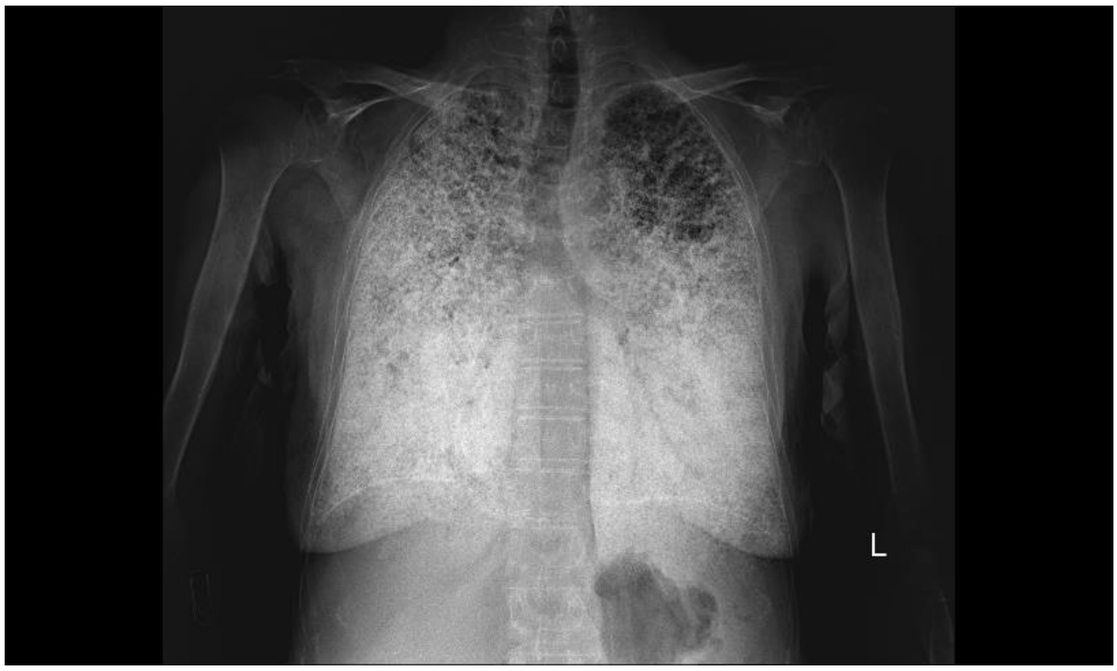
pH: 7.4,

pCO2: 38.4

pO2: 64.8 (L)

ECG- Sinus rhythm

Chest x-ray:



Treatment:

- Levoxa 500mg, Urbason, Novphyllin, Bromhexine
- Metoprolol
- Oxygen Therapy
- Inhalations

After 1 week in the hospital, the patient was discharged in a better general condition compared to the condition, she came into the hospital.

In-house referrals:

- Symbicort 160/4.5 inhalations 2x2
- Spiriva Respimat 2.5 microgram inhalation solution
- Ventolin Solution (Per need)
- Home Oxygen Therapy (more than 16 hours per day, mandatory when sleeping)

The patient was told to come for a follow-up appointment in two weeks, but she did not attend the scheduled appointment.

Conclusion:

Pulmonary alveolar microlithiasis is very rare lung disease, the diagnosis usually is made when imagine examination for some other condition is made. The patients usually are younger than 40 years. It is rare to be diagnosed in older patient.

Effective medical therapy for this condition is not reported yet. The prognosis is poor, and the cause of death usually is respiratory failure.

The patients with Pulmonary alveolar microlithiasis need lung transplantation at some stage of the disease.

References:

[1] Jönsson ÅLM, Bendstrup E, Mogensen S, Kopras EJ, McCormack FX, Campo I, et al. Eight novel variants in the *SLC34A2* gene in pulmonary alveolar microlithiasis [Internet]. Eur Respir J. 2020. Feb;55(2):1–11. 10.1183/13993003.00806-2019

[2] Castellana G, Lamorgese V. Pulmonary alveolar microlithiasis. World cases and review of the literature. Respiration. 2003;70:549–55.