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Clinical image

Pulmonary Alveolar Microlithiasis: Case Report

Microlitiasis alveolar pulmonar: reporte de un caso

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Pulmonary alveolar microlithiasis (PAM) is a rare condition characterized by the presence of microscopic calcium phosphate granules at the alveolar level, known as microliths or calcospherites. It is an autosomal recessive congenital disease caused by a mutation in the SLC34A2 gene, which encodes the type IIB sodium-phosphate cotransporter, expressed in type II alveolar cells. From an epidemiological perspective, most cases are concentrated in Asia (56.3%) and Europe (27.8%), with Turkey, China, Japan, India, and Italy being the most affected countries. In Spain, as of 2017, 38 cases had been reported, representing 3.7% of the total cases worldwide.

We present the case of a 52-year-old man from Morocco who sought medical attention in 2016 due to a one-week history of cough with hemoptysis and dyspnea. A chest X-ray showed bilateral micronodular opacities with blurring of the costophrenic and cardiophrenic angles, initially suggesting miliary tuberculosis. On physical examination, the patient was eupneic while breathing ambient air, with an oxygen saturation between 96 and 98%. Pulmonary auscultation revealed fine bilateral crackles, predominantly at the lung bases.

As part of the diagnostic workup, microbiological tests were performed, ruling out *Mycobacterium tuberculosis*. Chest computed tomography revealed extensive, diffuse bilateral lung parenchymal involvement with multiple calcified pulmonary nodules and confluent calcified areas with air bronchograms, primarily in the posteroinferior regions of both lower lobes, the paracardiac areas, and both upper lobes. These findings were highly suggestive of pulmonary alveolar microlithiasis, prompting a transbronchial cryobiopsy. Pathological analysis revealed alveoli filled with laminated calcified bodies with lateral striations, confirming the diagnosis (Fig. 1b).

After nine years of follow-up, the patient has developed digital clubbing (Fig. 1c) and progressive respiratory decline, exhibiting a restrictive ventilatory pattern, reduced carbon monoxide transfer capacity (DLCO), resting hypoxemia, and significant desaturation during exertion. Consequently, portable oxygen therapy has been prescribed for ambulation.

PAM was first described in 1686 by the Italian scientist Marcello Malpighi. Although it can affect both sexes, it predominantly occurs in males, except in Spain, Italy, and France, where incidence is higher in women. The disease is typically diagnosed between the second and third decades of life, though cases have been reported in newborns with fatal outcomes within hours.[1]

In the early stages, patients may be asymptomatic, with symptoms developing slowly and progressively. The most common symptoms include exertional dyspnea and dry cough, although asthenia, chest pain, and hemoptysis have also been reported. On physical examination, crackles and digital clubbing may be observed, as in our case.

The diagnosis can be confirmed through transbronchial cryobiopsy, although in the presence of typical radiological findings, it is often unnecessary. Recently, thoracic ultrasound has been proposed as a diagnostic tool, as it reveals characteristic findings in PAM patients, such as pleural thickening, hyperechoic irregularities, and echogenic foci without prominent acoustic shadows in subpleural and basal regions (the “comet tail” phenomenon).[2]

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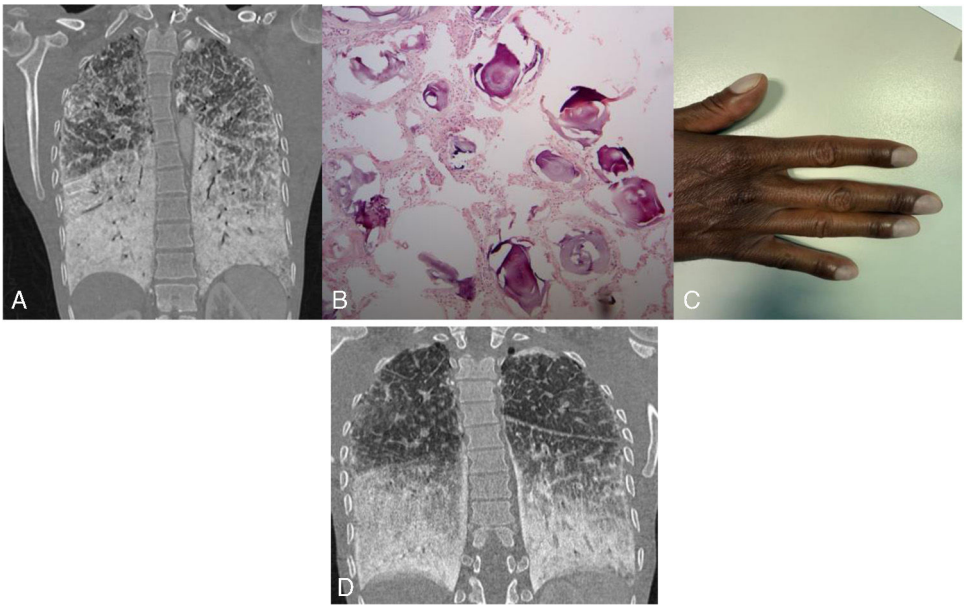

Fig. 1. (A) CT coronal section 2025: extensive bilateral and diffuse pulmonary parenchymal involvement due to the presence of multiple calcified pulmonary nodules and large confluent areas of consolidation with air bronchogram in the upper lobes, middle lobe, and lower lobes. A "crazy paving" pattern is observed in the remaining parenchyma, along with calcification of the interlobular septa and fissures. (B) Histological section of cryobiopsy: in the histological section, alveoli filled with spherical calcified bodies are observed, some with concentric psammomatous calcifications. (C) Clubbing: clubbing secondary to lung disease is evident. (D) CT coronal section 2016: shows extensive bilateral and diffuse pulmonary parenchymal involvement due to the presence of multiple calcified pulmonary nodules and large confluent areas of consolidation to a lesser extent than in 2025.

From a functional perspective, initial studies are often normal. However, as the disease progresses, a reduction in DLCO and the development of a restrictive ventilatory disorder are observed.[2]

The differential diagnosis includes diseases associated with pulmonary calcifications and dense or ground-glass micronodular opacities, such as miliary tuberculosis, pulmonary alveolar proteinosis, sarcoidosis, varicella pneumonia, metastatic calcification, and pneumoconioses (e.g., silicosis), among others.[2]

Declaration of generative AI and AI-assisted technologies in the writing process

The authors declare that artificial intelligence (ChatGPT) was used in the preparation of this manuscript solely for assistance grammatical review, and text translation. All ideas, analyses, interpretations, and conclusions presented in this work are the exclusive intellectual effort of the authors.

The accuracy of the AI-generated information has been carefully verified to ensure its reliability and consistency with the available scientific evidence. The authors take full responsibility for the content of the manuscript and its originality.

Informed consent

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Authors' contributions

All authors have contributed significantly to the research and preparation, revision and final production of the manuscript and approve its submission.

Conflicts of interest

The authors declare to have no conflict of interest directly or indirectly related to the manuscript contents.

References

1. Cuesta Lujano L, Gutiérrez Domingo Á, Fernández Ollero L. Microlitiasis alveolar y su peculiar disociación clínico-radiológica. Arch Bronconeumol. 2018;54:493–4, http://dx.doi.org/10.1016/j.arbres.2017.12.012.
2. Gaudó Navarro J, Quezada Loaiza CA, Jiménez Castro D. Microlitiasis alveolar. In: Manual de Neumología y Cirugía Torácica. 4ª ed. Madrid: Editorial SEPAR; 1998 [capítulo 46].