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Pulmonary Alveolar Microlithiasis Complicated by Pulmonary Hypertension: A Case Report

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Abstract **Case Report**

Pulmonary alveolar microlithiasis (PAM) is an ultra-rare diffuse lung disease caused by biallelic pathogenic variants in the SLC34A2 gene, resulting in defective phosphate clearance by type II pneumocytes and progressive intra-alveolar deposition of calcium phosphate microliths. The disease classically evolves with clinico-radiological dissociation: radiographic abnormalities may be striking long before severe respiratory symptoms appear. We report the case of a 42-year-old Moroccan man with diabetes who presented with gradually progressive exertional dyspnea over several months. Chest radiography revealed diffuse bilateral micronodular opacities resembling a sandstorm pattern. Contrast-enhanced chest computed tomography demonstrated innumerable bilateral calcified micronodules with predominantly subpleural and peribronchovascular distribution, associated with bilateral subpleural cystic lesions and paraseptal emphysema. The examination also showed dilation of the main pulmonary artery, enlargement of the right-sided cardiac chambers, a right-to-left ventricular ratio greater than 1, and moderate pericardial effusion, raising strong suspicion for pulmonary hypertension complicating advanced PAM. This report highlights the decisive role of thoracic imaging in suggesting the diagnosis, especially in settings where genetic or histopathological confirmation may not be immediately available. The discussion places the present case in the context of the largest available reviews and recent case reports, with emphasis on pathophysiology, characteristic imaging findings, functional decline, major complications such as pulmonary hypertension and pneumothorax, differential diagnosis, and current treatment limitations.

Keywords: pulmonary alveolar microlithiasis; sandstorm lung; chest CT; pulmonary hypertension; subpleural cysts; case report.

INTRODUCTION

Pulmonary alveolar microlithiasis is a rare hereditary lung disease characterized by diffuse deposition of minute calcium phosphate concretions within the alveolar spaces [1,9-11]. Although described by Puhr in 1933, PAM remains unfamiliar to many clinicians because of its very low prevalence and frequently indolent course [1]. The most comprehensive review published to date analyzed 1,022 cases reported worldwide and confirmed the rarity of the condition, its familial clustering in a substantial proportion of patients, and its highly variable clinical course [3].

The modern understanding of PAM changed after the identification of pathogenic variants in SLC34A2, the gene encoding the type IIb sodium-phosphate cotransporter expressed in alveolar type II cells [9-11]. Under normal conditions, this transporter contributes to phosphate recycling generated during surfactant turnover. When transporter function is lost, phosphate accumulates in the alveolar lining fluid and combines with calcium to form hydroxyapatite microliths [9-11]. This mechanism explains both the progressive mineralization of the lungs and the absence of a clearly effective anti-inflammatory or anti-infectious treatment.

A hallmark of PAM is the striking mismatch between symptoms and imaging. Many patients remain asymptomatic or only mildly symptomatic for years despite impressive radiographic abnormalities, which is why the disease is often discovered incidentally or during evaluation for unrelated respiratory complaints [1,3,7,8]. Over time, however, microlith accumulation, interstitial remodeling, impaired diffusion, and pulmonary vascular consequences may lead to exercise limitation, chronic

respiratory insufficiency, pulmonary hypertension, and cor pulmonale [3-6,8].

Imaging is central to diagnosis. Chest radiography typically shows diffuse, bilateral, fine calcific micronodules giving the classic sandstorm appearance, sometimes with obscuration of cardiac or diaphragmatic contours in advanced disease [1-3,7,8]. High-resolution or thin-section CT further refines the diagnosis by demonstrating the calcified nature of the nodules, subpleural and peribronchovascular predominance, septal calcification, dense consolidative changes in advanced stages, and associated subpleural cysts that may predispose to spontaneous pneumothorax [3,6,8].

CASE PRESENTATION

A 42-year-old Moroccan man, known to have diabetes mellitus, was admitted for progressively worsening exertional dyspnea evolving over several months. There was no reported history of previous tuberculosis, no relevant occupational or environmental exposure, and no known familial history of chronic interstitial or calcific lung disease. The patient did not describe chest pain, hemoptysis, recent infectious syndrome, or acute pleuritic symptoms. At presentation, the dominant complaint was limitation during physical effort, suggesting a chronic and slowly progressive respiratory process rather than an acute pulmonary event.

The initial chest radiograph demonstrated diffuse, bilateral, fine micronodular opacities arranged in clusters, producing a pattern reminiscent of a sandstorm. This appearance immediately raised the possibility of a diffuse micronodular lung disease; however, the density and extensive bilateral distribution strongly suggested a calcific process rather than purely inflammatory or infectious miliary dissemination.

A contrast-enhanced chest CT scan was then performed for lesion characterization. The examination showed innumerable diffuse calcified micronodules involving both lungs. Their distribution was predominantly subpleural and peribronchovascular. The calcific micronodular burden was bilateral and extensive, yielding the characteristic tomographic correlate of the sandstorm pattern described in PAM [2,3,7,8] (figure 1 and 2).

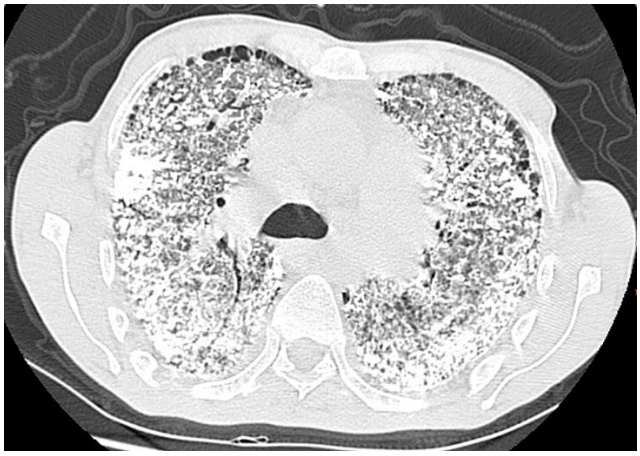

Figure 1: Chest CT scan parenchymal window with Typical “sandstorm” appearance

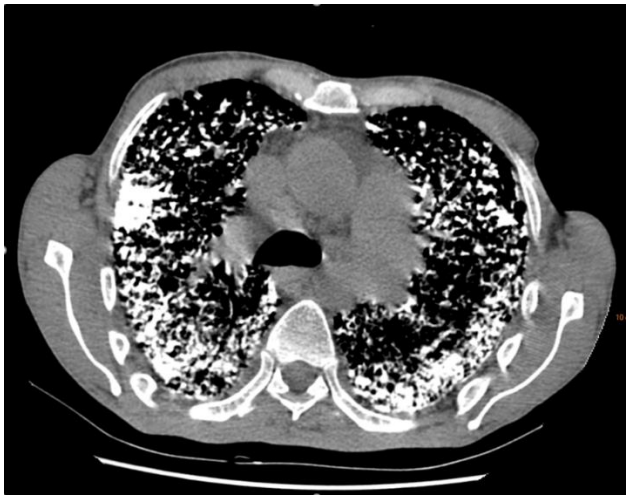

Figure 2: Chest CT scan in mediastinal window showing microdular, subpleural and septal calcifications

In addition to the diffuse calcified parenchymal lesions, CT revealed bilateral subpleural cystic changes and associated paraseptal emphysema. In the context of PAM, subpleural cysts are clinically relevant because they reflect chronic peripheral parenchymal remodeling and may create a substrate for spontaneous pneumothorax, a complication reported in advanced disease and specifically emphasized in recent case literature from Morocco [6].

The cardiovascular component of the CT examination was also notable. The main pulmonary artery measured 38 mm, while the right and left pulmonary arteries measured 29 mm and 27 mm, respectively. The pulmonary artery-to-aorta ratio was greater than 1 (figure 3). Furthermore, the right ventricle appeared enlarged with a right ventricle/left ventricle ratio greater than 1. Cardiomegaly was present predominantly at the expense of the right-sided chambers, and a moderate pericardial effusion was identified. Taken together, these findings strongly favored associated pulmonary hypertension with right heart strain in the setting of advanced chronic parenchymal lung disease [5,8].

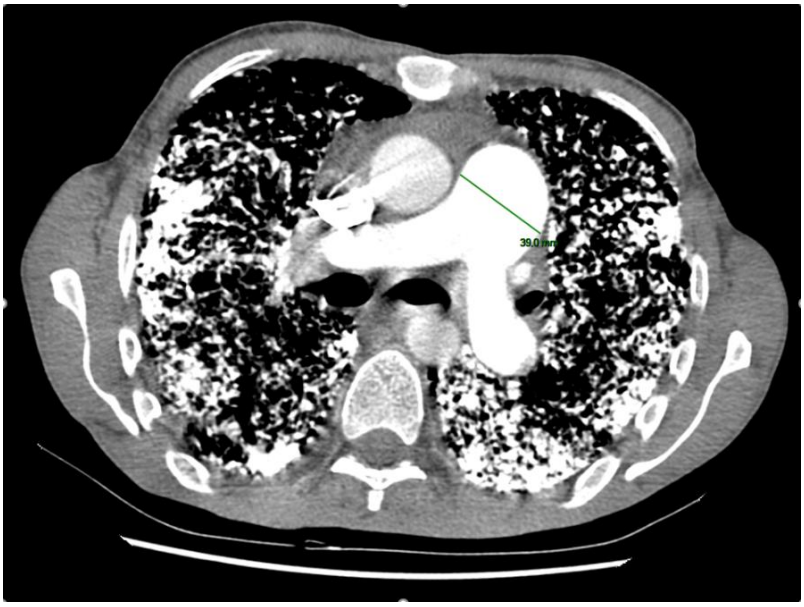

Figure 3: contrast-enhanced chest CT scan showing dilation of the main pulmonary artery

No mediastinal or axillary lymphadenopathy was detected. The absence of significant thoracic adenopathy, combined with the diffuse calcified micronodular pattern and subpleural cystic changes, made diagnoses such as sarcoidosis or infectious granulomatous disease less likely.

Based on the overall imaging pattern, the diagnosis most strongly suggested was pulmonary alveolar microlithiasis complicated by pulmonary hypertension. Even in the absence of immediate genetic testing or lung biopsy, the imaging constellation was highly characteristic and sufficiently specific to orient diagnostic reasoning toward PAM [1,3,8].

DISCUSSION

PAM is now recognized as a monogenic disorder of phosphate handling within the distal airspaces. The identification of SLC34A2 as the causal gene was a major advance because it provided a unifying explanation for the microliths seen pathologically and radiologically [9,10]. Loss of function of the sodium-phosphate cotransporter impairs phosphate reuptake by alveolar type II cells, leading to progressive precipitation of calcium phosphate within alveoli. This mechanism aligns with the largely intra-alveolar location of the microliths and the tendency for slow but relentless mineral accumulation over years [1,9-11].

From an epidemiological standpoint, the 2015 review by Castellana et al. remains the cornerstone reference [3]. That review emphasized the geographic clustering of cases, the importance of familial occurrence, and the broad spectrum of clinical severity, ranging from incidental discovery to progressive respiratory disability. The same review also reinforced the concept that imaging, especially CT, often provides near-pathognomonic clues when the disease is sufficiently advanced [3]. More recent overviews, including the European Respiratory Review article by Kosciuk et al., have refined current knowledge on genetics, biomarkers, and disease monitoring, but they continue to confirm the centrality of the classic imaging phenotype [11].

The present case fits the classic radiological framework of PAM. The chest X-ray demonstrated diffuse sandstorm-like opacities, and CT revealed innumerable calcified micronodules with subpleural predominance. Similar findings were described by Madhala et al. in 2022, who reported a young man with long-standing dyspnea and characteristic bilateral

sandstorm opacities on radiography and CT [2]. Chu et al. likewise highlighted the typical clinico-radiological dissociation in a case labeled 'stone lungs,' underscoring that dramatic radiographic calcification can coexist with symptoms that are initially less severe than the images might suggest [8].

An important feature in our patient was the presence of subpleural cystic lesions. These lesions are well recognized in PAM and may reflect chronic alveolar destruction, interstitial remodeling, and mechanical fragility of peripheral lung tissue [3,6,8]. Their importance is not only descriptive: they have direct prognostic implications because they increase the likelihood of pleural complications. The recent case by Oujaber et al., reported in 2026, is particularly relevant because spontaneous pneumothorax was the revealing event in a Moroccan patient with PAM [6]. The resemblance between that report and the present case lies in the presence of peripheral cystic changes, although our patient had not yet presented with pneumothorax.

The signs of pulmonary hypertension in this case deserve special emphasis. Enlargement of the main pulmonary artery, pulmonary artery-to-aorta ratio greater than 1, right ventricular enlargement, right-sided cardiomegaly, and associated pericardial effusion together support advanced cardiopulmonary involvement. Pulmonary hypertension in PAM is considered a late and ominous complication resulting from chronic hypoxemia, parenchymal destruction, vascular remodeling, and increased pulmonary vascular resistance [3,5,8]. Shyllesh et al. described a 2025 case in which severe pulmonary hypertension coexisted with rheumatic heart disease, demonstrating how PAM can participate in complex hemodynamic deterioration [5]. Even when no second cardiac disorder is present, pulmonary hypertension may emerge as the dominant determinant of functional decline and prognosis.

The functional trajectory of PAM is variable but usually unfavorable over the long term. Mari et al. drew attention in 2024 to measurable decline in lung function over time, reinforcing the need for serial monitoring rather than simple static diagnosis [4]. Although some patients remain stable for prolonged periods, others progress to restrictive ventilatory impairment, diffusion limitation, chronic hypoxemic respiratory failure, and eventual need for advanced respiratory support or transplantation [3,4,11]. The current patient already demonstrates imaging signs of advanced disease, which is clinically relevant even in the absence of full pulmonary function testing data in the present file.

The differential diagnosis of diffuse micronodular pulmonary opacity depends heavily on whether calcification is evident. Miliary tuberculosis, fungal dissemination, healed varicella pneumonia, sarcoidosis, pulmonary alveolar proteinosis, talcosis, hemosiderosis, and metastatic pulmonary calcification may all enter the discussion depending on context [3,8,11]. However, PAM becomes the leading hypothesis when the nodules are innumerable, bilateral, calcified, relatively symmetric, and associated with subpleural cysts and dense peripheral involvement. The absence of significant adenopathy in our patient also argues against some inflammatory and granulomatous alternatives.

Histological confirmation is traditionally possible by transbronchial or surgical biopsy, which reveals concentrically laminated calcospherites filling alveolar spaces [1,3,11]. Genetic confirmation through SLC34A2 testing is increasingly desirable because it establishes the molecular diagnosis and may enable family counseling [9,10]. In practice, however, the imaging appearance may be so characteristic that clinicians can reach a high-confidence diagnosis before invasive confirmation, especially in resource-limited settings. This is particularly true when chest radiography and CT findings are concordant, as in the present observation.

Therapeutic options remain limited. No medical treatment has consistently altered the natural history of PAM. Corticosteroids, serial bronchoalveolar lavage, calcium-chelating strategies, and bisphosphonates such as disodium etidronate have been attempted, but evidence remains inconsistent and insufficient to recommend them as standard therapy [3,11]. Supportive care therefore focuses on symptom relief, oxygen therapy when indicated, vaccination, management of respiratory infections, surveillance for pulmonary hypertension, and careful follow-up of pleural complications.

For end-stage disease, lung transplantation remains the only established definitive treatment [3,11]. Published reports suggest that transplantation can be successful and recurrence in the graft is not a major issue because the fundamental defect is localized to the native pulmonary epithelium rather than being driven by a circulating factor [11]. This therapeutic reality makes early recognition important: once signs of significant pulmonary hypertension or chronic respiratory insufficiency appear, referral to specialized centers should be considered sooner rather than later.

The Moroccan origin of the present patient adds contextual value. The recent report by Oujaber et al. described what the authors considered the 15th reported Moroccan case [6], underscoring that PAM remains exceptionally uncommon in the country. Publishing additional well-documented North African cases is therefore useful not only for educational reasons but also for expanding the regional literature on presentation patterns, complications, and imaging phenotypes.

Overall, the present case illustrates several features that make PAM particularly instructive for radiologists and pulmonologists: a classic sandstorm

pattern on chest radiography, diffuse calcified micronodules on CT, subpleural cystic remodeling, and radiological evidence of pulmonary hypertension. The case also exemplifies why PAM should be considered whenever a diffuse bilateral micronodular pattern appears unusually dense or calcific, especially when the clinical history lacks a convincing infectious or occupational explanation.

CONCLUSIONS

Pulmonary alveolar microlithiasis is a rare but highly characteristic diffuse lung disease in which radiology often drives the diagnosis. In our patient, the association of diffuse bilateral calcified micronodules, sandstorm chest radiograph, subpleural cystic lesions, and CT signs of pulmonary hypertension strongly supported the diagnosis of advanced PAM.

This case reinforces three practical messages. First, CT can be decisive when the calcific nature and distribution of lesions are recognized. Second, pulmonary hypertension should be actively sought because it marks advanced disease and worsens prognosis. Third, although pharmacological treatment remains unsatisfactory, early identification is still valuable because it guides surveillance, counseling, and timely referral for specialized management including transplant evaluation when appropriate.

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