Original Article

Pulmonary alveolar microlithiasis: high-resolution computed tomography findings in 10 patients*

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Abstract

Objective: To present the high-resolution computed tomography (HRCT) findings of pulmonary alveolar microlithiasis. **Methods:** The HRCT scans of 10 adult patients (seven females and three males; mean age, 38.7 years) were retrospectively analyzed. The films were studied independently by two radiologists. **Results:** The most common tomographic findings were ground-glass attenuation and linear subpleural calcifications, which were seen in 90% of the patients. Other relevant findings were small parenchymal nodules, calcification along the interlobular septa, nodular cissures, subpleural nodules, subpleural cysts, dense consolidations, and a mosaic pattern of attenuation. **Conclusions:** The HRCT findings presented by individuals with pulmonary alveolar microlithiasis are distinct. In most cases, such findings can form the basis of the diagnosis, eliminating the need to perform a lung biopsy.

Keywords: Tomography, X-Ray Computed; Lithiasis/lung; Lung diseases.

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Introduction

Pulmonary alveolar microlithiasis (PAM) is a rare, familial disease of chronic evolution that affects young adults and generates special interest in radiological studies due to its pathognomonic imaging pattern, which, in the opinion of many authors, precludes the need for a lung biopsy in most cases.[1-5]

More than half of all patients are asymptomatic at the time of diagnosis.[5] The disease remains silent for a long period, with subsequent occurrence of dyspnea, chest pain, and dry cough, and can result in cyanosis, respiratory failure, and cor pulmonale.[2,4,5] The scarcity of clinical symptoms contrasts with the extensive alterations seen on chest X-rays.[1,4] In most patients, the disease is diagnosed in routine examinations, although the diagnosis can be made during the investigation of the family history in an individual with alveolar microlithiasis.[5] High-resolution computed tomography (HRCT) has helped to characterize the imaging of the disease and can be very useful in the diagnosis of PAM.[1-4,6,7]

The objective of this study was to describe the alterations identified in the tomography scans of 10 patients with PAM.

Methods

In this study, the chest tomography scans of ten patients with PAM, from seven different institutions, located in four states of the federation of Brazil (Rio de Janeiro, São Paulo, Goiás, and Paraná), were retrospectively analyzed. Of these patients, seven were female and three were male. Patient ages ranged from 22 to 59 years (mean, 38.7 years).

The diagnosis was based on the patterns observed on X-rays and tomography scans, which are considered pathognomonic of the disease.[1,3-5] However, six patients were also submitted to open lung biopsies, which allowed histopathological confirmation.

Tomography scans of the chest were performed in various tomography scanners, axial slices ranging from 1 to 2 mm in thickness, in 10-mm increments, performed during a deep inspiration, from the apices to the lung bases. Tests were performed using a parenchymal window, with a width of 1000-1500 Hounsfield Units (HU) and a center between −650 and −750 HU. Tests were also performed using a mediastinal window, with a width of 350-400 HU and a center between 40 and 60 HU.

The HRCT analysis was performed by two independent observers, and discordant results were resolved by consensus. The study included the assessment of the lung parenchyma, regarding small parenchymal nodules, ground-glass opacities, mosaic pattern of attenuation, and calcification along the interlobular septa, subpleural linear calcifications, subpleural nodules, nodular fissures, dense consolidation, subpleural cysts, and apical bullae. Criteria for definition of these findings are those indicated in the Fleischner Society Glossary of Terms.[8] The terminology used is that which is recommended by the Terminology Consensuses of the Brazilian College of Radiology and the Brazilian Thoracic Society.[9,10]

Small parenchymal nodules were characterized as round focal opacities of less than 10 mm in diameter.

Ground-glass opacity was defined as greater pulmonary attenuation, albeit with visible bronchial or vascular margins. Consolidations were defined as a homogeneous increase in the attenuation coefficient of the lung parenchyma, leading to the loss of the individualization of the vascular structures and of the airway wall. Consolidations were designated dense when they presented density greater than that of the soft tissues, when assessed at the mediastinal window. A mosaic pattern of attenuation was defined as areas of ground-glass opacity with thickening of interposed interlobular septa.

Calcification along the interlobular septa was characterized when thin linear opacities, designing the periphery of secondary pulmonary lobes, were visualized using mediastinal windows.

Subpleural linear calcifications were thus described in patients who presented calcified, juxtapleural, continuous linear opacities, also visible at the mediastinal window.

Subpleural cysts were defined as round hypodense structures, with a diameter of less than 10 mm, aligned next to the pleura, whereas subpleural nodules were defined as nodular opacities along the pleural surface, and nodular fissures were defined as nodules distributed along the fissure lines. Apical bullae were defined as well-delineated thin-walled >10-mm air spaces observed in the lung apices.

Results

The most common alterations revealed in the assessment of tomography scans of the chest of the ten patients diagnosed with PAM were ground-glass opacities and subpleural linear calcifications, observed in nine patients each (90%).

Small parenchymal nodules (Figure 1) were found in seven patients (70%).

Calcification along the interlobular septa was found in five patients (50%). Of these, four also presented associated ground-glass attenuation, configuring the mosaic pattern of attenuation (Figure 2).

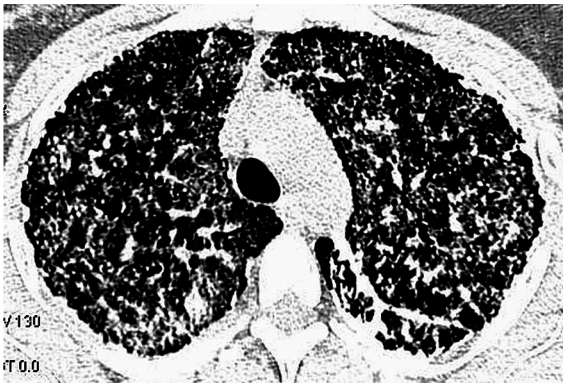

Figure 1 - Lung window slice showing small disseminated nodules and ground-glass opacities. Note also the nodulations along the pleural surface.

Dense consolidation (Figure 3), subpleural nodules, nodular fissure and subpleural cysts (Figure 4) were observed in five cases each (50%). Apical bullae were identified in only one patient (10%).

Tomographic alterations were predominant in the inferior and posterior portions of the lungs, and were found in nine patients (90%). Apical predominance occurred in only one of the cases studied. These findings are represented in Table 1.

Discussion

A rare disease, PAM presents chronic evolution, poorly defined etiology and pathogenesis, basically characterized by numerous small calculi (denominated calciferites, calcospherites or microlites) within air spaces.[1-3,5] In the literature, studies involving larger patient samples have found no predominance of gender.[5] In the present study, seven of the ten patients analyzed were female. Age bracket varies considerably, from newborns to patients in the ninth decade of life, the mean age being 35 years at the time of diagnosis.[4,5,11] In the present study, ages ranged from 22 to 59 years (mean, 38.7 years).

The disease presents a high incidence of familial occurrence (approximately one-third of the cases), suggesting an autosomal recessive inheritance pattern.[1-3,5] In this patient sample, there were two cases of monozygotic twins. However, the families of the other patients were not adequately investigated in order to exclude other familial cases.

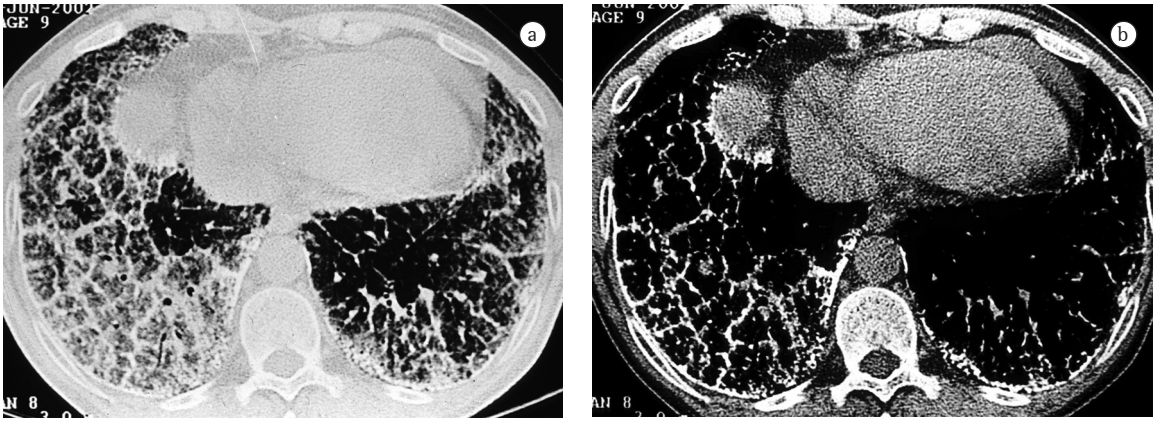

Figure 2- a) Intermediate window slice revealing ground-glass opacities with thickening of interposed interlobular septa, characterizing a mosaic pattern of attenuation; and b) Mediastinal window slice showing calcification along the interlobular septa and subpleural regions.

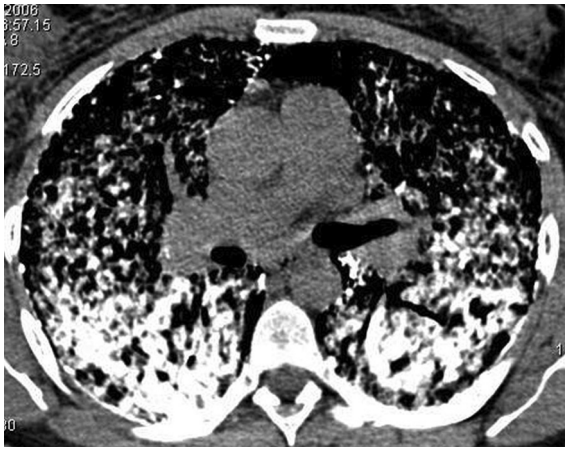

Figure 3 - Mediastinal window slice showing a confluence of nodules forming consolidations with calcium density, predominant in the posterior regions of the lungs. Note also the calcified nodules along the pleural surface.

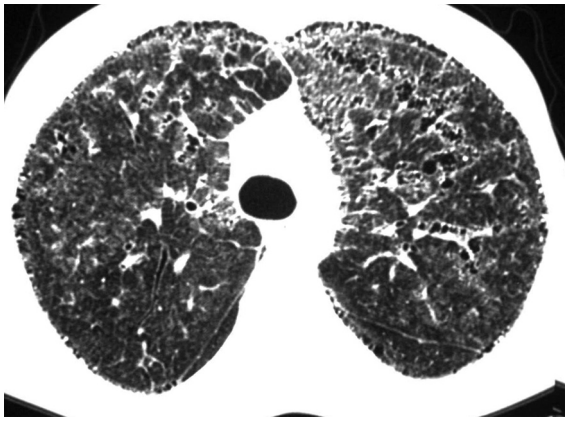

Figure 4 - Lung window slice showing ground-glass opacities and thickening of interlobular septa. Note also the cysts and subpleural nodules.

Table 1 - Distribution of tomographic findings.

	Number of cases	Percentage (%)
Ground-glass opacities	9	90
Linear subpleural calcifications	9	90
Small parenchymal nodules	7	70
Subpleural nodules	5	50
Nodular fissure	5	50
Calcification along the interlobular septa	5	50
Dense consolidations	5	50
Subpleural cysts	5	50
Mosaic pattern	4	40
Apical bullae	1	10

The HRCT findings in patients with alveolar microlithiasis vary considerably, and most publications refer to reports of isolated cases.

Ground-glass opacities are a common finding in the literature.[1,3,4,6,7,11,12] This was the major finding in this sample and was present in 90% of the cases. Ground-glass attenuation is probably due to small calculi in the air space.

Subpleural linear calcifications were also found in 90% of the patients analyzed in this study. This finding, although commonly seen in tomography scans of patients with PAM, is controversial in the literature. Although some authors describe them as pleural calcifications,[13,14] no research has reported the histopathological confirmation of pleural calcification. Therefore, the best explanation seems to be the accumulation of intra-alveolar calculi in the periphery of secondary pulmonary lobes, demarcating the pleural surface, and producing the aspect of pseudo-pleural calcifications.[4,11,15-17]

The calcification aspect along the interlobular septa was also a common point shared by this study and the data in the literature. This aspect was observed in five patients, corresponding to 50% of the sample. The calcium aspect of the septa also occurs due to the accumulation of calculi in the periphery of secondary pulmonary lobes, since there is no histopathological confirmation of calcifications in the interlobular septa or of interstitial involvement in the initial phases of the disease.[4,7,11]

The mosaic pattern of attenuation occurred in four patients, which represents 40% of our sample. In this study, this aspect can also be observed in the mediastinal window images, since there is calcification along the interlobular septa. There is no report in the literature of any other disease with similar tomographic manifestation. This finding is considered very specific and even pathognomonic of PAM on the HRCT scan.[7]

Small nodules were identified in 70% of the patients. The small nodules seen on the HRCT scan correspond to dense micronodules (<1 mm), on chest X-rays and are very thin, well-defined, and diffusely spread throughout the parenchyma, making the lungs homogeneously hypotransparent, character-

izing the pattern described by some authors as a 'sandstorm' aspect, which is considered typical of the disease. When this pattern is identified in chest X-rays, lung biopsy is needed only in exceptional cases.[1-4] On the HRCT scan, it is often impossible to define the calcium density of the nodules. This is probably due to their small dimensions. When they converge, calcium can be better characterized.

Converging small nodules can form areas of parenchymal consolidation. Due to the presence of calcium, consolidations can have greater density than that of the soft tissues. These consolidations predominantly occur along the heart borders in the lower posterior region of the lungs and tend to be symmetrical.[1,2,4,6,7,11,12] Dense consolidations containing interposed air bronchograms were found in 50% of the patients studied, especially in the lower posterior regions. The literature hardly mentions this finding.[6] Therefore, PAM should be considered a differential diagnosis for dense consolidation, together with pulmonary toxicity by amiodarone, metastatic pulmonary calcification, silicoproteinosis, talcosis and amiloidosis.[18]

Another finding often mentioned in studies that used HRCT imaging is that of the subpleural cysts. These small thin-walled cysts, located in subpleural spaces, can provoke the formation of a peripheral radiotransparent band, between the calcified parenchyma and the ribs, described in chest X-rays as the sign of 'black pleura'.[1,3,4,7,11] In the present study, we observed subpleural cysts in 50% of the cases. In addition, one patient presented apical bullae.

In conclusion, HRCT scans of the chest facilitate the diagnosis of PAM, since they can demonstrate patterns characteristic of the disease. The most common tomographic findings were diffuse ground-glass attenuation and subpleural linear calcifications, which were present in 90% of the patients studied. Other alterations found on the HRCT scan, in decreasing order of frequency, were small parenchymal nodules, nodular fissures, subpleural nodules, calcifications along the interlobular septa, dense consolidations, and subpleural cysts. The finding of calcifications along the interlobular septa, associated with ground-glass attenuation, configuring a mosaic pattern of attenuation, observed in the mediastinal window, can be considered pathognomonic of PAM, since this aspect, present in 40% of the cases of our sample, has not been described in any other disease.

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