Pulmonary cysts associated with calcified nodules

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A 57-year-old woman presented with a two-month history of cough and dry mouth. A CT scan of the chest showed multiple cysts and calcified nodules (Figure 1). The patient presented with two basic CT patterns: multiple pulmonary cysts and calcified pulmonary nodules. The differential diagnosis of diffuse pulmonary cysts is extensive and includes neoplastic, inflammatory, and infectious diseases. Primary causes of diffuse pulmonary cysts include lymphangioleiomyomatosis, Langerhans cell histiocytosis (LCH), Birt-Hogg-Dubé syndrome, *Pneumocystis jirovecii* pneumonia, and lymphocytic interstitial pneumonia (LIP). Multiple pulmonary nodules have numerous etiologies. However, the combined presence of multiple pulmonary nodules and calcifications narrows the diagnostic possibilities, which include calcified metastases, amyloidosis, hyalinizing granulomas, epithelioid hemangioendothelioma, rheumatoid nodules, and multiple chondromas, as well as calcifications resulting from residual granulomatous lesions, particularly tuberculous lesions.[1-3]

By defining morphological features and cyst distribution, CT plays an extremely important role in the differential diagnosis of pulmonary cysts associated with calcified nodules, as do associated findings. In most cases, diagnosis is made by combining CT findings and clinical findings (particularly extrapulmonary manifestations), without the need for lung biopsy. CT findings of nodules and cysts in the same patient are consistent with LCH and LIP associated with amyloidosis. Given that patients with LCH present with nodules that are small and do not calcify, a presumptive diagnosis of LIP associated with amyloidosis was made. The fact that some of the calcifications were within the cystic lesions constituted further evidence of LIP associated with amyloidosis. In addition, our patient presented with xerostomia. Evaluation of clinical and laboratory data led to a diagnosis of Sjögren's syndrome. The final diagnosis was Sjögren's syndrome with LIP associated with amyloidosis.

The association of lymphoproliferative disorders (particularly LIP) with amyloid deposits, cystic lung formation, and Sjögren's syndrome is widely recognized. However, the exact nature of this relationship remains unclear.

A rare lymphoproliferative disorder, LIP is most common in patients with immunodeficiency or autoimmune disease, particularly Sjögren's syndrome. In addition to nodules and cysts, other CT findings include ground-glass opacities, peribronchovascular thickening, and ill-defined centrilobular nodules. Patients can be asymptomatic or present with dyspnea, cough, fatigue, and chest pain. In many cases, cysts are incidental findings on routine CT scans or are findings associated with complications such as spontaneous pneumothorax.

Although histopathological examination is usually required for diagnosis, our patient was definitively diagnosed on the basis of imaging and clinical findings, without the need for lung biopsy.

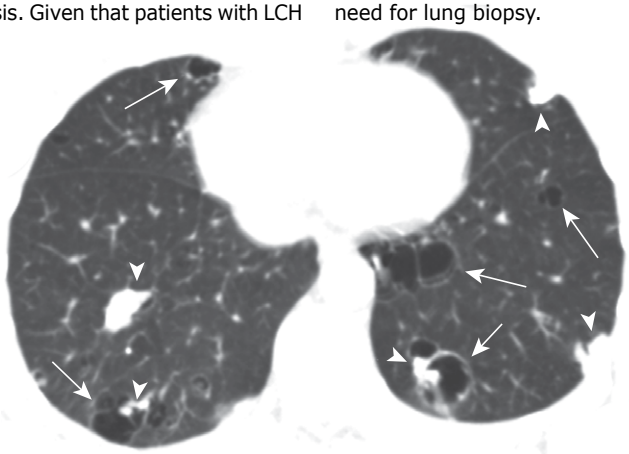

Figure 1. Axial CT scan of the chest with lung window settings at the level of the lung bases, showing multiple cysts (arrows) and calcified nodules (arrowheads). Note that some of the calcifications are within the cystic lesions.

REFERENCES

1. Baldi BG, Carvalho CRR, Dias OM, Marchiori E, Hochhegger B. Diffuse cystic lung diseases: differential diagnosis. J Bras Pneumol. 2017;43(2):140-149. https://doi.org/10.1590/s1806-37562016000000341
2. Ferreira Francisco FA, Soares Souza A Jr, Zanetti G, Marchiori E. Multiple cystic lung disease. Eur Respir Rev. 2015;24(138):552-64. https://doi.org/10.1183/16000617.0046-2015
3. Marchiori E, Zanetti G, Hochhegger B. Multiple calcified nodules. J Bras Pneumol 2016;42(3):164. https://doi.org/10.1590/S1806-37562016000000094

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