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An Unusual Case of Diffuse Pulmonary Meningotheliomatosis Accompanying with Cystic Lung Disease

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Introduction: Diffuse pulmonary meningotheliomatosis is a rare disease, presenting with diffuse bilateral pulmonary nodules on HRCT with unclear clinical significance.

Case Presentation: A 63-years-old male patient was admitted to our hospital with cough and mild exertional dyspnea. Thorax HRCT scan revealed bilaterally multiple, uniform, thin-walled cysts ranging between 0.5 and 2.5 cm in diameter, diffusely distributing from apex to basis of the lung suggesting lymphangioleiomyomatosis radiologically. Hence the patient was male and symptomatic; further evaluation was needed. Bronchoscopy was nondiagnostic which led to a diagnostic surgical procedure. The patient underwent diagnostic right lower lobe and middle lobe wedge resections. Histopathological examination revealed diffuse meningotheliomatosis unexpectedly. When HRCT findings were reviewed, micronodules smaller than 1 mm were identified. Although the patient only received symptomatic treatment, there were no changes in thorax CT findings through the three years follow up.

Conclusion: To our knowledge this case is the first in the literature, demonstrating diffuse meningotheliomatosis accompanied with cystic disease of the lung.

Keywords: Cystic lung disease, diffuse pulmonary meningotheliomatosis, interstitial lung disease