Diffuse Cavitating Pulmonary Meningotheliomatosis: A Rare and Elusive Pathology

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Diffuse pulmonary meningotheliomatosis (DPM) is an exceptionally rare parenchymal lung disease that predominantly affects young to middle-aged females. It is distinguished by extensive bilateral minute pulmonary meningothelial-like nodules (MPMNs) on imaging, which may seldomly cavitate. These findings are generally incidentally found on imaging due to its asymptomatic course, however, patients may present with dyspnea and/or cough. Cases of DPM are few and pathogenicity associations have not yet been established but some cases display an association with malignancy. Tissue biopsy is strictly necessary for diagnosis given its wide differential diagnosis. A 45-year-old female with a past medical history of hypothyroidism and hyperlipidemia is referred to pulmonary medicine clinics due to abnormal radiologic imaging findings. The patient refers to occasional bouts of dyspnea with no temporal association of years of evolution for which she uses budesonide/formoterol as needed. She is a nonsmoker, without overt exposure or occupational risk factors. A high-resolution chest CT scan revealed innumerable bilateral thin-walled pulmonary cysts with thickened interlobular septa, suggestive of lymphangioleiomyomatosis (LAM). Vascular endothelial growth factor (VEGF) levels and rheumatoid panel were nondiagnostic, and pulmonary function testing revealed normal FEV1/FVC, moderate air trapping, and increased airway resistance. An open biopsy of the right lung was performed and pathological examination revealed meningothelial nodules, with histologic findings supporting the diagnosis of cavitating diffuse pulmonary meningotheliomatosis. Literature describing DPM is very limited, with only 25 cases documented through 2016. Since the majority of patients are asymptomatic, further research into the pathophysiology and possible etiologies involving the development of DPM and other MPMNs may shed light on their clinical significance and prognosis once symptoms become evident. We aim to raise awareness of MPMNs to improve the identification of clinical patterns and findings related to disease progression as well as establish treatment options. The documentation of these cases is needed to help to establish correlations with other possible pulmonary and extrapulmonary manifestations of this elusive disease which may guide further treatment and prognosis.

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