Pulmonary Meningotheliomatosis - Rare or Underrecognized?

H. Anderson, M. Izquierdo;
Prisma Health, Greenville, SC, United States

Pulmonary meningotheliomatosis (PM) is an exceptionally rare condition characterized by the proliferation of mesothelial cells within the lung parenchyma, leading to nodular (minute pulmonary meningothelial-like nodules (MPMN)) or interstitial (diffuse pulmonary meningotheliomatosis (DPM)) changes. MPMN are generally asymptomatic and are often discovered incidentally, whereas DPM typically presents with dyspnea and a radiographic appearance consistent with a significant interstitial process. Here, we present the case of a 77-year-old female with multiple comorbidities who was incidentally diagnosed with pulmonary meningotheliomatosis.
A 77-year-old female with a history of pulmonary hypertension, severe obstructive sleep apnea on bilevel positive airway pressure (BiPAP), renal cell carcinoma (treated with nephrectomy 2 years prior), and pulmonary emboli, who to the hospital for previously planned video assisted thoracoscopic surgery (VATS) with left lower lobe wedge resection in setting of a 1.1 cm nodule. The nodule had been followed on serial imaging for two years prior. She was asymptomatic. Postoperatively, she required a prolonged ICU stay due to profound respiratory failure. Due to suspicion for pulmonary hypertension (no prior, formal diagnosis) as the causative factor, she underwent a right heart catheterization, which showed a mean PA pressure of 68 mmHg, PCWP of 19, and PVR of 11. Based on these findings and the patient's pertinent history, the presence of Groups 1, 2, and 3 pulmonary hypertension were diagnosed. She was started on a PDE5 inhibitor, an endothelin receptor antagonist, a prostacyclin agonist. Respiratory failure improved. Pathology results from the wedge resection indicated adenocarcinoma with negative margins as well as pulmonary meningotheliomatosis. PET scan obtained prior to admission was without evidence of metastatic disease, and, based on stage 1 disease (pT1b), surveillance imaging was planned.
This case is notable for the incidental finding of pulmonary meningotheliomatosis (MPMN subtype) in a patient with multiple complex comorbidities. The clinical relevance of this diagnosis is of question. Although no clearly defined risk factors have been elucidated, a loose association has been observed between female sex and a history of malignancy. The potential for development of DPM in those with a history of MPMN is not known. Given this patient's complex pulmonary history, the diagnosis of MPMN may provide valuable insight for future clinical management, as it represents a distinct etiology that could be identified in subsequent imaging studies. On a broader scale, this case offers a foundation for further research into the diagnosis and progression of MPMN.

This abstract is funded by: none