

Diffuse Pulmonary Meningotheliomatosis Diagnosed Concurrently With Pulmonary Adenocarcinoma

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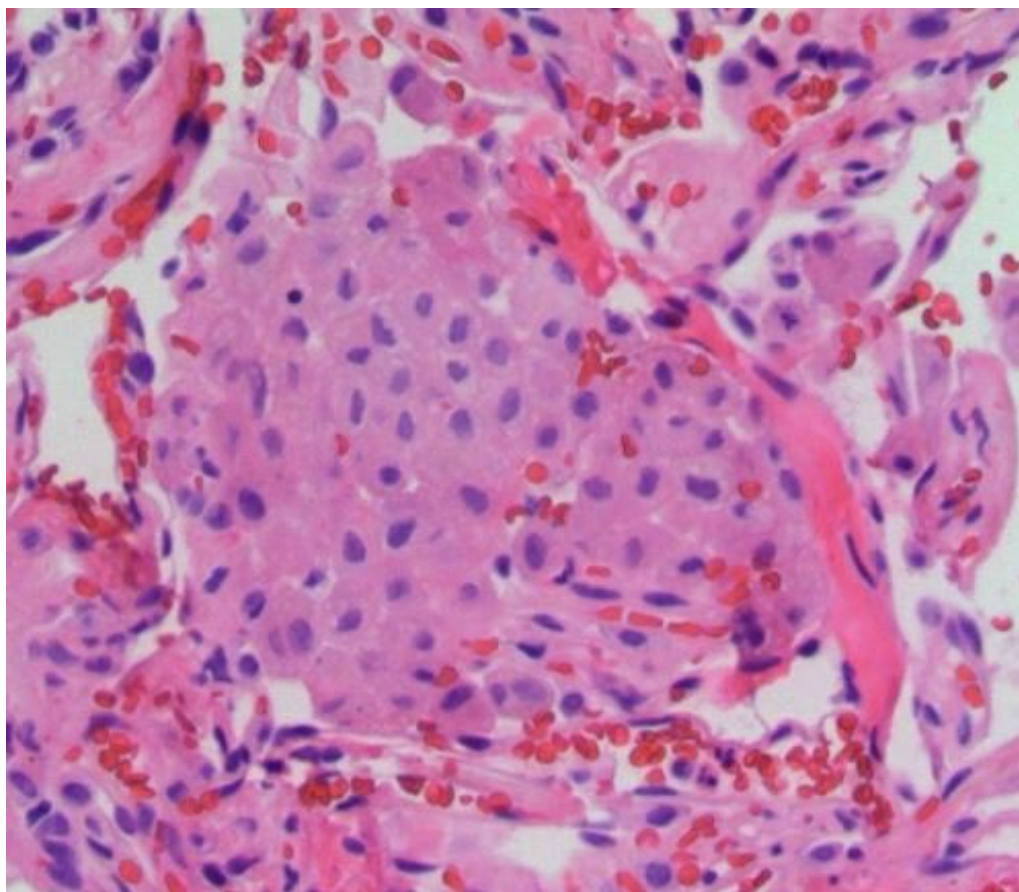
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The patient in this case is a 77-year-old female who was incidentally found to have diffuse pulmonary meningotheliomatosis (DPM). Prior to diagnosis, she had a past medical history which included tobacco use with greater than 45 pack year history, previous pulmonary embolism maintained on apixaban, renal cell carcinoma, polycythemia, obstructive sleep apnea, pulmonary hypertension, essential hypertension, and pulmonary nodule. Symptoms included dyspnea and significant hypoxemia without any other causative etiology. On CT imaging she was noted to have micronodular changes bilaterally. She underwent video-assisted thorascopic surgery with left lower lobe resection due to increasing size of a pulmonary nodule which subsequently resulted in the diagnosis of T1b adenocarcinoma as well as multifocal pulmonary meningotheliomatosis. On pathologic examination of the resected tissue, there was noted to be multifocal perivascular aggregates of spindle cells consistent with the diagnosis of DPM. These cells were diffusely immunoreactive for vimentin and EMA.

Minute pulmonary meningiothelial-like nodules (MPMNs) were identified in 1960 and initially called pulmonary chemodectomas. In 1988 terminology was changed to MPMN after evidence of meningothelial derivation was discovered. While MPMNs are relatively commonly identified incidentally as a single nodule, the term DPM was introduced in 2007 and is a rare diffuse pulmonary disease characterized by multiple lesions [1, 2, 3]. Radiographic features typically include randomly distributed ground glass attenuation or discrete micronodules sometimes with a near solid appearance due to alveolar compression [1,3]. Occasionally there is noted to be a cranio-caudal gradient. On light microscopy, cells are characterized by uniform oval or spindle shaped cells with abundant pale pink cytoplasm. Electron microscopy reveals features reminiscent of meningioma cells with absence of mitosis. Immunohistochemistry studies of MPMNs show them to be positive for progesterone receptor, epithelial membrane antigen (EMA), and vimentin, as in the case described above [1].

Both MPMN and DPM are most commonly identified in females and are often diagnosed in the 5th-7th decades of life. Classically patients are asymptomatic and the clinical significance remains unclear with patients typically having a benign clinical course [2]. The etiology of both remains a topic in need of further investigation. Various studies have suggested an association with thromboembolic, neoplastic, and smoking-related interstitial-lung disease - all factors that were present in this case [3]. Despite this, there has yet to be any clear identification of risk factors for development of MPMNs or DPM.



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