A Rare Case of Pulmonary Nodules: Diffuse Pulmonary Meningotheliomatosis

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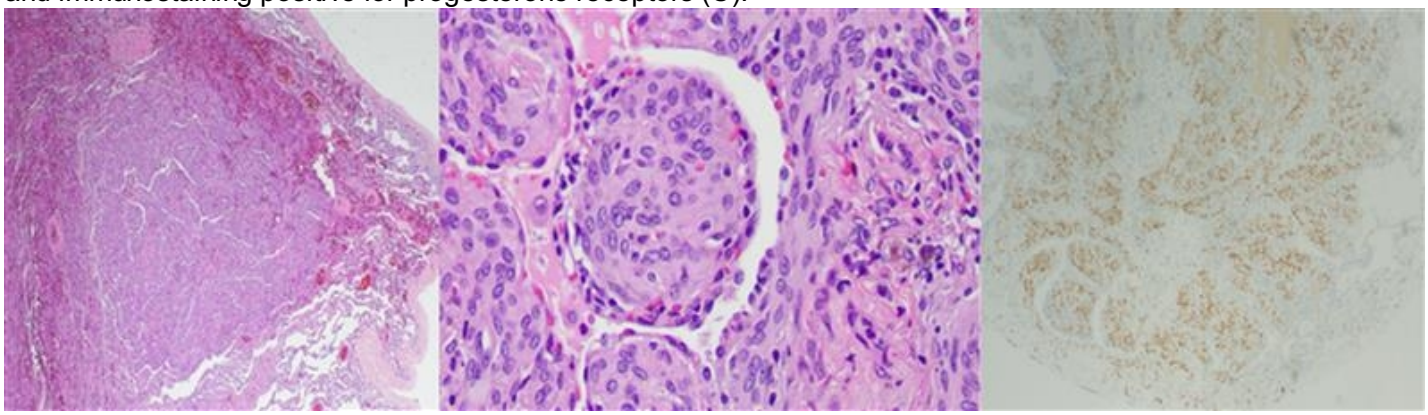
Introduction Minute pulmonary meningothelial-like nodules (MPMNs) were first described in 1960 and thought to be pulmonary chemodectomas. Further research showed their resemblance with meningothelial cells. MPMNs are typically found incidentally and in the majority of the time, they are solitary nodules, with fewer reported incidences of multiple lesions named diffuse pulmonary meningotheliomatosis. It is more common in women in their fifth to sixth decade of life. It is typically an asymptomatic disease, although there have been a few reports of patients who presented with shortness of breath and cough. Although most solitary nodules can be removed, little is known about treatment options for multiple lesions. Case Description A 51-year-old female, former smoker, with a past medical history of transient ischemic attack, hypertension, and childhood asthma on albuterol presented to the clinic for worsening dyspnea since December 2019. Her dyspnea originally started with cold symptoms including a cough; although her cough improved, her dyspnea persisted. Spirometry was consistent with a restrictive lung disease. A CT of the chest was obtained, which showed diffuse subcentimetric nodularities in random distribution. Fungal serology and connective tissue disease panels were negative. Quantiferon testing was negative. She subsequently underwent a bronchoscopy with bronchoalveolar lavage (BAL) and transbronchial lung biopsies (TBBX). Cultures were negative for bacterial, mycobacterial and fungal growth. Pathology was negative for malignancy. Since she had ongoing dyspnea with an unclear etiology of her numerous bilateral pulmonary nodules, she underwent a right thoracotomy with multiple lung biopsies. Pathology was significant for nested cells with immunostaining positive for epithelial membrane antigen (EMA), CD56, and progesterone receptor. The morphology and immunophenotype supported the diagnosis of diffuse pulmonary meningotheliomatosis. Discussion Diffuse pulmonary meningotheliomatosis is a rare diagnosis, and the etiology is still unclear, with the leading theory being that it originates from meningothelial cells. Patients have been found to have single lesions, usually found incidentally, as well as multiple lesions. It is currently unclear if these are a reactive proliferation, a neoplastic process, or a transition point between the two. This case illustrates the need for consideration of diffuse pulmonary meningotheliomatosis in the differential diagnosis for patients who present with dyspnea and are found to have multiple pulmonary nodules on imaging. Figure 1: Nested cells with meningothelial like morphology (A+B) and immunostaining positive for progesterone receptors (C).



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