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Diffuse Pulmonary Meningotheliomatosis: A Case Report

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Introduction/Objective: Diffuse pulmonary meningotheliomatosis (DPM) is a pulmonary disease characterized by the presence of widespread bilateral minute pulmonary meningothelial-like nodules (MPMNs). It is exceedingly rare and predominantly seen in females. The nodules are typically asymptomatic and detected incidentally on imaging.

Methods/Case Report: We present a case of a 57-year-old woman with a history of chronic cough and CT imaging findings of bilateral multiple ground-glass nodules. Wedge resections from the right upper and right lower lobes were obtained, which revealed gross changes suggestive of interstitial lung disease. Histologic evaluation of the wedges showed similar morphology, consisting of discrete areas with variable interstitial expansion of the stroma with ovoid/spindle cells, collagen, elastosis,

and mild chronic inflammatory cells, in a somewhat perivascular pattern. Some areas were also cystic. This appeared in a background of emphysematous change and focal foreign body giant cell reaction to non-polarizable material. Immunohistochemical stains were diffusely positive for vimentin, EMA, and PR, and CD163 highlighted increased cells that were S100 and CD1a negative. Additionally, the ovoid/spindle cells were negative for desmin, SMM, HMB-45, HHV8, pan-CK, chromogranin, and multiplex SOX10-MART1.

Results (if a Case Study enter NA): NA

Conclusion: Diffuse pulmonary meningotheiomatosis is a rare entity that requires sufficient histological evaluation to identify the ovoid/spindle cell component histologically and immunohistochemically. This disease should be considered in the differential for cases of diffuse interstitial pulmonary infiltrates. Few cases have been reported in the literature, requiring further research to understand the mechanisms of this disease and its clinical significance.

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