CHEST Annual Meeting
October 18-21
2020

Lung Pathology

SESSION TITLE: Medical Student/Resident Lung Pathology
SESSION TYPE: Med Student/Res Case Rep Postr
PRESENTED ON: October 18-21, 2020

DIFFUSE PULMONARY MENINGOTHELIOMATOSIS DIAGNOSED BY TRANSBRONCHIAL CRYOBIOPSY

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INTRODUCTION: Minute pulmonary meningothelial-like nodules (MPMNs) are typically asymptomatic, solitary lesions incidentally noted on pathologic evaluation of lung tissue. However, diffuse, bilateral pulmonary involvement, termed diffuse pulmonary meningotheliomatosis (DPM), is exceedingly rare and has only been described in a small number of case studies. We report a case of a patient diagnosed with DPM via transbronchial cryobiopsy.

CASE PRESENTATION: A 67-year-old non-smoking female with a history of scleroderma was referred for diffuse bilateral pulmonary nodules found on computed tomography (CT) scan of the chest with mosaicism concerning for diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH). Pulmonary function testing was notable for an isolated reduction in diffusing capacity at 58% with a normal echocardiogram. Two transbronchial cryobiopsy samples were obtained from the right lower lobe. Pathology revealed a meningothelial nodule. Immunohistochemistry was positive for epithelial membrane antigen (EMA) as well as progesterone receptor (PR) and negative for synaptophysin and chromogranin. Follow up imaging at 1 year demonstrated stability. Taken in context with her radiographic findings, the patient was diagnosed with DPM.

DISCUSSION: DPM is an extremely rare condition characterized by MPMNs diffusely involving lung parenchyma. It predominantly affects females (2.2:1) with a median age of 59-62. DPM has no predilection for specific lobes, and it can be mistaken for metastatic or interstitial disease. Histologically, MPMNs appear as epithelioid cells arranged in small nests in pulmonary interstitium. Immunochemically, MPMNs tend to be positive for vimentin, EMA and PR and negative for S-100 protein, chromogranin A, and synaptophysin. Though the clinical significance of DPM is still unclear, prior studies have noted higher incidence among patients with underlying lung disease, particularly thromboembolism and malignancy.

CONCLUSIONS: In summary, we present a case of DPM diagnosed by transbronchial cryobiopsy to highlight the importance of considering DPM in the differential of bilateral micronodular lung disease.

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DOI: https://doi.org/10.1016/j.chest.2020.08.1471