



## Lung Pathology

**SESSION TITLE:** The Path to the Final Diagnosis

**SESSION TYPE:** Rapid Fire Case Reports

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### RIDDLE ME THIS: A RARE CASE OF DIFFUSE PULMONARY MENINGIOTHIELIOMAS

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**INTRODUCTION:** Minute pulmonary meningotheelial-like nodules (MPMNs) are rare, small, and asymptomatic benign lesions that predominantly affect females due to meningotheelial cellular proliferation in the lung interstitium.<sup>1</sup> They are associated with many diseases, including pulmonary thromboembolism, interstitial lung disease, and lung adenocarcinoma, and can present as single or multiple ground glass nodules (GGNs) on computed tomography (CT). Here, we present a case of diagnostic dilemma in a patient with a history of malignancy and immunosuppression presenting with diffuse pulmonary meningotheiomatosus (DPM).

**CASE PRESENTATION:** A 61-year-old female with a history of ductal hyperplasia of the breast, common variable immunodeficiency syndrome (CVID), asthma, obstructive sleep apnea, and thyroid follicular cancer was diagnosed with diffuse pulmonary nodules incidentally via CT angiogram chest during inpatient admission for COVID-19 infection. Post-COVID her symptoms improved, but a follow-up CT scan chest 1 year later showed numerous sub 6 mm bilateral centrilobular GGNs and solid pulmonary nodules with mild upper lobe predominance. Labs demonstrated chronic neutropenia with lymphocytosis, positive perinuclear and cytoplasmic antineutrophil cytoplasmic (PANCA & CANCA) antibody, but negative antinuclear (ANA), myeloperoxidase (MPO), and proteinase 3 (PR3). A thyroid radioactive iodine uptake scan showed expected uptake in the thyroid gland with no abnormal uptake in the lungs, suggesting no evidence of pulmonary metastasis. The patient eventually underwent bronchoscopy with bronchioalveolar lavage (BAL) with transbronchial cryo biopsy of lung nodules, which showed MPMNs with further immunostaining negative for malignancy. The patient's case was discussed with a multidisciplinary ILD clinic, and given the complexity of the case, lack of symptoms, and stability of nodules, a decision was made for close follow-up with serial CT scans.

**DISCUSSION:** In our case, the patient was diagnosed with DPM, which is an exceptionally rare form of widespread MPMs with few cases reported in the literature. Differentials at the time of diagnosis included malignant nodules, atypical infection, autoimmune interstitial lung disease (ILD), and granulomatous lymphocytic interstitial lung disease (GLI-ILD). A systemic approach to lung nodules and transbronchial cryobiopsy led to a clinical diagnosis.

**CONCLUSIONS:** This case highlights the importance of lung biopsy for pulmonary nodules of unclear etiology and DPMs as a rare differential. Although MPMNs usually have an indolent course, they may mimic adenocarcinoma in situ, metastasis, or atypical infection, which may lead to misdiagnosis, especially in patients with confounding comorbidities

**REFERENCE #1:** Gleason J, Valentin R, Almeida P, Martinez N, Bejarano P. Diffuse pulmonary meningotheiomatosus: A literature review of a rare diffuse parenchymal lung disease with unclear clinical significance. Review Article. The Journal of Association of Chest Physicians. January 1, 2017, 2017;5(1):18-25. doi:10.4103/2320-8775.196647

**REFERENCE #2:** Alkurashi AK, Almodallal Y, Albitar HAH, Cheville JC, Iyer VN. Diffuse Pulmonary Meningotheliomatosus: A Rare Lung Disease Presenting with Diffuse Ground-Glass Opacities and Cavitation. Am J Case Rep. Nov 9, 2020;21:e926172. doi:10.12659/ajcr.926172

**REFERENCE #3:** Zhang Y, Wu J, Zhang T, Zhang Q, Chen Y-C. Minute pulmonary meningotheelial-like nodules: rare lesions appearing as diffuse ground-glass nodules with cyst-like morphology. Quantitative Imaging in Medicine and Surgery. 2021;11(7):3355-3359. doi:10.21037/qims-20-676

#### DISCLOSURES:

Speaker/Speaker's Bureau relationship with Boehringer-Ingelheim Please note: Sept 2021-Present Added 04/06/2023 by Briana DiSilvio, source=Web Response, value=Honoria

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