Diffuse Pulmonary Meningotheliomatosis and Turner's Syndrome: A Growing Area of Literature

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Minute pulmonary meningothelial-like nodules are typically asymptomatic solitary lesions that are incidentally noted on histopathologic review of lung tissue. Diffuse lung involvement is rare and termed diffuse pulmonary meningotheliomatosis (DPM). We report a case of a 57-year-old female with histologically confirmed DPM after ipsilateral lobal wedge resections. Her past medical history is notable for Turner syndrome, aortic coarctation and bicuspid aortic valve status post replacement, left vocal cord paralysis and heart failure with reduced ejection fraction. She was referred to outpatient pulmonology clinic for evaluation of chronic cough. Chest computed tomography (CT) demonstrated ground glass nodules bilaterally. Some nodules appeared to have central clearing or cavitation, and the largest nodule measured five millimeters. The patient had no prior history of tobacco use. Pulmonary function testing noted a mild obstructive ventilatory defect, no significant response to bronchodilators, air trapping, hyperinflation, and a mildly reduced diffusion capacity. The patient was recommended to use a fluticasone-salmeterol inhaler. Bronchoscopy with bronchoalveolar lavage and transbronchial lung biopsy was done. Bronchoscopic cultures demonstrated mixed respiratory flora; biopsy showed no malignant cells, mild acute inflammation, and pulmonary macrophages. The patient was prescribed Amoxicillin-Clavulanate for ten days with recommendation to repeat CT scan in three months. Although the patient felt better, the repeat CT demonstrated more ground-glass nodules scattered throughout the lungs. The maximum nodule size remained at five millimeters. Thoracic surgery was consulted, and right upper and right lower lobe wedge resections were performed. Immunohistochemical staining of spindled cells with indistinct borders demonstrated positive for Vimentin, EMA, PR, and CD163, and negative staining for S100, CD1a, desmin, chromogranin, cytokeratin, and HMB-45. In considering these immunohistochemical and pathological findings, the patient was diagnosed with DPM. At two years following the diagnosis of DPM, the number of lung nodules has not significantly increased, and the patient has not been diagnosed with malignancy. This is the third reported case in the literature of a patient with Turner Syndrome who later developed DPM. Though there is a known relationship of Turner syndrome with meningiomas, no similar association has been established for DPM. Further investigation is needed to determine if exogenous hormone exposure, particularly progesterone, may be linked to the development of DPM in specific genotypes of Turner Syndrome. This case also demonstrates the need for clinicians to consider a diagnosis of DPM in patients presenting with diffuse ground glass pulmonary nodules.

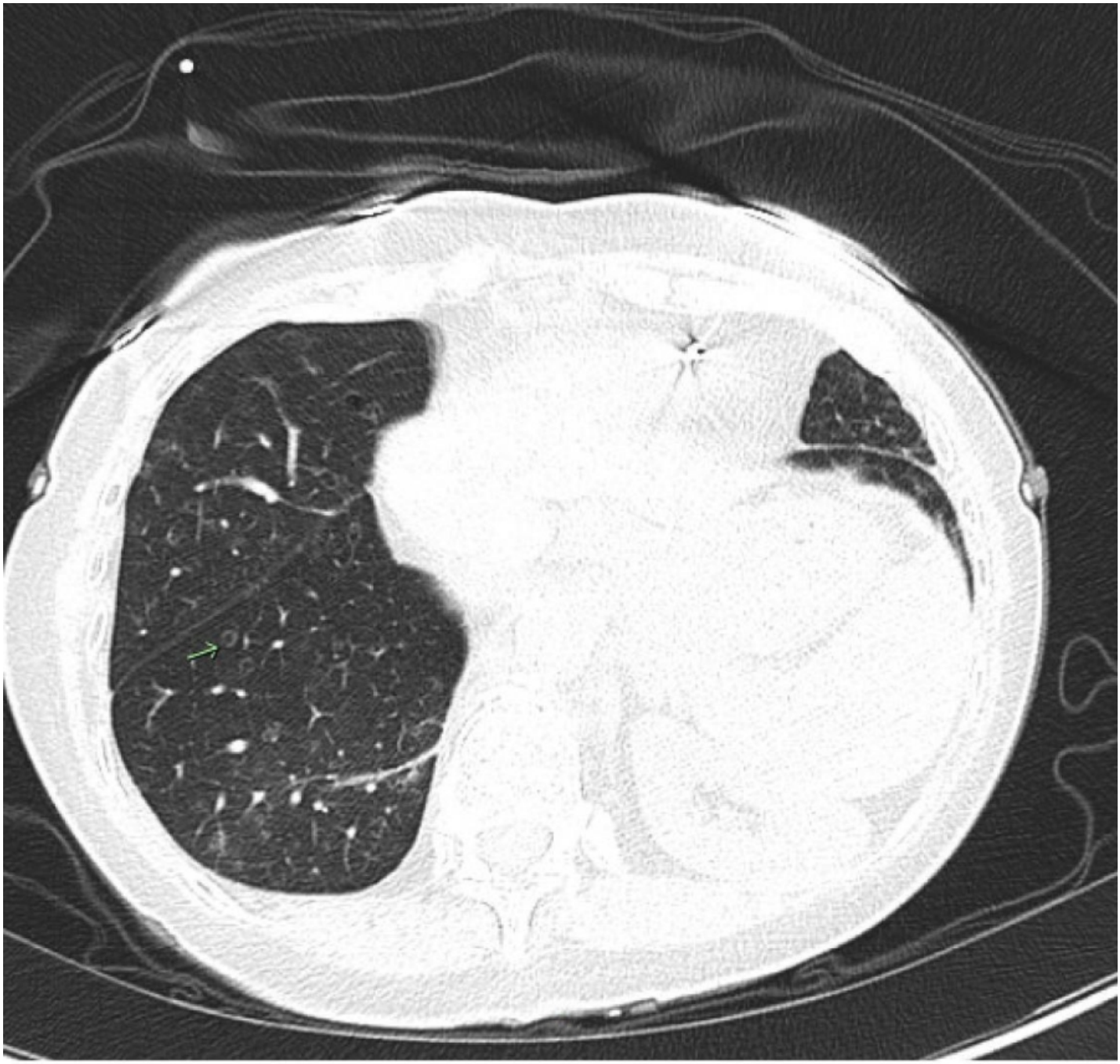

Image demonstrates small cavity lesions. A singular lesion is marked.

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