A Rare Case Of Diffuse Pulmonary Meningotheliomatosis

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INTRODUCTION:
Pulmonary meningothelial nodules are uncommon lesions consisted of epithelioid cells located inside the lung interstitium. A few cases of symptomatic diffuse pulmonary meningothelial nodules have been described.
CASE PRESENTATION:
A 54-year old female patient presented to pulmonary clinic with shortness of breath of one year duration that gradually progressed and was associated with cough. She denied fever, chest pain, sputum production and wheezing. No exposure to tobacco, asbestosis, birds and animals. Her medical problems were significant for hypothyroidism and gastroesophageal disease. Vital signs were blood pressure 130/85 mm Hg, respiratory rate 16/min, temperature 98.9 F and oxyhemoglobin saturation 90% on room air. Physical examination was remarkable for diffuse rhonchi posterior chest bilaterally. CT chest showed multiple centrilobular pulmonary nodules. Pulmonary function test showed normal spirometry but significantly reduced diffusing capacity of the lungs for carbon monoxide (DLCO) of 8.13 ml/min/mmHg (40% predicted). Bronchoscopy with bronchoalveolar lavage (BAL) and transbronchial biopsies were not diagnostic. Extensive workup including blood cultures, BAL cultures including fungal, viral, acid fast bacillus smear and cultures were negative. Serum Histoplasmosis antigen and antibodies, urine Histoplasmosis antigen, and Coccidioidomycosis antibodies (IgM, IgG) were negative. Immunologic workup was negative. Patient continued to have cough and shortness of breath. Wedge resection of left lower lobe was performed. Pathology showed multiple pulmonary meningothelial-like nodules; consistent with diffuse pulmonary meningotheliomatosis. Patient remianed symptomatic, required oxygen supplementation and was referred for evaluation of lung transplant.
DISCUSSION:
Pulmonary meningothelial nodules first defined by Korn et al are usually asymptomatic and are typically found incidentally during autopsies. Common in females between 54 to 75 years of age. Exact etiology and pathogenesis remains unclear. Previously reported cases described mild restrictive disease. The patient described in this case was symptomatic but with normal spirometry and severely decreased DLCO. Treatment of diffuse pulmonary meningotheliomatosis is not well defined. Patient is referred to lung transplant evaluation for the further management.
CONCLUSION:
Diffuse pulmonary meningotheliomatosis should be considered in the differential diagnosis of patients with unexplained dyspnea and diffuse interstitial nodules. Lung transplant may be a feasible option in patients with symptomatic patients with hypoxemia and reduced DLCO<40.
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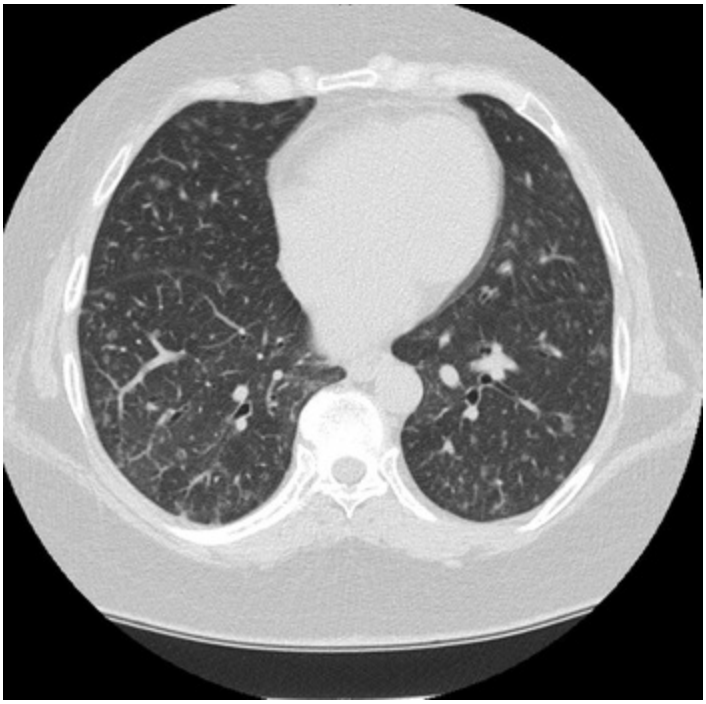

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