Diffuse Pulmonary Meningotheliomatosis: A Rare Lung Disease Presenting with Diffuse Ground Glass Opacities and Cavitation

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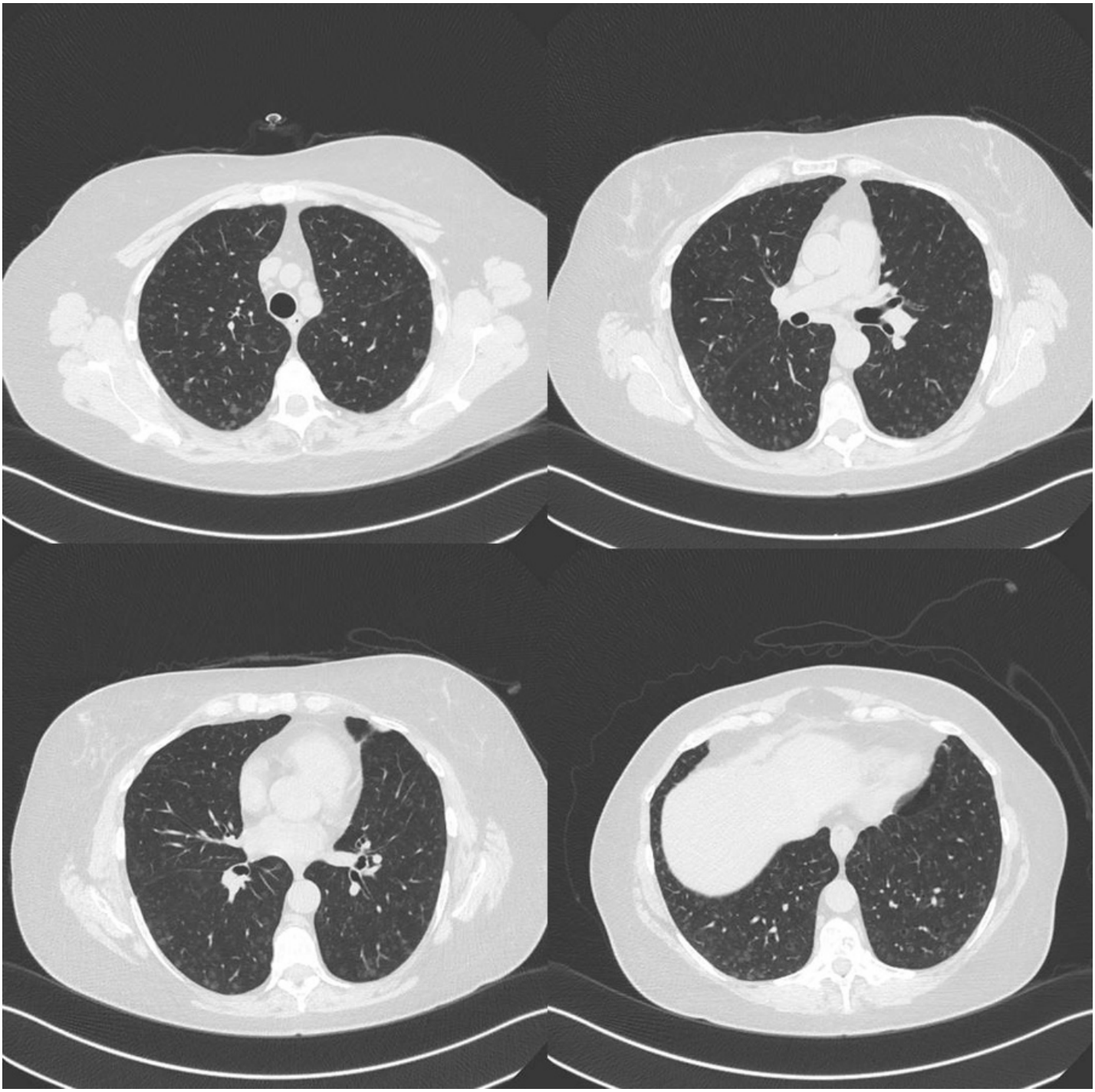
Rationale: Diffuse pulmonary meningiotheliomatosis (DPM) is an exceedingly rare diffuse pulmonary disease characterized by the presence of widespread bilateral minute pulmonary meningiothelial-like nodules (MPMNs). We present a 55-yeard-old woman with DPM presenting with chest pressure. Case Report: A 55-year-old woman presented to the clinic with non-exertional chest pressure and dry cough of 4-month duration. She was a lifelong non-smoker and denied any other symptoms. Physical examination did not reveal any abnormalities. Twelve-lead electrocardiogram was normal. High resolution chest computed tomography (CT) showed innumerable faint diffuse small ground-glass nodules, some of which were cavitary in nature. Laboratory tests including complete blood count, inflammatory markers, ANA, anti-dsDNA, anti scl-70 and RNA polymerase III antibodies, rheumatoid factor and anti CCP antibody, in addition to ANCA and extractable nuclear antibody panels were all negative. Hypersensitivity pneumonitis panel, cystic fibrosis genetic mutation testing and fungal serologies were all negative as well. Pulmonary function tests did not reveal any abnormalities. The patient ultimately underwent uncomplicated right video-assisted thoracoscopic surgery (VATS) with multiple lung biopsies. Pathology showed multiple well-circumscribed interstitial meningothelial-like nodules in perivenular distribution with occasional whorling of cells consistent with diffuse pulmonary meningotheliomatosis (figure 3). One year following diagnosis, the patient continued to complain of non-exertional chest pressure without pulmonary complaints. Repeat imaging with high-resolution chest CT scan showed stable findings. Discussion: MPMNs typically present as isolated pulmonary nodules [1, 2]. However, DPM is an exceptionally rare form of widespread MPMNs of unclear clinical significance with few cases reported in literature [2]. It is more common in middle-aged females and in patients with history of malignancy [2]. Most patients are asymptomatic and those with symptoms usually present with nonspecific cardiopulmonary symptoms [2]. Diagnosis is typically established with surgical lung biopsy. This case highlights that DPM should be considered in patients presenting with diffuse ground-glass nodules and that further research is needed to better understand this disease and its clinical significance. References: [1]: Gleason J, Valentin R, Almeida P, Martinez N, Bejarano P. Diffuse pulmonary meningotheliomatosis: A literature review of a rare diffuse parenchymal lung disease with unclear clinical significance. The Journal of Association of Chest Physicians. 2017;5(1):18-25. [2]: Suster S, Moran CA. Diffuse pulmonary meningotheliomatosis. The American journal of surgical pathology. 2007;31(4):624-631.



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