Rethinking Chronic Multinodular Interstitial Disease in a 70-year-old Woman: A Case of Diffuse Pulmonary Meningotheliomatosis

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Background: Diffuse pulmonary meningotheliomatosis (DPM) is a rare, benign proliferation of minute meningothelial-like nodules ranging from 100mm - 11mm.[1] The condition presents primarily in women (9:1) in middle age.[2] Discovered incidentally or with mild symptoms such as dyspnea and dry cough, the imaging findings can be concerning for miliary metastatic, atypical infectious, and immune mediated diffuse parenchymal lung disease thereby complicating the diagnostic process.
Case Description: A 70-year-old female never smoker presented to our pulmonary clinic for evaluation of dyspnea and dry cough. She described evaluation roughly 20 years prior that had revealed innumerable small ground glass pulmonary nodules on CT imaging. Pulmonary function testing at that time was unremarkable, and laboratory studies had only revealed a weakly (+) SSA antibody. She was referred for a right mini-thoracotomy wedge resection, with pathology demonstrating small nodules partially involving the pleura, suspected to represent hypersensitivity pneumonitis (HP), as well as, several incidentally noted pulmonary “chemodectomas”. She was trialed on inhaled therapies, antibiotics, and even a two-year course of steroids for suspected HP without notable clinical or radiologic improvement. Interestingly, she was subsequently found to have an intracranial meningioma which was treated with external beam radiation. Upon presentation at our institution, a repeat high resolution chest CT showed mosaic attenuation with innumerable groundglass nodules and micronodules in a centrilobular and peri-lymphatic distribution. There was no significant change in appearance or distribution from prior imaging (Figure 1, A-B). Rigid bronchoscopy with transbronchial lung cryobiopsy was performed with pathology showing multiple meningothelial-like nodules (Figure 1, C). This pathologic finding, in conjunction with her clinical history, including past surgical biopsy, led to a diagnosis of DPM.
Discussion: This case adds to about 50 cases of DPM and the clinical significance of diffuse perivenular and interstitial meningothelial-like cell proliferations in the lung without clear malignant potential. Additionally, it provides another case to a notable collection of cases with metachronous intracranial meningioma, suggesting possible shared pathogenesis or genetic predisposition.[2]
While the clinical course of DPM is generally indolent, it poses diagnostic challenges due to overlapping imaging characteristics with more aggressive pulmonary pathologies. The differential may include sarcoidosis, miliary infectious and malignant etiologies, DIPNECH, and follicular bronchiolitis.
Conclusions: A systematic approach to this patient’s ground-glass nodules with clinical re-evaluation given ineffective empiric treatment ultimately led to the correct diagnosis. It highlights the importance of biopsy for the diagnosis of pulmonary nodules of unclear etiology even with benign clinical progression.

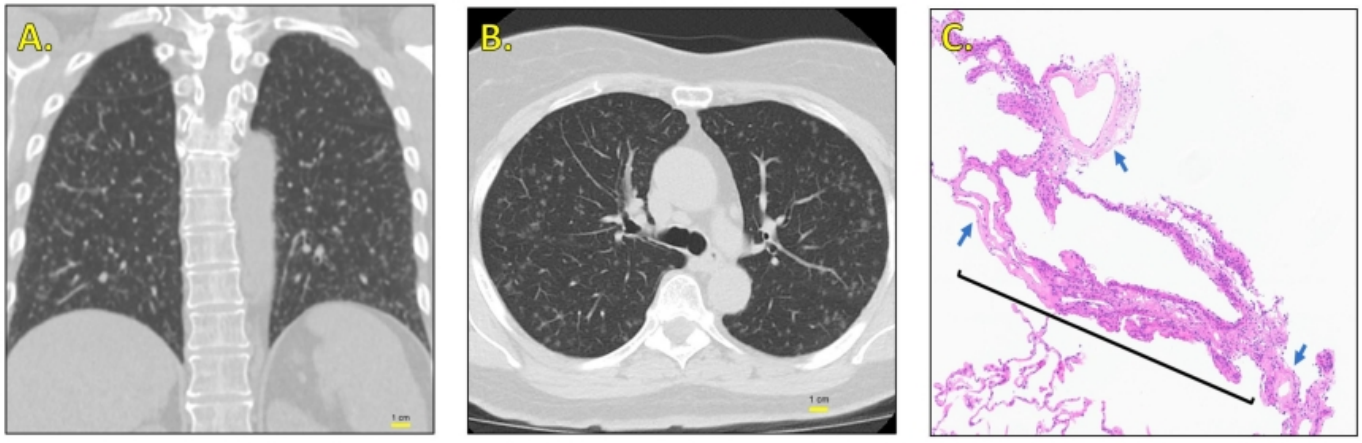

Figure 1: A. Coronal plane (slice width 1 mm) high resolution commuted tomography image demonstrating diffuse, innumerable bilateral minute centrilobular and perilymphatic ground glass nodules of varying sizes. B. Axial plane (slice width 1 mm) demonstrating the same. C. Histologic sections demonstrating well-circumscribed meningothelial like nodule (black bracket) with perivenular distribution (blue arrows).

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