Diffuse Pulmonary Meningotheliomatosis: A Scary, Yet Benign Picture

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Introduction

Pulmonary Meningothelial-like nodules (MLNs) comprise epitheloid cell nests arranged around small veins and usually manifest as incidentally detected, asymptomatic, solitary or multiple nodules in lung biopsy. Rarely, these lesions can be profuse enough to manifest as diffuse bilateral pulmonary nodules on radiographic imaging. Here we describe such an instance of 'diffuse pulmonary meningotheliomatosis (DPM).

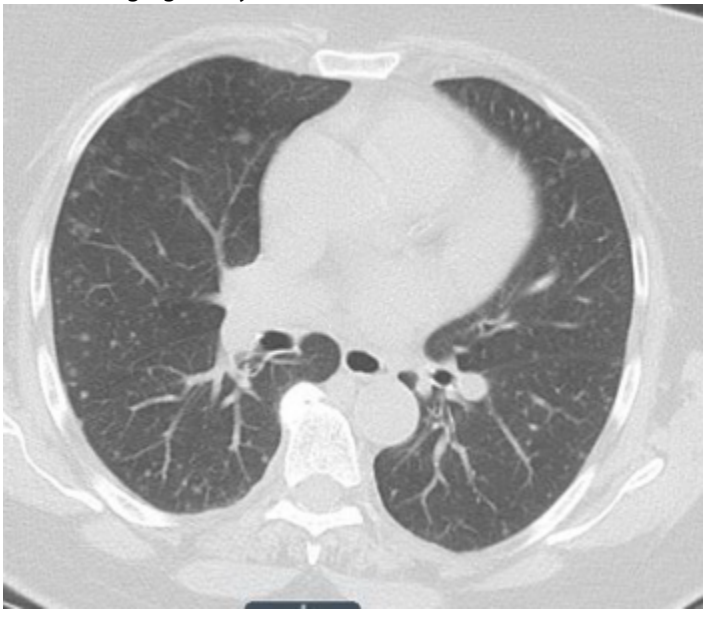
Case Presentation

A 74-year-old, non-smoking female, was referred to pulmonology clinic for evaluation of diffuse pulmonary nodules that were incidentally detected on CT scan. A comprehensive review of systems, physical exam, basic labs and PFT were normal. Review of the chest CT showed innumerable small solid and ground glass nodular opacities, 5mm or less in diameter, distributed throughout the lungs, with a predominant centri-lobular distribution and with central cavitation in some nodules. A repeat CT chest re-demonstrated the pulmonary nodules, with few ground glass nodules having mildly progressed in size over the six-week period. The patient underwent trans-bronchial lung biopsy. Biopsy demonstrated collection of cytologically bland epithelioid cells with occasional cytoplasmic intranuclear pseudoinclusions, situated within the interstitium and arranged in a distinctly nested, whorled pattern typical of meningothelial-like nodules. Immunohistochemical staining patterns supported the diagnosis of meningoendothelioma.

Discussion

The clinical significance of DPM is unclear as data is mostly limited to case reports. While pathogenesis of solitary MLNs is agreed to be reactive in nature, DPM may represent a transition between reactive and neoplastic process. The cases reported thus far in literature have varied in terms of cigarette exposure, presence of symptoms, PFT results etc. The factors that make our case stands out out from the 11 previously reported DPM cases are 1) the presence of a normal pulmonary function test 2) presence of cavitation in DPM nodules which can erroneously suggest granulomatous or infectious processes. This was noted in only a few previous cases. 3) the fact that the diagnosis was made by transbronchial biopsy while others relied mostly on transthoracic biopsy.

Since the radiographic presentation in DPM can mimic metastatic malignancy, miliary tuberculosis, idiopathic interstitial pneumonia etc., a tissue diagnosis is important to obtain. Management of DPM is usually conservative with observation and follow up imaging. In our patient, we did a 3 month follow up CT and another one at 1 year mark, both of which showed stable nodules. The plan is to continue annual imaging for 2 years and then discontinue if lesions remain stable



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