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

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF 1 **Adham K. Alkurashi**
ABCDE 2 **Yahya Almodallal**
ABCDEF 3 **Hasan Ahmad Hasan Albitar**
AB 4 **John C. Cheville**
ABCDEF 5 **Vivek N. Iyer**

1 Division of Cardiovascular Diseases, Mayo Clinic, Rochester, MN, U.S.A.
2 Department of Pediatrics, Mayo Clinic, Rochester, MN, U.S.A.
3 Department of Internal Medicine, Mayo Clinic, Rochester, MN, U.S.A.
4 Department of Laboratory Medicine and Pathology, Mayo Clinic, Rochester, MN, U.S.A.
5 Division of Pulmonary and Critical Care Medicine, Mayo Clinic, Rochester, MN, U.S.A.

Corresponding Author: Vivek N. Iyer, e-mail: iyer.vivek@mayo.edu

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Patient: Female, 55-year-old
Final Diagnosis: Diffuse pulmonary meningotheliomatosis (DPM)
Symptoms: Chest discomfort • dry cough
Medication: —
Clinical Procedure: —
Specialty: Pulmonology

Objective: Rare disease

Background: Diffuse pulmonary meningotheliomatosis (DPM) is an exceedingly rare diffuse pulmonary disease with a female predominance. It is characterized by the presence of widespread bilateral minute pulmonary meningothelial-like nodules (MPMNs) on chest imaging. Patients are generally asymptomatic or may present with nonspecific symptoms such as dyspnea. The nodules are typically detected incidentally on imaging for other indications. Here, we present a rare case of DPM in a 55-year-old woman.

Case Report: A 55-year-old woman presented to the clinic with non-exertional chest pressure and dry cough of 4-month duration. She had a history of hypertension, hypercholesterolemia, hypothyroidism, gastroesophageal reflux disease, and impaired fasting blood glucose and was a lifelong nonsmoker. Physical examination was unremarkable. High-resolution chest computed tomography (CT) showed innumerable diffuse small ground-glass nodules. An extensive laboratory workup was negative for autoimmune and infectious etiologies. The patient underwent uncomplicated right video-assisted thoracoscopic surgery, and lung biopsy showed multiple well-circumscribed interstitial meningothelial-like nodules in perivascular distribution with occasional whorling of cells. The diagnosis of diffuse pulmonary meningotheliomatosis (DPM) was confirmed. The patient continued to complain of non-exertional chest pressure without pulmonary complaints, and a repeat chest CT showed stable findings 1 year after the diagnosis.

Conclusions: DPM should be considered in the differential diagnosis for patients presenting with diffuse bilateral pulmonary nodules. Patients are typically asymptomatic and it is most commonly detected incidentally. Further research is needed to better understand this disease and its clinical significance.

MeSH Keywords: Cough • Multiple Pulmonary Nodules • Pulmonary MedicineFull-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/926172>

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Background

Diffuse pulmonary nodules on chest computed tomography (CT) can signify airspace or interstitial disease processes. The differential diagnosis is broad and examples include benign neoplasms such as hamartomas and pulmonary papillomatosis, malignant neoplasms such as metastases and bronchoalveolar cell carcinoma, infections such as miliary tuberculosis, inflammatory disorders such as granulomatosis with polyangiitis, pulmonary arteriovenous malformations, and hypersensitivity pneumonitis, among others [1,2].

Diffuse pulmonary meningiotheliomatosis (DPM) is an exceedingly rare diffuse pulmonary disease that presents as diffuse pulmonary nodules on imaging. It is characterized by the presence of widespread bilateral minute pulmonary meningothelial-like nodules (MPMNs). It is more common in middle-aged females and patients with a history of malignancy [3]. Diagnosis is typically established with a lung biopsy. Here, we present a case of DPM in a 55-year-old woman presenting with non-specific cardiopulmonary symptoms.

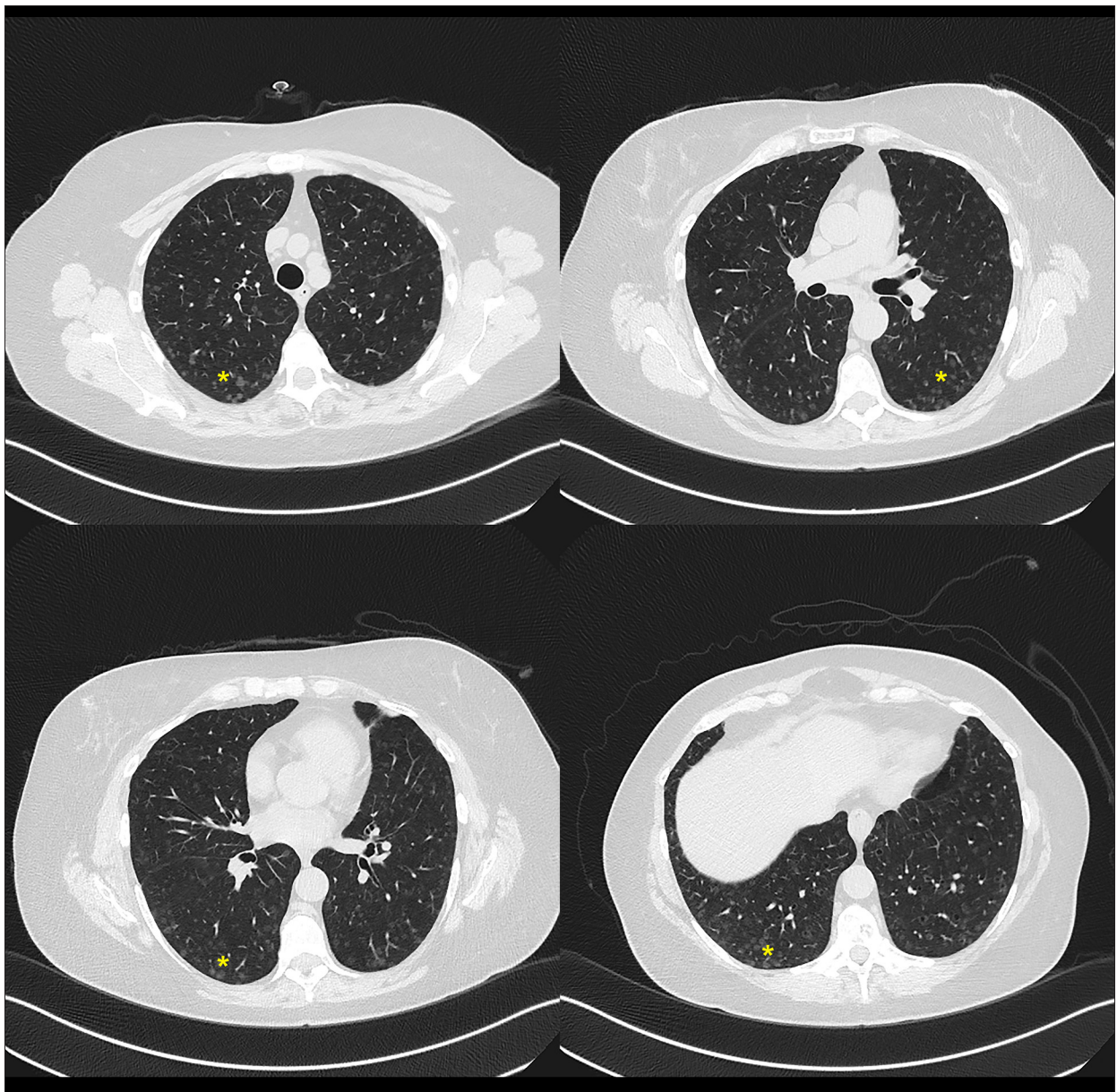


Figure 1. High-resolution chest computed tomography showing innumerable diffuse small ground-glass nodules (asterisks), some of which demonstrate central cavitation.

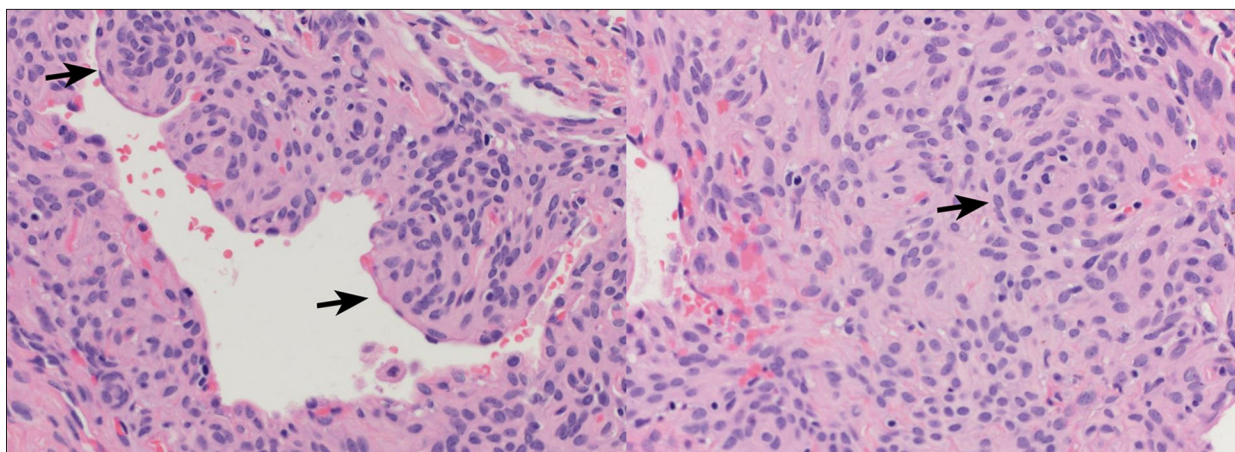


Figure 2. Histologic sections showing multiple well-circumscribed interstitial meningothelial-like nodules (arrows) with the predominant perivascular distribution. The nodules are composed of oval-to-spindle cells with indistinct cellular borders and uniform oval nuclei. Whorling of the tumor cells can also be appreciated. Overall, the morphologic features are consistent with diffuse pulmonary meningotheliomatosis.

Case Report

A 55-year-old woman presented to the clinic with non-exertional chest pressure and dry cough of 4-month duration. The patient was a lifelong nonsmoker and worked as a saleswoman. Her past medical history was significant for hypertension, hypercholesterolemia, hypothyroidism, gastroesophageal reflux disease, and impaired fasting blood glucose, on lisinopril 10 mg, levothyroxine 125 mcg, and omeprazole 20 mg daily. She denied any other symptoms. A physical examination did not reveal any abnormalities. A 12-lead electrocardiogram was normal. High-resolution chest computed tomography (CT) showed innumerable diffuse small ground-glass nodules, some of which were cavitory in nature (Figure 1).

Laboratory tests, including complete blood count, inflammatory markers, antinuclear antibody (ANA), anti-dsDNA, anti-scl-70 and RNA polymerase III antibodies, rheumatoid factor, and anti-cyclic citrullinated peptide (CCP) antibody, in addition to antineutrophil cytoplasmic antibodies (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 perivascular distribution with occasional whorling of cells, consistent with diffuse pulmonary meningotheliomatosis (DPM) (Figure 2). One year after 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

Minute pulmonary meningothelial-like nodules (MPMNs) were initially described in 1960 and were thought to be chemoreceptors due to the close proximity to blood vessels and their resemblance to chemodectomas [3,4]. However, this was later refuted as ultrastructural and immunohistochemical analysis showed a strong resemblance to meningothelial cells; hence, they were called MPMNs [5]. Patients with MPMNs are usually asymptomatic or may present with nonspecific cardiopulmonary symptoms such as in the case described above [3]. MPMNs are typically detected on chest CT scans as isolated pulmonary nodules and can be discovered incidentally on imaging performed due to other indications [3].

On the other end of the spectrum is DPM, which is an exceptionally rare form of widespread MPMNs with few cases reported in the literature [6–9]. DPM has a strong female preponderance (female: male ratio of 9: 1) and is classically seen in middle-aged females [10]. Radiographic features of DPM include diffuse innumerable bilateral pulmonary nodules on chest CT, some of which may have central cavitation [3]. Transbronchial or surgical (VATS or open) lung biopsies can be used to confirm the diagnosis of DPM.

This rare entity is considered to be of unclear clinical significance. However, it is worth mentioning that malignancy can be seen in a significant number of patients. This was highlighted by Gleason et al. in their review paper, where 44% of patients had a history of or active malignancy at the time of DPM diagnosis [3]. There is a scarcity of data on DPM, and further studies are needed to better understand this disease, its natural course, and clinical implications.

Conclusions

This case highlights that DPM should be considered in patients presenting with diffuse pulmonary nodules and that further research is needed to better understand this disease and its clinical significance.

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Conflicts of interest

None.