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CLINICAL IMAGE

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# Diffuse pulmonary meningotheliomatosis

**Anthony Nadrous**[1] | **Sushilkumar Sonavane**[2] | **Andras Khoor**[3] | **Alejandra Yu-Lee Mateus**[1] | **Augustine Lee**[1]

[1]Division of Pulmonary, Allergy, and Sleep Medicine, Mayo Clinic Florida, Jacksonville, Florida, USA

[2]Department of Radiology, Mayo Clinic Florida, Jacksonville, Florida, USA

[3]Department of Laboratory Medicine and Pathology, Mayo Clinic Florida, Jacksonville, Florida, USA

**Correspondence**
Augustine Lee, Division of Pulmonary, Allergy, and Sleep Medicine, Mayo Clinic Florida, 4500 San Pablo Rd S, Jacksonville, FL 32224, USA.
Email: lee.augustine@mayo.edu

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**Key message**

Diffuse pulmonary meningotheliomatosis (DPM) is an ultra-rare pulmonary disease characterized by innumerable bilateral minute meningothelial-like nodules, sometimes presenting a characteristic 'cheerio-sign' on imaging. Most patients with DPM are asymptomatic and experience no disease progression. Although little is known about its nature, DPM may be associated with pulmonary malignancies, mostly lung adenocarcinoma.

**KEYWORDS**

chest computed tomography, pulmonary meningotheliomatosis, pulmonary nodule

## CLINICAL IMAGE

Diffuse pulmonary meningotheliomatosis (DPM) is a rare pulmonary disease of unclear aetiology. It is characterized by innumerable bilateral minute pulmonary meningothelial-like nodules (MPMN's) and is most common in females between 50 and 70 years old. Most patients are asymptomatic, and the nodules are typically non-progressive. A 49-year-old woman, never smoker, and on hormone replacement therapy (HRT), presented with a nonproductive cough and dyspnea. Chest computed tomography showed numerous, randomly distributed, bilateral, solid and ground glass micronodules of 2–4 mm (shown in Figure 1A). Several nodules exhibited the characteristic 'cheerio-sign' marked by central lucency (shown in Figure 1B, arrows). A wedge biopsy showed multiple perivenular nodules (shown in Figure 1C) composed of nests of bland cells containing a moderate amount of cytoplasm, round to oval nuclei, and finely granular chromatin (shown in Figure 1D). The patient's symptoms improved after the withdrawal of her HRT, and the nodules have remained stable for the last 2 years. Less than 30 cases of DPM are reported in the literature. Although DPM is considered benign, there may be associations with other malignancies, such as lung adenocarcinoma,[1] and potential hormonal influence since most cases are in women and many MPMN's are immunoreactive with progesterone receptors.[2] As lung cancer screening increases, the 'cheerio-sign' can be helpful in identifying DPM, with a broader differential including adenocarcinoma.

## AUTHOR CONTRIBUTIONS

Mr. Nadrous was responsible for writing the manuscript. Dr. Sonavane and Dr. Khoor submitted images and descriptions of chest CT and histological findings, respectively. Dr. Yu Lee-Mateus and Dr. Lee reviewed and contributed significantly to the manuscript, and Dr. Lee was responsible for the study design and conceptualization. The final manuscript was reviewed and approved by all authors.

## CONFLICT OF INTEREST STATEMENT

None declared.

## DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

## ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

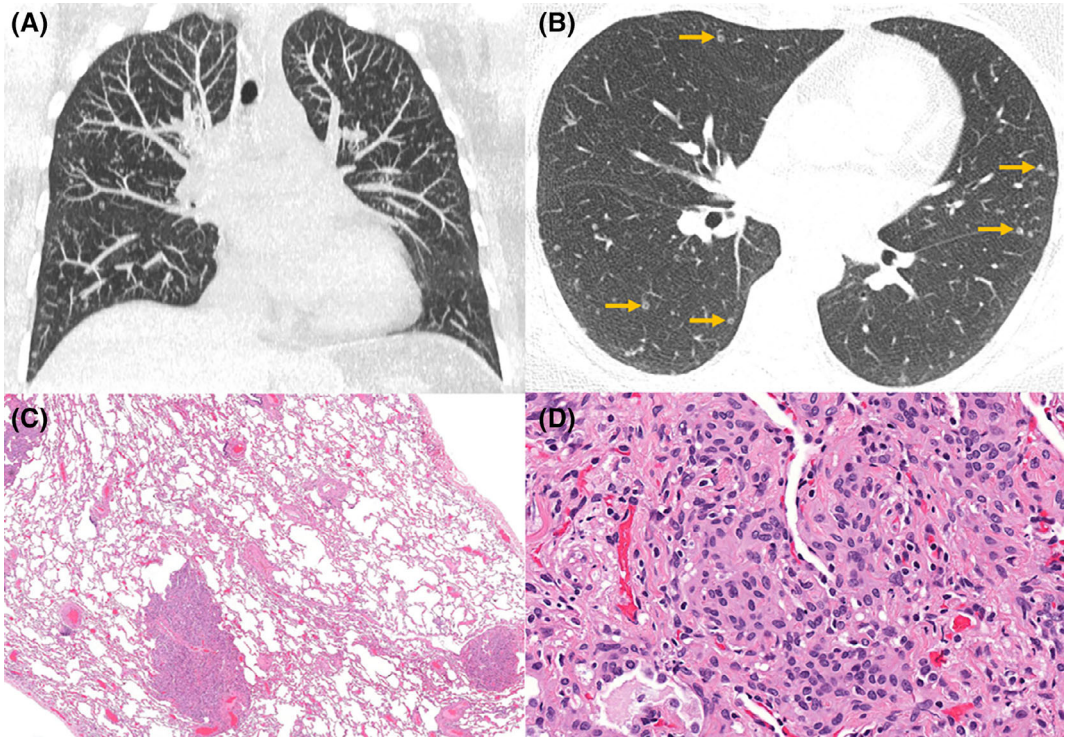


**FIGURE 1** (A) Computed tomography (CT) of the chest in coronal view showing numerous, randomly distributed, bilateral, solid and ground glass micronodules of 2–4 mm. (B) CT of the chest in axial view showing several nodules exhibiting the characteristic 'cheerio-sign' marked by central lucency (yellow arrows). (C) A thoracoscopic wedge biopsy showing multiple (3) perivenular nodules (haematoxylin & eosin, original magnification ×20). (D) High magnification of DPM biopsy revealing nests of bland cells containing a moderate amount of cytoplasm, round to oval nuclei, and finely granular chromatin (haematoxylin & eosin, original magnification ×400).

## ORCID

*Anthony Nadrous* https://orcid.org/0009-0009-6062-9269

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