

Diffuse Pulmonary Meningotheliomatosis

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Diffuse pulmonary meningotheliomatosis (DPM) is defined as numerous minute pulmonary meningothelial-like nodules (MPMNs), which are proliferations of meningothelial-like cells within the lung interstitium that manifest diffusely throughout the lungs (1–3). DPM is most commonly diagnosed in women in their 6th or 7th decade of life (4). Patients are typically asymptomatic but may present with dyspnea or cough (1,2,5). DPM is associated with various lung diseases including pulmonary embolism and primary adenocarcinoma; its exact cause is un-

known (1,4,5). At imaging, DPM manifests as numerous small (<6 mm) ground-glass or solid nodules throughout the lungs in a random distribution (4). Some nodules may have a central lucency (“Cheerio” sign) (Figs 1, 2) (2,5,6). The differential diagnosis for the Cheerio sign includes metastatic mucinous adenocarcinoma and Langerhans cell histiocytosis. The diagnosis of DPM is often made at imaging, but if histopathologic confirmation is required, surgical biopsy, or less commonly transbronchial biopsy, can be performed (Fig 3) (1,3,5).

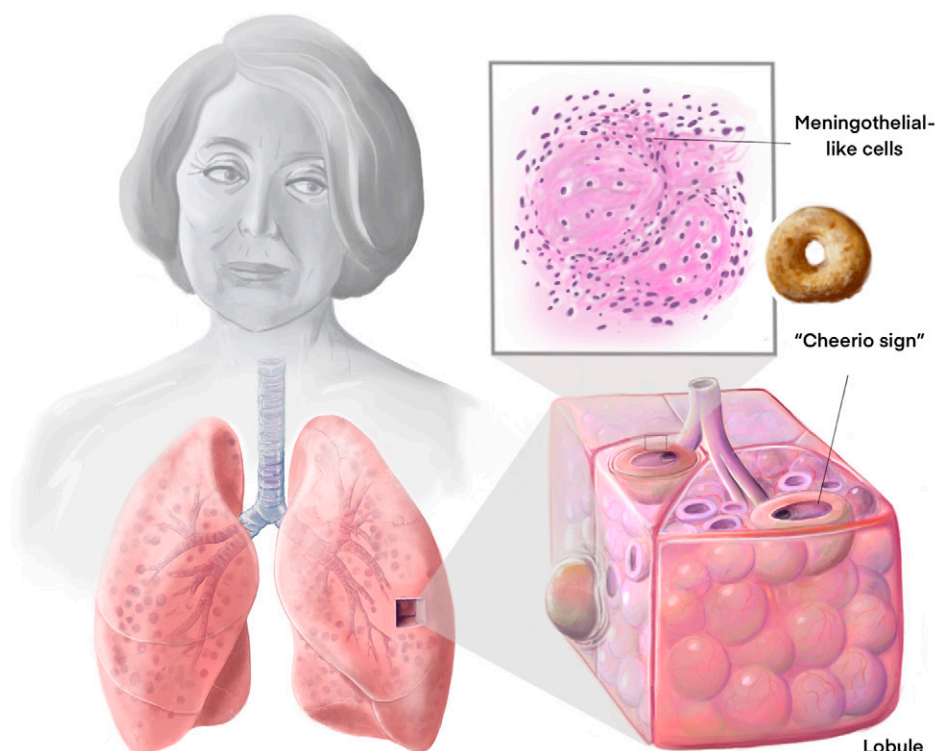


Figure 1. Medical illustration depicts DPM. DPM primarily affects older women. The main imaging feature is the presence of small pulmonary nodules randomly distributed throughout the lung tissue. Some of these nodules display a central lucency, commonly referred to as the “Cheerio” sign due to its resemblance to the O-shaped breakfast cereal. Histologically, these nodules consist of clusters of spindle cells that resemble meningothelial cells. The formation of a Cheerio sign occurs when these clusters surround airspaces.

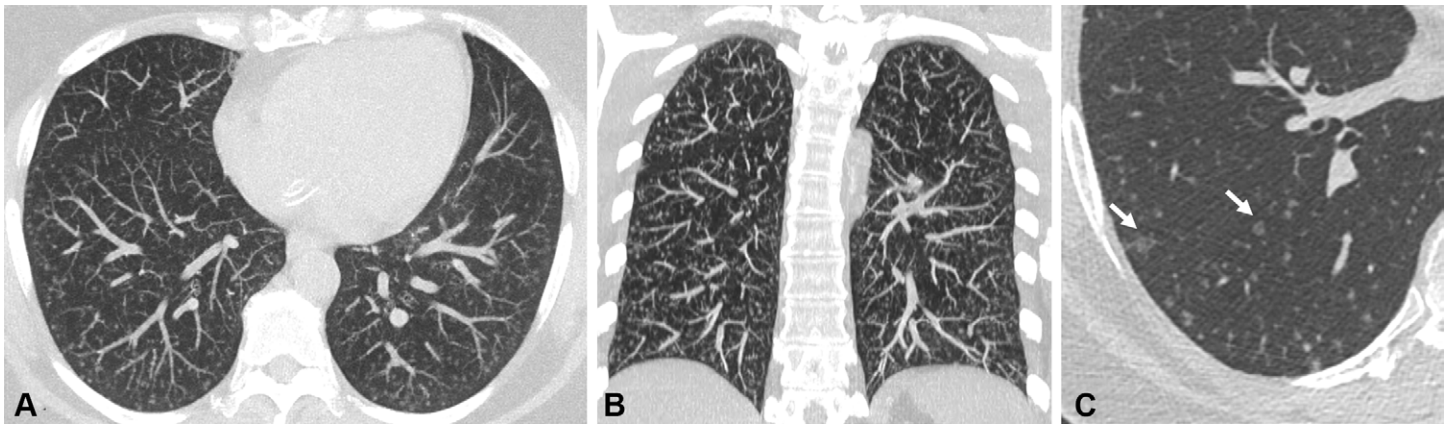


Figure 2. DPM in a 52-year-old woman with chronic exertional dyspnea. The patient had no history of smoking, illicit drug use, occupational exposure, or pets. Axial (A) and coronal (B) maximum intensity projection CT images demonstrate numerous small nodules throughout both lungs in a random distribution. Close-up axial CT image (C) shows that some nodules (arrows) have central clearing (Cheerio sign).

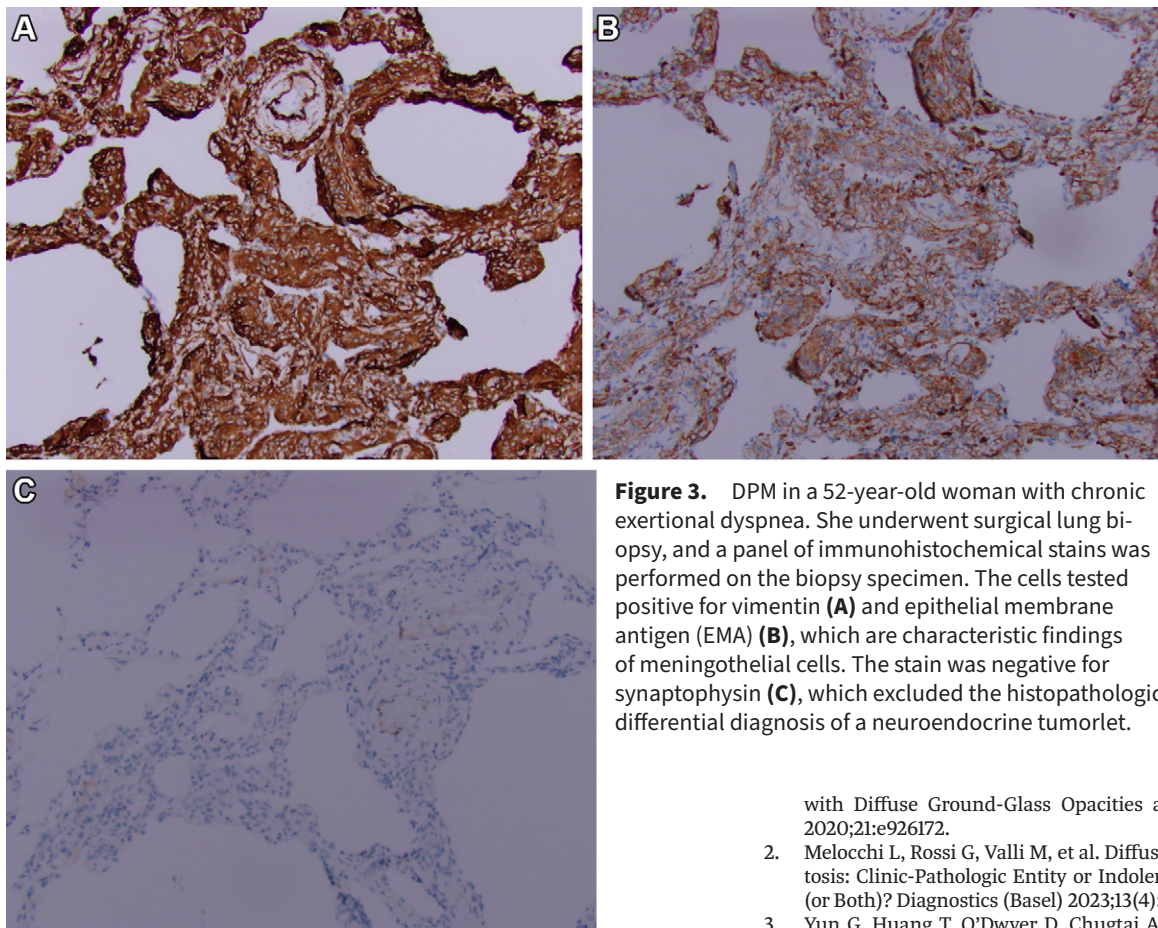


Figure 3. DPM in a 52-year-old woman with chronic exertional dyspnea. She underwent surgical lung biopsy, and a panel of immunohistochemical stains was performed on the biopsy specimen. The cells tested positive for vimentin (A) and epithelial membrane antigen (EMA) (B), which are characteristic findings of meningotheelial cells. The stain was negative for synaptophysin (C), which excluded the histopathologic differential diagnosis of a neuroendocrine tumorlet.

Acknowledgment.—Pathology images courtesy of Jon H. Ritter, MD, Department of Pathology and Immunology, Washington University School of Medicine in St Louis, St Louis, Mo.

Disclosures of conflicts of interest.—All authors have disclosed no relevant relationships.

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