Lung Cancer

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LONG-TERM FOLLOW-UP OF PATIENTS WITH BIOPSY CONFIRMED DIFFUSE PULMONARY MENINGOTHELIOMATOSIS

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PURPOSE: Diffuse pulmonary meningotheliomatosis (DPM) is a rare disease characterized by innumerable minute pulmonary meningothelial-like nodules (MPMN's). DPM is more common in females, presenting between 50-70 years old. Although DPM is considered benign, there may be associations with other malignancies. Given its rarity, long-term prognosis and optimal management strategies remain uncertain.

METHODS: A retrospective study of patients with biopsy confirmed DPM at Mayo Clinic sites.

RESULTS: All 11 patients identified were female with a median age of 58 years (IQR 52.3, 63.8). Most patients reported either cough, dyspnea, or chest discomfort, but in 2 the nodules were found incidentally. Five (46%) patients had a prior history of cancer, and one (9%) was diagnosed with lung adenocarcinoma concurrently with DPM. Four (36%) were former smokers. Three (27%) were receiving hormone replacement therapy (HRT) at the time of diagnosis. Chest computed tomography (CT) showed innumerable small (<5mm) bilateral ground glass nodules and three (27%) exhibited central cavitation ("cheerio"-sign), typical of DPM. Histology showed meningothelial nodules characterized by oval to spindle cells with indistinct cellular borders and some whorling. Immunochemical staining was positive for vimentin, EMA, and CD56 in one patient, and ER in another. In a median follow-up of 3.5 years (IQR 1, 5.75), 80% showed no progression of disease. One subject showed slight growth in size and number of MPMN's 7 years later, while another developed a new 9x12 mm nodule 6 years after the initial DPM diagnosis. One patient noted symptomatic improvement after discontinuing HRT.

CONCLUSIONS: Although little is known about this disease, our cohort represents the largest single center series of patients with biopsy confirmed DPM, contributing to less than 30 cases reported in the literature. We confirm a female predominance, mostly occurring after the age of 50, with a possible association with cancer (54.5%). Most were symptomatic, and nodules remained stable on long-term follow-up, though one presented a slow progression of the MPMN's. A third of the women were on HRT with one symptomatic patient improving after discontinuing HRT, suggesting a pathophysiologic link to estrogens/progestins. Whether DPM's relation to a neoplastic propensity is coincidental or indicative of a shared mechanism predisposing these patients to cancer remains unclear.

CLINICAL IMPLICATIONS: These findings confirm prior observations linking cancers and the hormonal system to DPM and provide reassuring long-term prognostic data. With increased lung cancer CT screening efforts, the "cheerio"-sign may facilitate its recognition and management. Further studies should provide additional insight into the pathophysiology of DPMs and suggest possible interventions in the rare progressive or symptomatic patient.

DISCLOSURES:

No relevant relationships by Andras Khoor

No relevant relationships by Augustine Lee

No relevant relationships by Anthony Nadrous

No relevant relationships by Sushilkumar Sonavane

No relevant relationships by Alejandra Yu Lee Mateus

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