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Diffuse Lung Disease

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Diffuse Pulmonary Meningothelial-Like Nodules: A Case Series

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PURPOSE: Diffuse pulmonary involvement by meningothelial-like nodules (PMNs) is an often incidentally-detected rare condition that is challenging due to unclear diagnostic and prognostic significance. We sought to assess frequency of diffuse PMN and its pathologic and clinical correlates to aid in advising management.

METHODS: We queried medical records at the Mayo Clinic, MN for PMNs noted on lung pathology specimens (2000-2016).

RESULTS: 46 patients (43 females; 93.5%) had 1 or more PMNs noted on lung pathology specimens. The average age at diagnosis was 61 (range: 27-77). In 7 patients, biopsy was performed for evaluation of diffuse micronodular disease with/without cavitation detected on imaging. Of these patients with 'diffuse pulmonary meningotheliomatosis', 5 patients had micronodular disease as the sole radiologic abnormality - 2 had no additional pulmonary pathology detected on biopsy, and 3 patients had findings suggestive of chronic hypersensitivity pneumonitis (follow up period (f/u): 1-6 mo). 1 patient with an associated GGO on imaging was found to have an area of atypical adenomatous hyperplasia (f/u: 38 mo). 1 patient who had biopsy for increased micronodule size and number was diagnosed with adenocarcinoma in situ in addition to diffuse PMN (f/u: 95mo). 1 or more PMNs were reported in other patients who underwent biopsy for known or suspected malignancy (26), interstitial lung disease (7), lung surface irregularity noted intra-operatively (5), and on a bullectomy specimen (1). 23 of these 39 patients had 5 or more PMNs. Reported size of nodules was between 1-6mm. Frequent associations of PMNs noted in non-neoplastic specimens (27) included granulomas (7), chronic bronchiolitis (10), atypical adenomatous hyperplasia (AAH) (3), and emphysema (4).

CONCLUSIONS: Our patients were predominantly female, and older - a trend observed in other analyses as well (Kuroki et al., 2002). When present incidentally, PMNs are noted in specimens with underlying inflammation, injury or tumor, and are likely a reaction to these, as opposed to precursors or etiologic agents of disease or malignancy in themselves, and may not be reported. Other series have reported association with thromboembolism, smoking-related bronchiolitis, interstitial lung disease, cardiac disease and malignancy (Mukhopadhyay at al., 2009). There may be a genetic or hormonal predisposition to the formation of PMNs. In the uncommon cases of patients with diffuse micronodular disease detected on imaging and confirmed as diffuse PMN on biopsy as the sole abnormality, there is inadequate experience or evidence to make recommendations regarding management.

CLINICAL IMPLICATIONS: A diagnosis of diffuse PMN as a cause for micronodular disease should only be made after biopsy. One or more PMN first detected in association with lung inflammation or tumor are likely markers of injury. For the patients with multiple PMNs detected incidentally and confirmed pathologically as the sole abnormality, these are likely benign. However, until such time that adequate longitudinal follow-up data or further evidence is available with regards to their etiology and pathogenesis, clinical judgement based on risk factors should be utilized in recommending follow-up, oncologic work-up or further imaging.

DISCLOSURE: The following authors have nothing to disclose: Shireen Mirza, Anja Roden, Charles Thomas

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