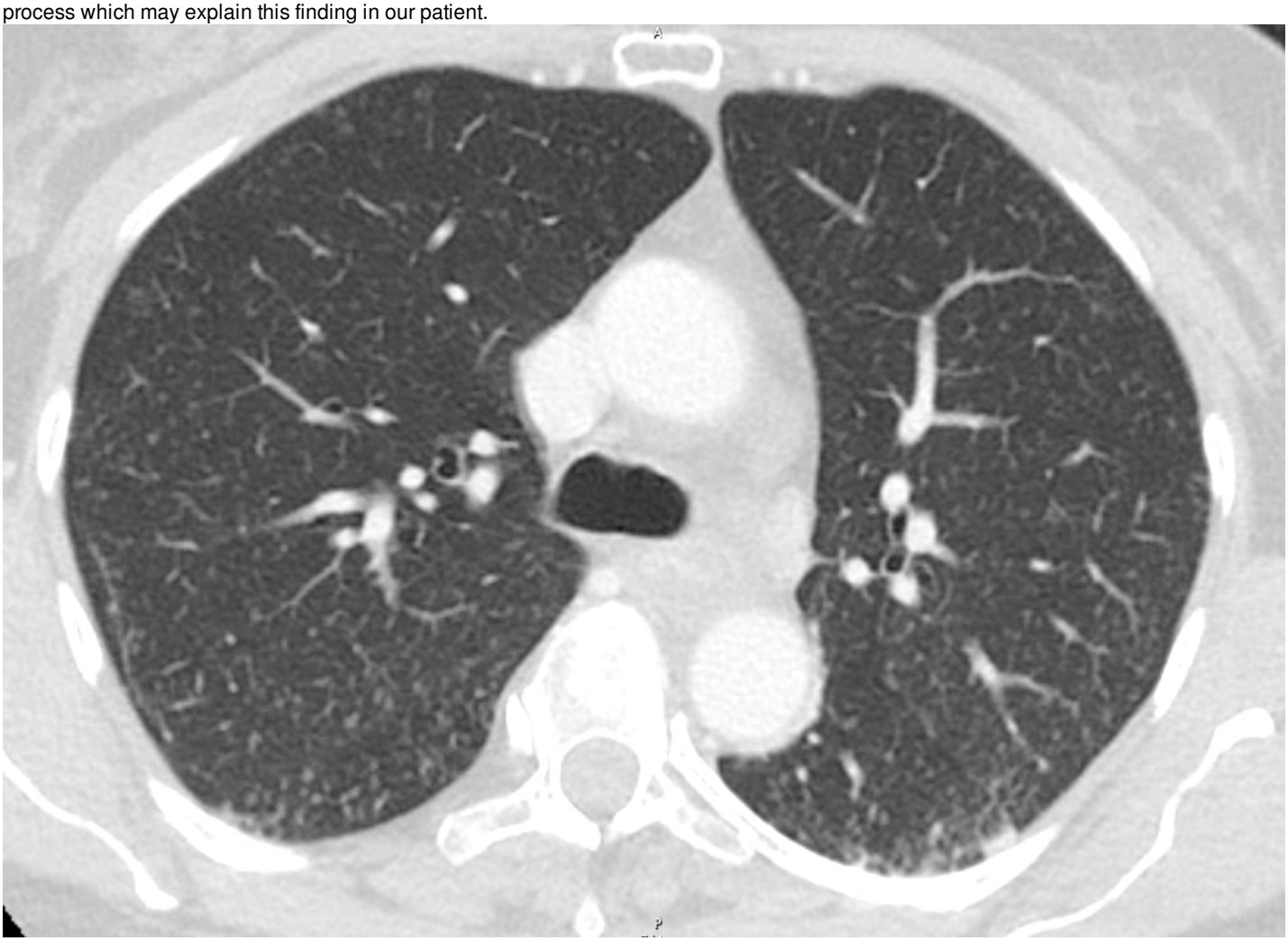
Meningotheliomatosis Presenting as Miliary Nodules in a Patient with Polyarteritis Nodosa

P. T. Neilan[1], R. Reddy[2]; [1]Internal Medicine, Univeristy of Florida, Gainesville, FL, United States, [2]Division of Pulmonary, Critical Care and Sleep Medicine, Gainesville, FL, United States.

Introduction: Diffuse pulmonary meningotheliomatosis is a rare disease of unknown etiology that is often found incidentally and displays heterogeneity on imaging from single nodules to diffuse micronodules. These usually benign findings are an important consideration in the evaluation of miliary pattern of lung nodules. We report a case of a meningotheliomatosis presenting as diffuse miliary nodules in a 54 year old woman with polyarteritis nodosa. Case Presentation: A 54 year old woman with a history of hypothyroidism presented for rapidly progressing lower extremity weakness secondary to a nontraumatic epidural hematoma and underwent T11/12 laminectomy and expansile duraplasty and L4 laminectomy. Subsequent imaging with CT Chest and MRA of the abdomen demonstrated miliary pulmonary nodules and luminal contour irregularities suspicious for aneurysms in the kidney and hepatic arteries. The patient underwent an extensive infectious workup including respiratory viral panel, serologies for fungal infections, RAST allergy panel, HIV, sputum culture with AFB x3, bronchoalveolar lavage, hypersensitivity pneumonitis panel, RPR, and Hepatitis C and B all of which were negative. Autoimmune workup including ANCA was negative. CRP was 144 and ESR was 53. Rheumatology was consulted and given the clinical and radiographic findings, the patient was diagnosed with polyarteritis nodosa. Transbronchial biopsy revealed meningothelial like micronodules. Further workup with wedge resection demonstrated diffuse meningotheliomatosis evidenced by multiple, patchy scattered foci of EMA+, PR+ whorled epithelioid cells with negative staining for chromogranin and synaptophysin. The patient was started on corticosteroids and discharged to rehabilitation on cyclophosphamide, dosed per the French Vasculitis Study Protocol. She improved clinically and was started on methotrexate. At follow-up nearly 2 years post discharge she was walking without a walker, her inflammatory markers normalized, and she had no pulmonary symptoms. Discussion: Miliary nodules have a large differential including tuberculosis, sarcoidosis, pneumoconiosis, metastatic cancer, histoplasmosis, varicella infection, and Langerhans cell histiocytosis. Meningotheliomatosis is a rare etiology of military nodules but should be considered. Meningothelial-like nodules have been described predominantly affecting females and those with a history of malignancy and may confer a significantly higher risk of malignant pulmonary tumors, most commonly lung adenocarcinomas. Histologically, these nodules are often positive for progesterone, EMA, vimentin and negative for myosin, actin, S-100, keratin, chromogranin A and synaptophysin. When compared to meningioma, meningothelial-like nodules lack consistent monoclonal expansion. Because they lack mutational damage, meningotheliomatosis may be the result of reactive proliferation rather than a neoplastic process which may explain this finding in our patient.



This abstract is funded by: Nonr

Am J Respir Crit Care Med 2020;201:A6648
Internet address: www.atsjournals.org