Diffuse Pulmonary Meningotheliomatosis in a Patient With Neurodermatitis With Prurigo Nodularis

To the Editor:

Diffuse pulmonary meningotheliomatosis (DPM) is a rare cause of diffuse small pulmonary nodules with ground glass opacifications and cavitations. The clinical significance remains unclear, and the disease may be initially mistaken for malignancy or interstitial lung disease (ILD) on thoracic imaging. The diagnosis of DPM should be considered in middle-aged female patients presenting with diffuse bilateral ground glass opacifications on computerized tomography (CT) of the chest with an otherwise non-diagnostic workup. We present a unique case of this unusual diagnosis.

CASE REPORT

A 55-year-old woman presented to our interventional pulmonology clinic for evaluation of 9 months of shortness of breath, exertional dyspnea, and abnormal thoracic imaging. She was a non-smoker, had no second-hand smoke exposure, and worked in an office with no occupational risk factors. She described symptomatology consistent with Raynaud's phenomenon in her distal extremities and skin pruritus. Her past medical history was notable for obesity and a hysterectomy 25 years prior. Her vital signs and pulmonary exam were normal, but her physical exam was notable for a perioral rash and a blanching maculopapular rash on the legs, arms, and buttocks with central ulceration previously felt to be consistent with neurodermatitis with prurigo nodularis, and a palpable cervical lymph node. The rash was treated with topical steroids without any response. A left posterior neck lymph node biopsy showed dematopathic lymphadenitis. A CT of her chest showed innumerable small ground glass nodules, the largest of which was 5mm, and some of which demonstrated cavitation (Fig. 1, images 1 – 4). There were also scattered intra-pulmonary calcified granulomas. There was no evidence of pulmonary embolism. A CT of her abdomen and pelvis showed left groin lymphadenopathy. A positron emission tomography whole body CT demonstrated only faint metabolic activity in the left groin region.

Laboratory studies were all normal, including a complete blood count, complete metabolic panel, C-reactive protein, erythrocyte sedimentation rate, antinuclear antibody, rheumatoid factor, cyclic citrullinated peptide, anti-centromere antibody, myeloperoxidase and proteinase 3 (PR3) anti-neutrophil cytoplasmic antibody, beta-2 glycoprotein antibody IgG and IgM, anticardiolipin antibody IgG and IgM, cryoglobulin, soluble interleukin-2 receptor (s-IL-2), and myositis panel. A broad infectious workup was negative, including a QuantiFERON gold, histoplasma antigen, and coccidioidomycosis serology and complement fixation. Pulmonary function testing was unremarkable.

The differential diagnosis included inflammatory (connective tissues disease, vasculitis, hypersensitivity pneumonitis, Langerhans cell histiocytosis), infectious (atypical pneumonia, viral pneumonia, tuberculosis, fungal pneumonia), and malignant (atypical adenomatous hyperplasia, adenocarcinoma in situ) causes of diffuse pulmonary nodules. Malignancy and infection were felt to be less likely than inflammatory, as it was suspected that the lung nodules, Raynaud's phenomenon, and neurodermatitis with prurigo nodularis were an interconnected underlying process.

After a multidisciplinary tumor board assessment, infection was felt to be unlikely, and there was a greater concern for interstitial lung disease (ILD) or malignancy. The nodules were too small for a CT-guided biopsy, and a tissue diagnosis was felt to be required. She underwent uncomplicated surgical robotic-assisted thoracic surgery (RATS) with multiple left lung biopsies. Pathology showed meningothelial-like nodules with no evidence of malignancy. Immunohistochemistry showed positive staining for epithelial membrane antigen (EMA), vimentin, and progesterone (Fig. 1, image 5–7), low staining for Ki-67, and negative staining for AE1/AE3, thyroid transcription factor-1 (TTF-1), S-100, Langerin, and CD1a. On the basis of the pathology report and the radiographic imaging demonstrating diffuse disease, a diagnosis of DPM was made. She was seen in follow-up 1 year after diagnosis with seemingly stable disease by thoracic CT imaging and resolving left groin lymphadenopathy.

Pulmonary meningothelial tumors are a rare benign neoplasm of unclear significance. They were first described in 1960 by Korn et al[1] as a chemodectoma due to histologic morphology and proximity to nearby blood vessels. These nodules were then more descriptively named in 1988 by Gaffey et al as minute pulmonary meningothelial-like nodules (MPMNs) after verification with immunohistochemical analysis showing reactivity for EMA and vimentin and no reactivity

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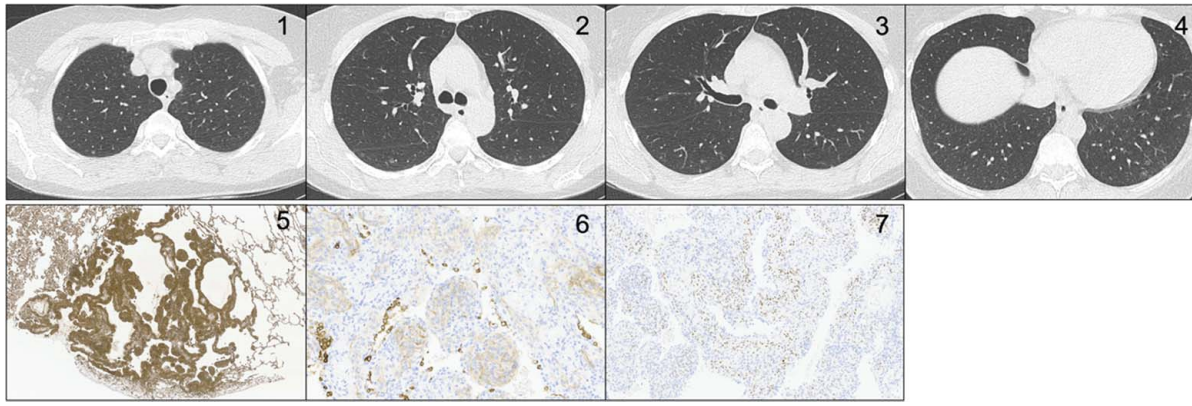

FIGURE 1. Thoracic CT 1.3 mm cuts (image 1 to 4) demonstrating diffuse tiny intraparenchymal nodules, some of which are cavitating. Biopsy showing positive histology for vimentin (image 5), EMA (image 6), and progesterone (image 7). EMA indicates epithelial membrane antigen.

to S-100.[2] These were later further characterized after the nodules were observed to contain endocrine granules and histologically resemble meningothelial cells.[3,4] However, a mutational analysis comparing intracranial meningiomas to MPMNs demonstrated a lack of monoclonal cell lineage and lack of mutational damage, suggesting that MPMNs are a reactive process rather than a neoplastic one.[5,6] Some patients present with few or solitary MPMNs, while others present with diffuse parenchymal lung disease, which is now termed DPM. Whether MPMNs and DPM are a spectrum of the same underlying process or altogether separate diseases remains uncertain, and their clinical significance likewise is unclear.

Most reported cases of DPM are incidentally diagnosed in middle-aged females, with a median age of 59.5 years.[7] A minority of reported cases have been diagnosed due to unexplained respiratory symptoms of dyspnea and/or cough. Radiographic imaging will demonstrate bilateral diffuse tiny ground glass nodules (< 3 mm in size) in a miliary pattern, some of which may have central cavitation. Some reports of DPM have noted an apical distribution.[7] As a result, it is a mimicker of lung malignancy and ILD. Biopsy is almost always done by surgical biopsy, but some cases have been diagnosed by transbronchial biopsy. Immunohistochemistry will show reactivity for EMA and vimentin and no reactivity to S-100. In a review of DPM by Gleason et al,[7] 44% of patients identified with DPM had either active or previous malignancy.

This case presents another layer of consideration due to the ulcerating skin findings and Raynaud's phenomenon in this patient. To the best of our knowledge, there are no other reports on DPM associated with cutaneous manifestations, and although we suspect a relationship between the cutaneous disease and her DPM, this is speculation based on an otherwise diagnostically negative workup. We suspect that an underlying reactive inflammatory process is driving her DPM, neurodermatitis with prurigo nodularis, and lymphadenopathy. She will continue serial yearly imaging to monitor the DPM, but we are not sure how long to follow this disease, and further research is required to address the longitudinal management of DPM.

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A Pilot Clinical Evaluation of a New Single Use Bronchoscope

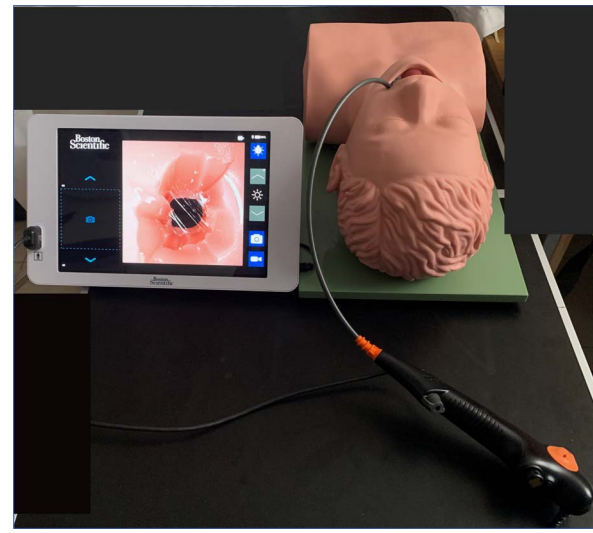

FIGURE 1. EXALT model B (Boston Scientific Corporation, Marlborough, MA) single use bronchoscope with image of distal airway from a low fidelity simulator on the accompanying EXALT monitor.

To the Editor:

A recently published article outlines the immense superiority in suction of the EXALT Model B (Boston Scientific Corporation, Marlborough, MA) single use bronchoscope (SUFB) (Fig. 1) in comparison to a commonly used SUFB using a "pseudo-mucus" compound.[1] This superiority is a key advantage in bronchoscopic procedures such as suctioning out thickened secretions, patients with haemoptysis, foreign body removal and suction around instruments during procedures. Our initial clinical experience supports this preclinical study.[1] In our pilot clinical study, with ethical approval, data was prospectively collected on 24 patients (mean age: 61, 50% female). Fifteen procedures were performed with the larger 2.8 internal diameter scope and 9 with the 2.2 scope. Other than bronchoalveolar lavage (n = 22) procedures (8/24 = 33%) included endobronchial biopsy (n = 3), endobronchial cryobiopsy (n = 1), transbronchial needle aspiration (n = 1), and transbronchial biopsy (n = 1). Two debulking of malignant airway obstruction procedures were performed in theater through a rigid bronchoscope including biopsy, mechanical debulking, argon plasma coagulation and electrocautery using the 2.8 EXALT SUFB. It was noted that the scope was rigid enough to mechanically debulk an endobronchial tumor and allowed electrocautery without loss of image. Our recent experience with SUFBs support their use through rigid bronchoscopy when teaching trainees due to no risk of damage of a reusable scope through the rigid scope. In both theater cases and in another patient having transbronchial lung biopsies for query recurrence of lung cancer, we noted the ability to suction blood around an instrument which we hypothesise is related to the unique working channel shape.[1]

We used a Likert scale to quantify user satisfaction with a maximum score of 5 recorded in 23/24 cases. There were no scope complications, however one technical issue was noted where there was an inability to pass a forceps into the anterior segment of the right upper lobe. We have previously noted loss of angle related to instrumentation in reusable scopes.[2]

The advantages of SUFBs have been postulated which include portability, lack of staff and personal protective equipment required to clean and sterility which are all advantages during the COVID-19 pandemic.[3,4] Other advantages include cost and potentially a reduction in carbon footprint.[5,6] The cost effectivity of SUFBs not only depends on this risk of infection but also the number of procedures a year (the more procedures, the more cost-effective reusable scopes are). The largest analysis to date (a microcosting analysis with 2.8% risk of infection) of the cost effectiveness of SUFBs in comparison to reusable scopes concluded that in a tertiary referral ICU center undertaking 1000 bronchoscopic procedures per year the cost of SUFB was <50% of reusable

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