A 42 year-old woman with recurrent pneumothoraces and dyspnea
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Lymphangioleiomyomatosis (LAM) is a rare multisystem disorder of unknown etiology characterized by the proliferation of atypical smooth cells in the lungs, pleura, mediastinum, thoracic duct and retroperitoneum. The occurrence is 1 case per 1,000,000 persons, but the actual incidence may be higher due to misdiagnosis. LAM occurs either sporadically or as a complication of tuberous sclerosis complex. Renal angiomyolipomas can occur in as many as 50-80% of all patients with LAM. We report a case of a woman who presented with a spontaneous pneumothorax and was ultimately diagnosed with LAM.

A 42-year-old woman with a history of asthma presented to her primary care physician with acute onset of shortness of breath, chest discomfort and productive cough. She was diagnosed with bronchitis but with worsening symptoms she subsequently presented to the Emergency Department. A chest x-ray revealed a left sided tension pneumothorax, for which a chest tube was urgently placed. The patient’s inpatient recovery was uncomplicated and she was discharged home.

Five years later the patient developed left sided chest pain. A chest-computerized tomography (CT) was negative for pneumothorax or pulmonary embolism but did reveal emphysema and a solid left renal mass measuring 3.8 X 2.5 cm. A left nephrectomy was performed and the biopsy was consistent with an angiomyolipoma. A year later a CT scan of the abdomen for follow up of previous incidental liver lesions confirmed thin walled cysts in the lung. LAM was suspected, and she was referred to a pulmonologist. An alpha-1 antitrypsin level, anti-neutrophilic cytoplasmic antibody, and magnetic resonance imaging of the brain were all normal. The patient developed another pneumothorax and was admitted to the hospital. A left video-assisted thoracoscopic exploration, along with mechanical pleurodesis and left lung biopsies were performed. Histologic results were consistent with LAM.

LAM is often misdiagnosed due to nonspecific presenting symptoms. Although there are currently no proven therapies for LAM, new treatments are on the horizon. LAM may be more common than previously recognized and the clinical course may be indolent and non-specific, easily obscuring its diagnosis.

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