Lung biopsy with proliferation of placed in fascicles surrounding venous structures (16).

B) of 2 days of sharp, left sided chest pain radiating to her left neck and upper back associated with shortness of breath and an episode of presyncope. Past medical history significant for chronic sinusitis and an episode of bronchitis, treated 2 months prior to her presentation. Family history was insignificant. Patient was a never smoker and lived a very healthy, active lifestyle. She worked as a sales representative for a technology company requiring frequent travel across the country.

**Physical Exam**

<table>
<thead>
<tr>
<th>Vital</th>
<th>Date: 36.5 °C (97.7 °F), Heart Rate: 82, BP (Non-Invasive): 115/71, Respiratory Rate: 20, SpO2: 91% on room air</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Constitutional:</strong></td>
<td>Appears in respiratory distress</td>
</tr>
<tr>
<td><strong>Head:</strong></td>
<td>Normocephalic and atramatic.</td>
</tr>
<tr>
<td><strong>Neck:</strong></td>
<td>Normal range of motion. No JVD.</td>
</tr>
<tr>
<td><strong>Cardiovascular:</strong></td>
<td>Normal rate and regular rhythm. No murmur heard.</td>
</tr>
<tr>
<td><strong>Pulmonary/Chest:</strong></td>
<td>Tachypneic, Lung sounds absent in left lower lobe and decrease in left upper lobe, lung sounds clear in right lobes</td>
</tr>
<tr>
<td><strong>Abdominal:</strong></td>
<td>Soft, non-tender, positive bowel sounds</td>
</tr>
<tr>
<td><strong>Musculoskeletal:</strong></td>
<td>Normal range of motion and strength. No edema on bilateral lower extremities.</td>
</tr>
<tr>
<td><strong>Neurological:</strong></td>
<td>She is alert and oriented to person, place, and time.</td>
</tr>
<tr>
<td><strong>Skin:</strong></td>
<td>Skin is warm and dry. No rash noted. No jaundice.</td>
</tr>
</tbody>
</table>

**Imaging and Pathology**

![Image](image1.png)

Figure 2: Histological staining of a patient with LAM.

C) Lung biopsy with proliferation of spindle-shaped myoid cells placed in fascicles surrounding venous structures (10).

**Figure 1: Actual CT Chest with IV contrast of our presented patient. A and B) Left sided pneumothorax shown with severe emphysema**

**Hospital Course and Management**

The patient appeared pale, diaphoretic and in respiratory distress with an oxygen saturation of 91% on room air upon presentation to the ED. Physical exam was significant for diffusely diminished lung sounds especially on her left lung fields. Chest x-ray and CT chest was significant for large left spontaneous pneumothorax, for which urgent pigtail thoracostomy was performed, and severe emphysema. She was admitted with a left sided chest tube on nasal cannula oxygen for further management and workup of her spontaneous pneumothorax and severe emphysema. Pulmonology and Cardiothoracic Surgery was consulted for assistance in this case. Workup included a renal ultrasound negative for angiomyolipoma, negative HIV screen, no alpha-1-antitrypsin deficiency, normal ACE levels, and absent anti-SSA or anti-SSB antibodies but a mildly positive ANA titer of 1:40.

Patient was unable to be transferred to a higher level of care due to insurance issues. Thus, the patient opted to proceed with surgical management at the community hospital. She underwent chemical pleurodesis to prevent further pneumothoraces with Video-Assisted Thoracoscopic Surgery (VATS) of the left lower lobe with wedge excision, which was sent to pathology, and pleural tent procedure to prevent air leaks postoperatively. She was extubated successfully onto nasal cannula oxygen and her postoperative course was uncomplicated. Pathology showed prominent cystic structures composed of glands on bland epithelium to spindle eosinophilic cells, which were strongly positive for Actin and Progesterone receptors with significant HMB45 and MITF staining. These findings were consistent with lymphangioleiomyomatosis (LAM).

She was later discharged in stable condition with nasal cannula oxygen and set up with a LAM specialty clinic. She was started on Sirolimus treatment to decrease the rate of progression of disease. About 7 months after her discharge, she successfully underwent bilateral lung transplantation.

**Lymphangioleiomyomatosis Management**

Patients who experience a spontaneous pneumothorax have higher risk of recurrence and therefore pleurodesis is a viable option to prevent further complications through lung re-expansion (4, 5). Transplant is recommended for patients with severe alveolar degression, when DLCO is < 40% of predicted and VO2max is < 50% of predicted (6). This can be managed pharmacologically with Sirolimus (immunosuppressant that targets MTOR and interrupts T cell activation downstream of the IL-2 receptor).

**Lymphangioleiomyomatosis Pathophysiology**

LAM typically occurs in females of childbearing age with pulmonary manifestations. Patients who undergo exercise testing may exhibit hypoxemia with poor ventilation and gas exchange (1). Pulmonary function testing may be significant for a decrease in forced expiratory volume in 1 second (FEV1) and diffusion capacity for carbon monoxide (DLCO) (3). Radiographic findings can be notable for hyperinflated lungs and diffuse thin-walled cysts evident in lung parenchyma (3). Workup includes diagnosis of exclusion with specific antibodies or titers such as anti-SSA antibodies, anti-SSB antibodies, ACE levels, alpha-1-antitrypsin levels, and VEG-F antibodies with definitive diagnosis limited to tissue confirmation.

**References**