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# Pneumothorax Secondary to Lymphangioleiomyomatosis

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## INTRODUCTION

Lymphangioleiomyomatosis (LAM) is a rare multisystem disease characterized by abnormal proliferation of smooth muscle-like cells, leading to cystic lung disease, abdominal angiomyolipomas, and lymphatic involvement, which may result in lymphadenopathy and chylothorax.[1] It occurs almost exclusively in women and may be sporadic or associated with tuberous sclerosis complex.[2]

Patients may be asymptomatic and diagnosed incidentally on imaging or present with progressive dyspnea or spontaneous pneumothorax.[3] Diagnosis is primarily clinical, based on characteristic imaging findings in conjunction with associated disease features.[4]

Due to its rarity, evidence guiding management is limited, and recommendations largely are based on expert opinion and case reports. Treatment for moderate to severe disease typically involves the mammalian target of rapamycin (mTOR) inhibitor sirolimus, while advanced cases may require lung transplantation.[5]

I present a case of a patient who developed an incidental contralateral spontaneous pneumothorax following Port-A-Cath placement and was subsequently diagnosed with LAM.

## CASE REPORT

A 44-year-old woman with no significant past medical history was diagnosed with triple-negative breast carcinoma after palpating a left-sided breast mass. She had experienced a multi-year delay in screening mammography due to the COVID-19 pandemic. Following biopsy and staging, she began neoadjuvant chemotherapy with plans for subsequent surgical resection.

She underwent right subclavian Port-A-Cath placement, and a post-procedure chest radiograph revealed a moderate left-sided pneumothorax (Figure 1). She was asymptomatic, hemodynamically stable, and maintaining adequate oxygen saturation on room air. She was transferred from the outpatient surgery center to The University of Kansas Hospital for further evaluation and management.

Computed tomography (CT) of the chest demonstrated a moderate left hydropneumothorax, right pleural effusion, and innumerable thin-

walled cysts throughout both lungs, along with scattered bilateral pulmonary nodules suggestive of LAM (Figures 2 and 3). She remained clinically stable during admission and was discharged without intervention.

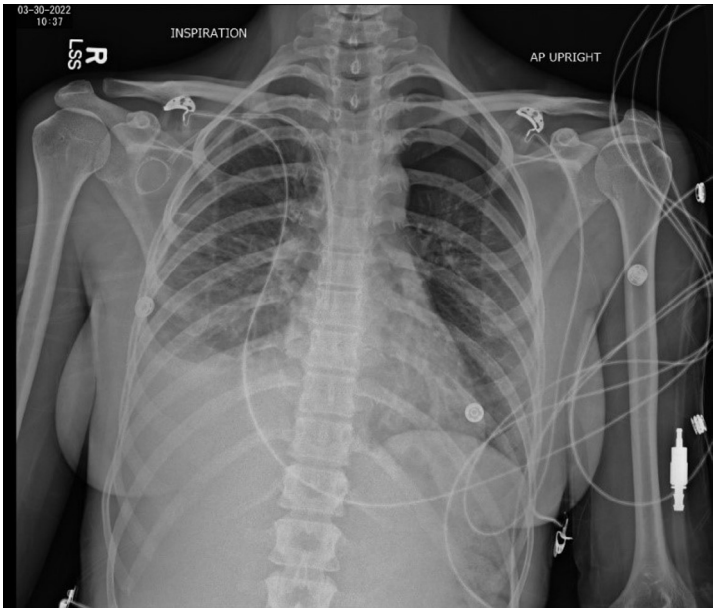


**Figure 1.** Chest x-ray with moderate left pneumothorax.

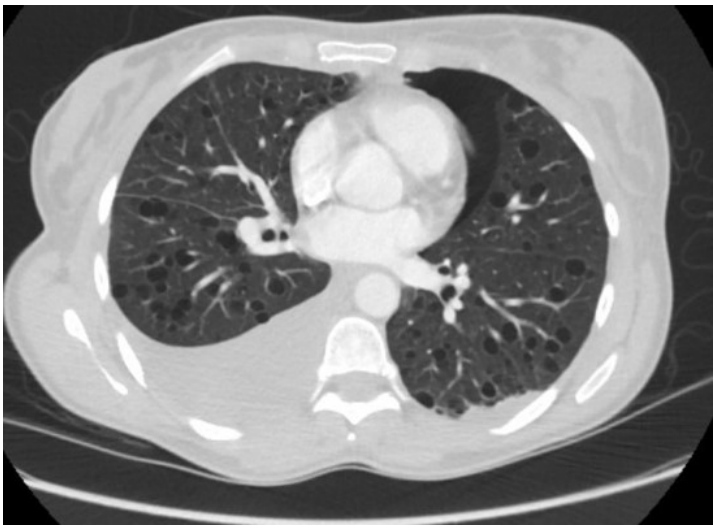

**Figure 2.** CT chest with contrast showing moderate left hydropneumothorax, right pleural effusion, thin-walled cysts, and scattered bilateral pulmonary nodules.

Given concern for LAM versus metastatic disease, additional evaluation was performed. CT of the abdomen and pelvis was negative for renal angiomyolipomas. Thoracentesis revealed chylothorax, and cytology was negative for malignancy. A repeat chest radiograph one month later showed improvement of the pneumothorax.

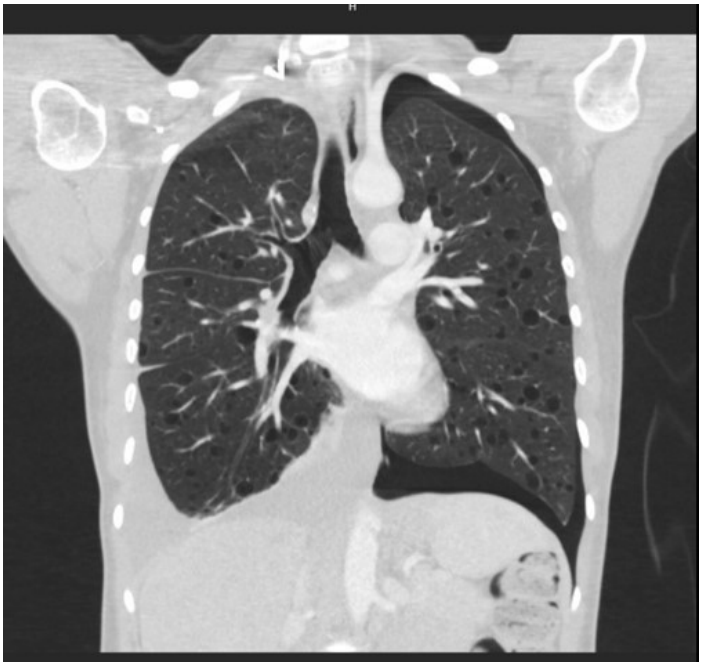

**Figure 3.** CT chest with contrast showing moderate left hydropneumothorax, thin-walled cysts, and scattered bilateral pulmonary nodules.

After completing neoadjuvant chemotherapy, she underwent total mastectomy with axillary lymph node dissection. Due to an elevated risk of pneumothorax, bilateral chest tubes were placed preoperatively. She subsequently received adjuvant chemotherapy and radiation therapy.

Nine months later, she developed a small spontaneous right-sided pneumothorax. Pleurodesis was recommended but declined. She has since completed breast cancer treatment and currently is in remission.

## DISCUSSION

This case describes a patient with triple-negative breast cancer diagnosed with LAM after a spontaneous pneumothorax contralateral to a recent port placement. LAM typically affects women aged 20-40 years but can occur in postmenopausal women.[6] Most cases are sporadic, with an estimated prevalence of 19 per million women, though recognition is increasing.[7]

Tuberous sclerosis complex (TSC), an autosomal dominant disorder characterized by hamartomas, is strongly associated with LAM; approximately 34% of women with TSC have radiographic evidence of LAM.[8,9] Mutations in TSC genes lead to dysregulated cell growth via mTOR pathway activation.[10]

LAM commonly presents with dyspnea on exertion and may include pneumothorax, hemoptysis, chylothorax, chylous ascites, and renal angiomyolipomas.[11] About one-third of cases are diagnosed following pneumothorax, and obstructive patterns are common on pulmonary function testing.[12] Evaluation includes pulmonary function testing and imaging, with differential diagnoses including other cystic lung diseases such as Birt–Hogg–Dubé syndrome and pulmonary Langerhans cell histiocytosis.[13]

Diagnosis should follow American Thoracic Society criteria using the least invasive approach.[4] In asymptomatic patients where diagnosis would not alter management, close clinical monitoring without invasive testing may be appropriate. Management depends on disease severity. Sirolimus, an mTOR inhibitor, stabilizes lung function and is recommended for patients with declining function or symptomatic chylous effusions.[15-17]

Spontaneous pneumothorax in LAM carries a high recurrence risk. Initial management includes chest tube placement, with consideration of blood patch for persistent air leak.[18] Pleurodesis or pleurectomy generally is recommended after the first episode, as recurrence rates are high and outcomes are similar between approaches.[1,19] Chylothorax may be managed with observation, drainage, or pleurodesis depending on severity.[20]

Exogenous estrogen may worsen disease and should be avoided; pregnancy may increase the risk of progression and complications.[21] Registry data suggest a median transplant-free survival exceeding 20 years, with better outcomes in those with preserved lung function.[22] Lung transplantation remains the definitive treatment for end-stage disease, with outcomes generally favorable compared to other indications, although recurrence in the transplanted lung has been reported.[23]

## CONCLUSIONS

LAM is a rare cystic lung disease that may occur sporadically or with TSC. It often presents with spontaneous pneumothorax, as in this case, and carries a high risk of recurrence, supporting early consideration of pleurodesis.

## ARTICLE INFORMATION

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