A CT Diagnosis of Lymphangioleiomyomatosis (LAM) in an Asymptomatic Postmenopausal Female

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LEARNING OBJECTIVES

- LAM should be considered for cystic findings on CT chest imaging, even in asymptomatic patients
- Effectiveness of early treatment with Sirolimus in LAM remains unclear

INTRODUCTION

Lymphangioleiomyomatosis (LAM) is a rare (5 per 1 million people) neoplastic disease characterized by

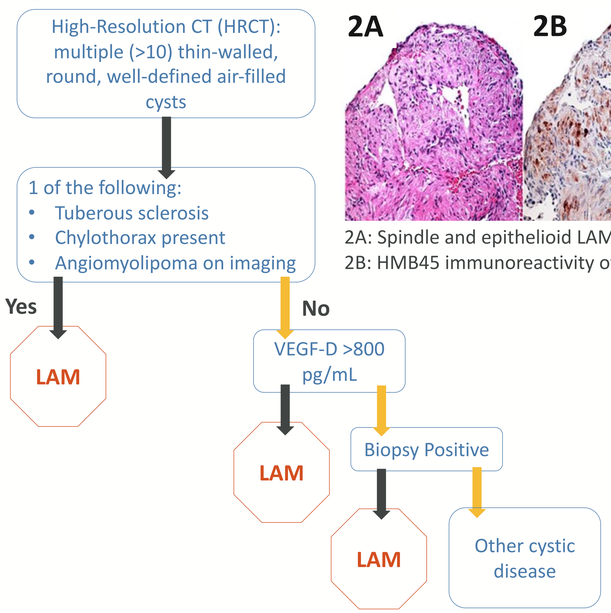
- Cystic lung destruction
- Preponderance for females of reproductive age
- Progressive respiratory failure

DISCUSSION

Radiographic Imaging Based Diagnosis:

- HRCT classic findings in ***all*** LAM patients exhibit are multiple round, thin-walled air-filled cysts that may vary in size and are evenly distributed throughout normal lung parenchyma[6]
- 76% of patients with LAM exhibit abnormal abdominopelvic imaging with 54% of these patients had renal angiomyolipoma[3]
- VEGF-D levels may be more reflective of active disease and lymphatic involvement[1], which may be why this asymptomatic patient had normal levels

Diagnosis Algorithm



2A: Spindle and epithelioid LAM cells[2]
2B: HMB45 immunoreactivity of LAM cells[2]

CASE REPORT

58-year-old never-smoking female presented to our pulmonary clinic with lung cysts found incidentally on CT Neck with no neck abnormalities.

Personal Medical History:

- Menopause from hysterectomy and bilateral oophorectomy
- On estrogen HRT
- Denies history of pneumothorax or autoimmune diseases

Physical exam:

- Vital signs stable with normal O_2 saturation on room air
- Cardiopulmonary and skin exam normal

Laboratory Results:

- ***BMP:*** Unremarkable
- ***Autoimmune Serologies:*** Negative
- ***Serum VEGF-D:*** <250 pg/mL

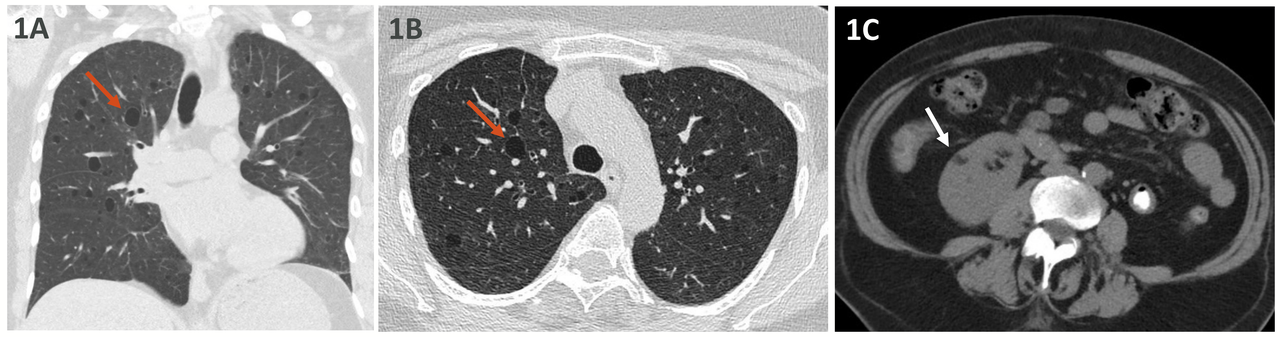
Pulmonary Function Test:

FEV_1: 82%
FVC: 83%
FEV_1/FVC: 100%
TLC: 91%
DLCO: 99%

Differential Diagnoses:

	Pulmonary Langerhans Cell Histiocytosis (PLCH)	Lymphocytic Interstitial Pneumonia (LIP)	Birt-Hogg-Dube (BHD)	Infectious Etiology	LAM
History/ Physical Exam Findings	Smoking history Skin rashes Diabetes insipidus	Autoimmune diseases	Folliculomas Renal tumors	PJP (immunocompromised)	Dyspnea Pneumothorax Chylothorax
HRCT Cyst Findings	Nodular and bizarrely-shaped cystic abnormalities in upper/middle zones	Randomly distributed cysts with internal structures, bordered by eccentric vessel	Round-lentiform thin-walled cysts of various sizes in basilar & subpleural regions	Numerous bilateral, upper lobe predominant cysts, variable in size and wall thickness	>10 thin-walled, round, well-defined air-filled cysts

Imaging Results:



Coronal (1A) and Axial (1B) are HRCT Chest images demonstrating multiple round, thin-walled air-filled cysts of various sizes, evenly distributed throughout normal lung parenchyma. The orange arrow in both images pinpoint to the same cyst.

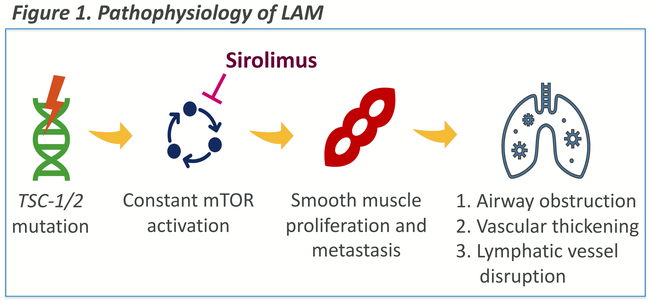
CT Abdomen/Pelvis (1C) demonstrates a 7.4mm macroscopic fat density (white arrow) with a Hounsfield reading of -68 to -23 present in the right kidney, findings supportive of an angiomyolipoma (AML).

DISCUSSION (CONT.)

An Unclear Disease Course:

- Common presenting symptoms of LAM include recurrent spontaneous pneumothorax, chylothorax, or worsening dyspnea in a reproductive-aged female as LAM cells can express estrogen hormone receptors[1]
- Gupta et. al (2019) characterized LAM's annual decline of FEV_1 as 89 +/- 53mL/yr regardless of baseline function
- Rate of FEV_1 decline slows post-menopause, but the decline is still notable at 74mL/yr[4]
- Major determinants of progression to death/transplant are menopausal status and disease baseline severity[4]
- Exogenous estrogens have been shown to cause exacerbation[5]

Figure 1. Pathophysiology of LAM



When to Treat:

- MILES clinical trial demonstrated Sirolimus, an mTOR inhibitor, improved quality of life and lung function measured by FEV_1 for patients with abnormal/declining lung function[1]
- Benefits unclear in asymptomatic patients with normal PFTs
- MILED study aims to determine if early, long-term sirolimus is effective in preventing disease progression and preserving quality of life

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