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*Clinical Image*

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# Asymptomatic presentation of a rare disease: Lymphangioleiomyomatosis

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**Abstract**

Lymphangioleiomyomatosis (LAM) is a rare disorder involving young female patients. We present a 35 years old female who presented to the emergency room for abdominal pain due to ureteral stone. Imaging study incidentally found pneumothorax as well as cystic lung disease. Further workup confirmed this to be LAM.

***Keywords:*** LAM; Pneumothorax; Cystic lung disease.

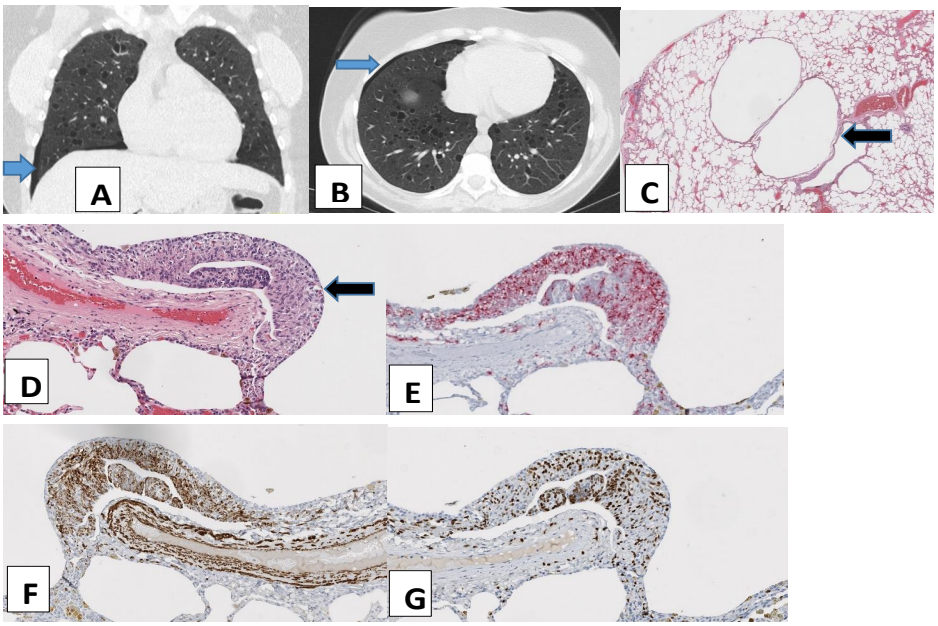


**Figure 1:** CT chest representative Coronal (A) and cross section (B) images show bilateral innumerable cystic lung lesions and right side small pneumothorax (arrows). Low power (C) and high power (D) hematoxylin and eosin stains show multiple cysts, with focal atypical thickened smooth muscle cyst lining (arrows). Image E shows positive for HMB45 ( seen as red color). Image F shows positive for desmin ( seen as brown color) and image G shows positive for progesteron receptor (seen as brown color)

## Clinical image description

A 35-year-old female with no previous diagnosed medical conditions presented to an emergency department due to complaints of right sided flank pain with nausea and vomiting. Patient delivered a healthy male child normally four months prior to presentation. CT scan of abdomen demonstrated right sided ureterolithiasis as well as a small right sided pneumothorax. Subsequent CT chest showed small pneumothorax and scattered multiple thin-walled pulmonary cysts bilaterally. Patient reported no pulmonary symptoms. The patient underwent video assisted thoracoscopic surgery procedure with right sided pleurodesis and surgical lung biopsy. Intraoperatively, multiple small cysts were visible on the surface of all three lobes of the right lung as well as some on the parietal pleura. The surgical lung biopsy specimen showed multiple cysts with an atypical smooth muscle lining, with variable spindled and epithelioid morphology. The atypical cells showed focal positivity for desmin (a smooth muscle marker), HMB45 (a melanocytic marker), and progesterone receptor. The radiographic and the staining on the pathology specimen pattern are typical of, and specific for lymphangioleiomyomatosis.

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