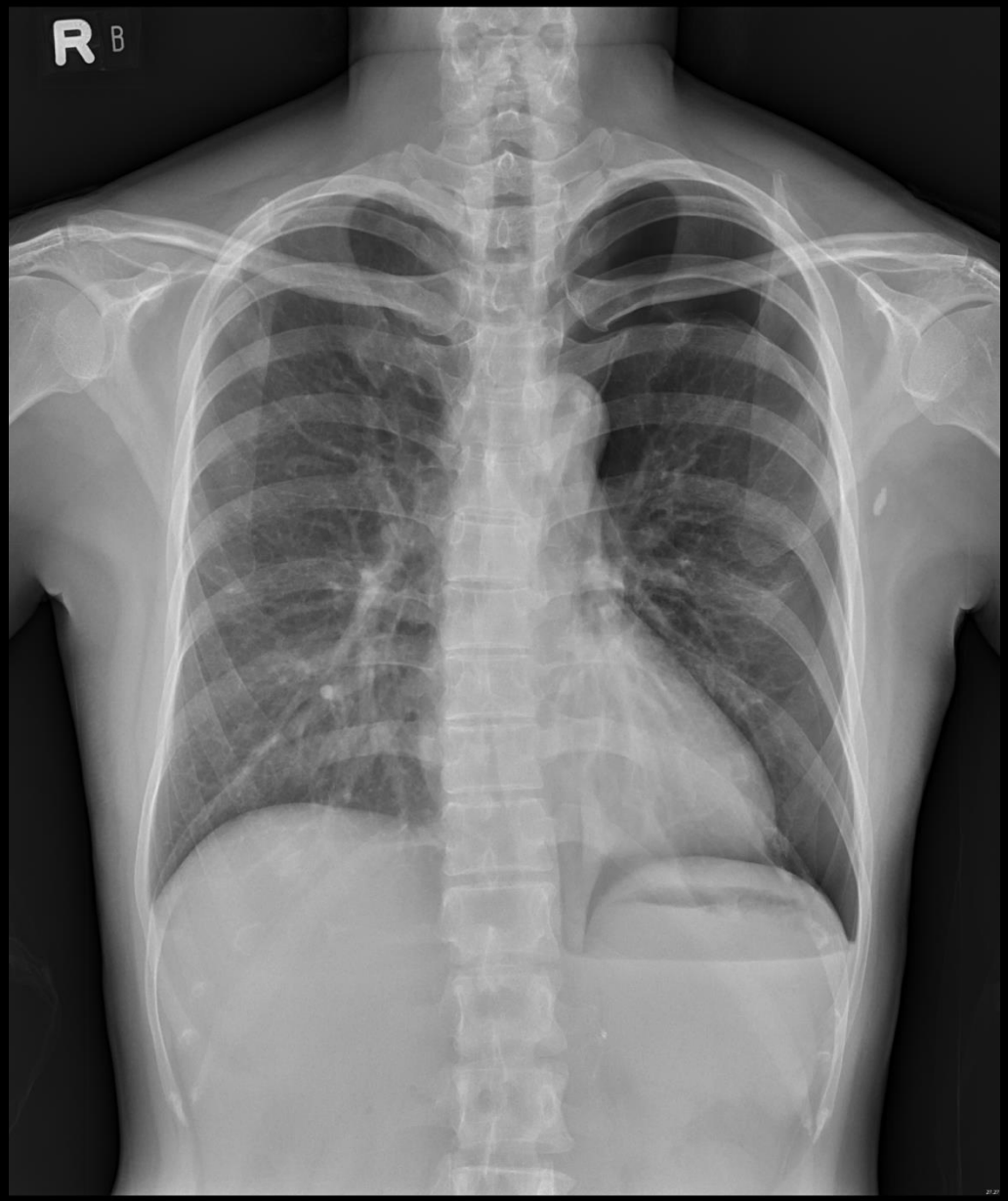
CASE 3

Case 3

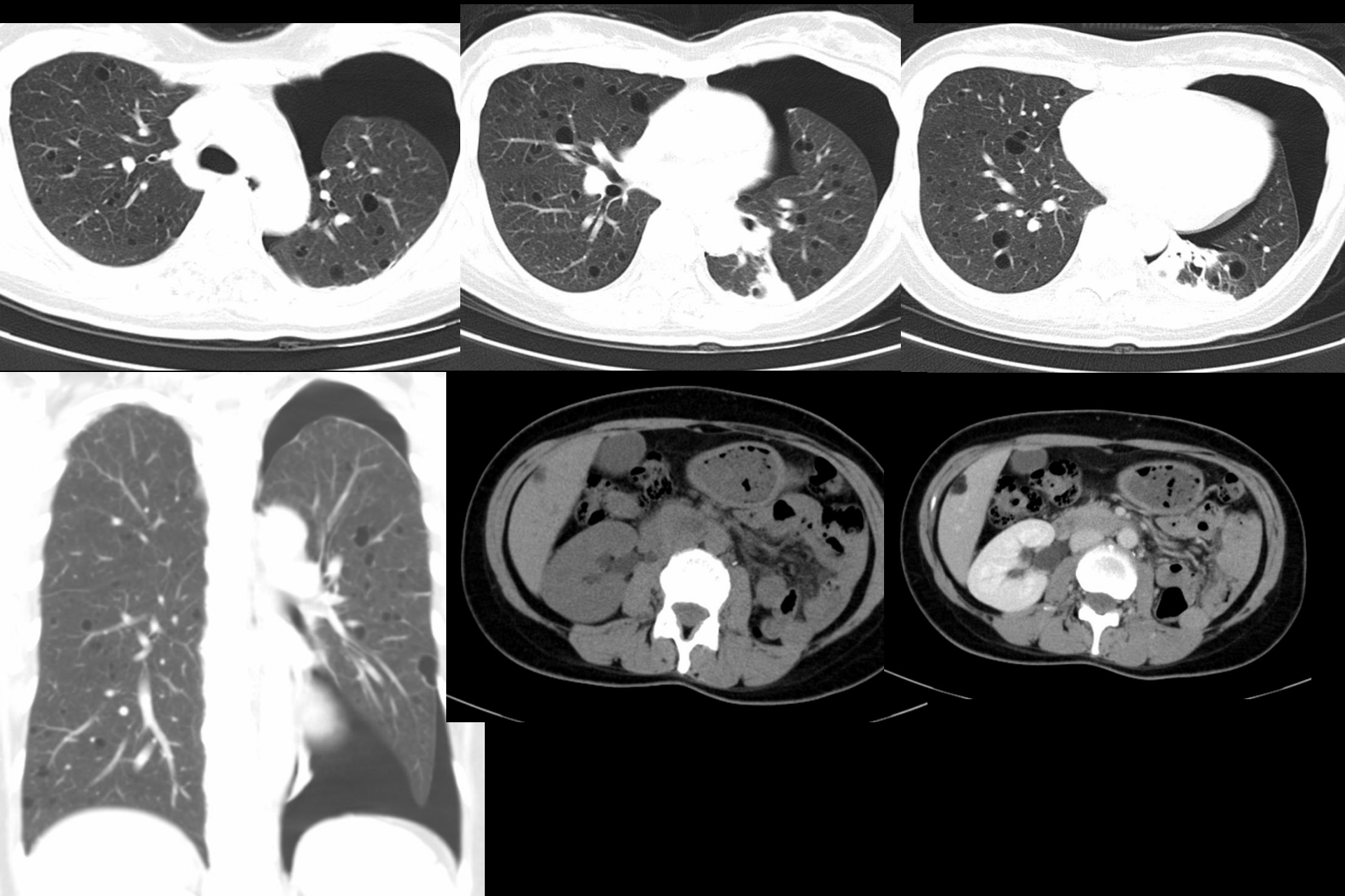
- 45 y/o female
- Non-smoker
- PHx: Left renal angiomyolipoma s/p left radical nephrectomy
- S: Chest tightness for 4 days
- O: Bloody tinged sputum

- 2017-03-23 CXR & Chest CT

2017-03-23 CXR



2017-03-23 Chest CT

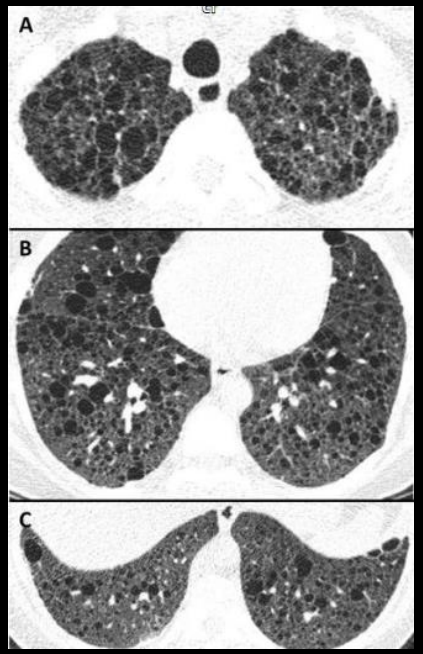


Differential diagnosis

- Lymphangioleiomyomatosis (LAM)
- Langerhans cell histiocytosis (LCH)
- Lymphocytic interstitial pneumonia (LIP)

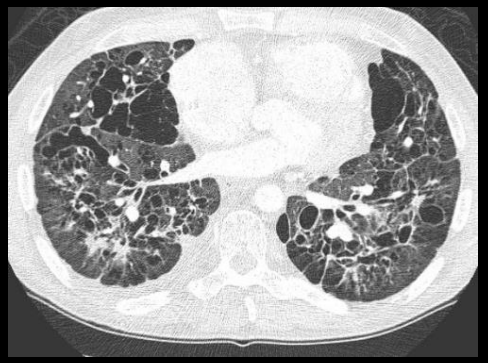
Lymphangioleiomyomatosis (LAM)

- Young women
- Thin walled cysts of variable sizes (mostly 2mm to 2cm) surrounded by normal lung parenchyma, diffuse distribution
- Recurrent spontaneous pneumothorax
- Chylothorax
- Sporadic or associated with tuberous sclerosis
  - ✓ Small lung nodules representing multifocal micronodular pneumocyte hyperplasia (MMPH) especially in tuberous sclerosis
  - ✓ Renal angiomyolipoma
  - ✓ Hepatic angiomyolipoma: Less strong than for renal AMLs.



Langerhans cell histiocytosis (LCH)

- Young smokers, no gender predilection
- Upper lung predominant, bizarre shaped cysts
- Nodules: Typical 1-5 mm
  - ✓ More pronounced early in the disease
  - ✓ Usually have irregular margins
  - ✓ Surrounding lung parenchyma appears normal



- Cysts (Thin or thick wall)
  - ✓ More pronounced later in the disease
  - ✓ Usually less than 10 mm in diameter
  - ✓ Confluence of 2 or more cysts results in bizarre shapes

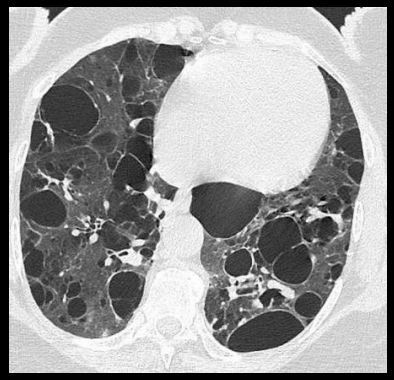
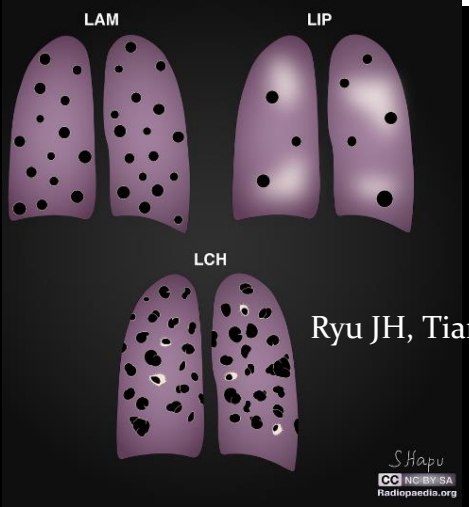


Table 2 Main characteristics of five major diffuse cystic lung diseases

	LAM	PLCH	LIP	BHD	Amyloidosis
Age at diagnosis	Adults	Younger adults, 3rd and 4th decades	Broad range	Adults	Adults
Gender	Nearly all women	No predominance	No predominance	No predominance	No predominance
Relevant history	Pneumothorax in majority; some have underlying TSC	Nearly all have smoking history; pneumothorax in 15%	Some have underlying CTD, immunodeficiency, etc.	Pneumothorax in 15%–25%; family history of BHD, renal tumor or pneumothorax	Some have systemic amyloidosis or underlying disease, e.g., CTD
Extrapulmonary manifestations	Renal angiomyolipoma, chylous ascites, stigmata of TSC	Bone lesions, diabetes insipidus	Signs of underlying disease, e.g., CTD, HIV infection, etc.	Benign skin tumors, renal neoplasms	Signs of underlying systemic amyloidosis, CTD, etc.
Laboratory testing	Elevated serum VEGF-D level, genetic testing (TSC)	NA	Dysproteinemia (polyclonal)	Genetic testing (*FLCN* gene mutations)	Dysproteinemia (monoclonal)
HRCT findings	Many round cysts, mostly 2 mm to 2 cm in size, with normal intervening lung parenchyma; diffuse distribution	Irregular cysts, often with nodules and architectural distortion of intervening parenchyma; relative sparing of lung bases	Cysts of varying sizes and shapes, often with ground-glass opacities, nodules, septal thickening and lymphadenopathy	Cysts of varying sizes and shapes; more prominent in lower lobes Disproportionately paramediastinal, elongated (floppy) cysts	Scattered cysts of varying sizes, often with nodules

BHD, Birt-Hogg-Dubé syndrome; CTD, connective tissue disease; HIV, human immunodeficiency virus; HRCT, high-resolution CT; LAM, lymphangioleiomyomatosis; LIP, lymphoid interstitial pneumonia; NA, not available; PLCH, pulmonary Langerhans' cell histiocytosis; TSC, tuberous sclerosis complex; VEGF-D, vascular endothelial growth factor-D.



Ryu JH, Tian X, Baqir M, Xu K. Diffuse cystic lung diseases. *Front Med*. 2013;7(3):316-327. doi:10.1007/s11684-013-0269-

Diagnosis

- Lymphangioleiomyomatosis (LAM)