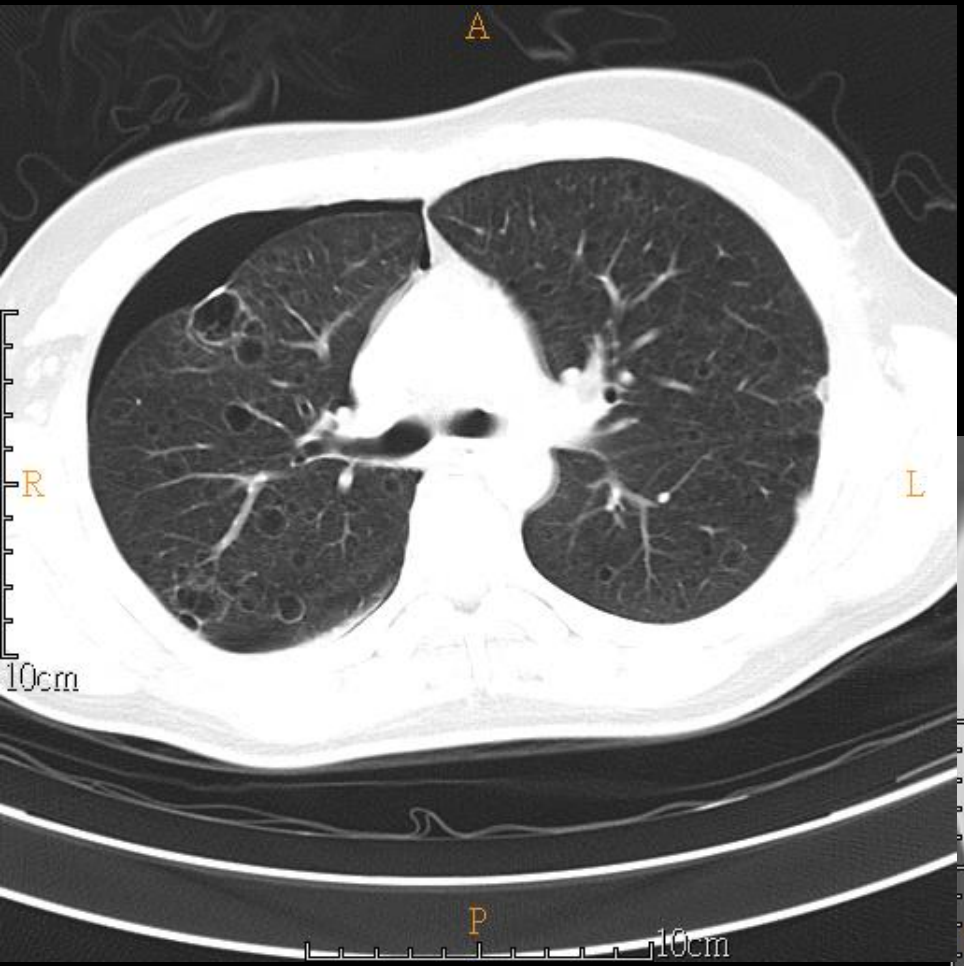
CASE 5
41 YEARS OLD WOMAN

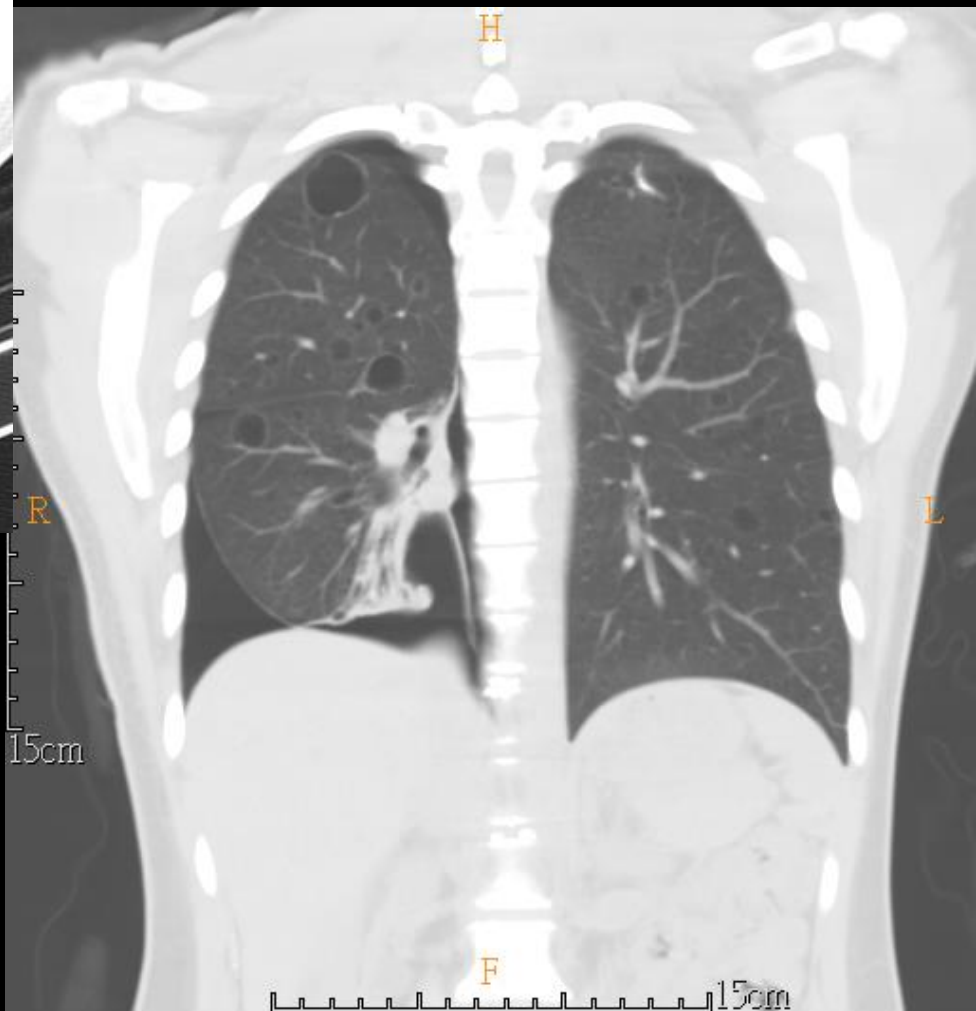
Brief history

- 41 years old woman
- Chief complain:
  - Short of breathe with right side chest pain

Imaging studies

- 2020/04/06 CXR
- 2020/04/07 Chest CT (NoC)


A
R
L
10cm
P
10cm


H
R
L
15cm
F
15cm

DDx:

- Lymphoid interstitial pneumonitis
  - Sjorgen disease etc.
- Lymphangioleimyomatosis

Lab

簽收日期	1090731	1090731	1090731
簽收時間	0958	0958	0958
工作單號	1201355645	1201355646	1201355646
試管編號	1201355645	1201355646	1201355646
報告日期	1090731	1090803	1090731
報告時間	1337	1425	1624
ANA [抗細胞核抗體法]		Negative[<1:80]	
anti-Ro [可抽出的核抗體測定— Ro/La 抗體]			<0.3 U/mL [<7.0]
anti-La [可抽出的核抗體測定— Ro/La 抗體]			<0.3 U/mL [<7.0]
anti-Ro 結果 [可抽出的核抗體測定— Ro/La 抗體]			Negative
anti-La 結果 [可抽出的核抗體測定— Ro/La 抗體]			Negative
RF (血液) [類風濕性關節炎因子試驗— 免疫比濁法]	10.4 IU/mL [<14.0]		

Pathology

病理診斷：Only tiny foci of cystic spaces focally surrounded by thicken septa composed of ovoid smooth muscle-like cells with eosinophilic to clear cytoplasm are noted at sections A1-2, B and C. Immunohistochemistry studies reveals these smooth muscle-like cells are positive for smooth muscle actin, Desmin and focally positive for HMB-45. Together with images studies, these features could be consistent with lymphangioleiomyomatosis.

Lung, upper lobe, right, VATS resection, bulla with emphysematous change and old hemorrhage
Lung, middle lobe, right, VATS resection, bulla with emphysematous change and old hemorrhage
Lung, lower lobe, right, VATS resection, bulla with emphysematous change and old hemorrhage

Cystic lung disease

	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	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.