The radiologist says my approach is...

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Cystic lung diseases =

Characterized by the presence of multiple lung cysts
RADIOGRAPHIC DEFINITION OF A CYST

• Thin-walled (<2 mm), air-filled, lucency with a well-defined lung/air interface

CYSTIC LUNG LESIONS: 1\textsuperscript{st} step

Am I really dealing with cystic lung lesions?

- Thin-walled lesions
- No bifurcation/connection to the bronchi

Cystic Interstitial Lung Diseases: Recognizing the Common and Uncommon Entities, Curr Probl Diagn Radio 2014
CYSTIC LUNG LESIONS

- Thin-walled lesions
  - ≠ emphysema: no discernable walls
CYSTIC LUNG LESIONS

– Thin-walled lesions

• ≠ cavities:
  (necrotizing lung mets, infection, vasculitis,..)

thicker wall
Comparison to previous CT helps
- No bifurcation/connection to the bronchi
  • ≠ Bronchiectasis
Diagnosis?

Mounier Kühn syndrome
Post processing minIP: sensitizes lung cyst detection
CYSTIC LUNG LESIONS: 2\textsuperscript{nd} step

- Are the cysts centrally located, with normal lung between them?

- Or subpleural, with several layers and no interposition of normal lung between the cysts?

• $\neq$ Honeycombing
To summarize differentials:

- No visible wall
  - **Emphysema**

- Sub pleural location, no interposition of normal lung
  - **Honey combing**

- Thick wall
  - **Cavities (mets, infection, vasculitis, ...)**

- Connection to the bronchi
  - **Bronchiectasis**
Diffuse cystic lung diseases

= Radiologists’ best friends
FIVE DIAGNOSIS TO CONSIDER

1. Pulmonary Langerhans Cell Histiocytosis (PLCH)
2. Lymphangioleiomyomatosis (LAM)
3. Lymphoid interstitial pneumonia (LIP)
4. Birt-Hogg-Dubé Syndrome (BHD)
5. Light chain deposition disease (LCDD)
Diagnostic Approach to DCLDs

• Repartition of the cysts
  – Upper predominance: LHC
  – Basilar and subpleural predominance: BHD

• Associated lesions
  – Nodules and cavitary nodules: LHC
  – Pleural effusion/chylothorax: LAM
  – Angiomyolipoma of the kidney: Tuberous sclerosis
  – Tumor of the kidneys: BHD

• Patient characteristics
  – Smoker: LHC
  – Dysimmunity (Sjogren, HIV): LIP
  – Female gender: LAM
  – Family history of pneumothoraces: BHD

Gupta et al. Diffuse Cystic Lung Disease. Am J Respir Crit Care Med. 2015
Pulmonary Langerhans Cell Histiocytosis (PLCH)
Clues

- **Upper lung predominance** of the cysts
- **Bizarre shape** of the cysts
- **Associated lung nodules** (cavitary and non cavitary)
- Patient characteristics
  - Smokers
Upper lung predominance / sparing of the costophrenic angles
Bizarre shape of the cysts
Thin-walled cysts + Thicker-walled cysts

NODULES + Cavitary NODULES
28 yo man, bilatéral pneumothorax

18 yo man, right pneumothorax

PLEURAL COMPLICATIONS
Nodules, some with central cavitation at the early phase of the disease

Upper lung predominance of the nodules

PLCH in a 39 yo man
PLCH in a 53 yo woman
CD1a staining on surgical biopsy specimen
38 yo woman
Breast cancer (N0)
at initial screening
Lung mets?

After smoking cessation
71 yo man heavy smoker, suspicious nodule left lower lobe

After smoking cessation

Upper lung predominance of lesions
LYMPHANGIOLEIOMYOMATOSIS
Clues

- Rounded cysts
- Homogeneous repartition and size
- Chylothorax possible
- Patient characteristics
  - Women
  - Tuberous sclerosis
    - LAM in 10-15% of men with TSC
ROUNDED CYSTS, VARIABLE NUMBER
DIFFUSE
Including costophrenic angles
Tuberous sclerosis = TSC-LAM: less symptomatic

Angiomyolipomas both kidneys
PLEURAL COMPLICATIONS

Pleural talcage after right pneumothorax

Left pneumothorax in a 46 yo woman with S-LAM
S-LAM with abundant chylothorax

S-LAM with previous chylothorax Band atelectasis
Lung cysts in a 49 yo woman
LAM?
No!
Bizarre shape, sparing of costophrenic angles = PLCH
Diagnosis

- Typical CT findings AND

  - Tuberous sclerosis, angiomyolipoma, lymphadenopathy, or chylothorax

or

- VEGF-D level > 800 pg/ml
LYMPHOCYTIC INTERSTITIAL PNEUMONIA & FOLLICULAR BRONCHIOLITIS
Clues

• No spatial predominance
• Variable size of the cysts
• Associated mosaic perfusion
• Patient characteristics
  – Dysimmunity
    • Sjögren (primary or secondary RA, SS,..) HIV
  – Few symptoms/normal PFTs
  – Pneumothorax rare
67 yo woman, systemic sclerosis with Sjogren
Post processing Min IP: associated mosaic perfusion

= redistribution of pulmonary blood flow in areas free of bronchial disease
= key finding in follicular bronchiolitis due to Sjogren syndrome
DISTRIBUTION

NO PREDOMINANCE
51 yo asymptomatic never smoker woman
HIV under triple combination therapy
48 yo smoker
HIV under triple combination therapy
Pathogenesis

• Cysts in FB/LIP may result from ischemia due to vascular obstruction, postobstructive bronchiolar ectasia, or bronchiolar compression by lymphoid tissues
LYMPHOMATOUS transformation should be suspected in case of CYST WALL THICKENING or focal consolidation.
Clues

- Round cysts, diffuse repartition
- Increasing number over time
- Association with nodules and mediastinal lymphadenopathies

- Patient characteristics
  - Lymphoproliferative disorder
80 yo man with myeloma
BIRT HOGG DUBE SYNDROME
Clues

• Sub pleural and basal predominance of the cysts
• Lentiform shape

• Patient characteristics
  - Family history of pneumothoraces / kidney tumors
  - Skin lesions (Fibrofolliculomas)

Birt-Hogg-Dube’s syndrome. State-of-the-art review with emphasis on pulmonary involvement, Respiratory Medicine, 2015
Lentiform shape
DISTRIBUTION

BASAL & SUB PLEURAL PREDOMINANCE
29 yo never smoker female, chest pain

- Family history of pneumothoraces:
  - Mother, brother, uncle & cousins
Un TDM sans injection est réalisé :

Subpleural cysts typical for BHD on CT

**CONSEQUENCES**

Family screening

MRI follow-up for renal tumor detection
63 yo man with previous renal carcinoma, 3 lung mets of right lower lobe
Subpleural, basilar cysts typical for BHD on CT
Pathogenesis

- Activation of the mTOR pathway, leading to cell dropout or adhesion protein defects or deficiencies
  - increase the vulnerability of the alveolar-septal junction to tearing by mechanical forces during the respiratory cycle
SUMMARY: the radiologist says my approach is......

1. Rule out other causes of lucencies (emphysema, bronchiectasis, cavities)
2. Shape and distribution of the cysts
3. Associated findings (angiomyolipoma, renal tumor, mosaic perfusion)
4. Patient characteristics+++  
   - Young smoker, bizarre shape, upper lung predominance and nodules = PLCH  
   - Woman+ round cysts+ diffuse repartition= LAM  
   - Dysimmunity+ round cysts, random distribution, mosaic perfusion= LIP/FB  
   - Family history of pneumothorax+ skin lesion+ lentiform shape, basilar and sub pleural predominance= BHD
• Thank you