CYSTIC LUNG DISEASE

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DEFINITIONS:

Are **air-filled areas** present within the lung parenchyma?

- ***Very Thin*** (<1mm) or ***no walls*** → EMPHYSEMA
- ***Thick*** walled (>4mm) → CAVITY
- ***Thin*** walled (1-4mm) → CYST

EMPHYSEMA
- COPD (UL)
- α1AT (LL)
- Ritalin Lung (LL predom)

CAVITY
- Malignancy
- Infection
- Septic emboli
- Vasculitis

Emphysema descriptors:
- **BLEBS** <2 cm
- **BULLAE** > 2 cm
- **GIANT BULLAE** > 30% of hemithorax

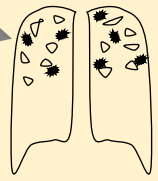
Cyst mimics:

CYSTIC BRONCHIECTASIS –
- Dilated airways (not real cysts)

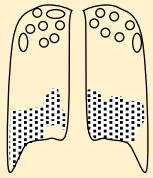
HONEYCOMBING –
- ≥3 adjacent air-filled spaces
- seen with emphysema, not with cysts

Upper Lobe predominant

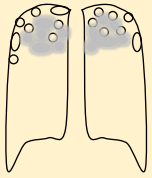
LANGERHANS CELL HISTIOCYTOSIS (LCH)
- **Smoking-associated** inflammation ♂>♀
- Chest: **UL** predominant ***'bizarre shaped'*** cysts & ***'stellate shaped'*** centrilobular nodules (1-5mm)



NEUROFIBROMATOSIS TYPE 1 (NF1)
- **Genetic**; neurofibromin (NF1)
- Skin: café-au-lait spots,axillary freckling, neurofibromas,
- Chest: **UL** predominant cysts, emphysema, & bullae, & LL fibrosis



PNEUMOCYSTIS JIROVECI PNEUMONIA (PJP)
- **Infectious**; occurs in individuals with severe **immunocompromise** (HIV CD4<200, BMT, etc)
- Chest: UL pneumotoceles & subpleural plebs, GGOs (**UL** if on PPx, **LL** if not; peripheral sparing GGOs)



PARACOCCIDIOIDOMYCOSIS
- **Infectious**; occurs in rural workers (immunocompentent) in S. America ♂>♀
- Causes diffuse LAD and can cause granulomas in many organs
- Chest: scattered cysts without lobar predominance, reverse halo sign, cavitations, and bronchiectasis.

HYPERSENSITIVITY PNEUMONITIS
- Inflammation due to inhaled antigens, forming granulomas
- Usually causes GGO and mosaicism rarely may cause **UL** cyst formation

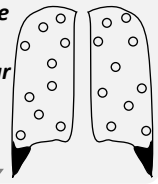
AMYLOIDOSIS
- Can occur with 1° or 2 ° amyloidosis
- Chest: diffuse peripheral thin-walled cysts, often also with nodules (including endobronchial) or masses

EHLERS-DANLOS SYNDROME
- **Genetic** connective tissue disease, rarely may develop *diffuse* cysts

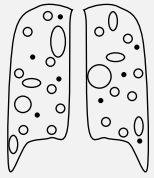
PROTEUS SYNDROME
- Rare **Genetic** syndrome (AKT1) that may present with *diffuse* cysts

Diffuse

LYMPHANGIOLYOMIOMATOSIS (LAM)
- **Genetic** or spontaneous, assoc. w/ tuberous sclerosis (TSC), ♀>>♂
- Skin: facial angiofibromas
- Kidney: Angiomyolipoma tumors
- Chest: ***Uniform size diffuse*** distribution of cysts, ***Intralobular thickening***, & pleural effusions (***chylothorax***)



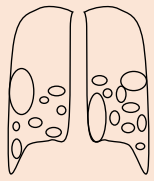
LIGHT CHAIN DEPOSITION DISEASE (LCDD)
- **Lymphoproliferative disease** assoc (esp multiple myeloma) causing non-amyloid deposition of Ab; ♀=♂
- Kidney: Proteinuria/nephrotic syndrome
- Chest: variable sized cysts, nodules, & LAD



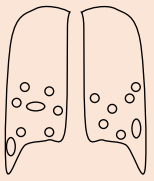
CYSTIC PULMONARY METASTATIC DISEASE
- **Metastatic malignancy** usually causes cavitary (thick walled) lesions.
- *Diffuse* cysts can be seen with epitheliod metastasis, & rarely with adenocarcinomas/sarcomas as reported here

Lower Lobe predominant

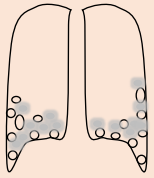
BIRT-HOGG-DUBE (BHD)
- **Genetic**; AD folliculin (FLCN) mutation, ♀=♂ prevalence
- Kidney: chromophobe RCC
- Skin: fibrofolliculomas and angiofibromas
- Chest: **LL** predominant *lentiform* cysts; often presents with PTX



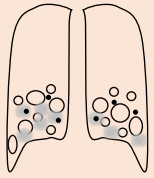
LYMPHOCYTIC INTERSITIAL PNEUMONITIS (LIP)
- **Autoimmune** (Sjogren's syndrome) & **immunodeficiency** (HIV) associated inflammation
- Dense lymphocyte infiltrates; overlap with FB
- Chest: **LL** Predominant cysts in bronchovascular distribution



DESQUAMATIVE INTERSTITIAL PNEUMONITIS (DIP)
- **Smoking-associated** ILD often associated with RB-ILD; ♂>♀
- Chest: **LL predominant** and Subpleural/basilar predominant cysts of uniform small size with associated **GGOs**



FOLLICULAR BRONCHIOLITIS (FB)
- Associated with **collagen vascular disease** & **immunodeficiency**
- Chest: centrilobular GGOs and nodules, sometimes with medium to large **LL** predominant cysts running along bronchovascular bundles



PULMONARY PAPPILLOSIS (PP)
- **Infectious**; vertically transmitted HPV infxn; very rare.
- Chest: usually endobronchial lesions, ***rarely*** *diffuse* pulmonary nodules that turn into cysts.

FIRE-EATERS LUNG
- Aspiration of flammable petroleum compounds causes inflammation; leading to cavitary or cystic disease

CONSTRICTIVE BRONCHIOLITIS
- Occurs due to **viral**, **autoimmune**, or GVHD. Typically causes mosaic attention & bronchiectasis. Rarely causes few small *diffuse* cysts

HYPER-IGE SYNDROME
- **Immunodeficiency**/STAT3 **mutation** causing sinopulmonary infections, & rarely pneumatoceles & cysts.