University Hospital Southampton
NHS Foundation Trust

Cystic Lung Disease – a Radiologist's Perspective

Wessex Respiratory Regional Training Day
21st May 2019
Orphan Lung Disease Day

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Pulmonary Langerhans Cell Histiocytosis

- Young or middle aged adults
- Smokers (90%)
- Present with cough and dyspnea
- Pneumothorax (20%)
- Disease regresses spontaneously (25%)
- Stabilizes (50%)
- Progression (25%)

Webb RW. Diffuse Cystic Lung Diseases. In: Webb, RW, Higgins CB (eds.). Thoracic Imaging Pulmonary and Cardiovascular Radiology. 3rd ed. Philadelphia: Wolters Kluwer; 2017. PP 645-698.

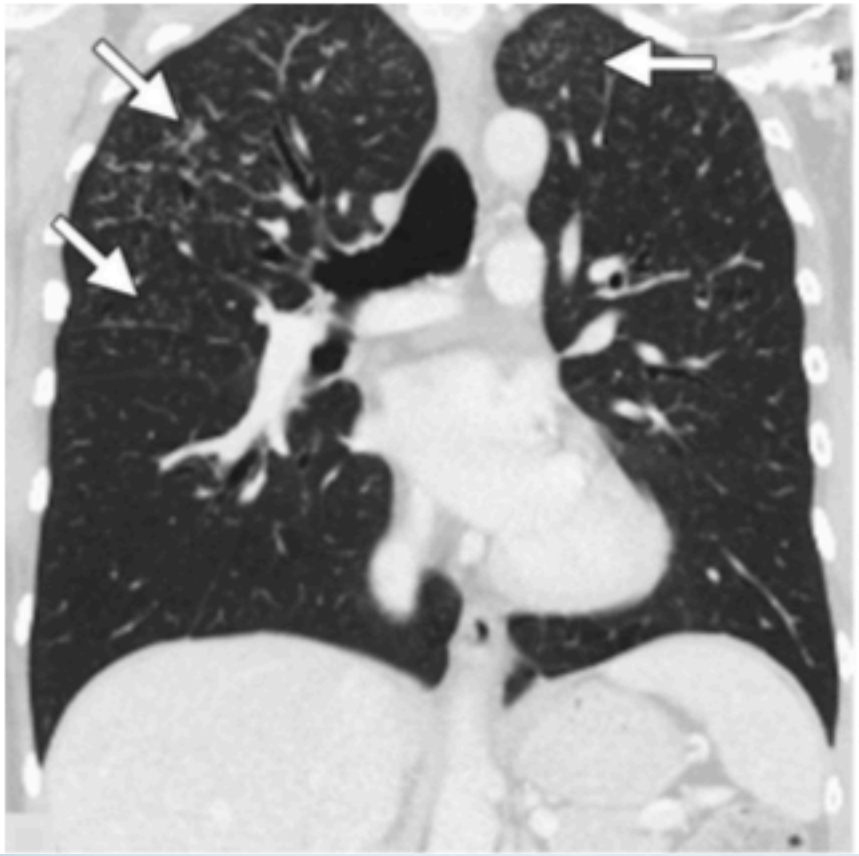
CXR Findings LCHx

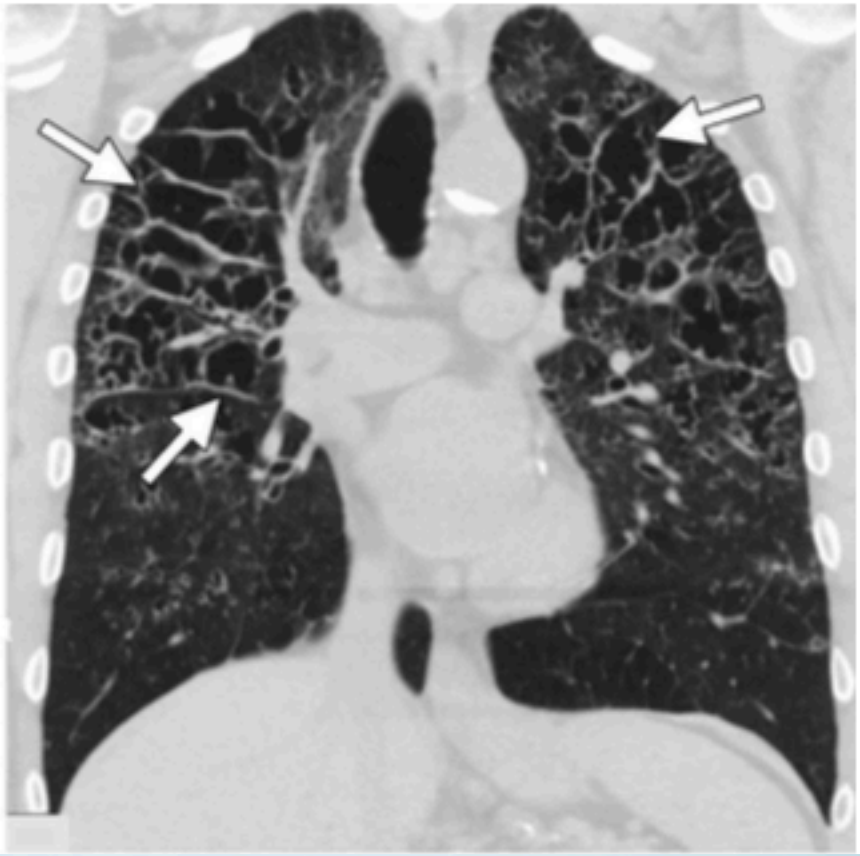
- Reticular
- Nodular
- Reticulonodular patterns
- Bilateral, predominately involving MZ + UZ with sparing of costophrenic angles
- Lung volumes – normal / increased

CT Findings LCHx

- Cysts < 10 mm diameter with distinct walls
- Spare costophrenic angles and bases
- Lung UL predominance
- Peribronchial distribution
- Progression from nodules to cavitating nodules to thick walled cysts to thin walled cysts
- Asso. with GGO
- Associated hilar and mediastinal adenopathy or lytic bone lesions

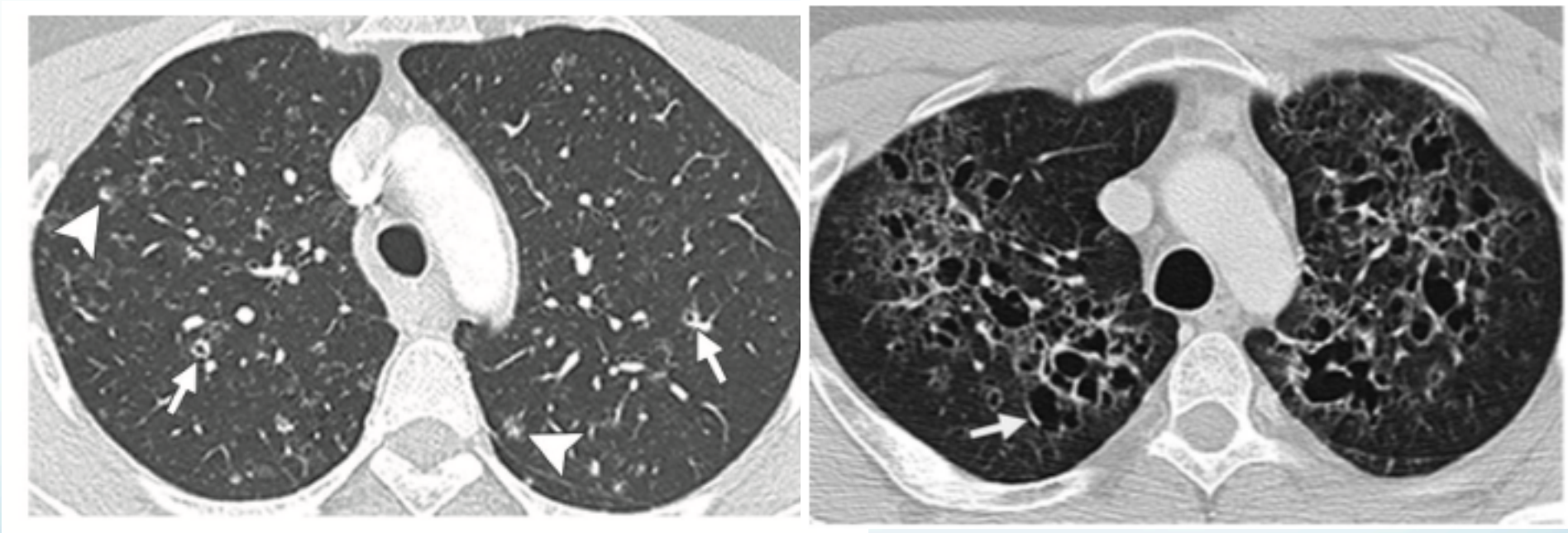
CT – LCHx





Zaveri J, La Q, Yarmksh G, Neuman J. More than Just Langerhans Cell Histiocytosis: A Radiologic Review of Histiocytic Disorders. RadioGraphics 2014 34:7, 2008-2024

CT - LCHx



Ahuja J et al. Histiocytic disorders of the chest: imaging findings. Radiographics. 2015 Mar-Apr;35(2):357-70.

Lymphangioleiomyomatosis (LAM)

- Proliferations of immature-appearing smooth muscle cells
- Sporadic LAM associated with gene mutations TSC2
- Women of childbearing age
- Identical to lung disease seen in Tuberous Sclerosis (TS)
- Cystic lung destruction
- Chylous effusions (60%)
- Pneumothoraces (80%)
- Renal angiomyolipomas (15%)
- Lung cysts
  - Round in shape
  - Diffuse distribution
  - Involve costophrenic angles
- Nodules seen occasionally

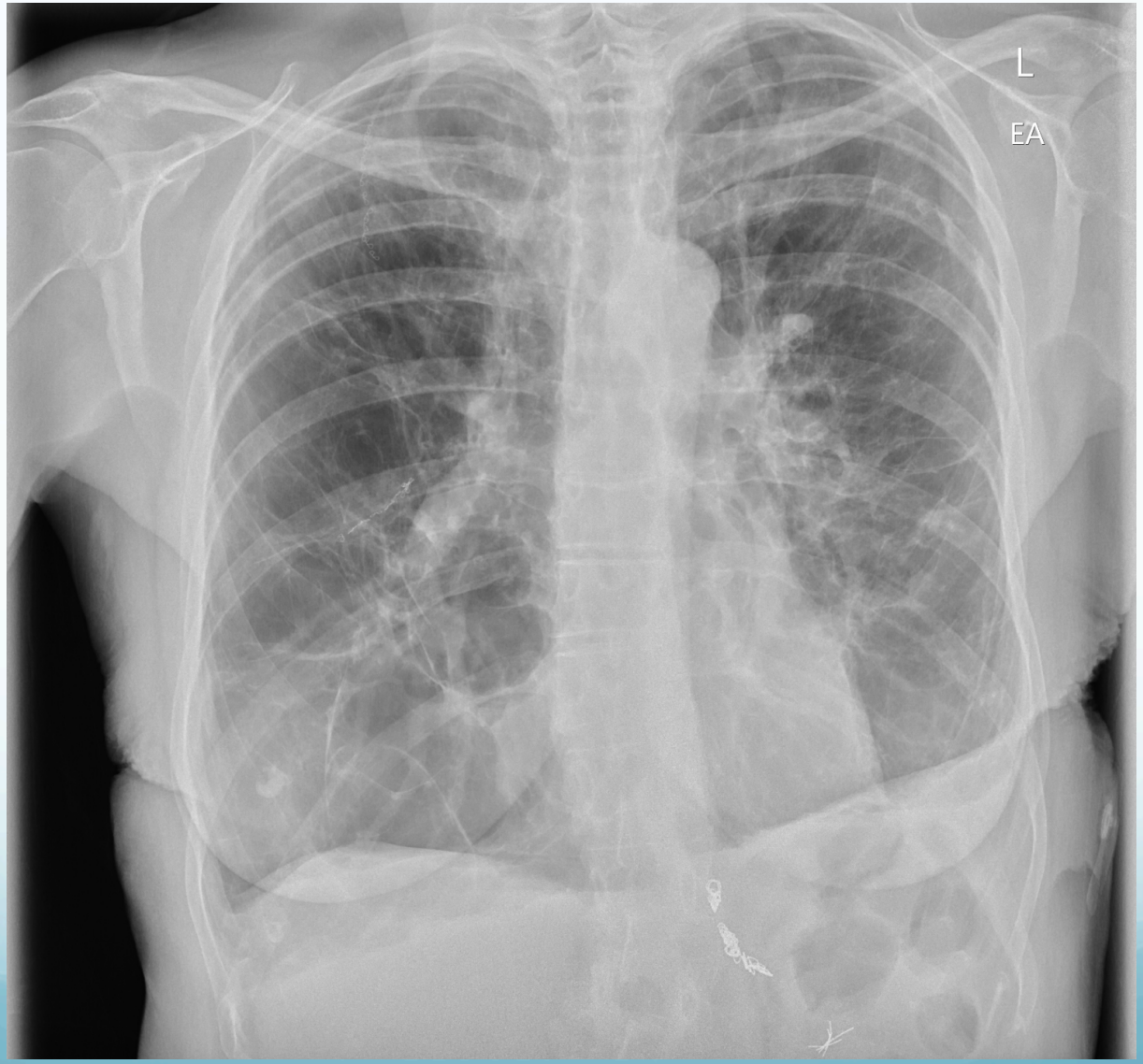
Tuberous Sclerosis

- LAM can occur in asso. with tuberous sclerosis
- Linked mutations in the tuberous sclerosis genes TSCI and TSC2

CXR - LAM / TS

- Fine reticular pattern
- Advanced disease mimicks honeycombing
- Lungs diffusely involved with bases involved to the same degree as the apices
- Pneumothorax (50%)
- Uni/Bilateral pleural effusions (10-20%)

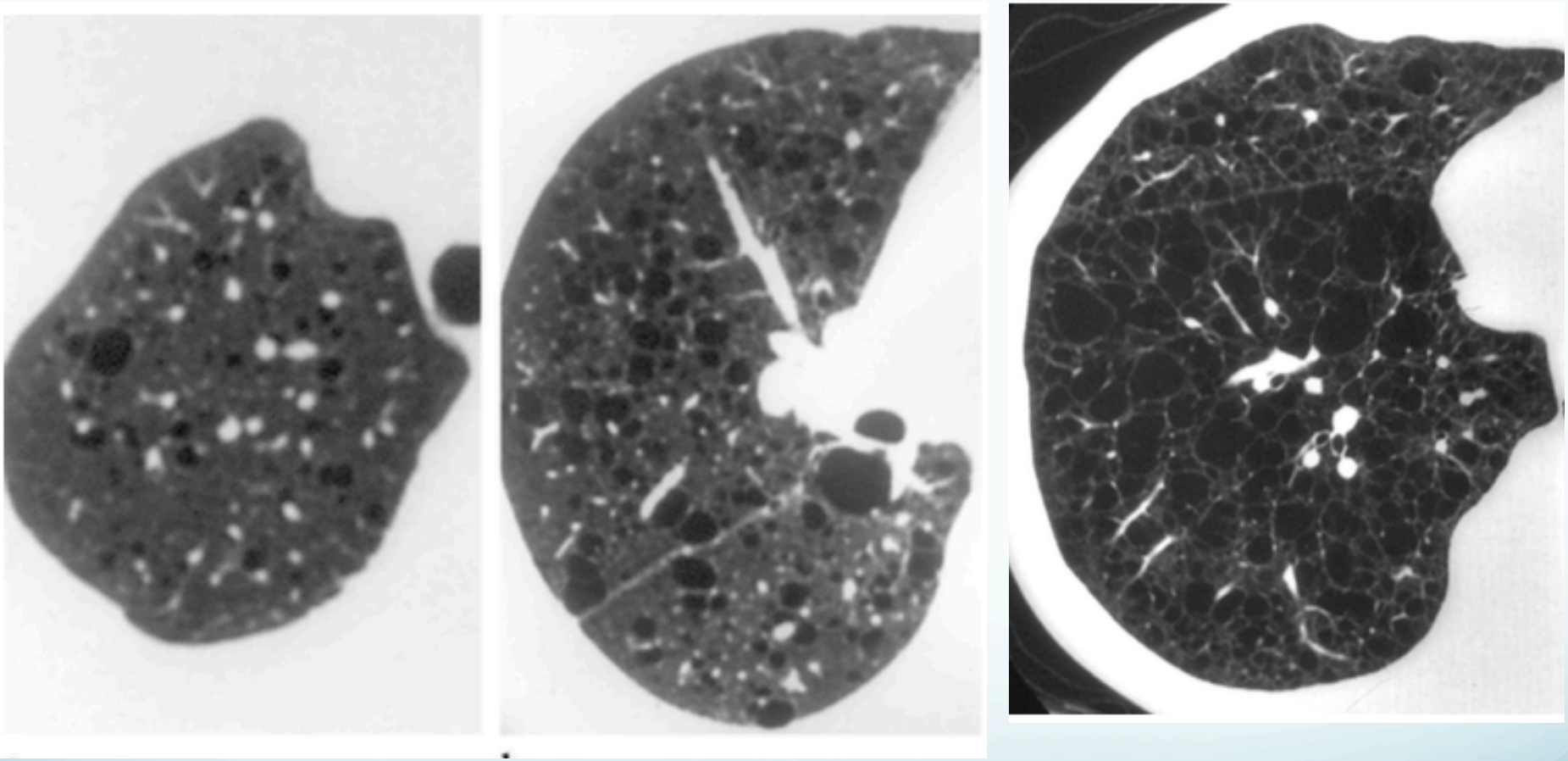
CXR – LAM



CT – LAM / TS

- Numerous, isolated, thin walled, rounded lung cysts
- Cysts 2-5 mm size
- Cyst size increases with progression of disease
- No lung zone spared
- TS-LAM small lung nodules representing pneumocyte hyperplasia (not see sporadic LAM)
- Other features:
  - Hilar, mediastinal & retrocural lymphdenopathy

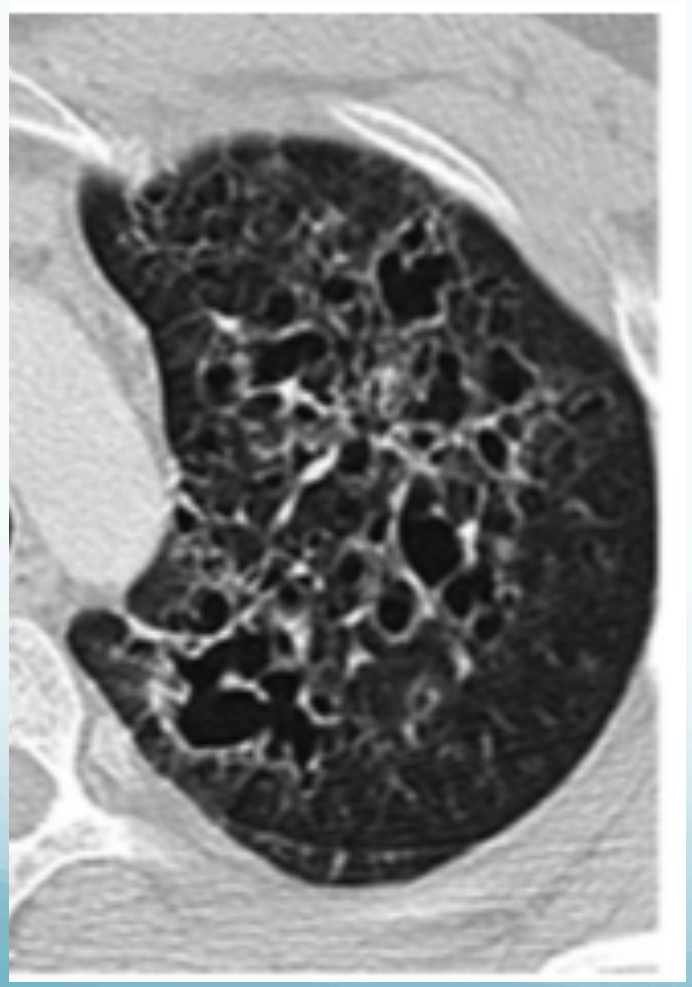
CT – LAM

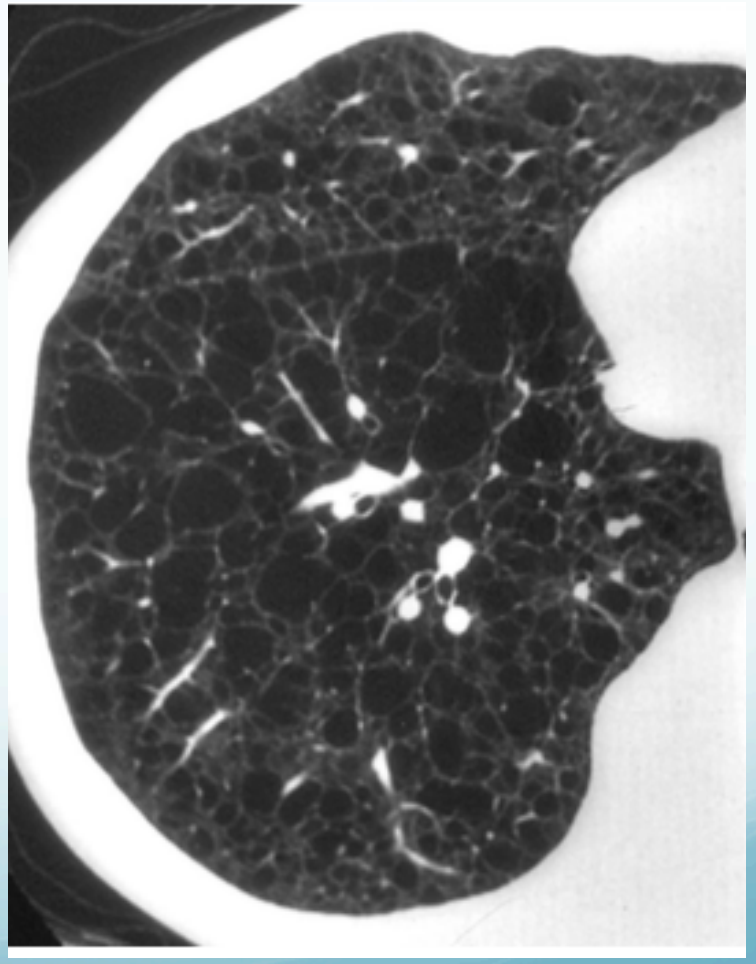


Abbot GF et al. From the archives of the AFIP: lymphangioleiomyomatosis: radiologic-pathologic correlation. Radiographics. 2005 May-Jun;25(3):803-28.

LAM

LCHx versus LAM

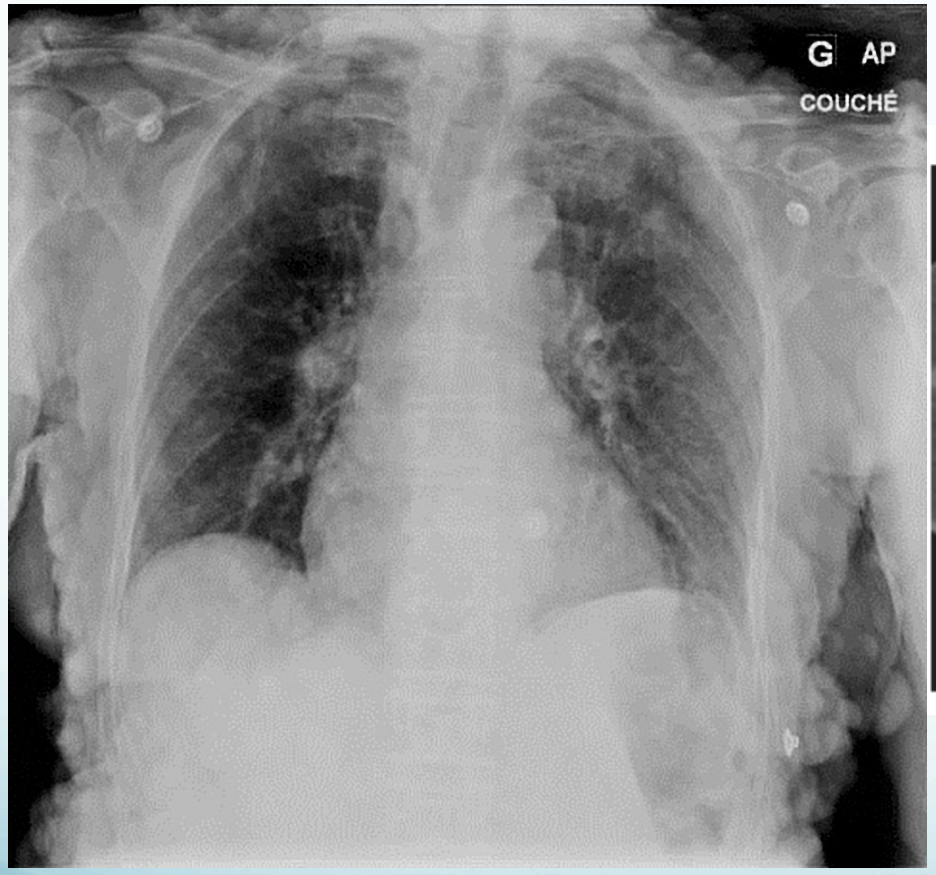


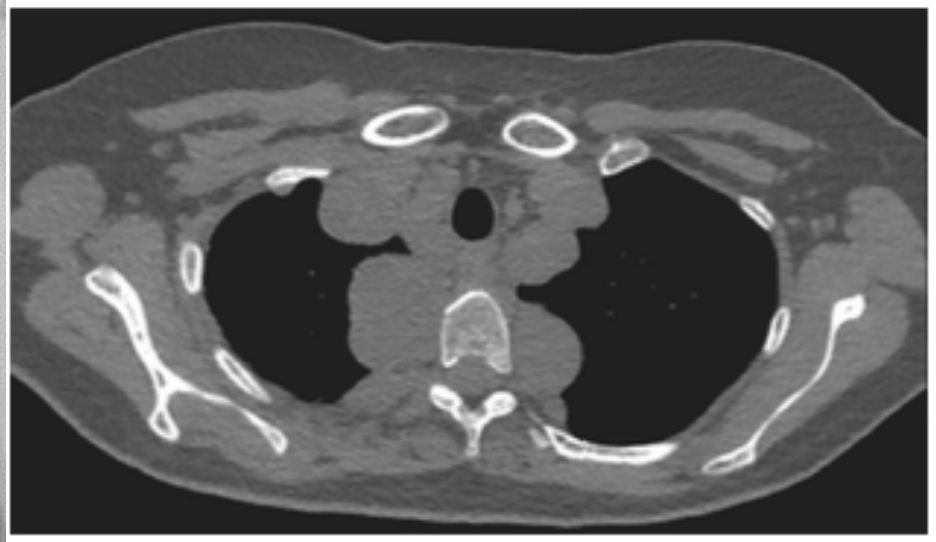


Neurofibromatosis

- Common genetic disorder (1 in 3000)
- Thoracic manifestations:
  - Rib abnormalities (rib notching and ribbon ribs)
  - Scoliosis
  - Cutaneous/subcutaneous neurofibromas mimicking the presence of lung nodules on CXR
  - Intercostal or mediastinal neurofibroma or schwannoma
  - Paraganglioma
  - Thoracic meningocele
- Lung disease (10-20%)
  - Bullae in UL and interstitial fibrosis at the lung bases

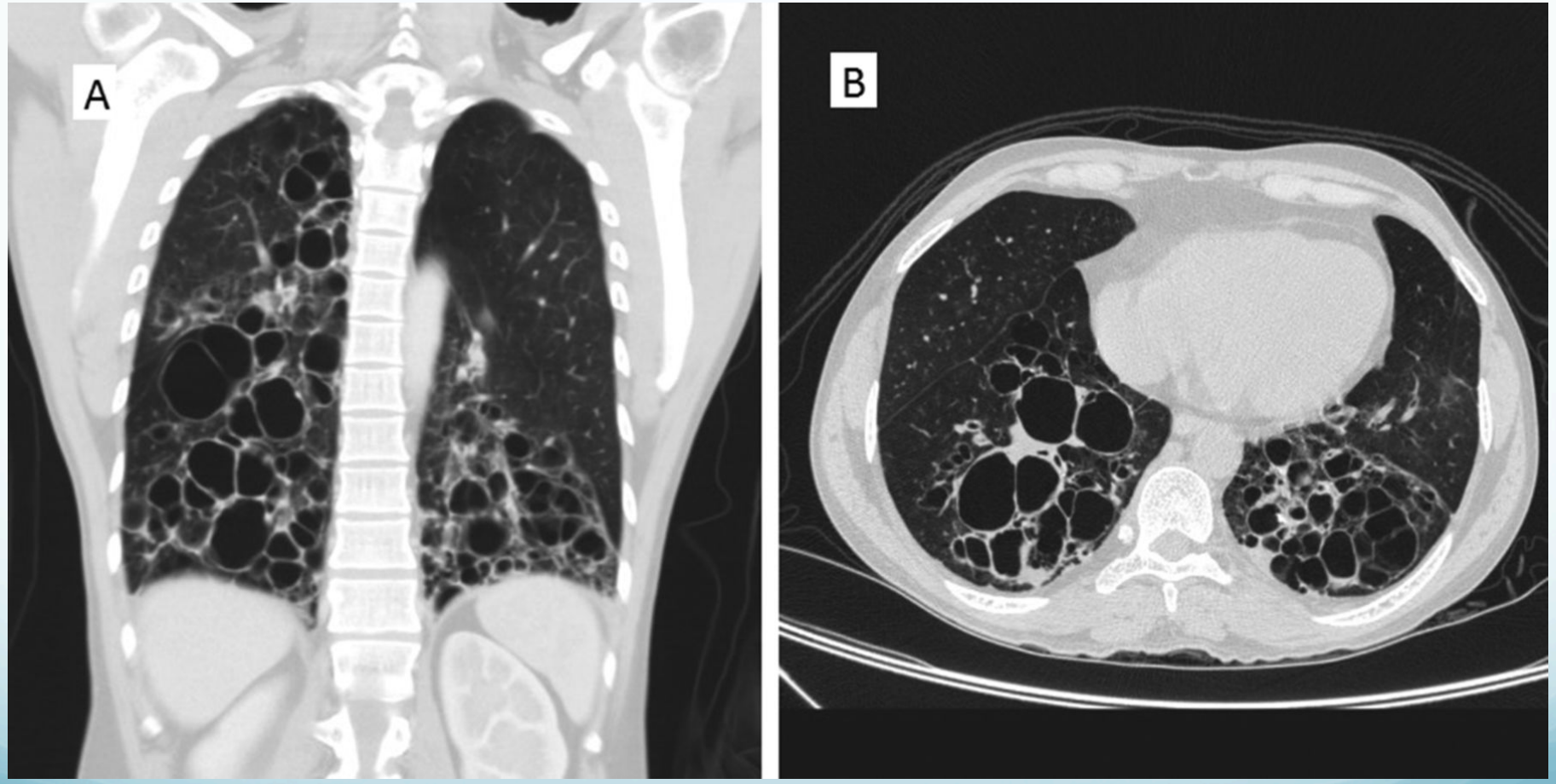
Imaging - NF





Chebib et al. Thoracic manifestations of segmental neurofibromatosis. BMJ Case reports. https://casereports.bmj.com/content/casereports/2017/bcr-2017-221253.full.pdf.
Fortin F. https://radiopaedia.org/cases/cutaneous-neurofibromas-in-neurofibromatosis-type-1

CT – NF

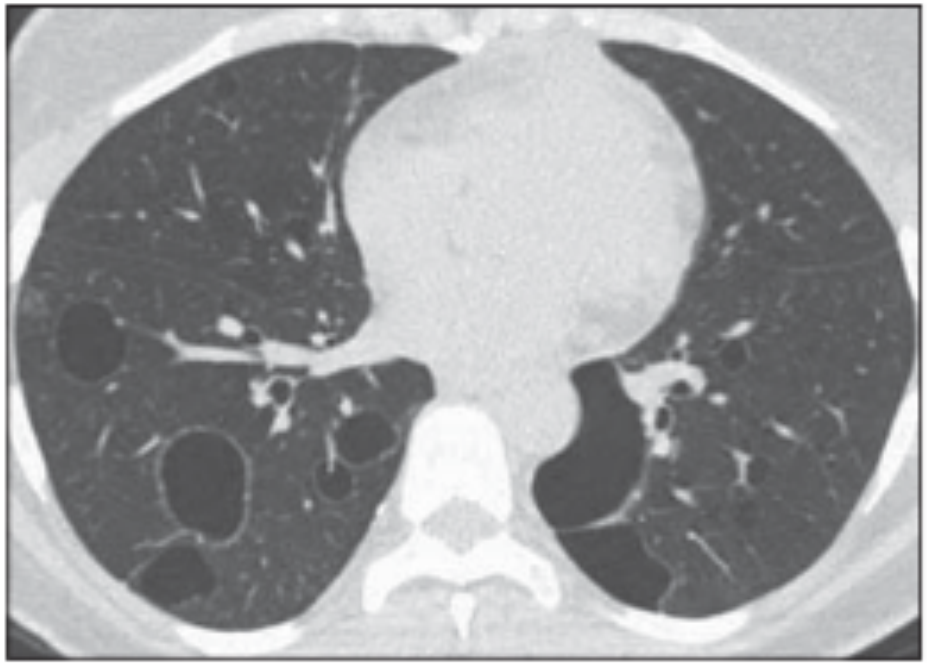


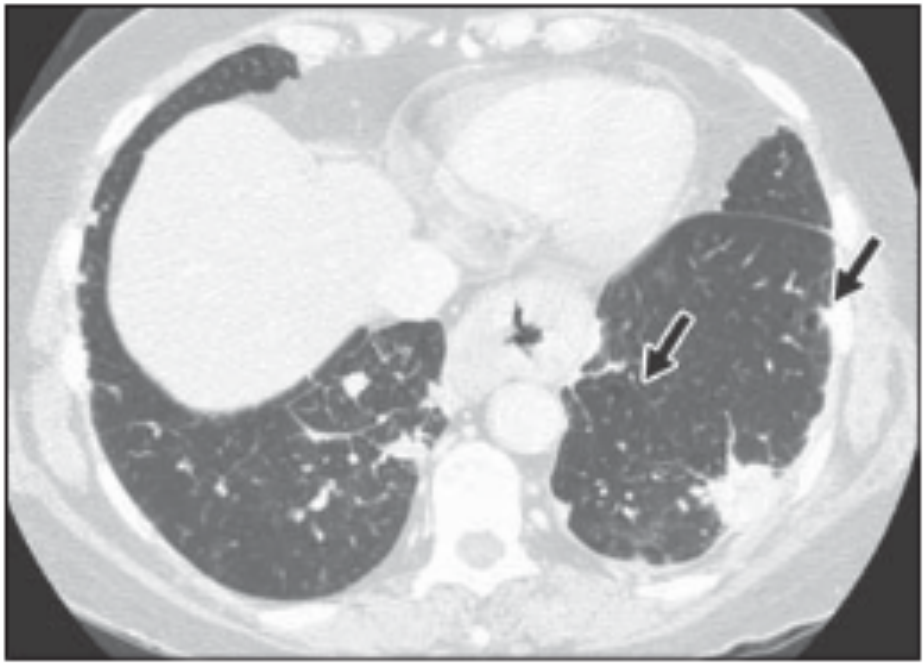
Madhyastha SP et al. BMJ Case Reports 2017;2017:bcr-2016-217774

Lymphoid Interstitial Pneumonia (LIP) and Sjögren's Syndrome

- LIP occurs in asso. with Sjögren's Syndrome
- Diffuse interstitial infiltrate of lymphocytes and plasma cells
- HRCT
  - Reticulonodular opacities
  - Patchy or diffuse GGO
  - Multiple thin walled cysts in relation to pulmonary vessels
  - Cysts thin walled – fewer in no. than LCHx or LAM
  - Poorly defined centrilobular nodules

CT - LIP



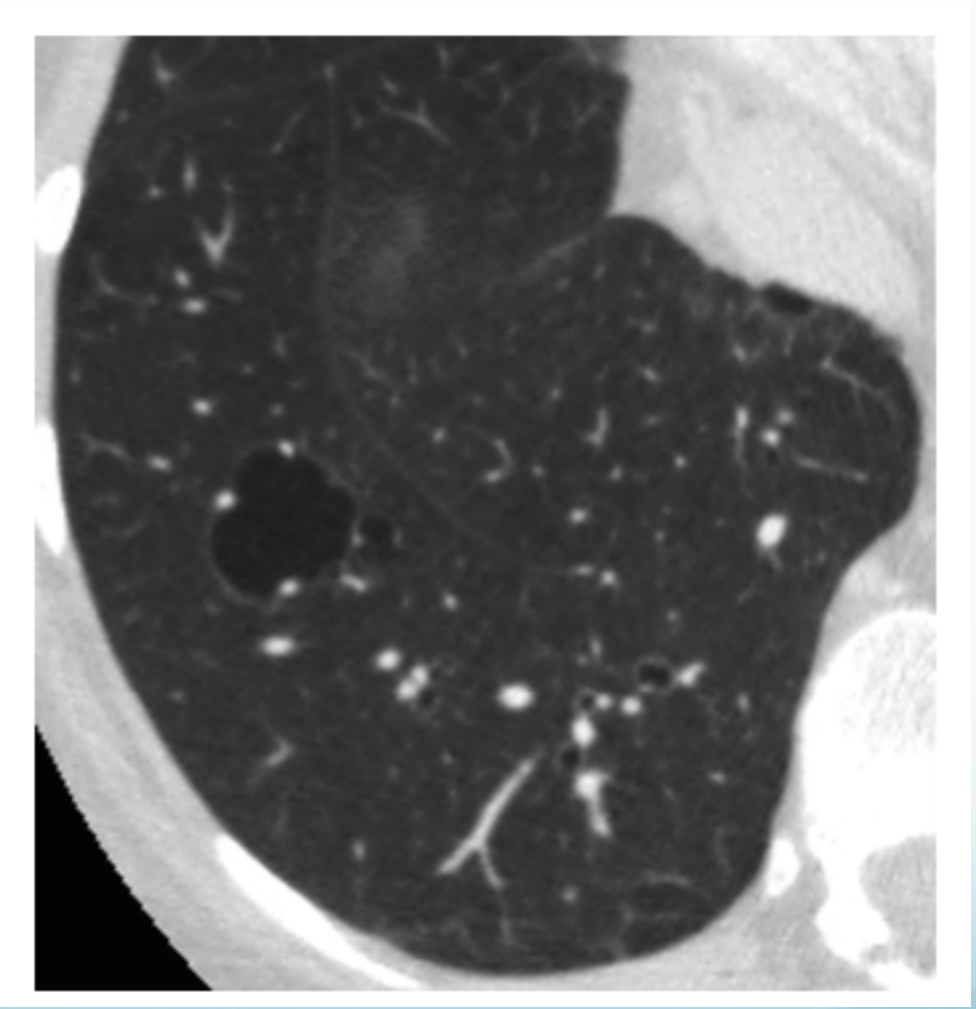


Seaman DM et al. Diffuse Cystic Lung Disease at High-Resolution CT. AJR. 2011; 196:1305–1311.

Light-Chain Deposition Disease

- Deposition of immunoglobulin light chains in various tissues and organs
- Asso. with Multiple Myeloma or lymphoproliferative disorders
- Renal involvement:
  - Proteinuria
  - With or without nephrotic syndrome
  - Renal failure
- HRCT:
  - Single or multiple nodular lesions of various sizes
  - Thin walled cysts >2cm with nodular thickening of cyst walls

CT – Light-Chain Deposition Disease



Sheard S et al. Pulmonary light-chain deposition disease: CT and pathology findings in nine patients. Clin Radiol. 2015 May;70(5):515-22. doi: 10.1016/j.crad.2015.01.002.

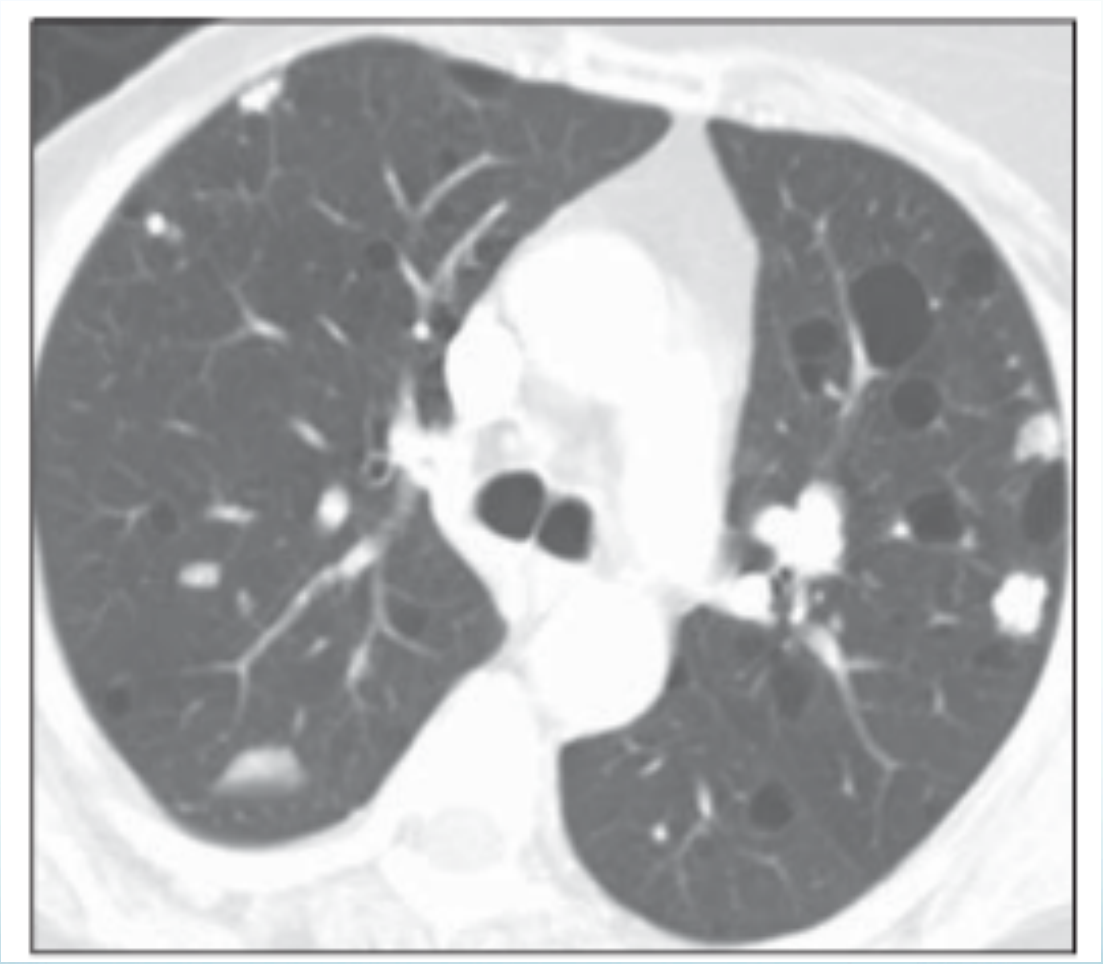
Amyloidosis

- Extracellular deposition of proteins in β-pleated sheets, localised (10-20%) or systemic (80-90%)
- Classified as:
  - Primary (i.e., asso. with multiple myeloma or macroglobulinemia) or
  - Secondary (i.e., asso. with rheumatoid arthritis, tuberculosis, Crohn disease, cystic fibrosis, or Mediterranean fever)

HRCT - Amyloidosis

- Nodules often calcified
- Interlobular septal thickening
- Honeycombing
- GGO
- Lymphdenopathy
- Pulmonary cysts (rare) most often described in asso. with Sjögren Syndrome

CT - Amyloidosis

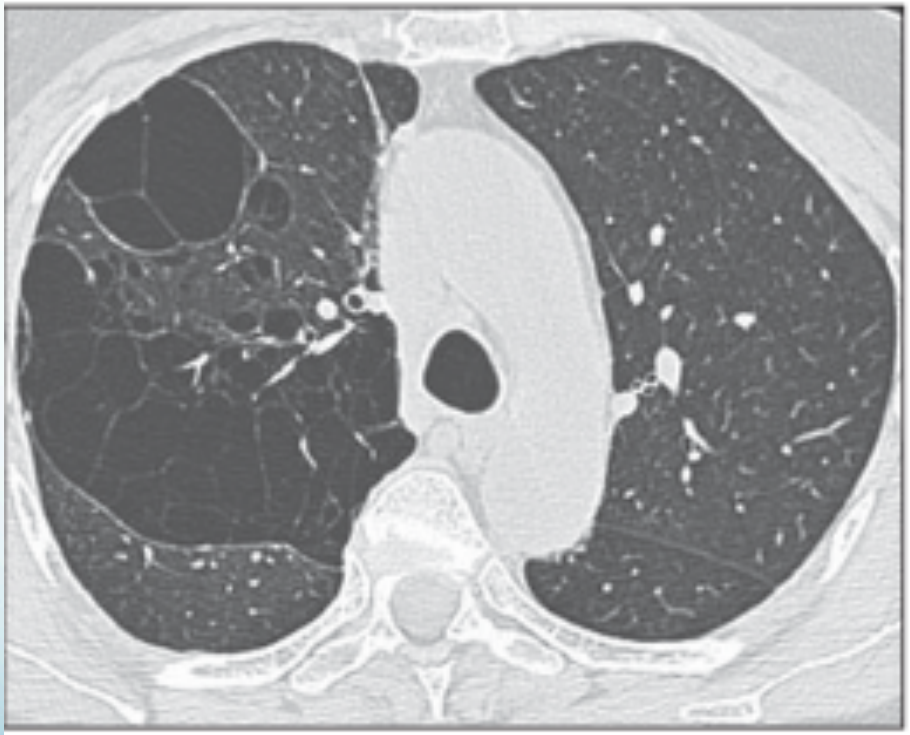


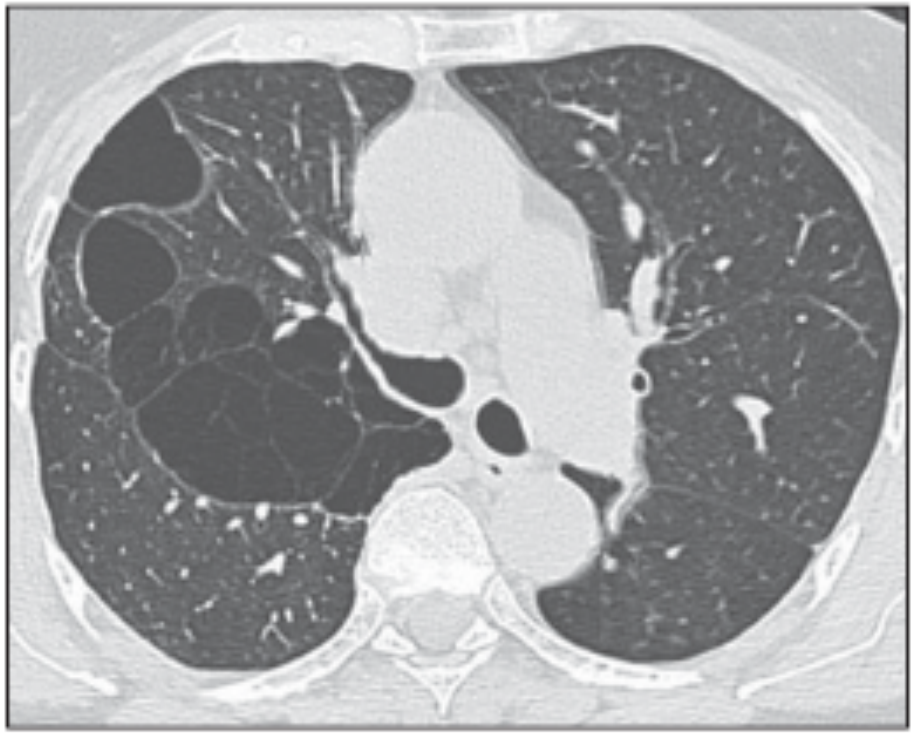
Seaman DM et al. Diffuse Cystic Lung Disease at High-Resolution CT. AJR. 2011; 196:1305–1311.

Birt-Hogg-Dubé Syndrome

- Rare – Autosomal Dominant disorder
- Characterised by:
  - Lung cysts
  - Fibrofolliculomas over face, neck and upper trunk
  - Renal tumours (ranging from benign oncocytoma to RCC)
  - Lung changes can predate other organ involvement
- Lung cysts limited in number
  - Thin walled, often subpleural, may involve fissures
  - Rounded or lenticular in shape
  - Cysts larger than LAM or LCHx (up to 8cm diameter)
  - Predominate in Lower + Medial LZ
  - Pneumothorax

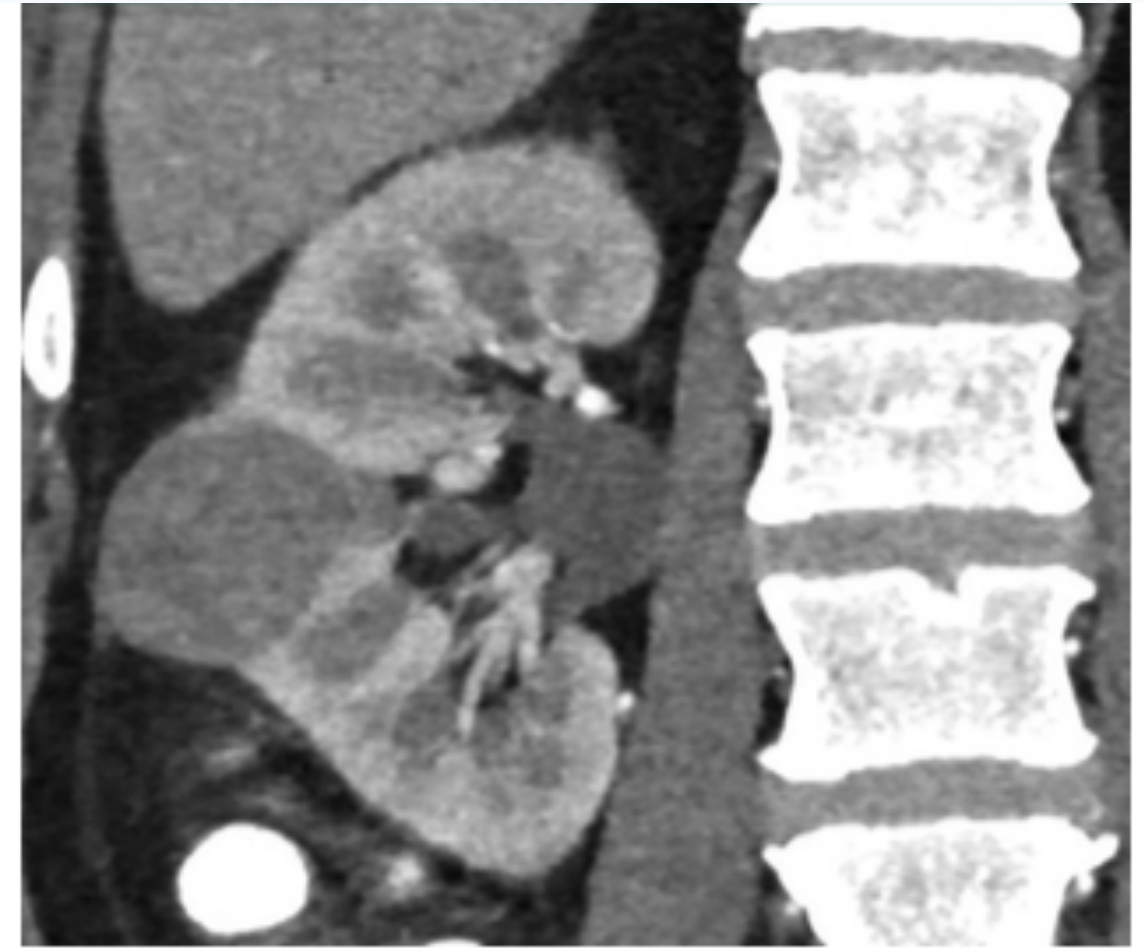
CT – Birt-Hogg-Dubé Syndrome





Agarwall PP et al. Thoracic CT findings in Birt-Hogg-Dube syndrome. AJR Am J Roentgenol. 2011 Feb;196(2):349-52. doi: 10.2214/AJR.10.4757.

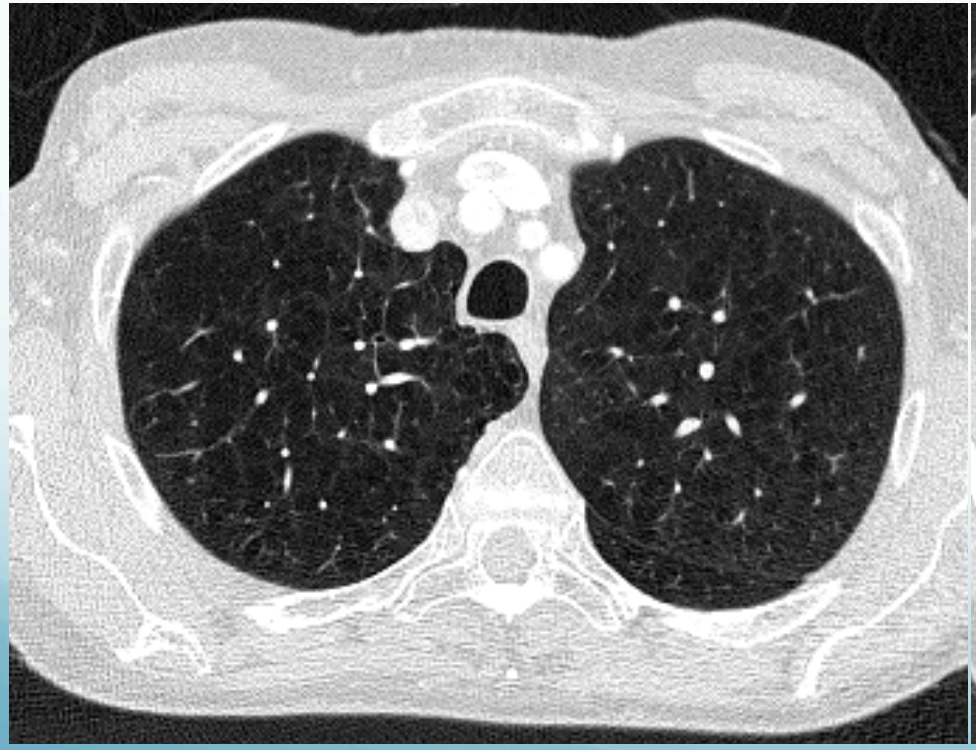
CT – Birt-Hogg-Dubé

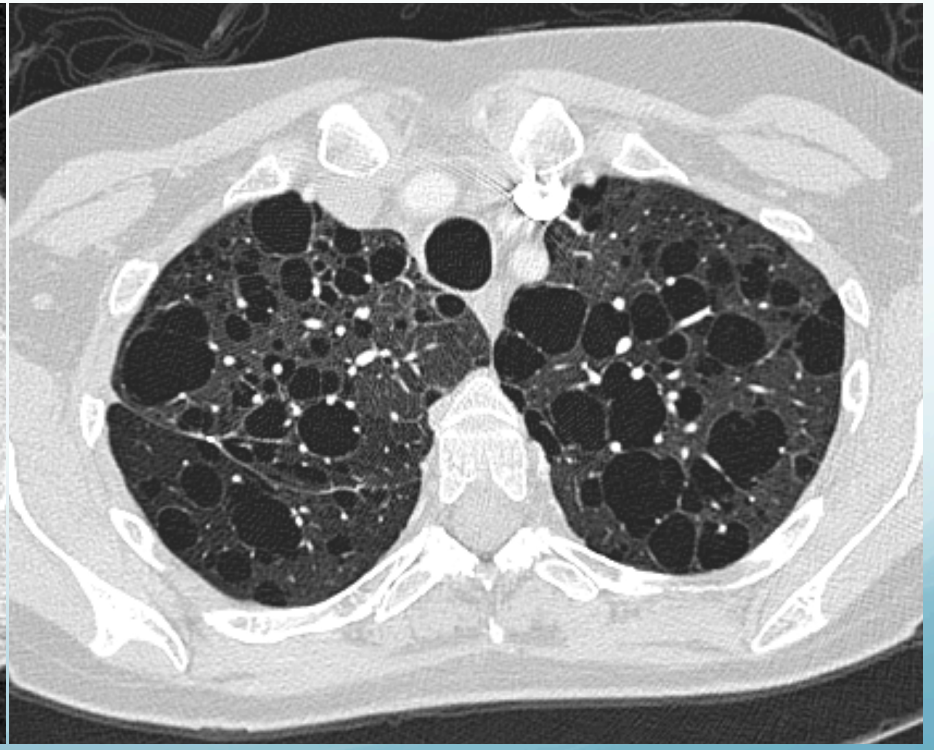


Zamora CA et al. Case 218: Birt-Hogg-Dubé Syndrome. RSNA. https://pubs.rsna.org/doi/full/10.1148/radiol.2015122724

Emphysema versus Cysts

- Emphysema = lack distinct walls
- Cysts = wall

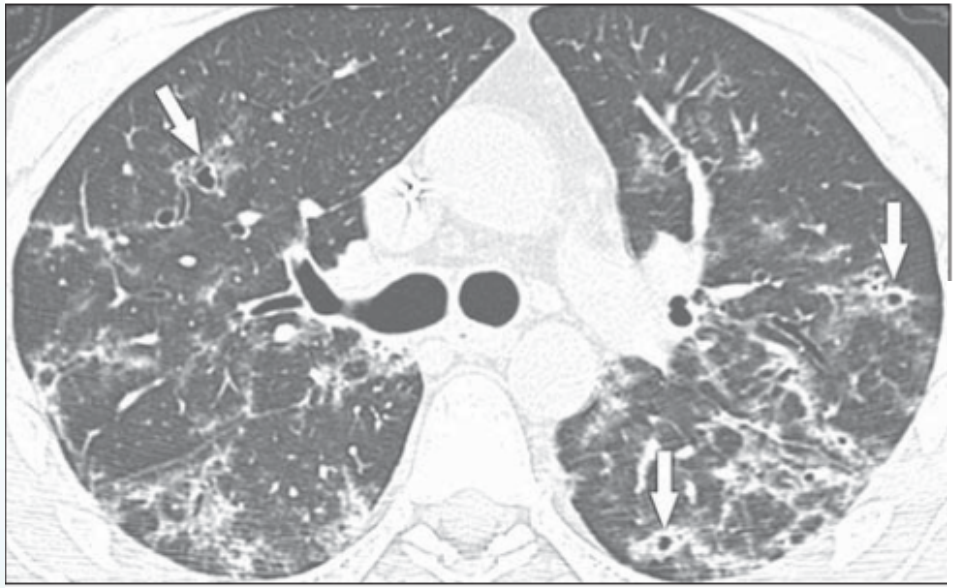


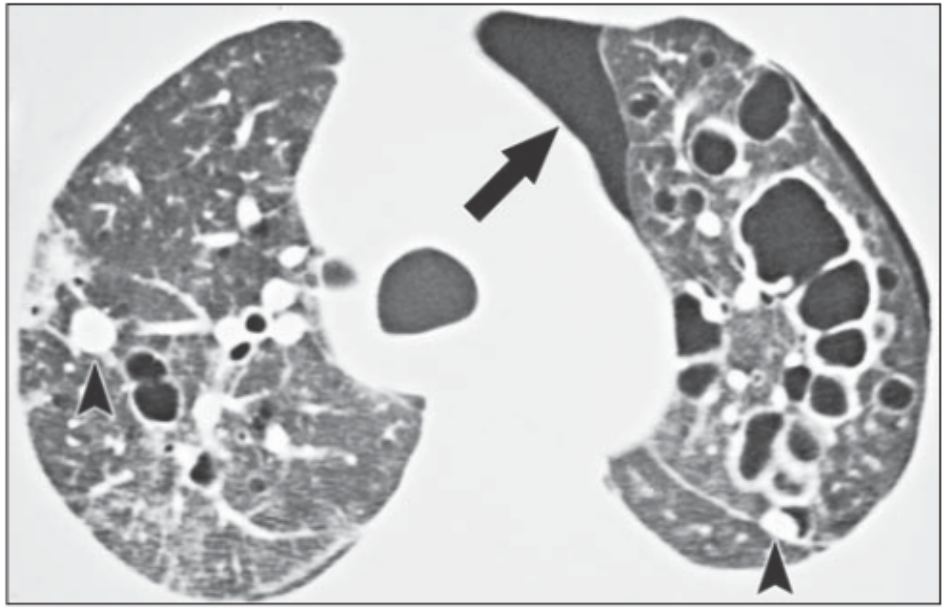


Infectious Diseases

- **Pneumocystis pneumonia (PCP) / pneumocystis jirovecii**
- Cystic changes on HRCT in pts with AIDS and PCP (20-35%)
- Cysts asso. PCP:
  - Variable in appearance – different sizes and shape with thick or thin wall
  - UL predominance is common (but can occur anywhere)

PCP cystic disease





Kanne JP et al. Pneumocystis jiroveci Pneumonia: High-Resolution CT Findings in Patients With and Without HIV Infection. *AJR* 2012; 198:W555–W561.

Pneumatocele

- Thin walled gas filled space
- Occuring in association with
  - Acute pneumonia
  - Trauma
- Combination of lung necrosis/laceration and bronchial obstruction
- Indistinguishable from a cyst or bulla on HRCT

Thank you

Any Questions?