

Diagnostic Approach to Diffuse Cystic Lung Disease: Radiologists' Perspectives

울산의대 서울아산병원 영상의학과
채 은 진

Lung Cysts

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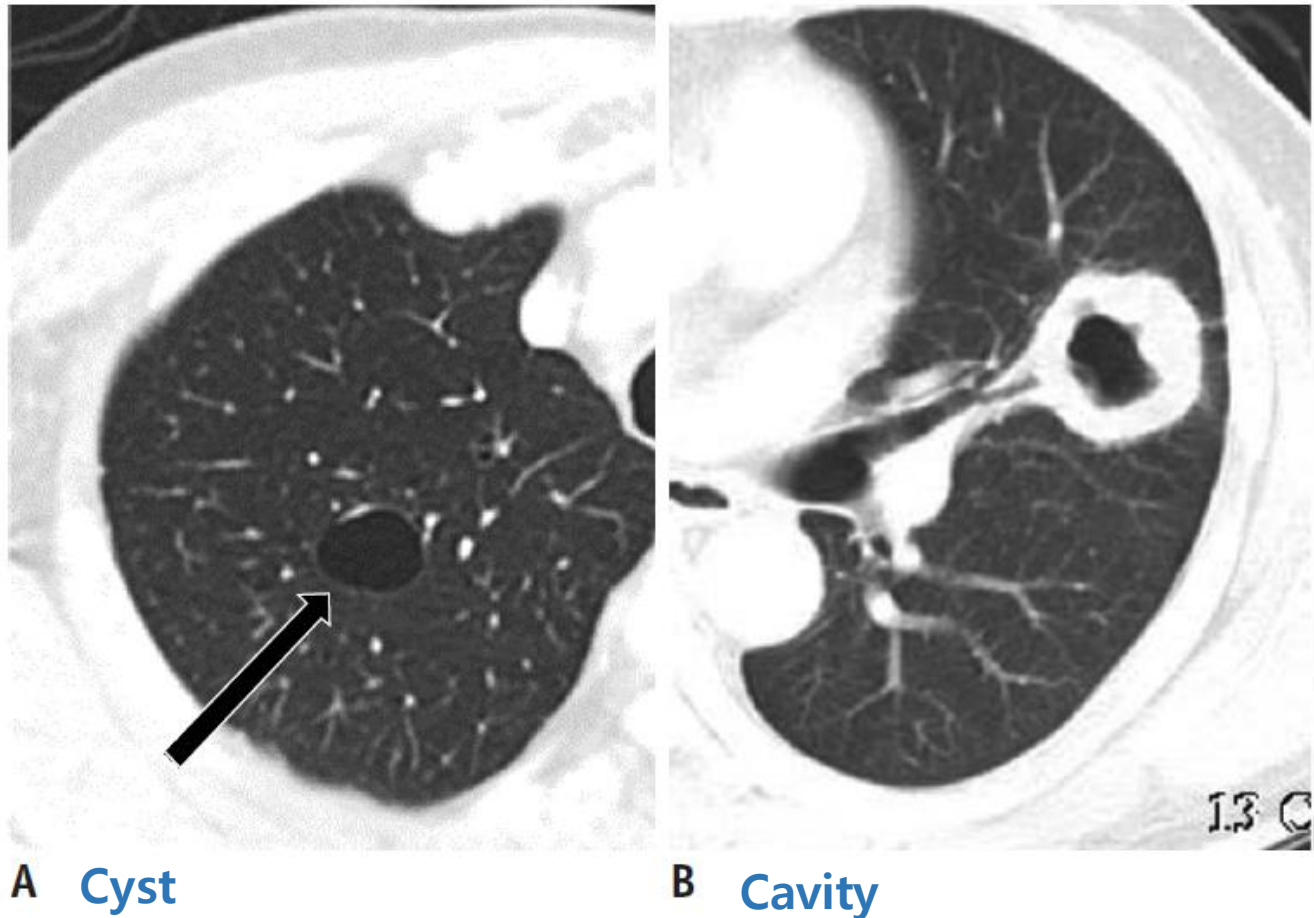


Cyst-like Lesions

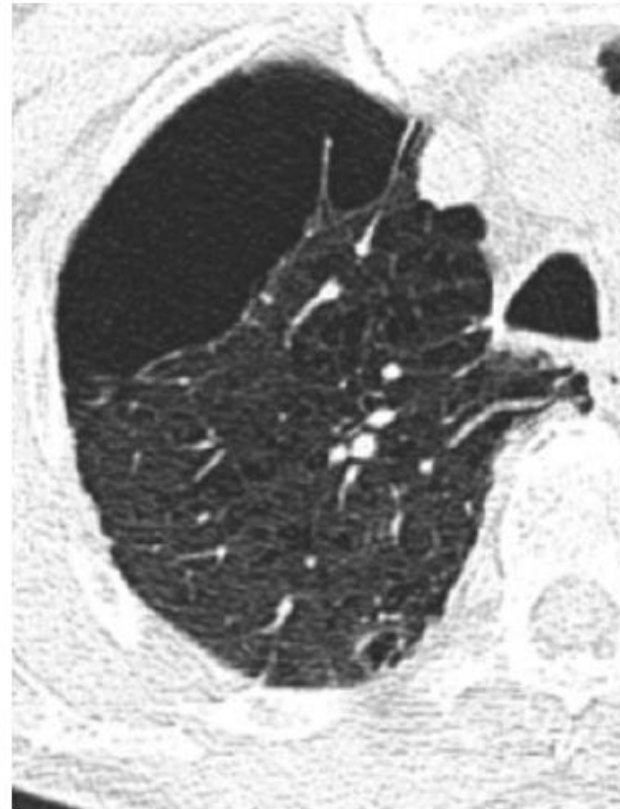
Table 1. Radiologic Distinctions for Air-Filled Lung Lesions

Air-Filled Lung Lesions	Air-Filled Lung Lesion Characteristics	Helpful Radiologic Findings
Cyst	Round Well-defined thin wall (< 2 mm)	Interfaced with normal lung
Cavity	Irregular Thick wall	Within consolidation, mass, or nodule
Bulla	More than 1 cm in size Imperceptible thin wall	Accompanied centrilobular and paraseptal emphysema
Pneumatocele	Round Thin-wall	Transient Adjacent consolidation or ground-glass opacity
Centrilobular emphysema	Usually without visible walls Central dot	Upper lung predominance
Honeycombing	Clustered 3–10 mm cystic lesions Well-defined 1–3 mm thickness walls One or more layers	Lower subpleural lungs Accompanied reticular pattern or traction bronchiectasis
Cystic bronchiectasis	Tubular rather than spherical	Branching pattern Associated bronchial wall thickening, centrilobular densities, air-trapping

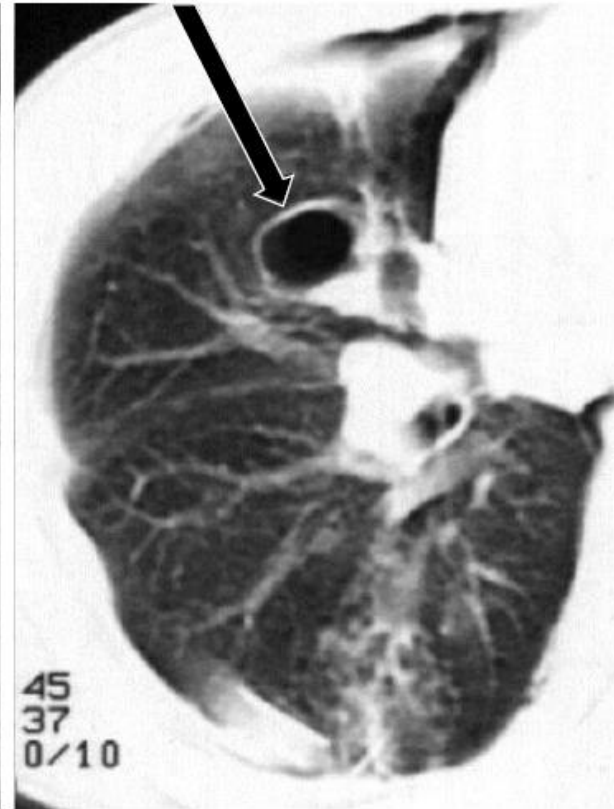
Cyst-like Lesions



Cyst-like Lesions



C Bulla

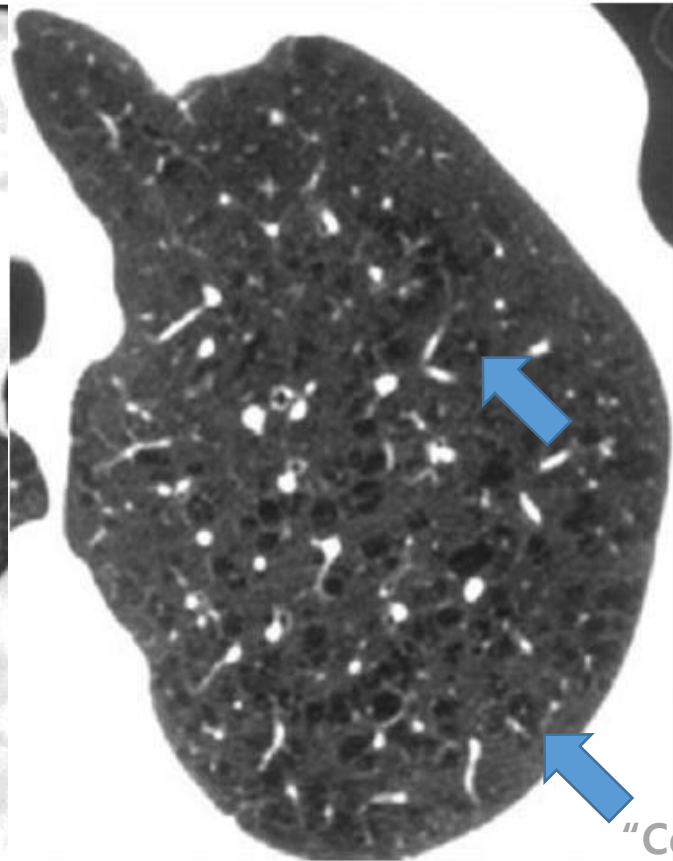


D Pneumatocele

Cyst-like Lesions



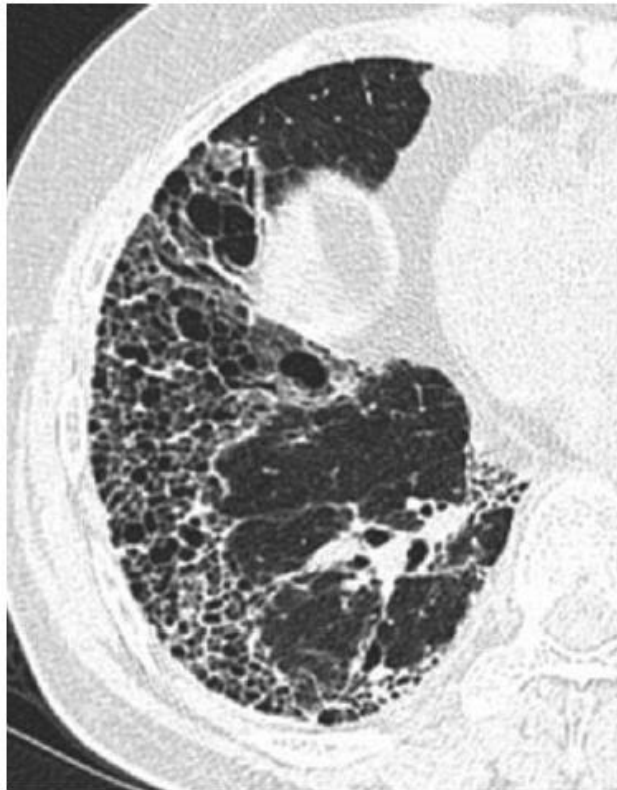
A Cyst



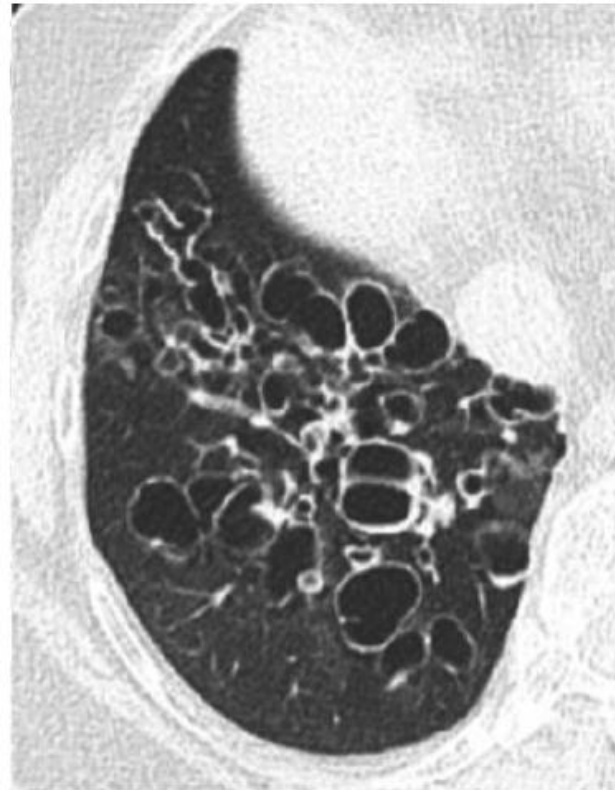
B Emphysema

"Central dot"

Cyst-like Lesions



C Honeycombing



D Bronchiectasis



Diffuse Cystic Lung Disease

Part I

Nishant Gupta^{1,2}, Robert Vassallo³, Kathryn A. Wikenheiser-Brokamp^{4,5,6}, and Francis X. McCormack^{1,2}

	Description
	Lymphangioliomyomatosis—sporadic as well as associated with tuberous sclerosis Pulmonary Langerhans cell histiocytosis, and non-Langerhans cell histiocytoses, including Erdheim Chester disease Other primary and metastatic neoplasms, such as sarcomas, adenocarcinomas, pleuropulmonary blastoma, etc.
2. Genetic/developmental/congenital	Birt-Hogg-Dubé syndrome Proteus syndrome, neurofibromatosis, Ehlers-Danlos syndrome Congenital pulmonary airway malformation, bronchopulmonary dysplasia, etc.
3. Associated with lymphoproliferative disorders	Lymphocytic interstitial pneumonia Follicular bronchiolitis Sjögren syndrome Amyloidosis Light-chain deposition disease
4. Infectious	<i>Pneumocystis jiroveci</i> Staphylococcal pneumonia Recurrent respiratory papillomatosis Endemic fungal diseases, especially coccidioidomycosis Paragonimiasis
5. Associated with interstitial lung diseases	Hypersensitivity pneumonitis Desquamative interstitial pneumonia
6. Smoking related	Pulmonary Langerhans cell histiocytosis Desquamative interstitial pneumonia Respiratory bronchiolitis
7. Other/miscellaneous	Post-traumatic pseudocysts Fire-eater's lung Hyper-IgE syndrome
8. DCLD mimics	Emphysema α_1 -antitrypsin deficiency Bronchiectasis Honeycombing seen in late-stage scarring interstitial lung diseases

Table 1. Classification of Diffuse Cystic Lung Diseases

Diffuse Cystic Lung Disease

Part I

Nishant Gupta^{1,2}, Robert Vassallo³, Kathryn A. Wikenheiser-B

Classification	Description
1. Neoplastic	Lymphangioliomyomatosis—sporadic as well as associated with tuberous sclerosis Pulmonary Langerhans cell histiocytosis, and non-Langerhans cell histiocytoses, including Erdheim Chester disease Other primary and metastatic neoplasms, such as sarcomas, adenocarcinomas, pleuropulmonary blastoma, etc.
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Radiologic Characteristics of Lung Cysts for Diagnostic Approach

- *Size*
- *Number*
- *Wall thickness*
- *Distribution*
- *Location*
- *Associated radiologic features – nodule, GGO, interstitial abnormalities, extrapulmonary lesion, etc*

Radiologic Diagnostic Approach to Cystic Lung Diseases

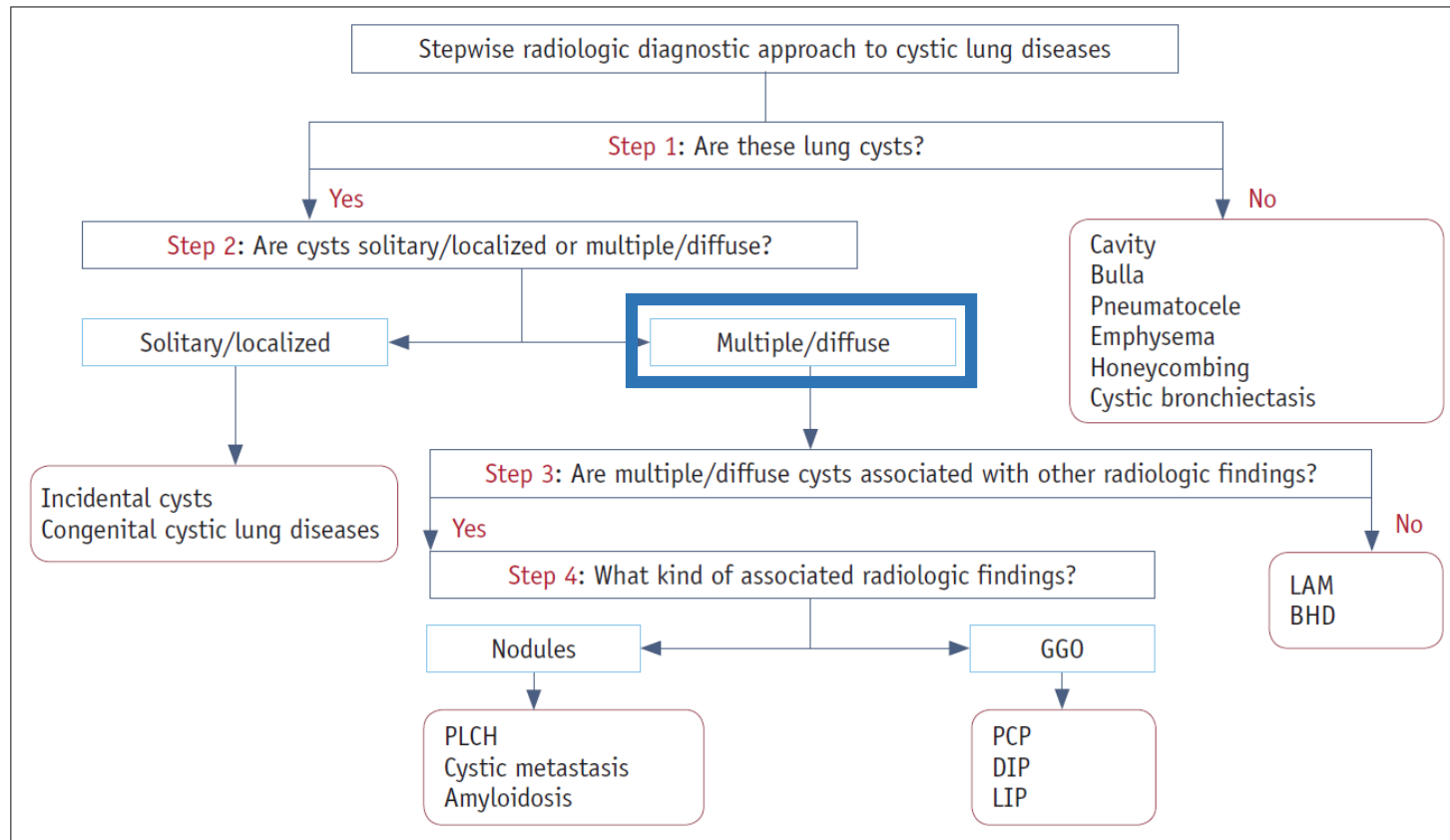


Fig. 1. Stepwise radiologic diagnostic approach to cystic lung diseases. BHD = Birt-Hogg-Dubé syndrome, DIP = desquamative interstitial pneumonia, GGO = ground-glass opacity, LAM = lymphangioleiomyomatosis, LIP = lymphoid interstitial pneumonia, PCP = *pneumocystis jirovecii* pneumonia, PLCH = pulmonary Langerhans cell histiocytosis

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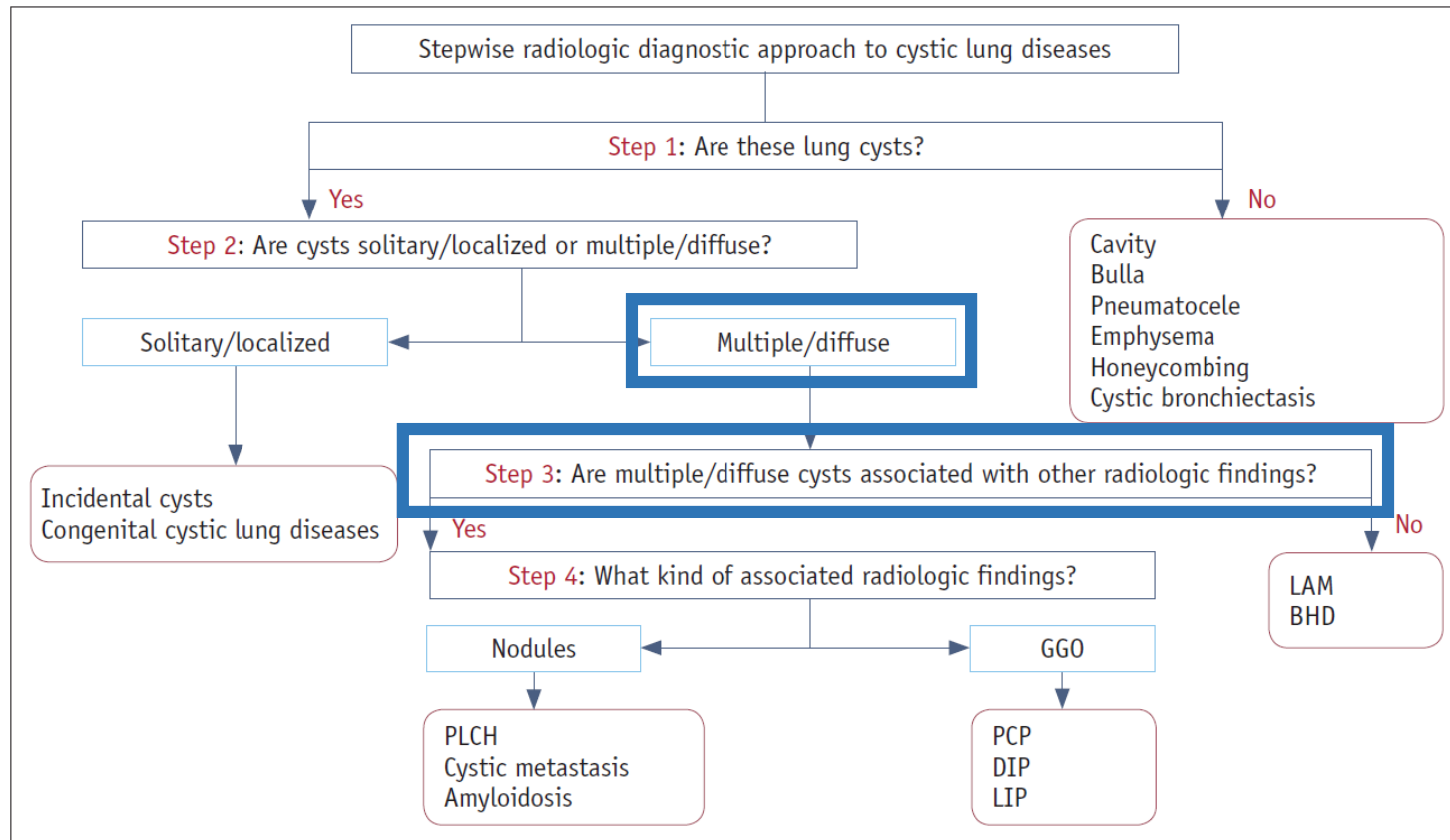


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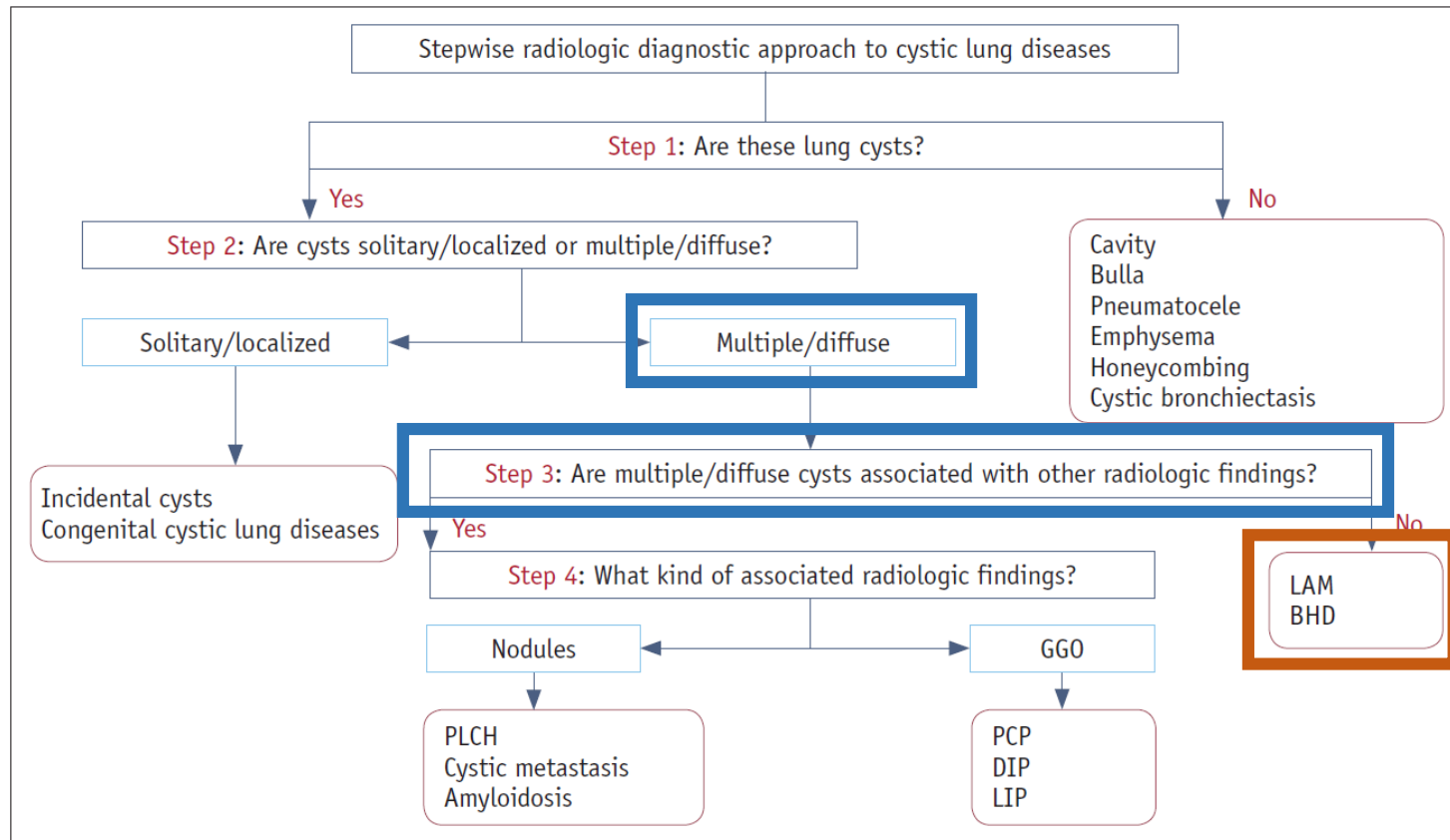
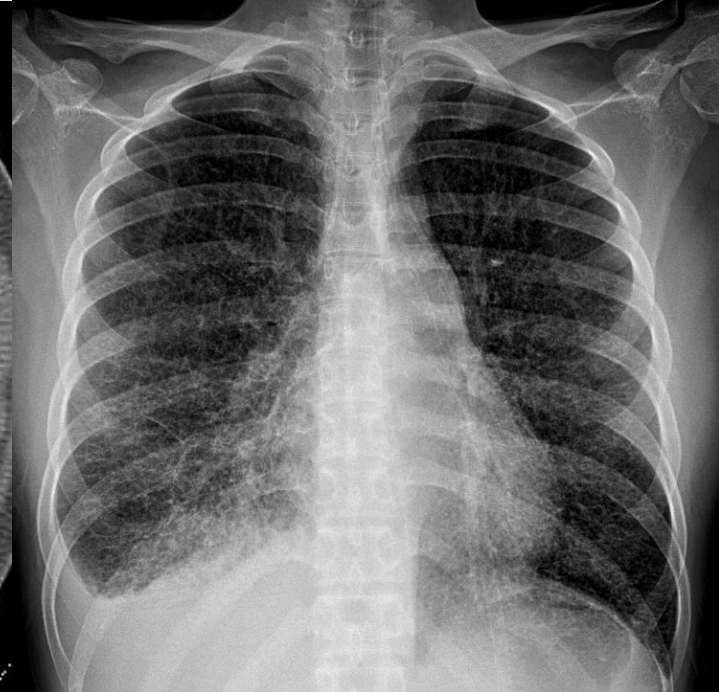
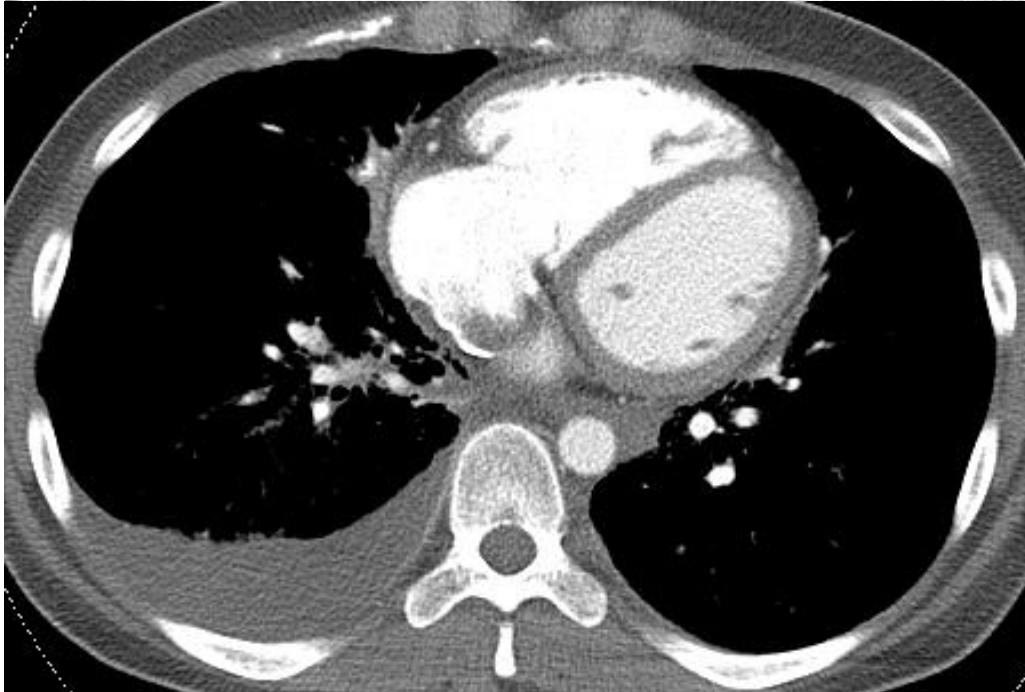
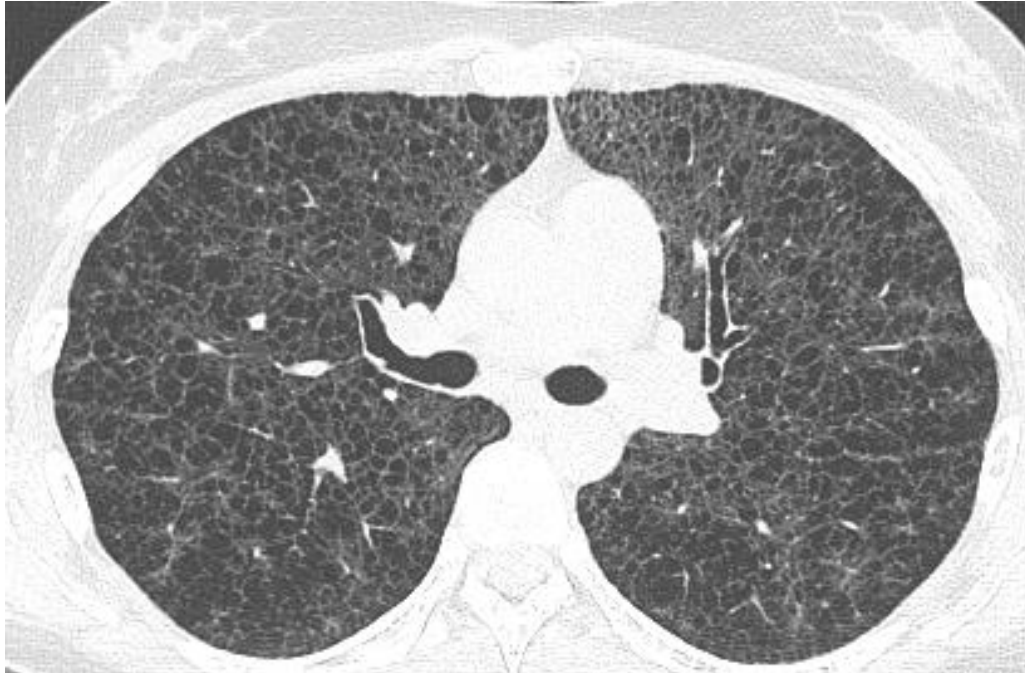
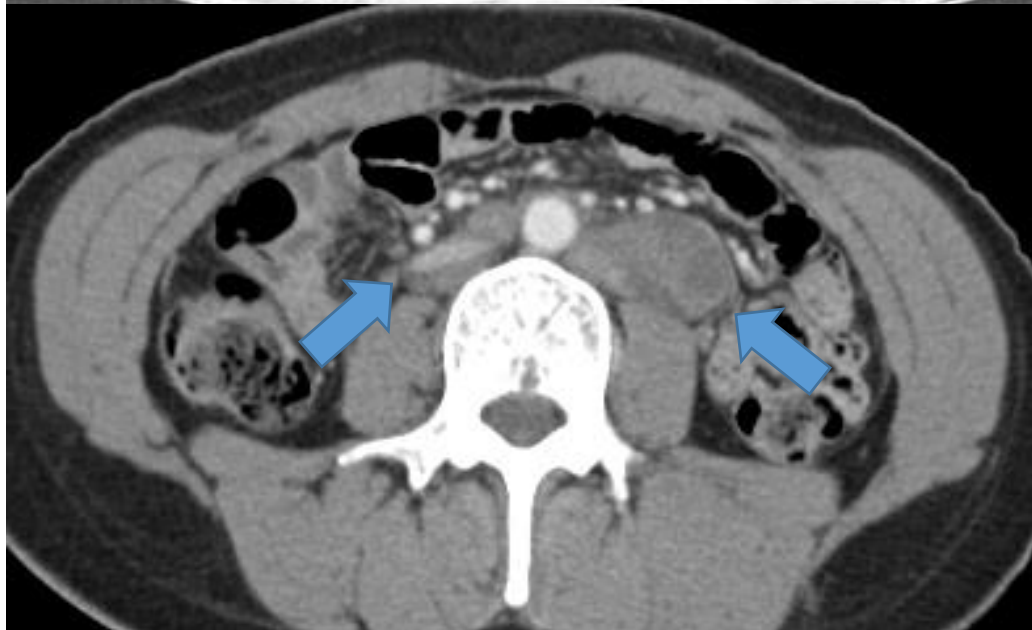
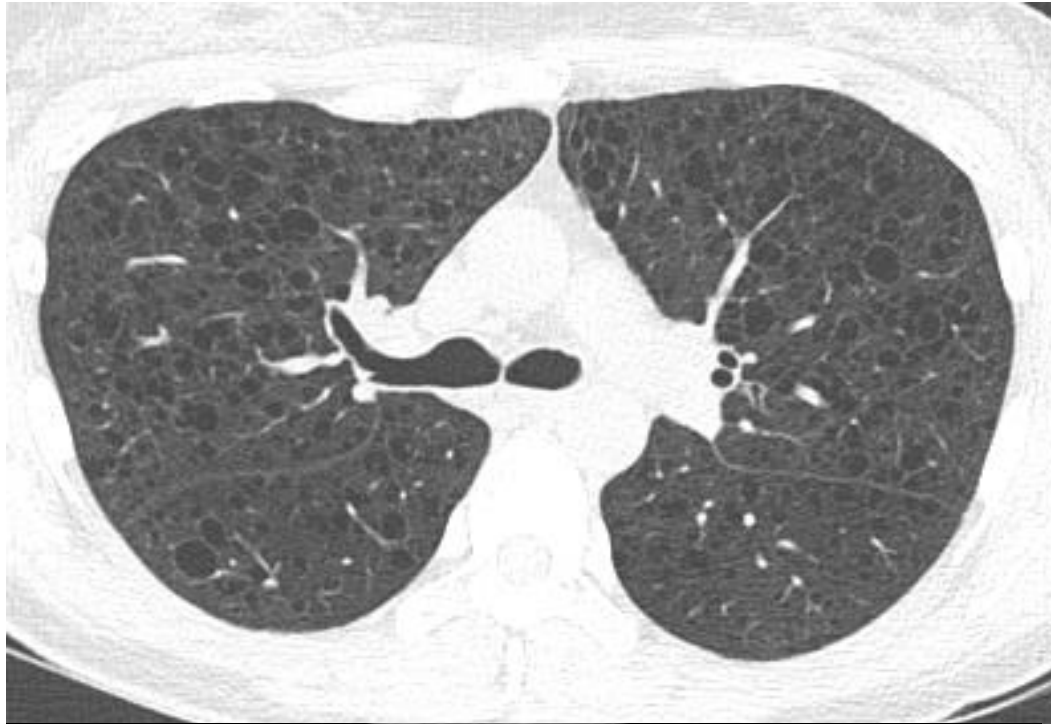


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Lymphangioliomyomatosis (LAM)



Lymphangioliomyomatosis (LAM) with Retroperitoneal Involvement



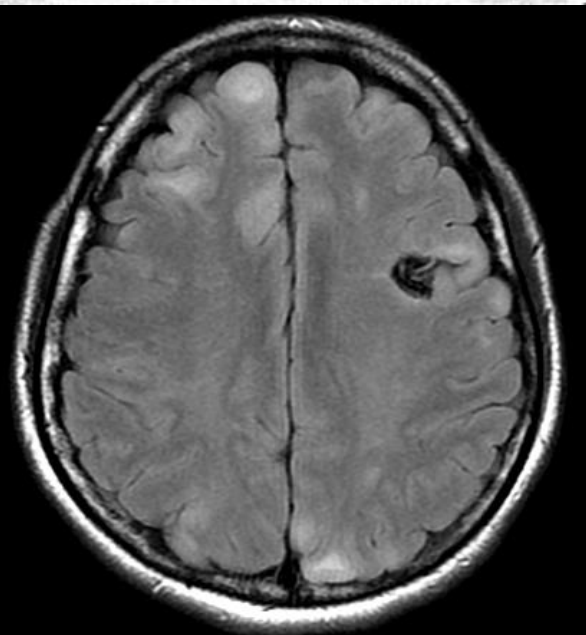
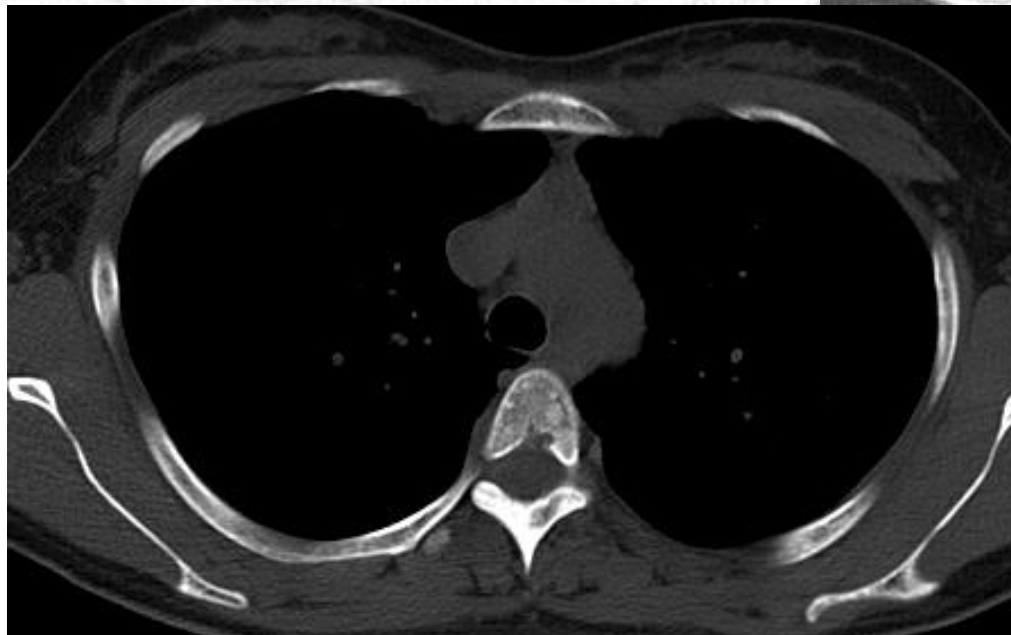
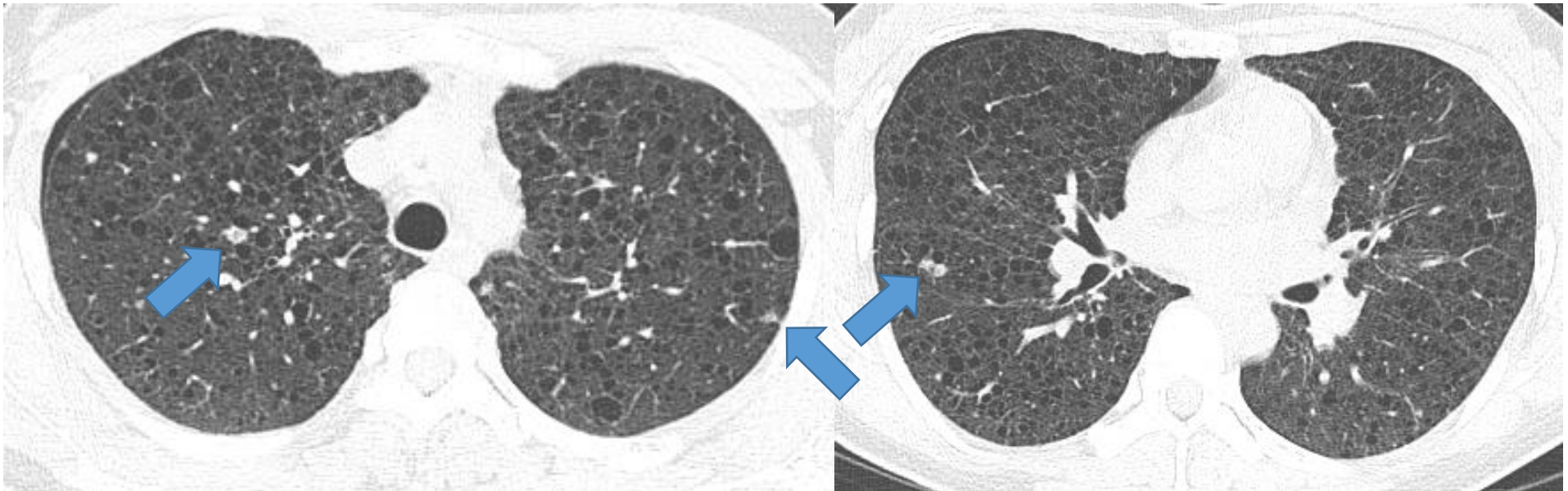
Extrapulmonary Manifestations in LAM

Abdominal lesions (~70%)

- Renal angiomyolipomas (20~54%)
- Lymphangiomyoma (~20%)
- Chylothorax / Chylous ascites
- Lymphadenopathy

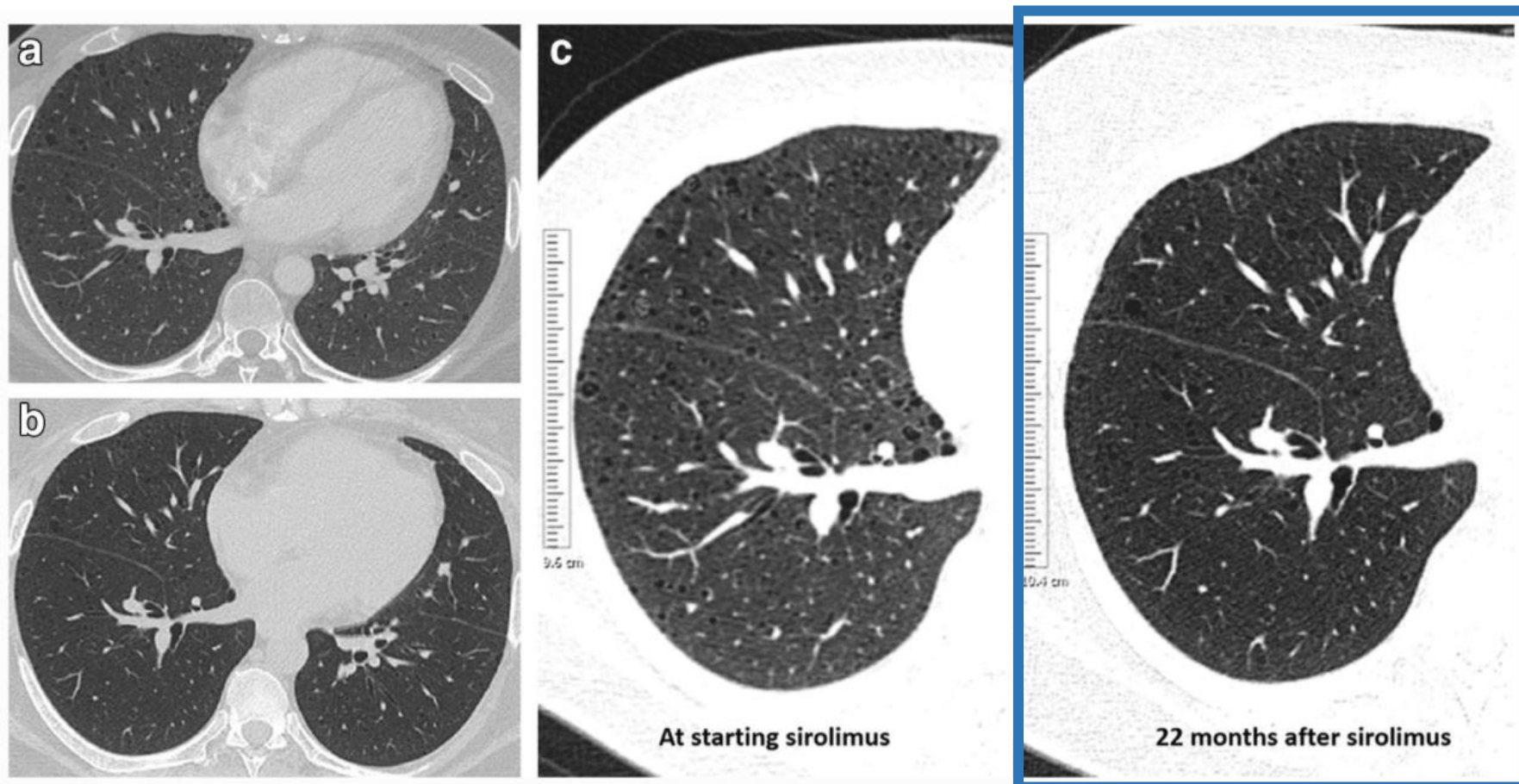
Tuberous Sclerosis Complex (TSC)

- With Multifocal Micronodular Pneumocyte Hyperplasia (MMPH)





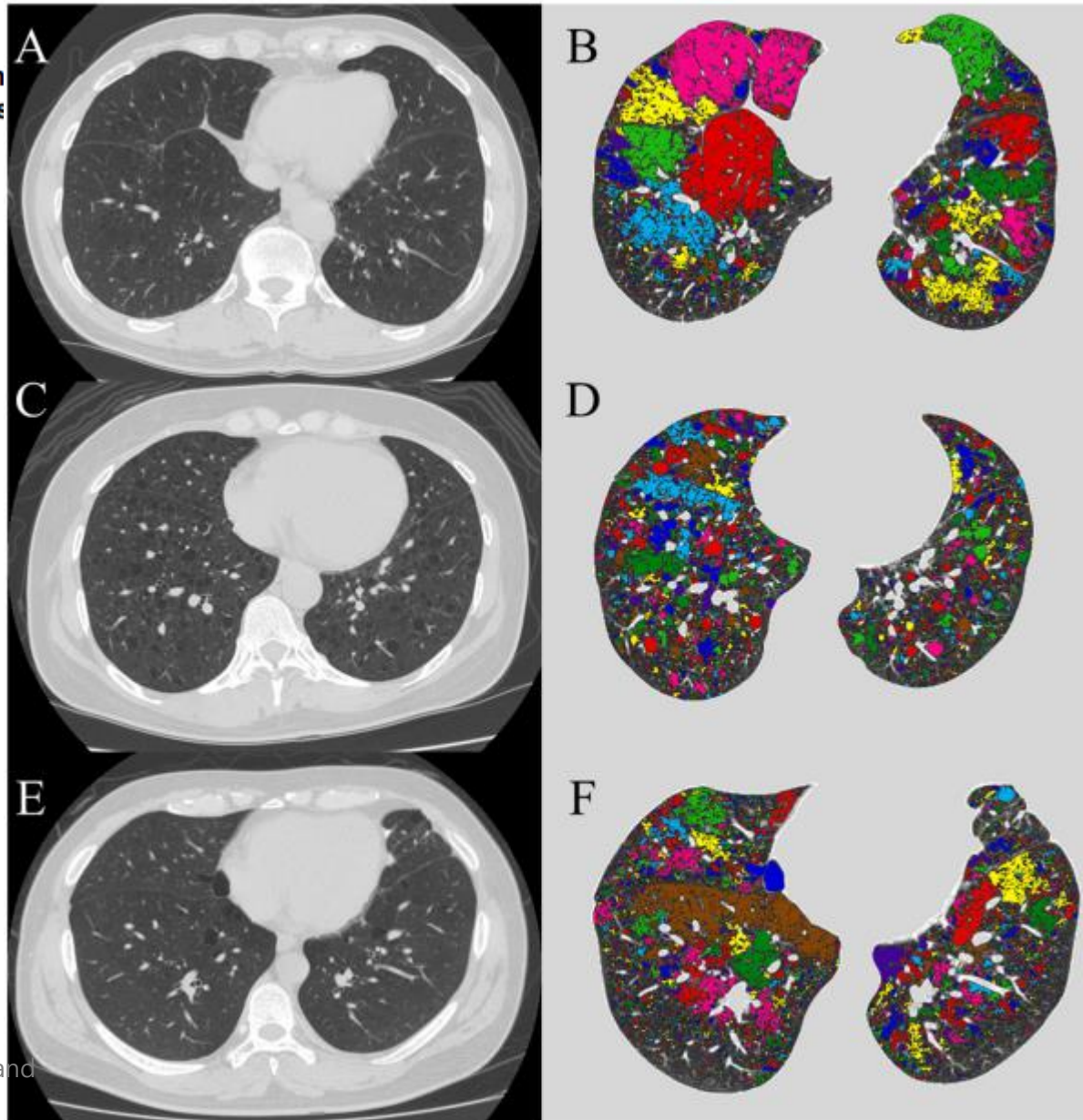
Effects of sirolimus in lymphangioleiomyomatosis patients on lung cysts and pulmonary function: long-term follow-up observational study



Chest CT images from a 37-year-old lymphangioleiomyomatosis patient with sirolimus therapy. The volume and the number of multiple thin-walled cysts decreased from the time of starting sirolimus therapy (a) to 22 months after the start of sirolimus therapy (b). In qualitative analysis, reader scores of both reviewers were “2” at the start of sirolimus, and their scores changed to “1” at 22 months after the start of sirolimus therapy. In quantitative analysis, masked lung cysts were shown in CT images at the start of sirolimus and at 22 months after starting sirolimus therapy (c). The cyst score at starting sirolimus was 16.2% and changed to 12.6% after sirolimus therapy

Difference of the progression of pulmonary cysts assessed by computed tomography among COPD, lymphangioleiomyomatosis, and Birt-Hogg-Dubé syndrome

Kazunori Tobino^{1,2,3*}, Toyohiro Hirai⁴, Takeshi Johkoh⁵, Kim Atsushi Kawaguchi⁷, Noriyuki Tomiyama⁸, Kazuhisa Takahas



Radiologic Diagnostic Approach to Cystic Lung Diseases

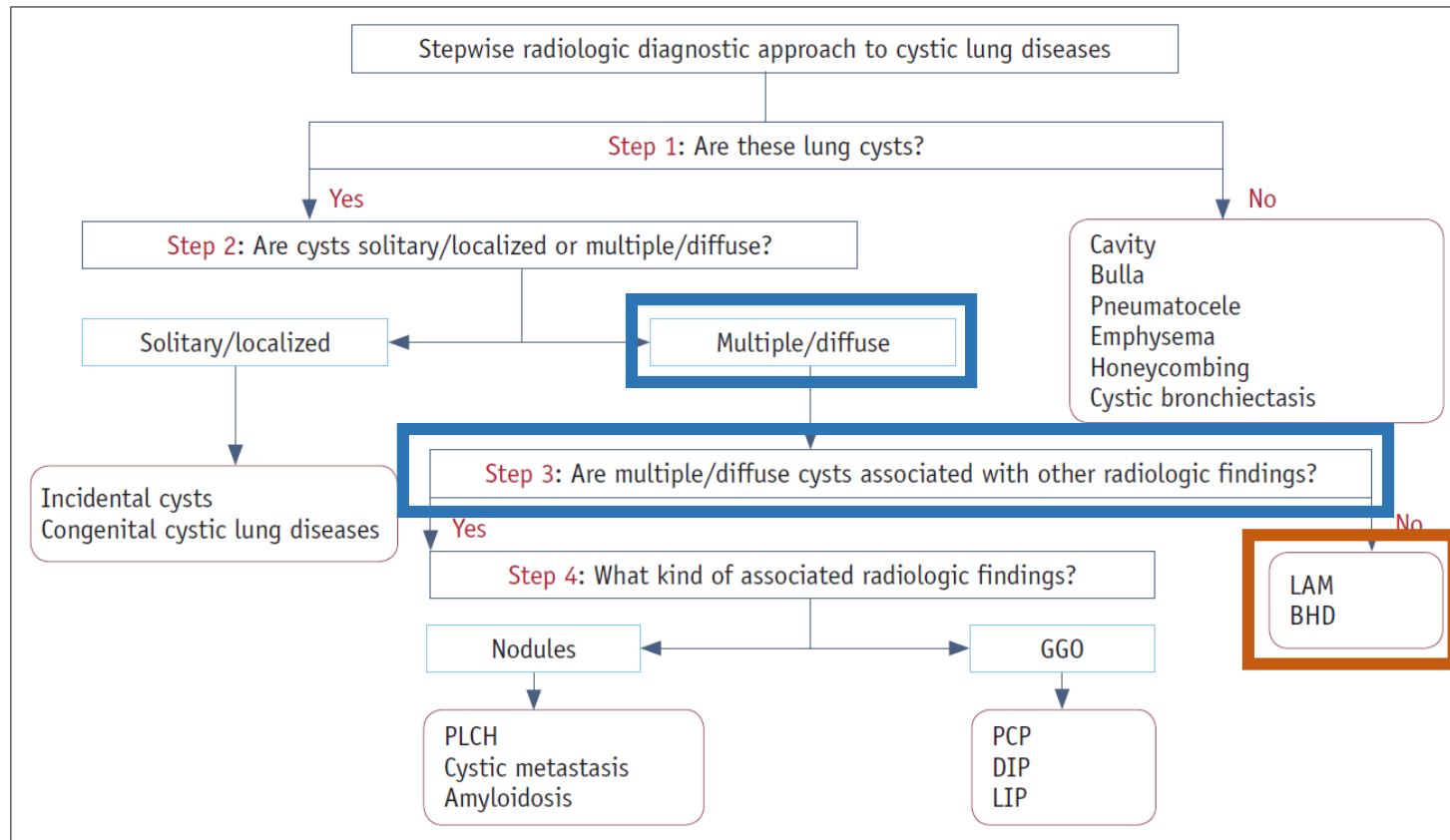


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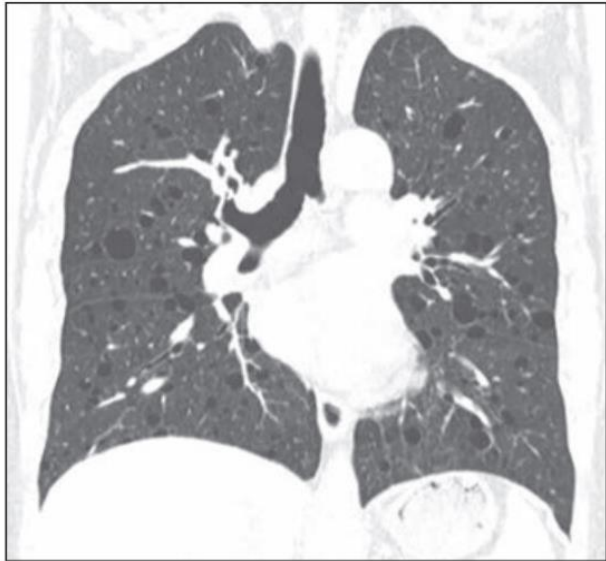
Birt-Hogg-Dubé (BHD) Syndrome



Differentiation Between Lymphangioliomyomatosis and Birt-Hogg-Dubé Syndrome: Analysis of Pulmonary Cysts on CT Images



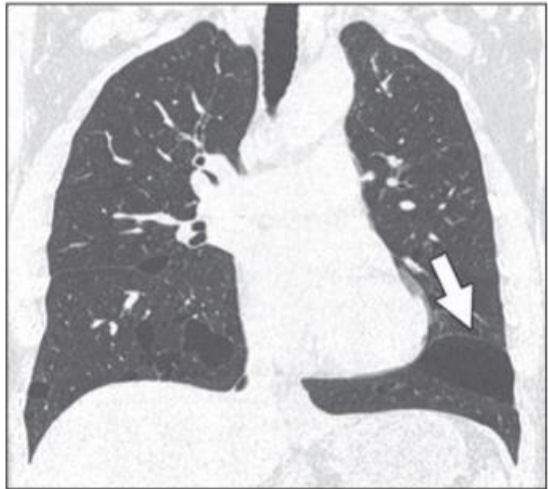
A



D



A

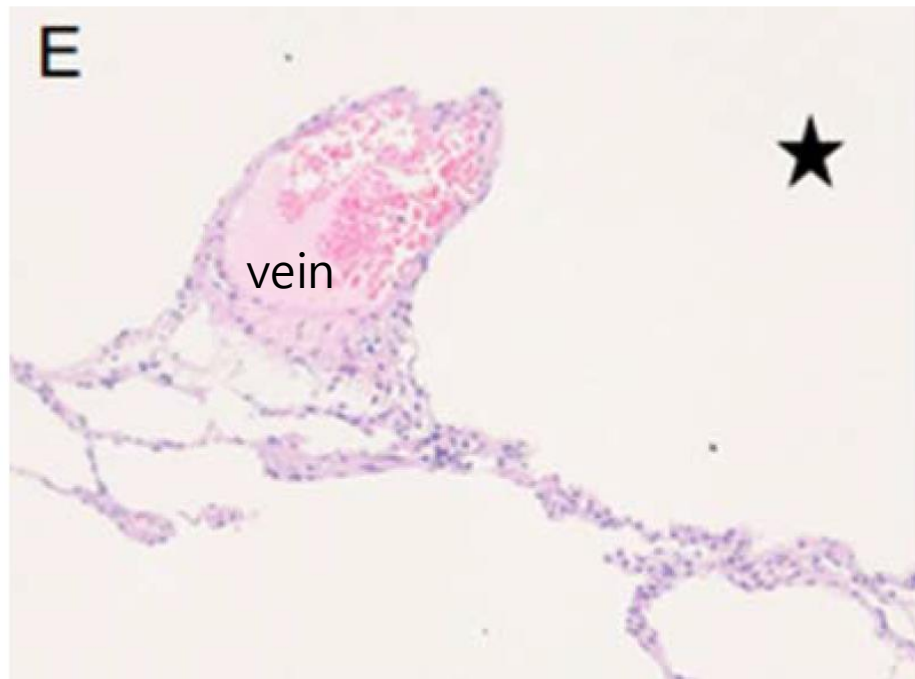
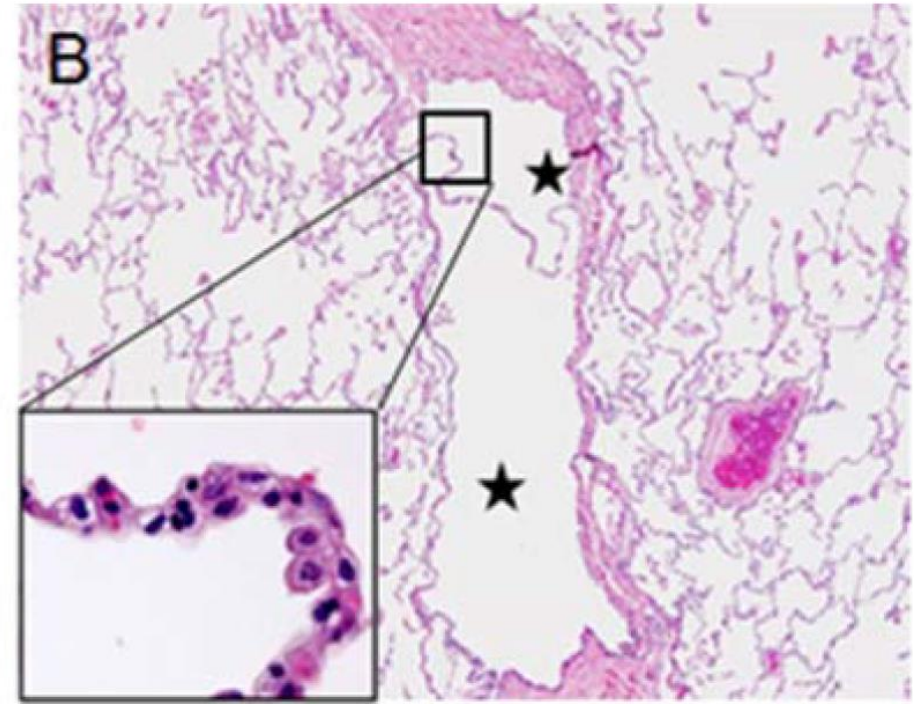
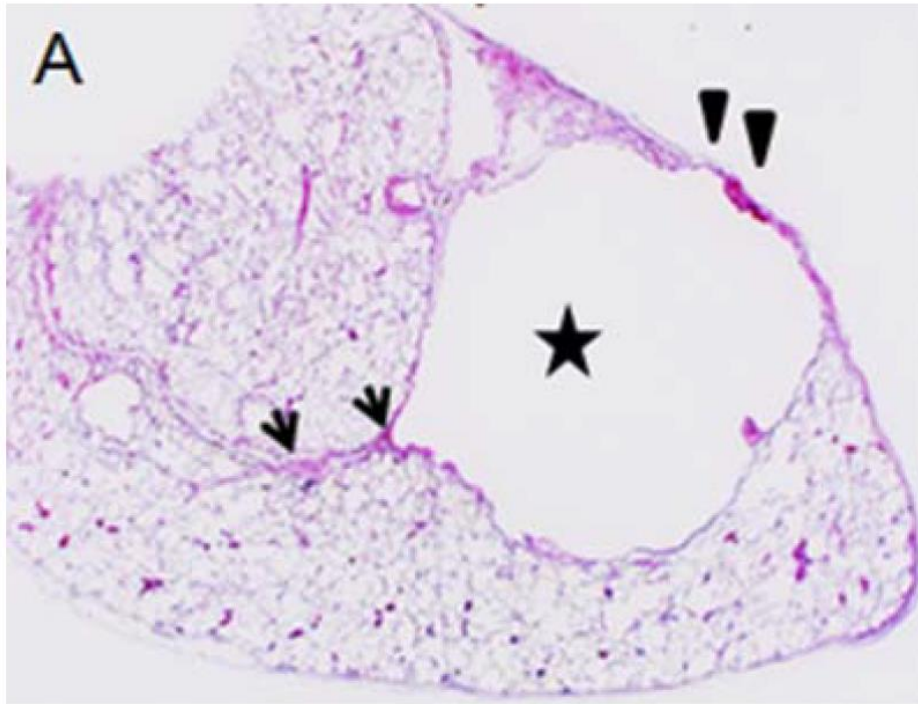


B

Air Cuff Sign - Presence of cysts encircling the bronchovascular bundle

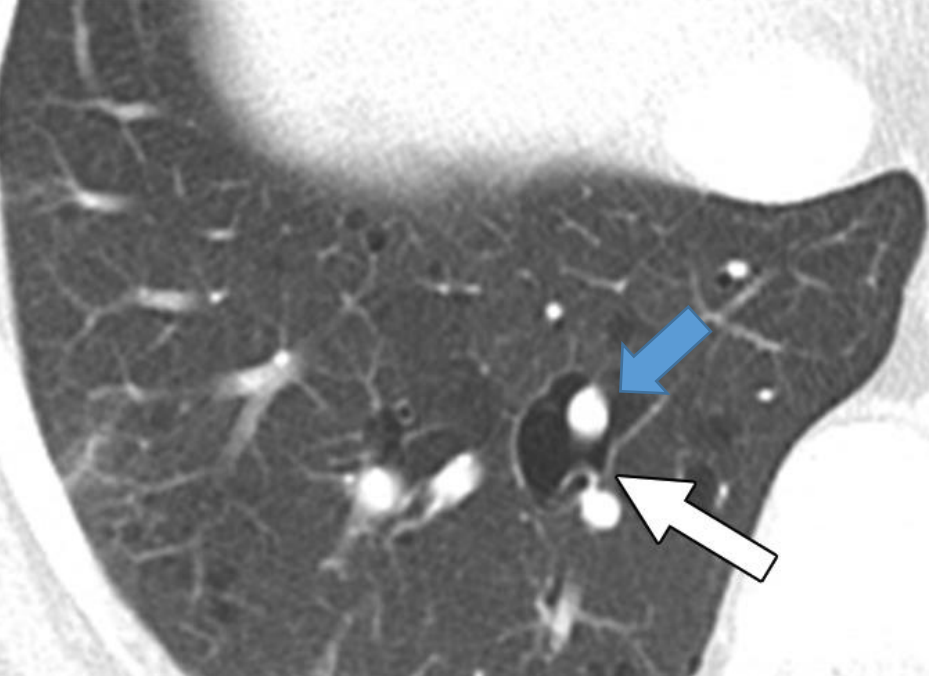
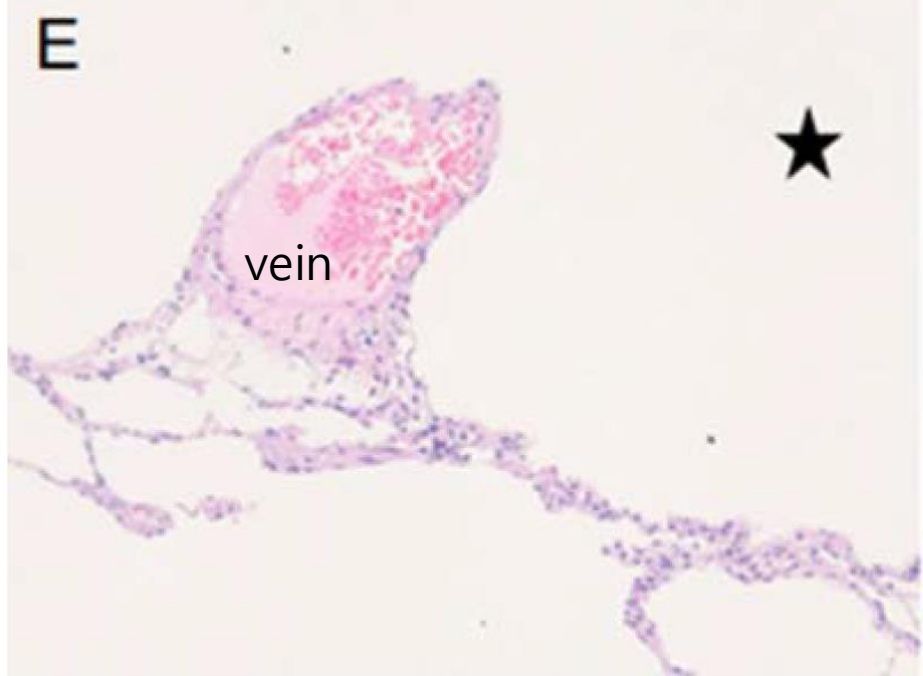
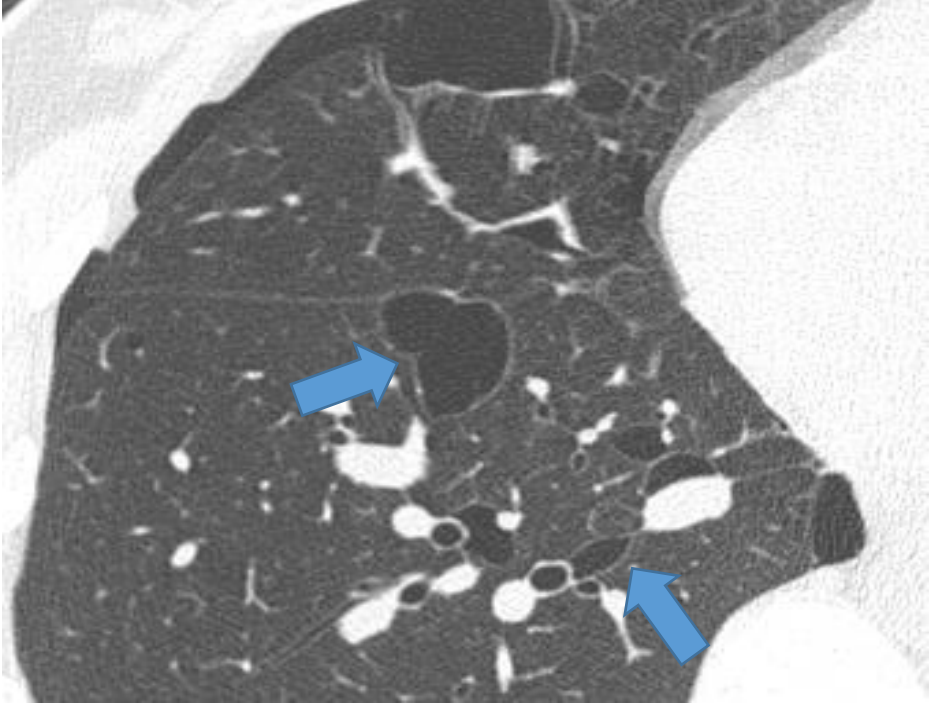
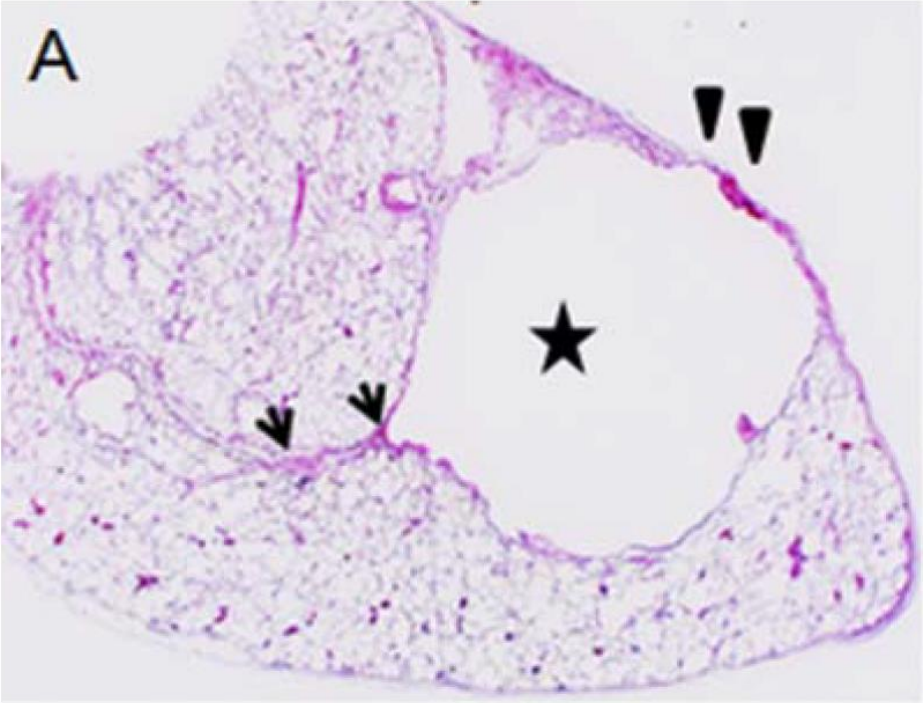


Air Cuff Sign – Cyst Incorporated into Interlobular Septum?

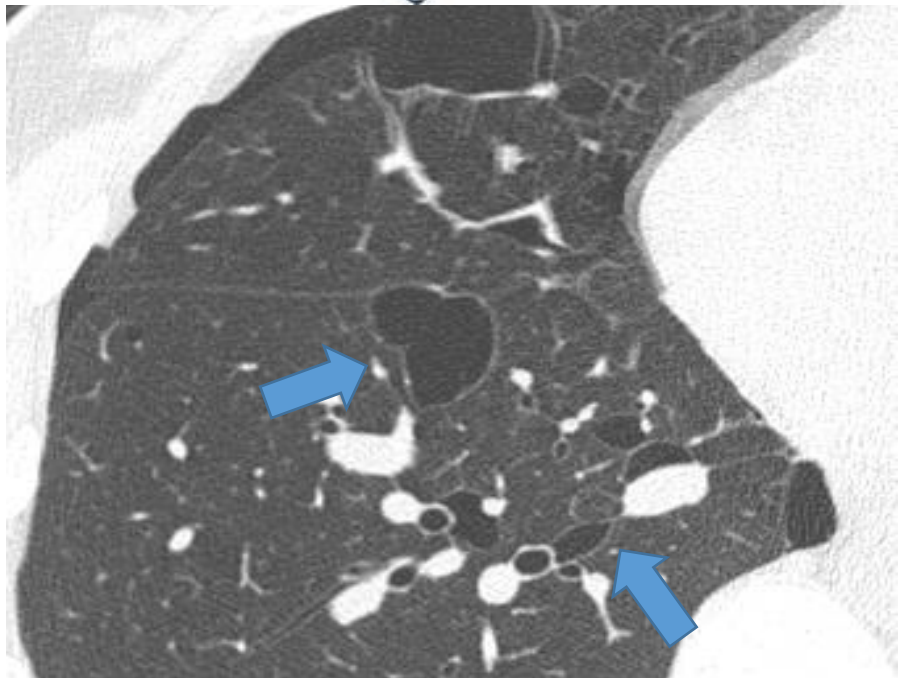
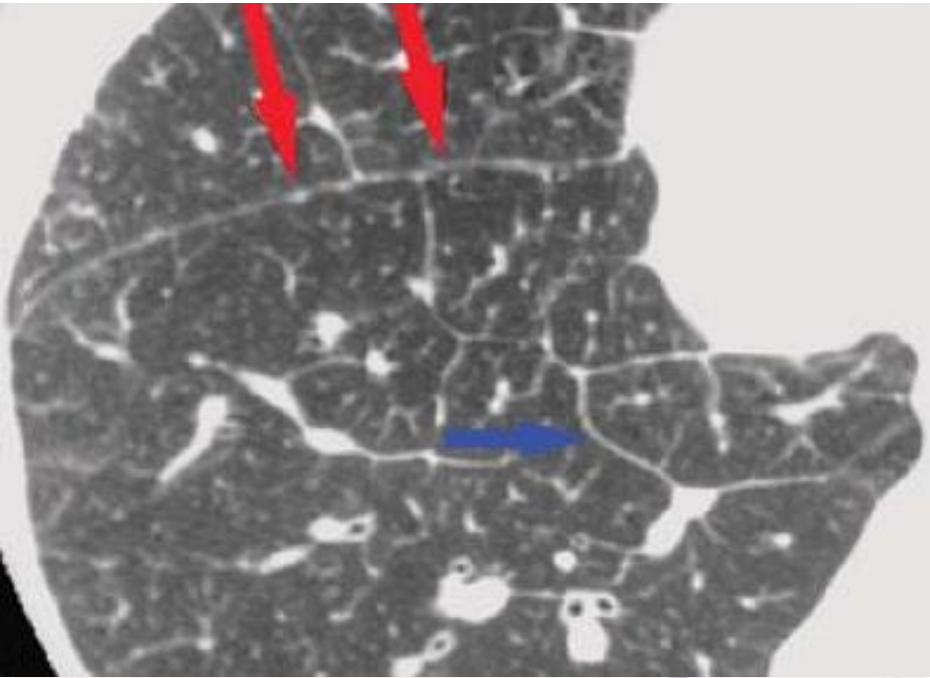
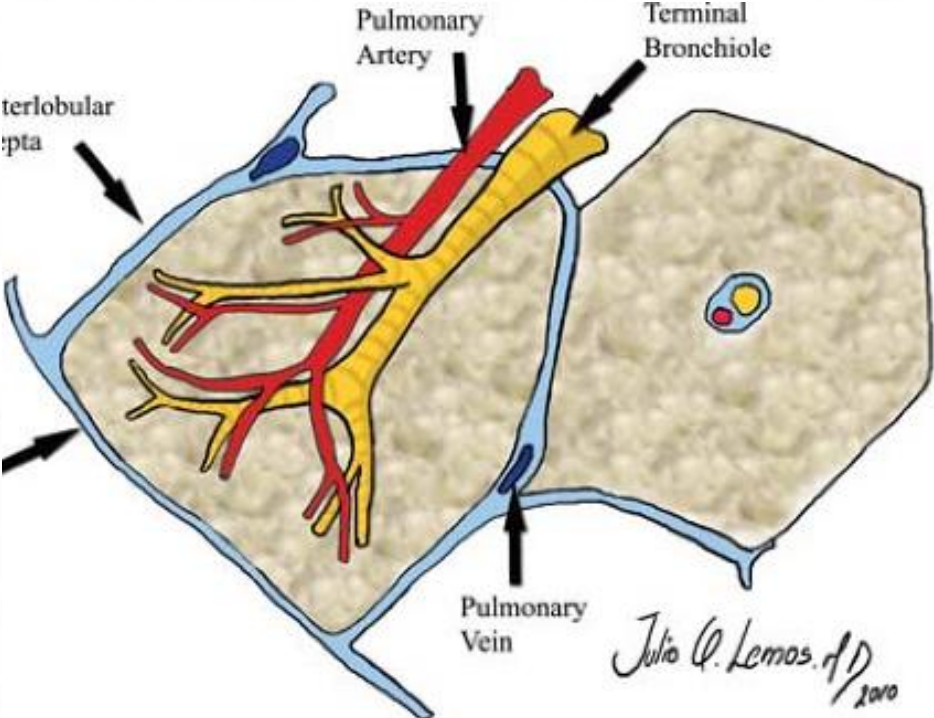
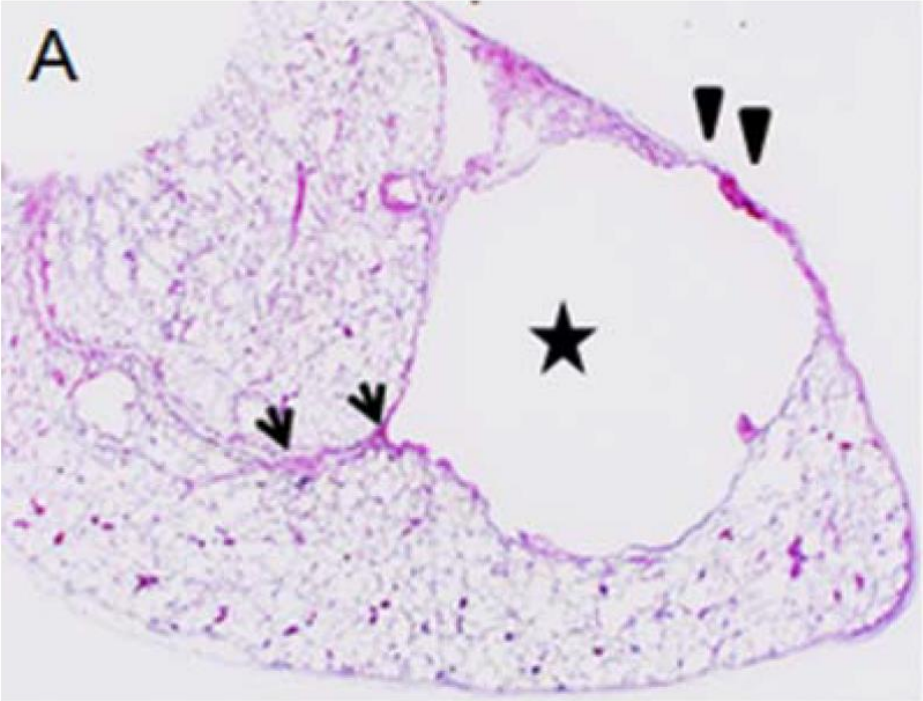


- (A) The cyst wall partially incorporates the pleura (arrowheads) and interlobular septum (arrows).
- (B) The cyst becomes deeply embedded in the interlobular septum.
- (E) Finger-like protrusion of a vein into the cystic space.

Air Cuff Sign – Cyst Incorporated into Interlobular Septum?



Air Cuff Sign – Cyst Incorporated into Interlobular Septum?



Course of Pulmonary Cysts in BHD



FLCN (-)
Skin fibrofolliculoma (+)
Family history of pneumothorax – 큰형님, 숙부2, 조카2
Renal tumor (+, Oncocytoma)
Ex- smoker (30 pyrs)

Radiologic Diagnostic Approach to Cystic Lung Diseases

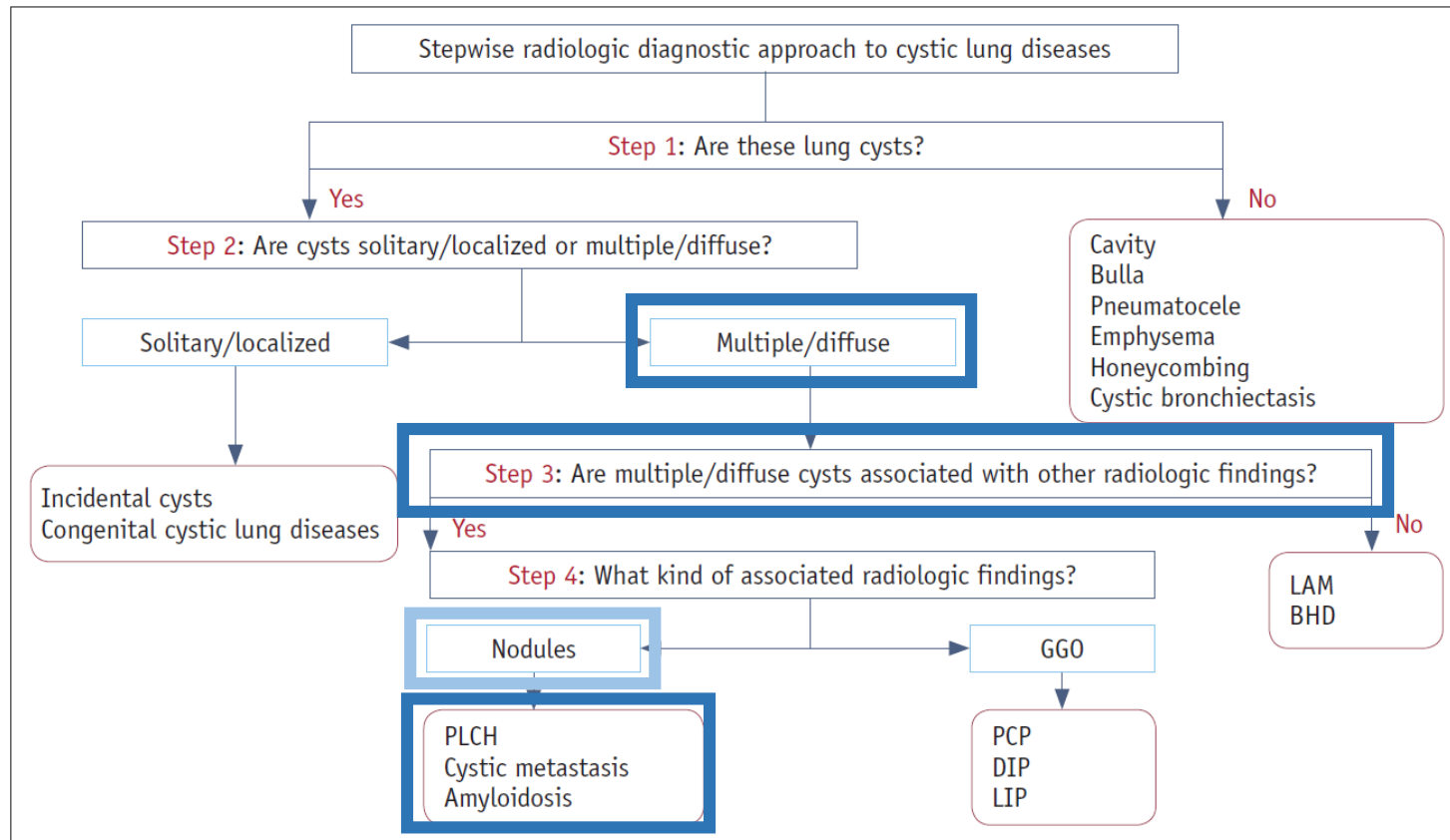
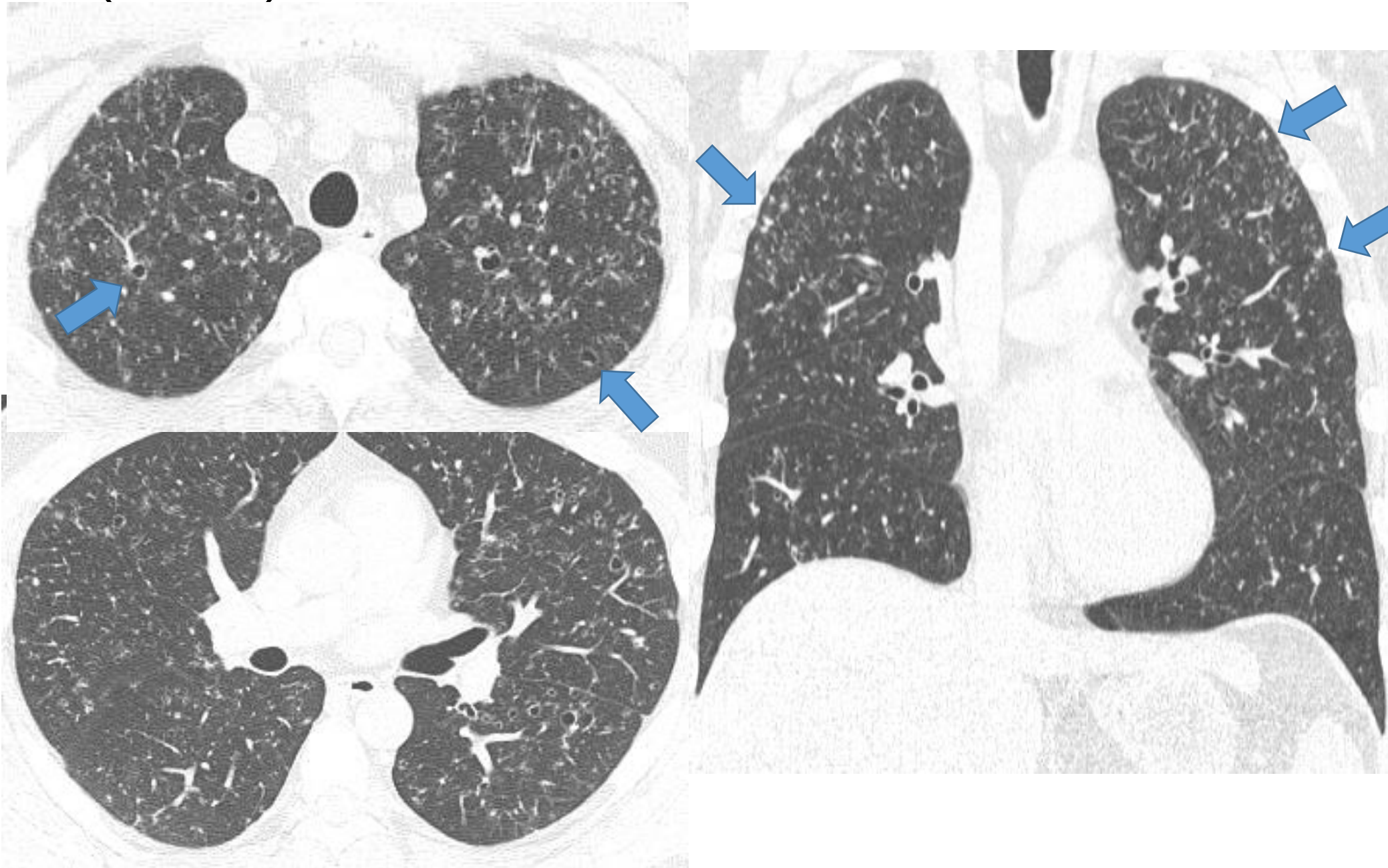
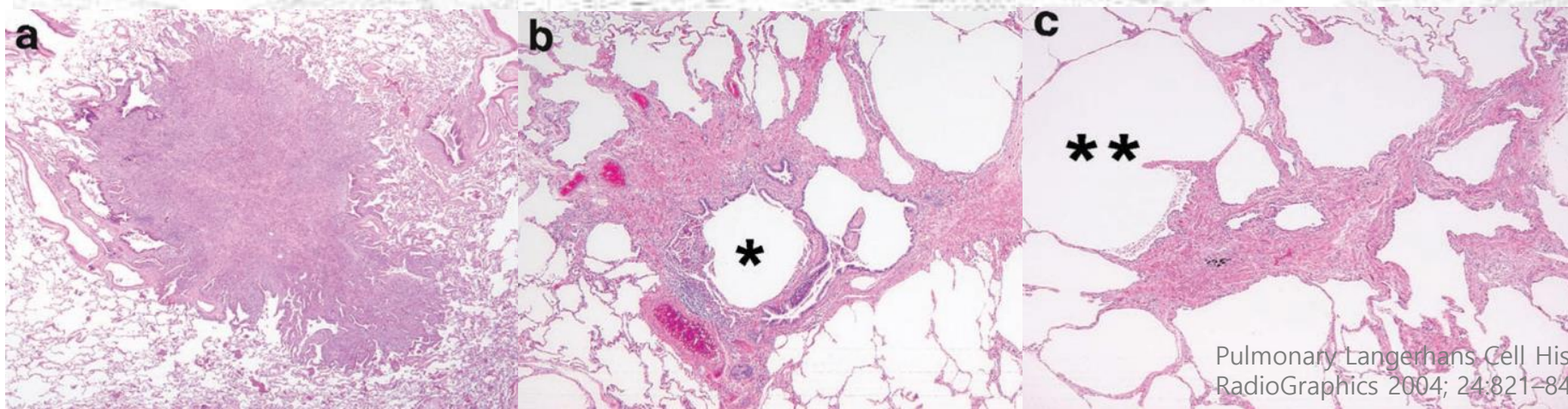
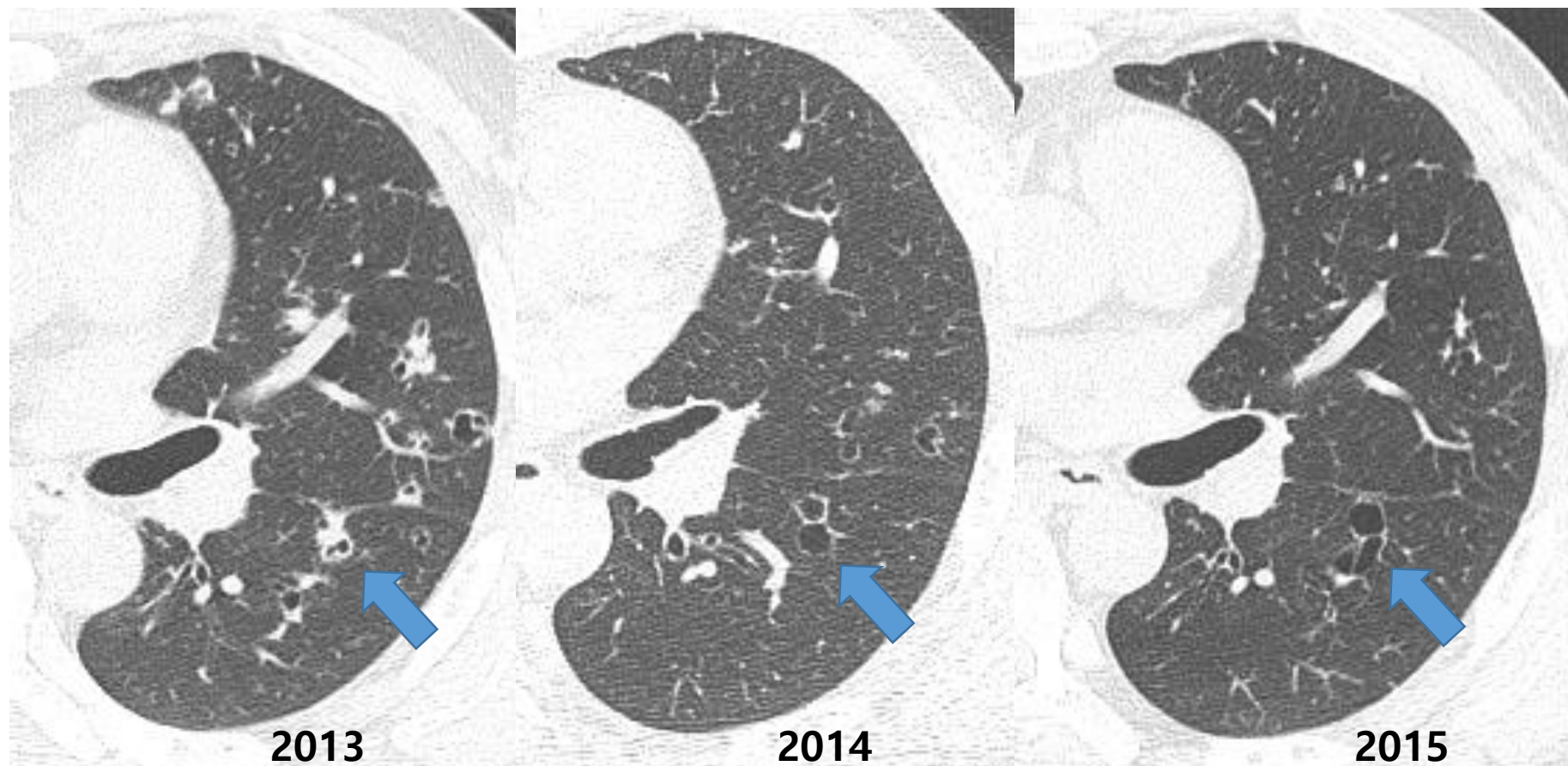


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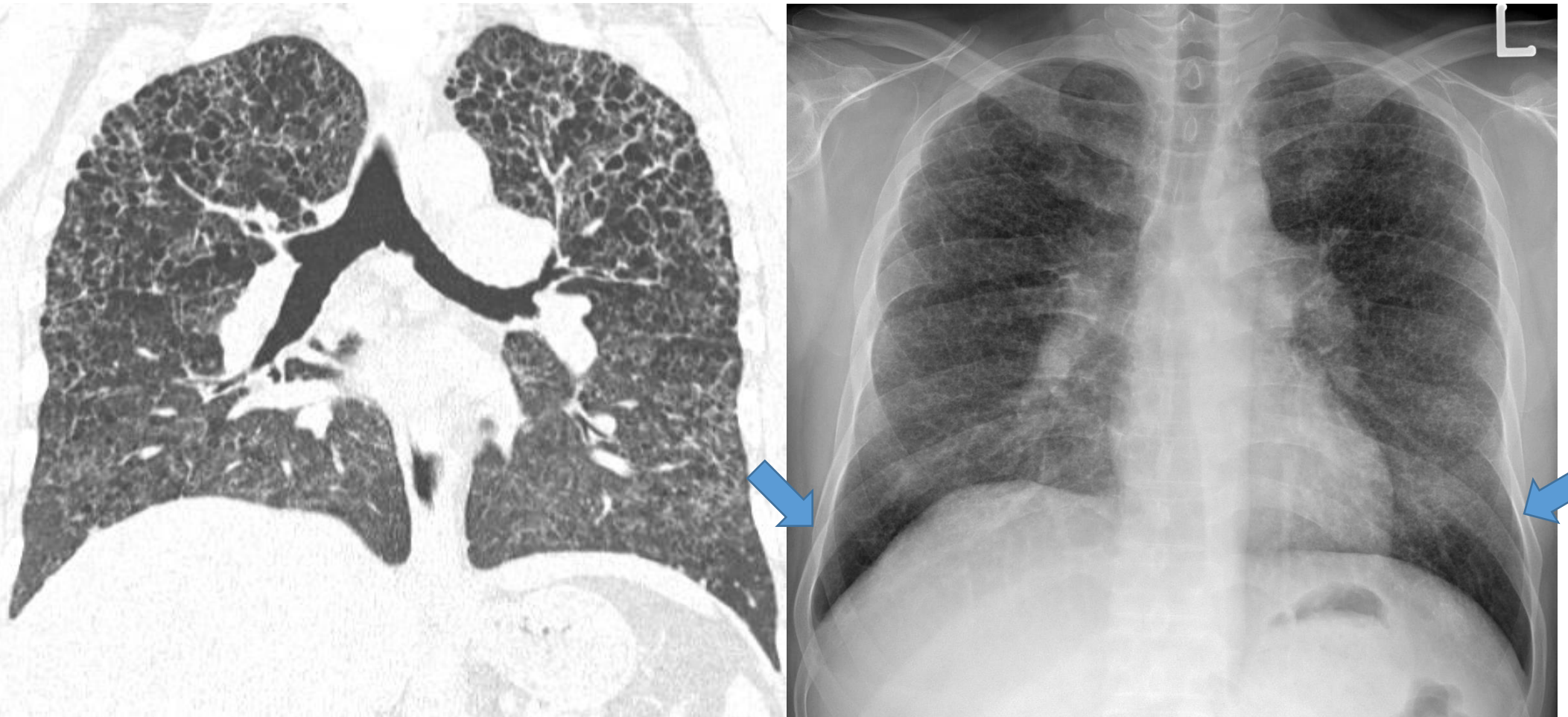
Pulmonary Langerhans Cell Histiocytosis (PLCH)



Course of PLCH



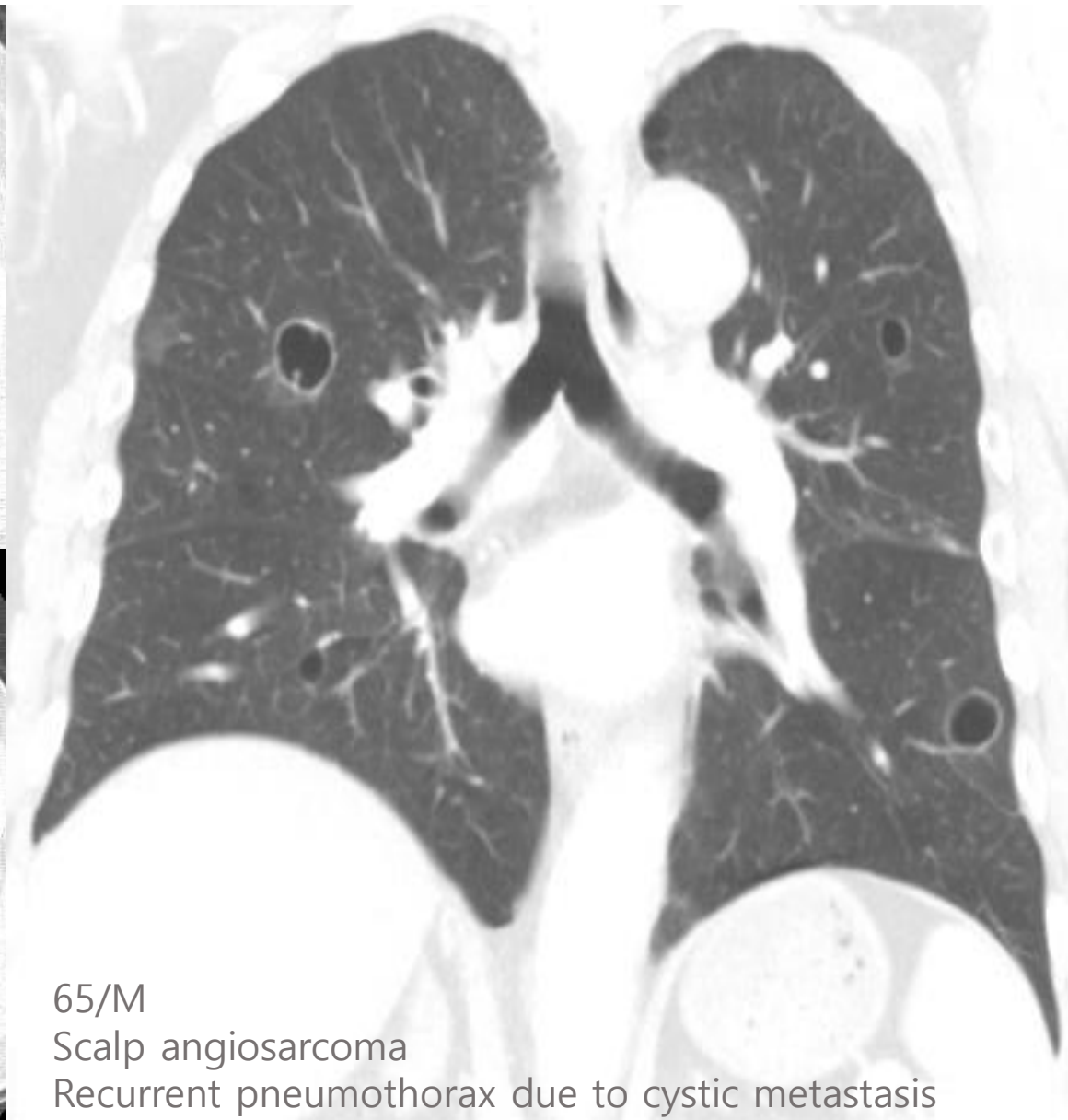
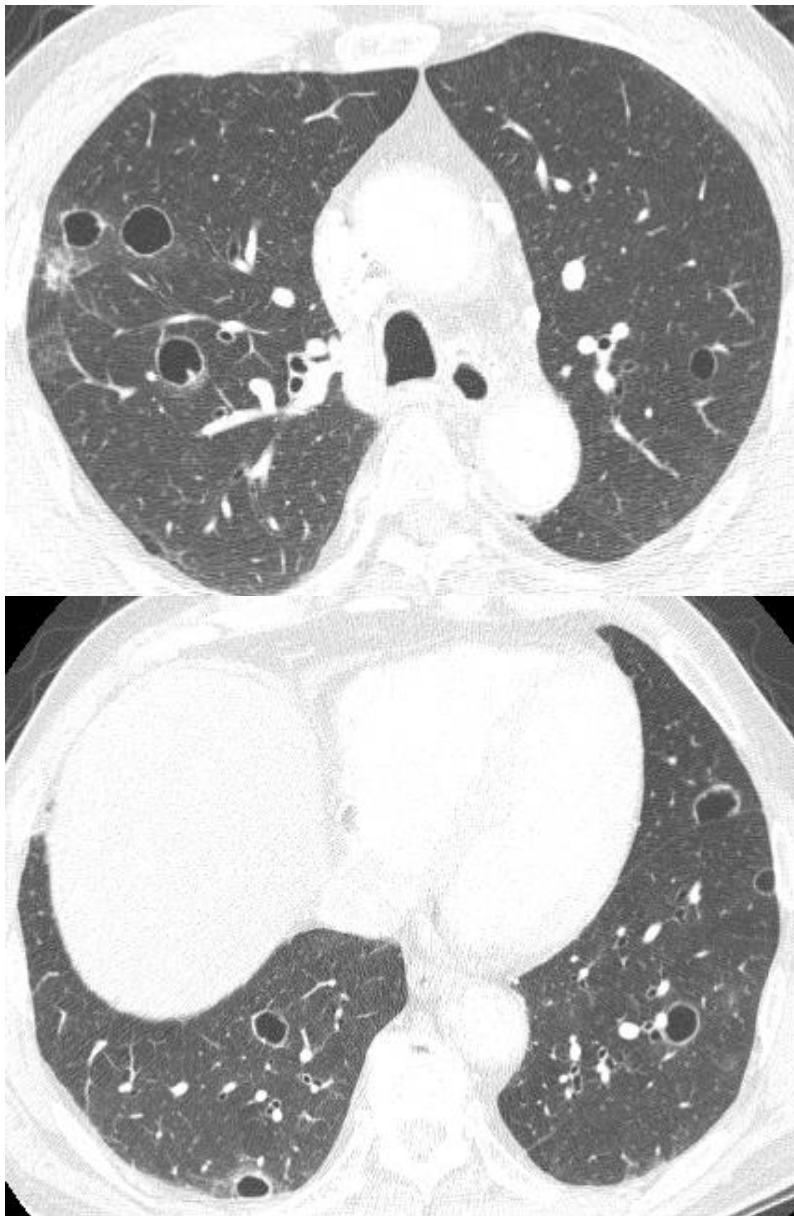
Upper lung dominance Sparing of costophrenic angle



Distribution - PLCH vs. LAM

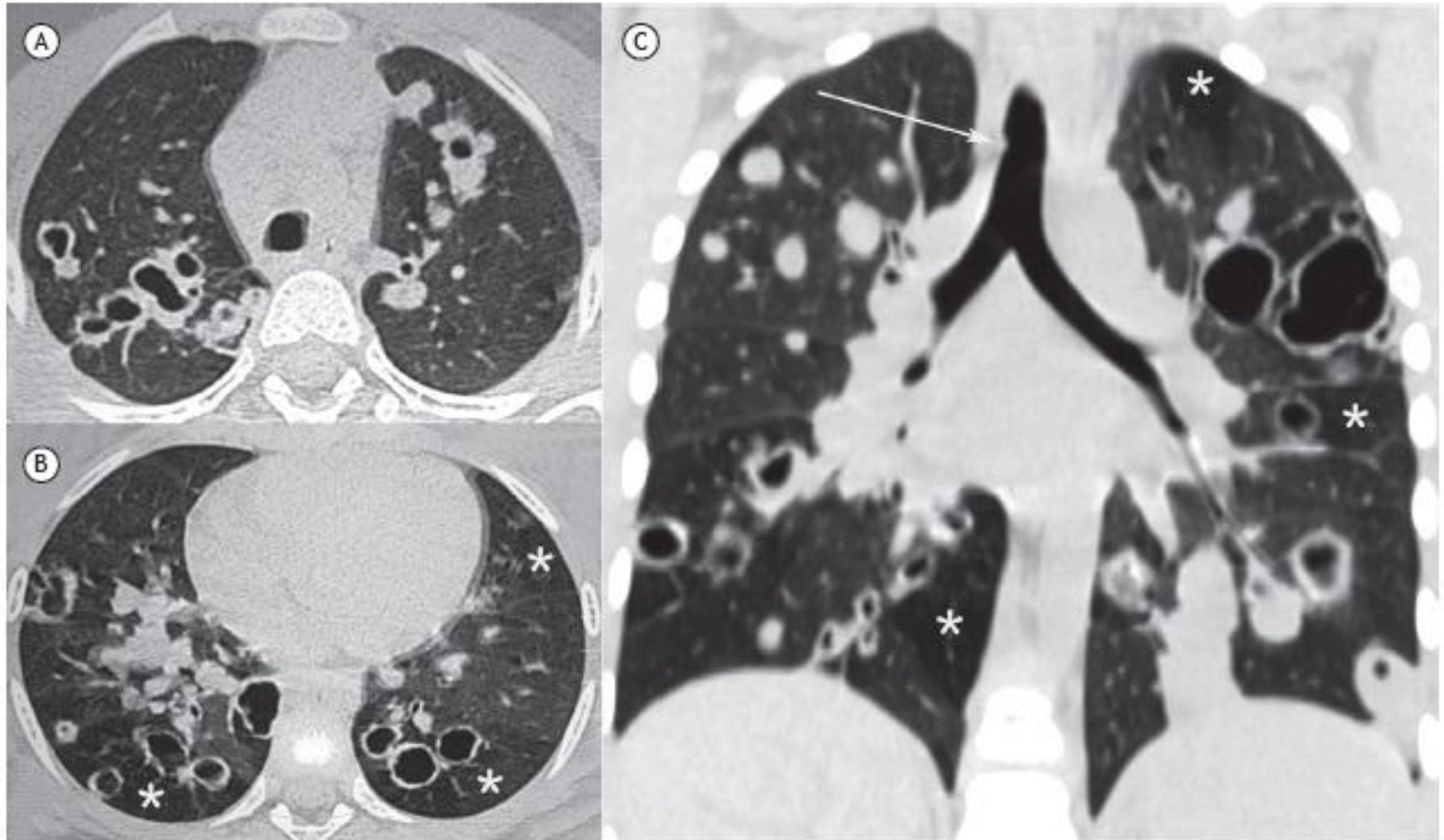


Cystic Metastasis



65/M
Scalp angiosarcoma
Recurrent pneumothorax due to cystic metastasis

Tracheobronchial Papillomatosis



Radiologic Diagnostic Approach to Cystic Lung Diseases

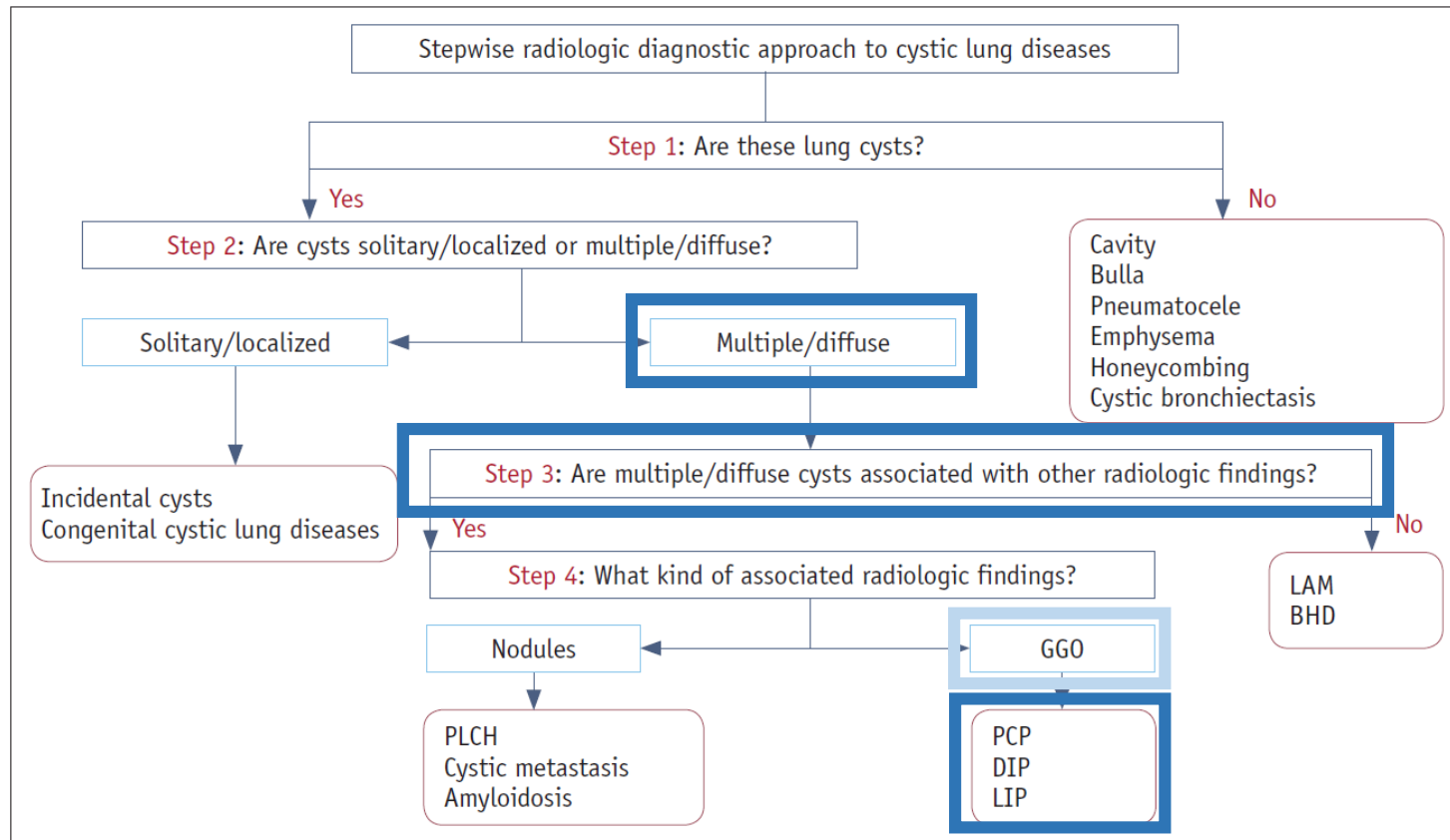
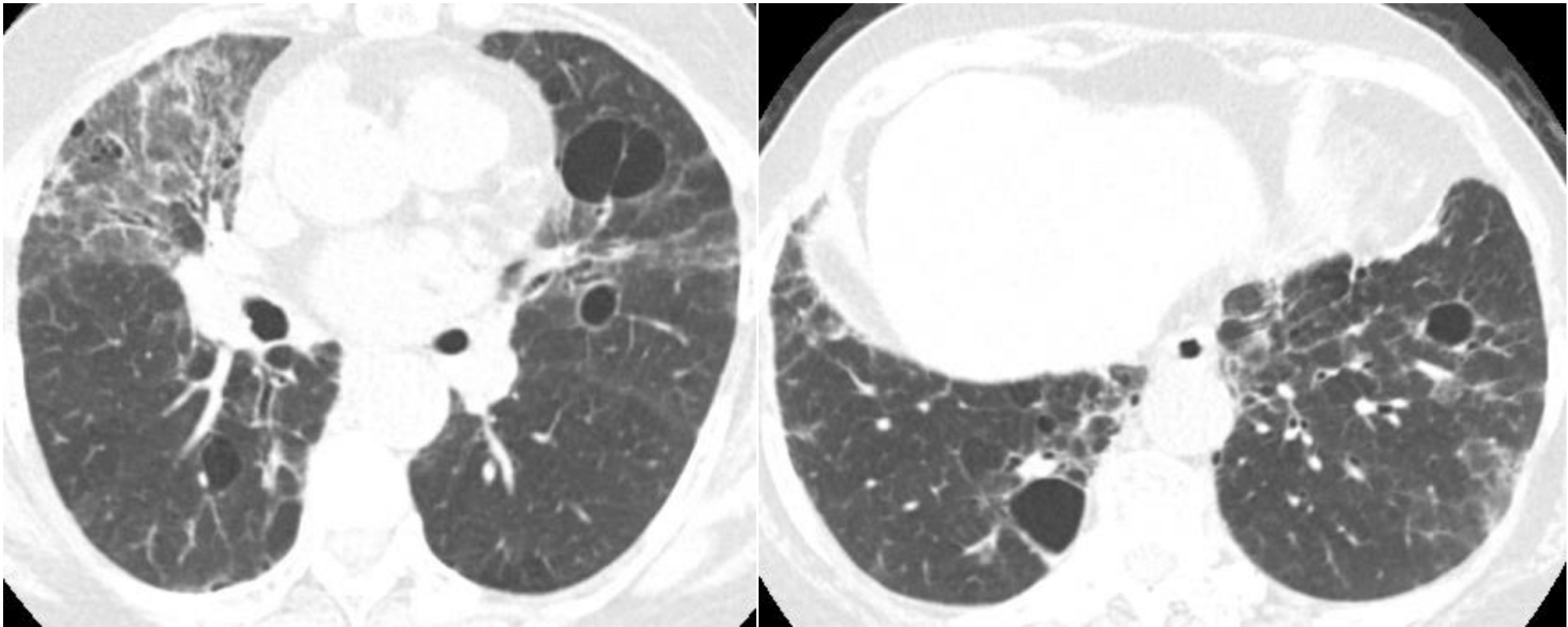
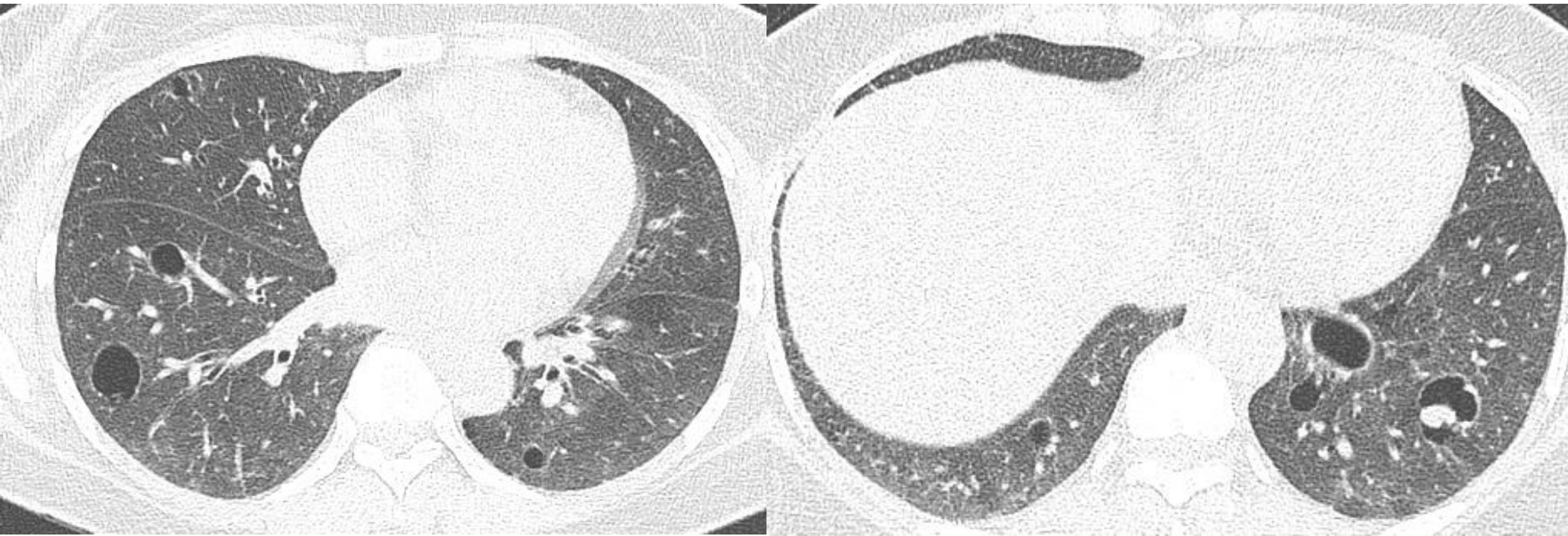


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Lymphocytic Interstitial Pneumonia (LIP) – with interstitial infiltration

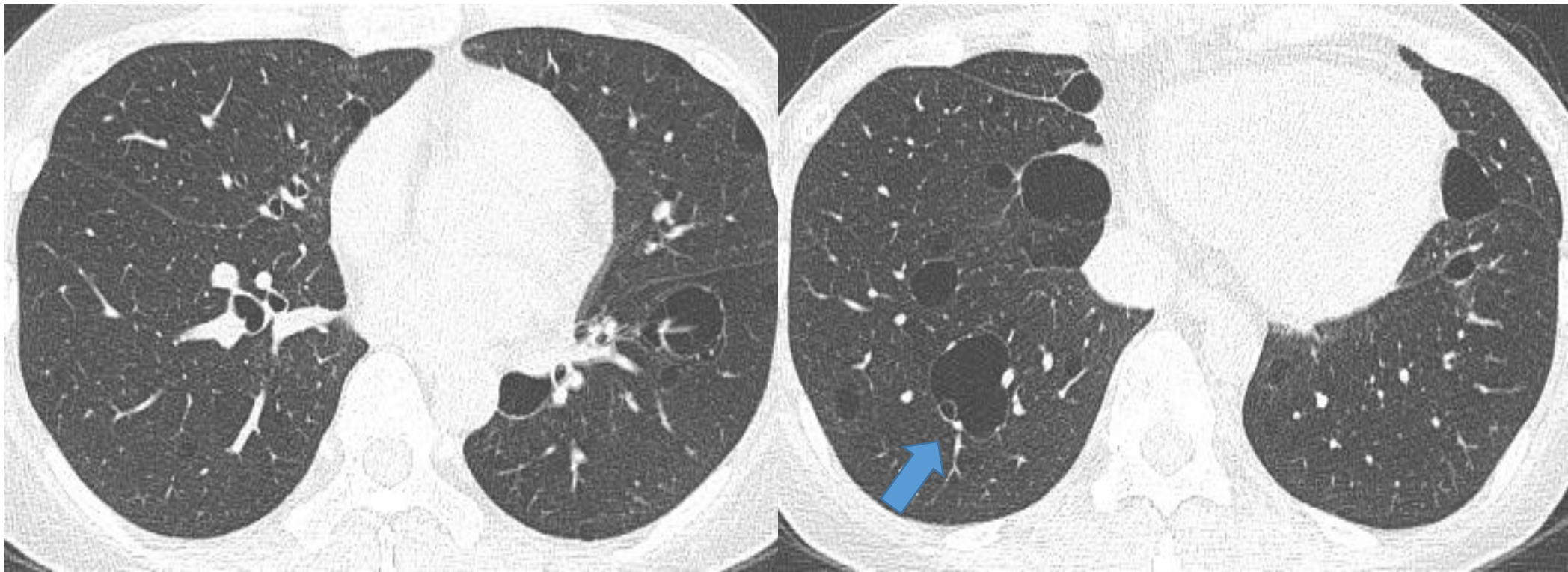


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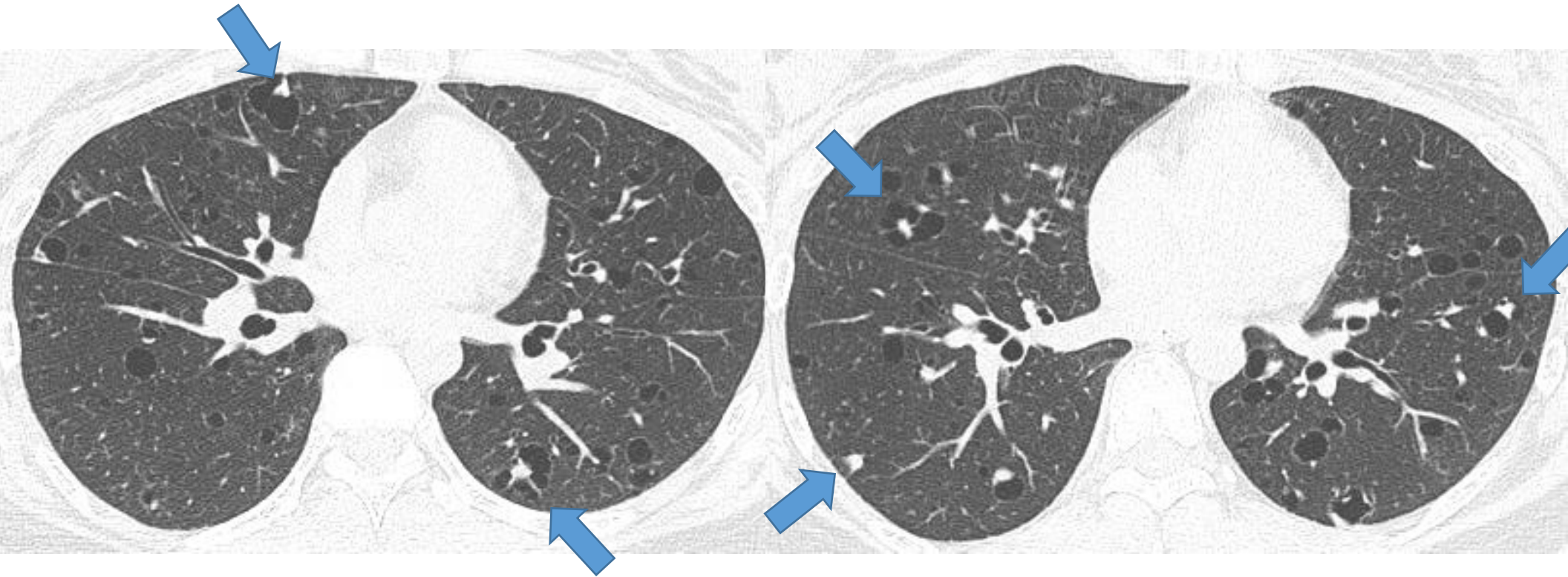


Lymphocytic Interstitial Pneumonia (LIP)

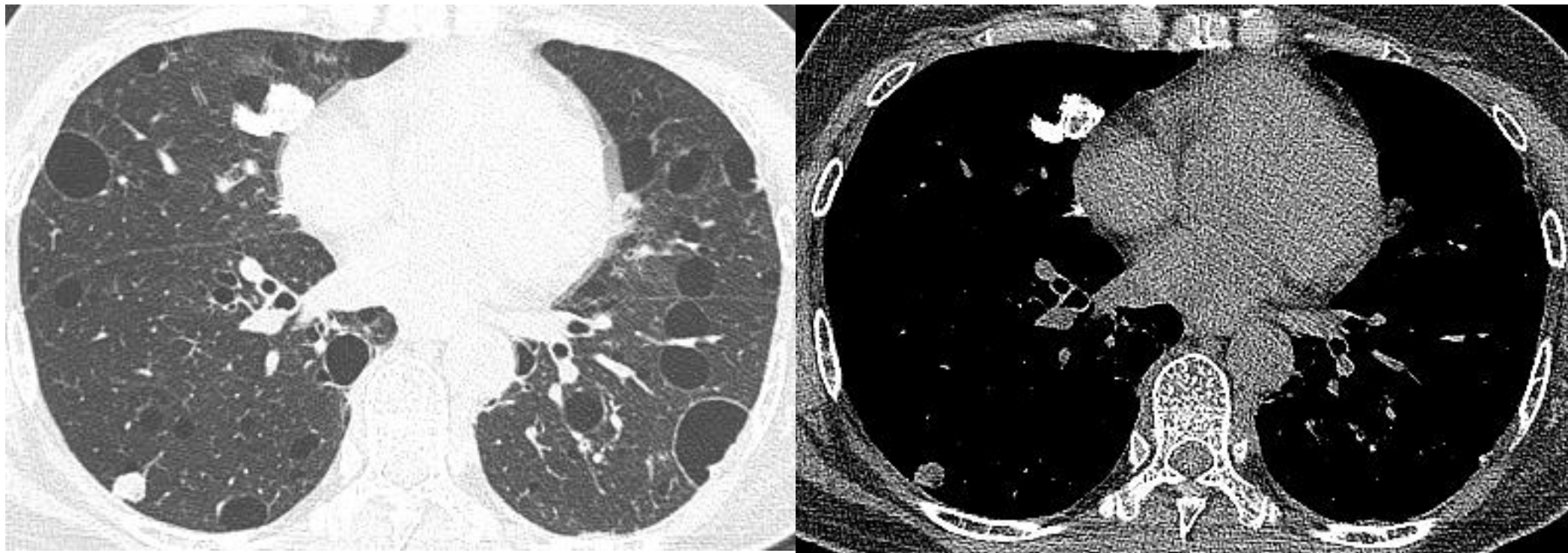
- Only cysts



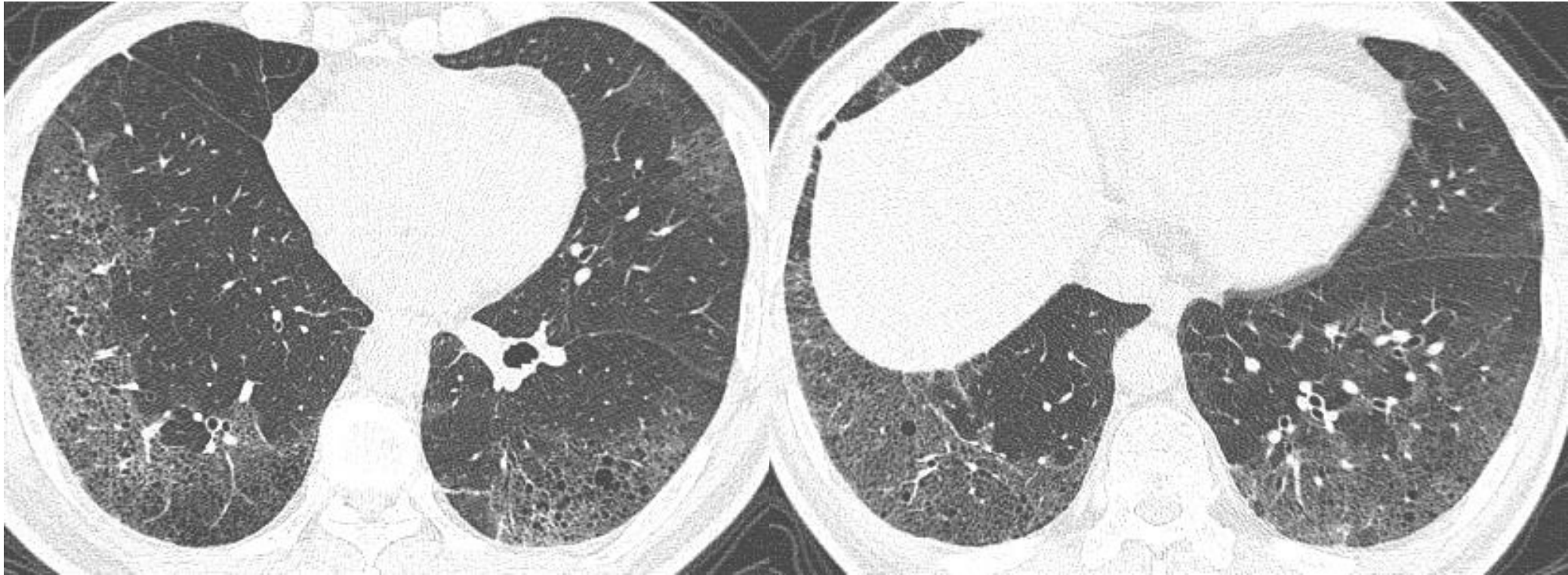
Lymphocytic Interstitial Pneumonia (LIP) – with amyloidosis and MALT lymphoma



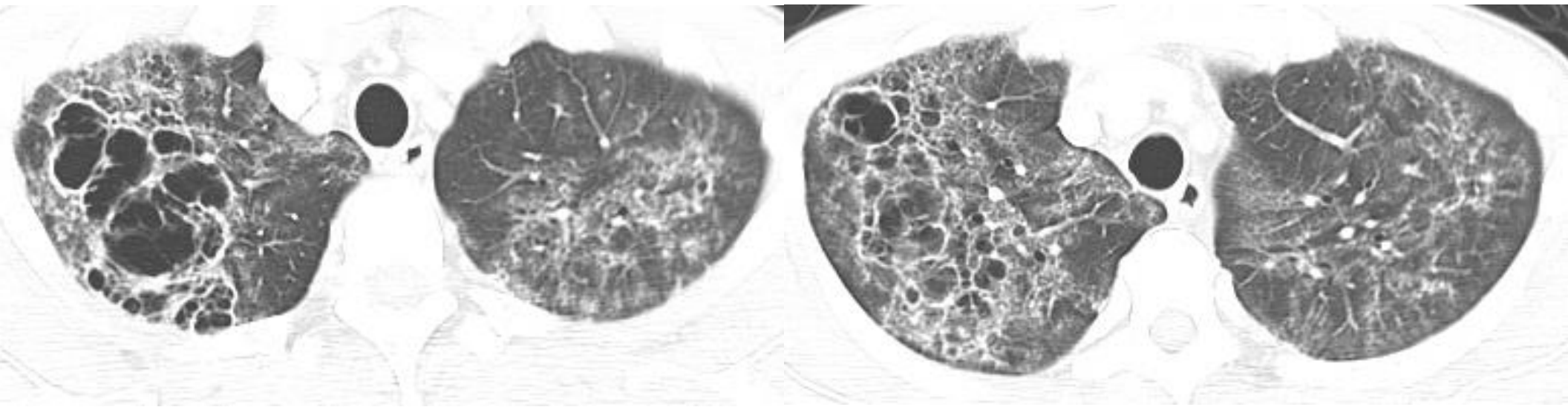
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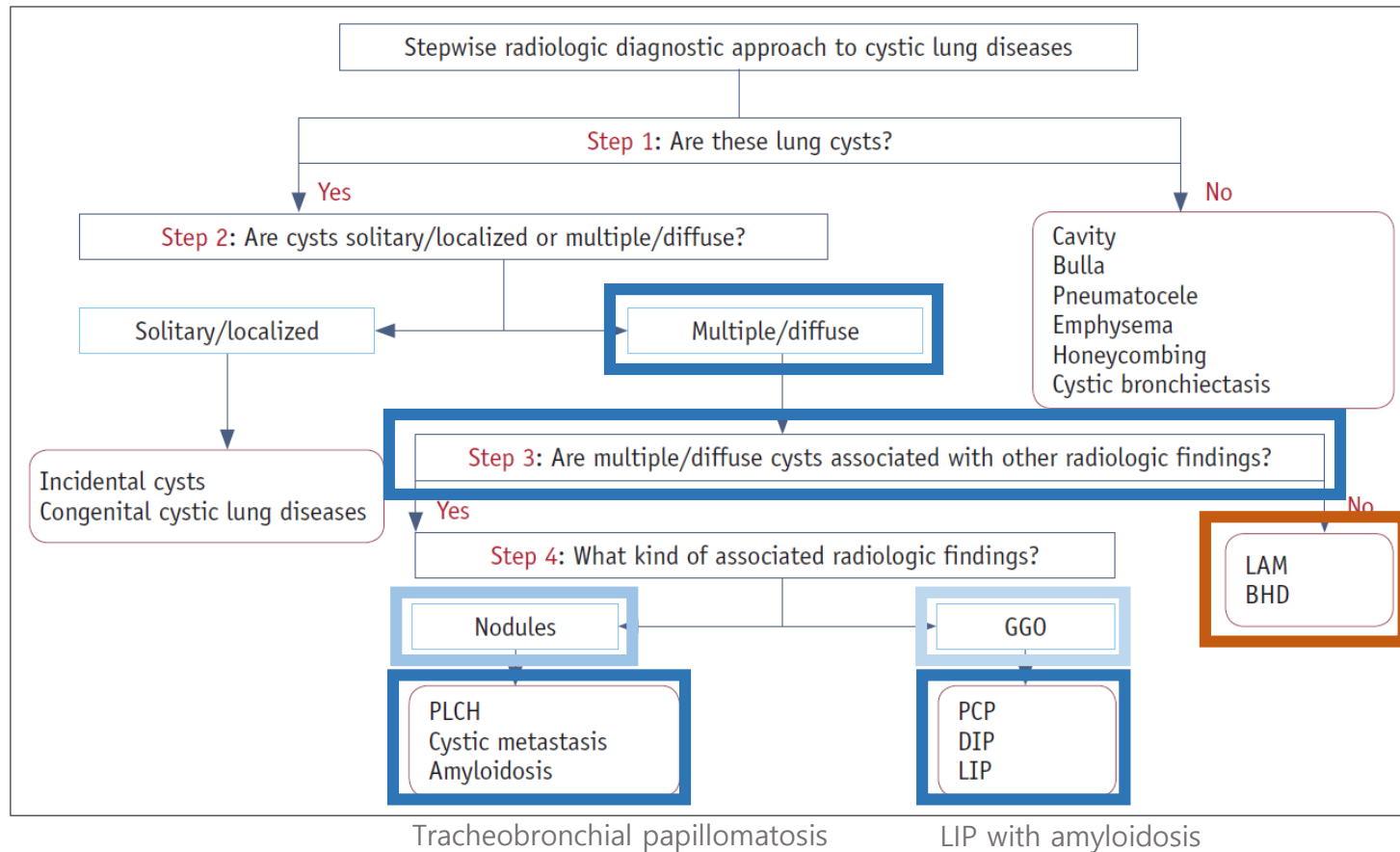
Desquamative Interstitial Pneumonia (DIP)



Pneumocystis Jirovecii Pneumonia (PCP)



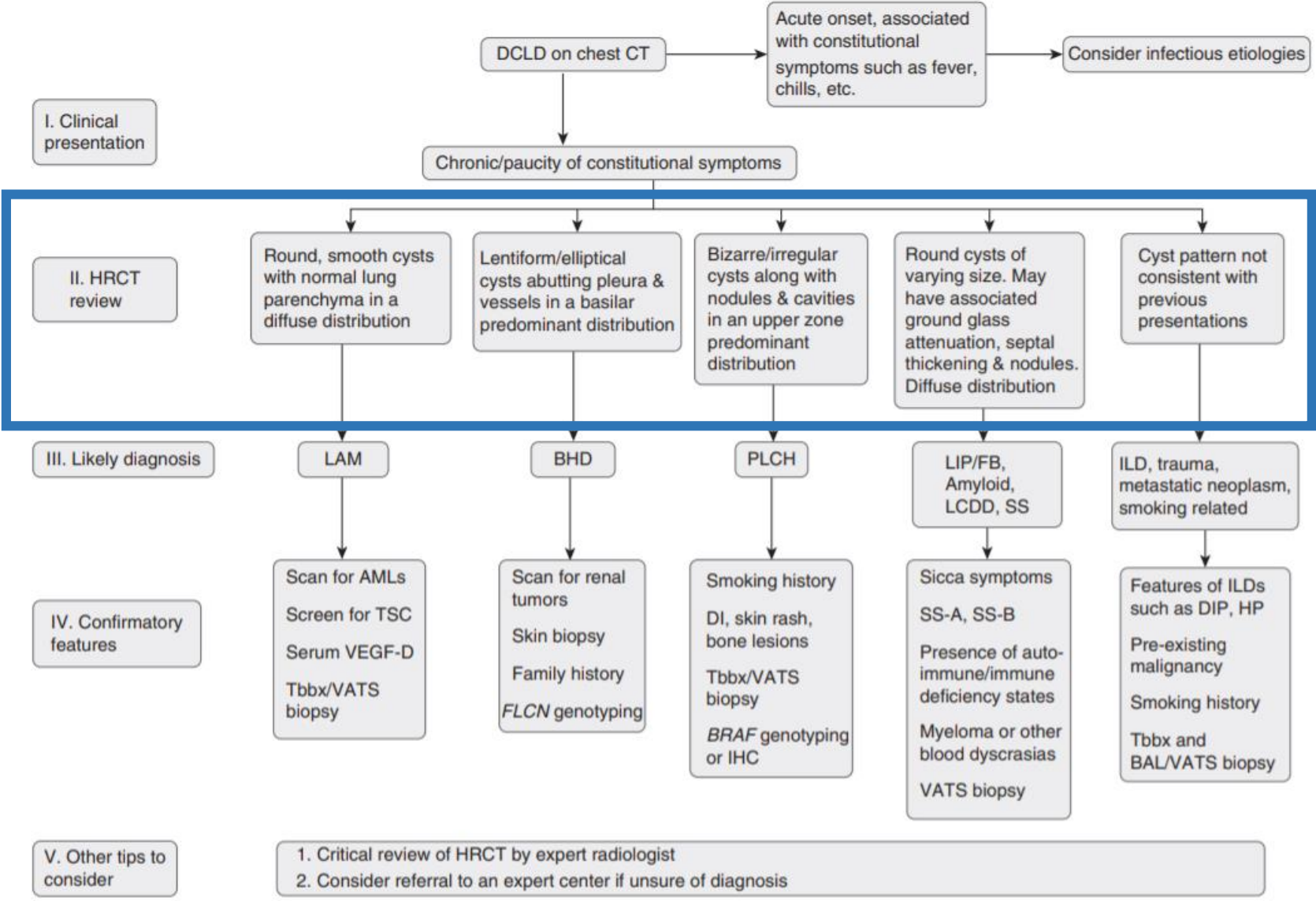
Radiologic Diagnostic Approach to Cystic Lung Diseases



CONCISE C

Diffuse Cystic L
Part II

Nishant Gupta^{1,2}, Robert V





ILD MDD Group in Asan Medical Center

Thank You for Your Attention!