Collagen Vascular Diseases: Thoracic Manifestations

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Goals

- Recognize common pulmonary manifestations of collagen vascular disease on CT.
- Understand the pulmonary complications of common collagen vascular diseases.
- List the most common pulmonary manifestations of specific collagen vascular diseases.

Collagen vascular diseases

- Autoimmune disorders characterized by the presence of autoantibodies
- Damage to connective tissues throughout the body
- Lung disease common: 15% of ILD is CVD related

Collagen vascular diseases

- Often asymptomatic
- ILD may be first manifestation of CVD
  - Some postulate that many cases of idiopathic ILD may be related to CVD
- CVD history often obviates biopsy
- HRCT imaging gold standard

Collagen vascular diseases

- Rheumatoid arthritis
- Progressive systemic sclerosis/scleroderma
- Dermatomyositis/polymyositis
  - Antisynthetase syndrome
- Systemic lupus erythematosus
- Sjögren syndrome
- Mixed connective tissue disease
- Undifferentiated connective tissue disease/IPAF
- *Will not discuss ANCA-associated vasculitis*

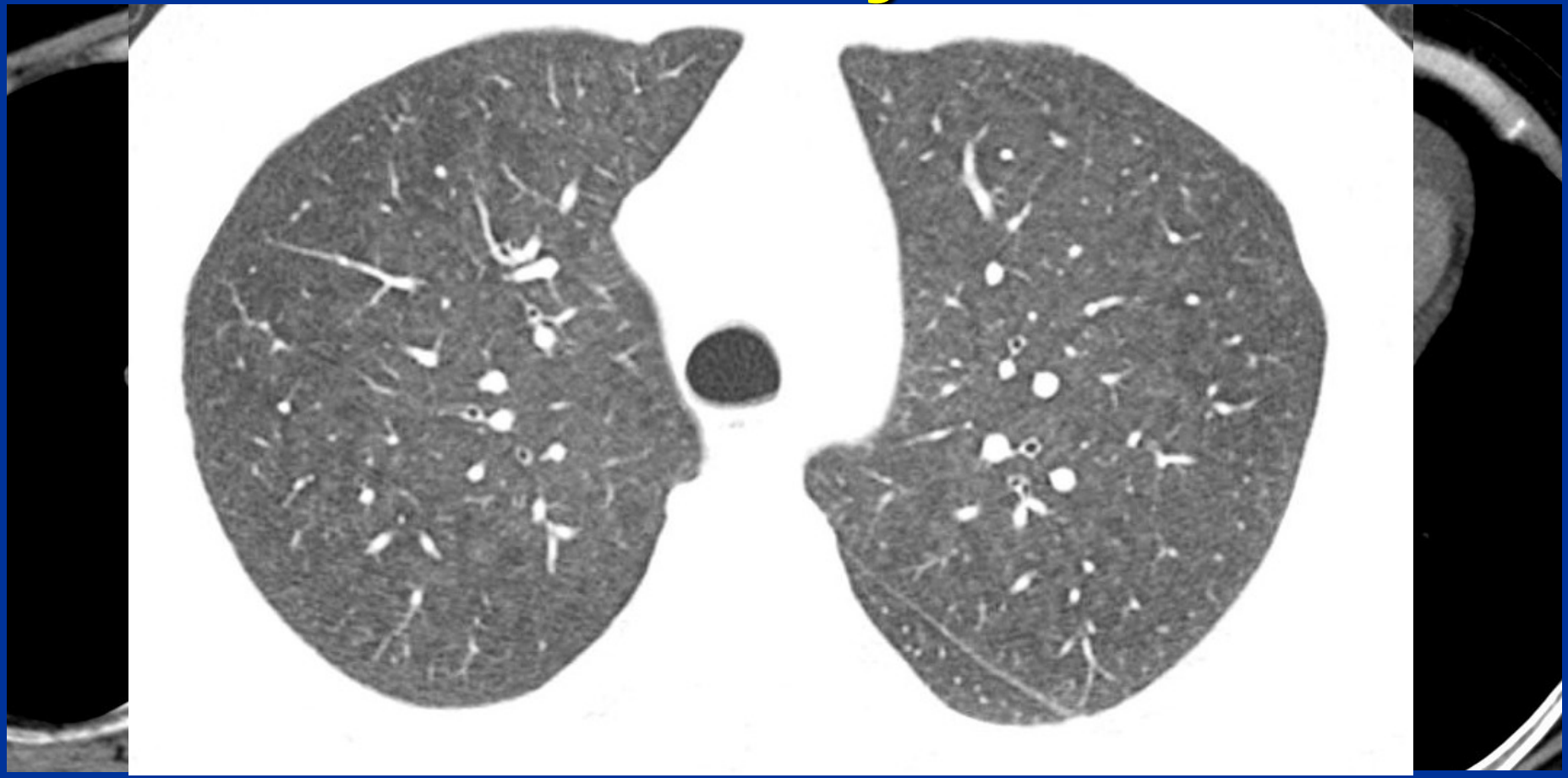
Autoantibodies

Rheumatoid arthritis	Rheumatoid factor, anti-CCP
Progressive systemic sclerosis	Anti-centromere antibody (limited PSS) Anti-SCL-70
Mixed connective tissue disease	Anti-ribonuclear protein
Dermatomyositis/polymyositis	Anti-Jo-1
Systemic lupus erythematosis	Anti double stranded (ds) DNA and anti-Sm Anti-nuclear factor (less specific) Anti-phospholipid antibodies
Sjogren syndrome	Anti-SS-A (Ro) Anti-SS-B (La)

Thoracic manifestations overview

- Pulmonary disease: IIP patterns
  - UIP,NSIP, OP, AIP, LIP
- Pleural disease
  - Effusions, thickening
- Airways
  - Bronchiectasis, OB, follicular bronchiolitis
- Vascular
  - Pulmonary HTN: Idiopathic, PVOD, PCH

Pulmonary HTN



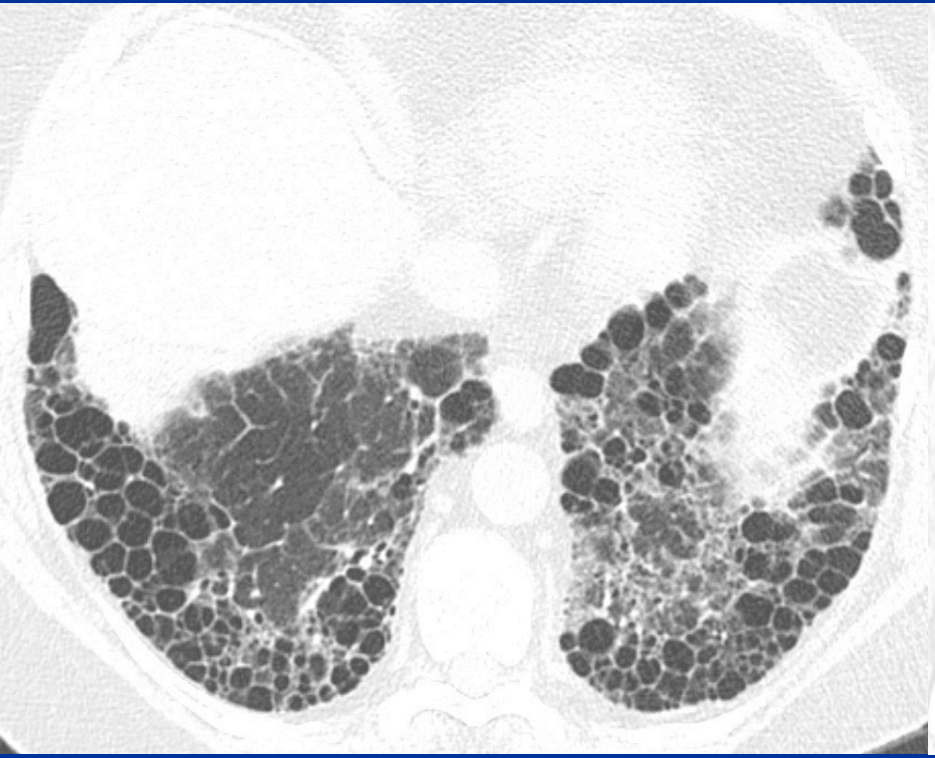
Rheumatoid arthritis

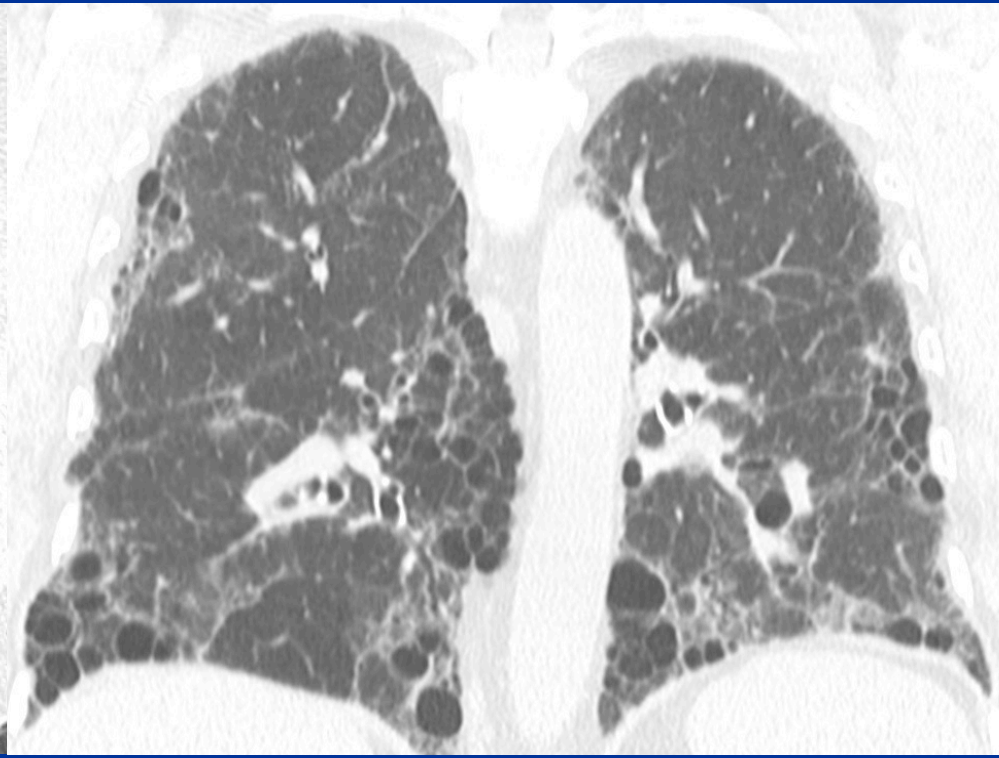
- Interstitial lung disease
  - UIP; OP or NSIP
- Airways disease
  - OB, bronchiectasis; follicular bronchiolitis
- Pleural effusion/thickening
- Pulmonary HTN
- Necrobiotic nodules

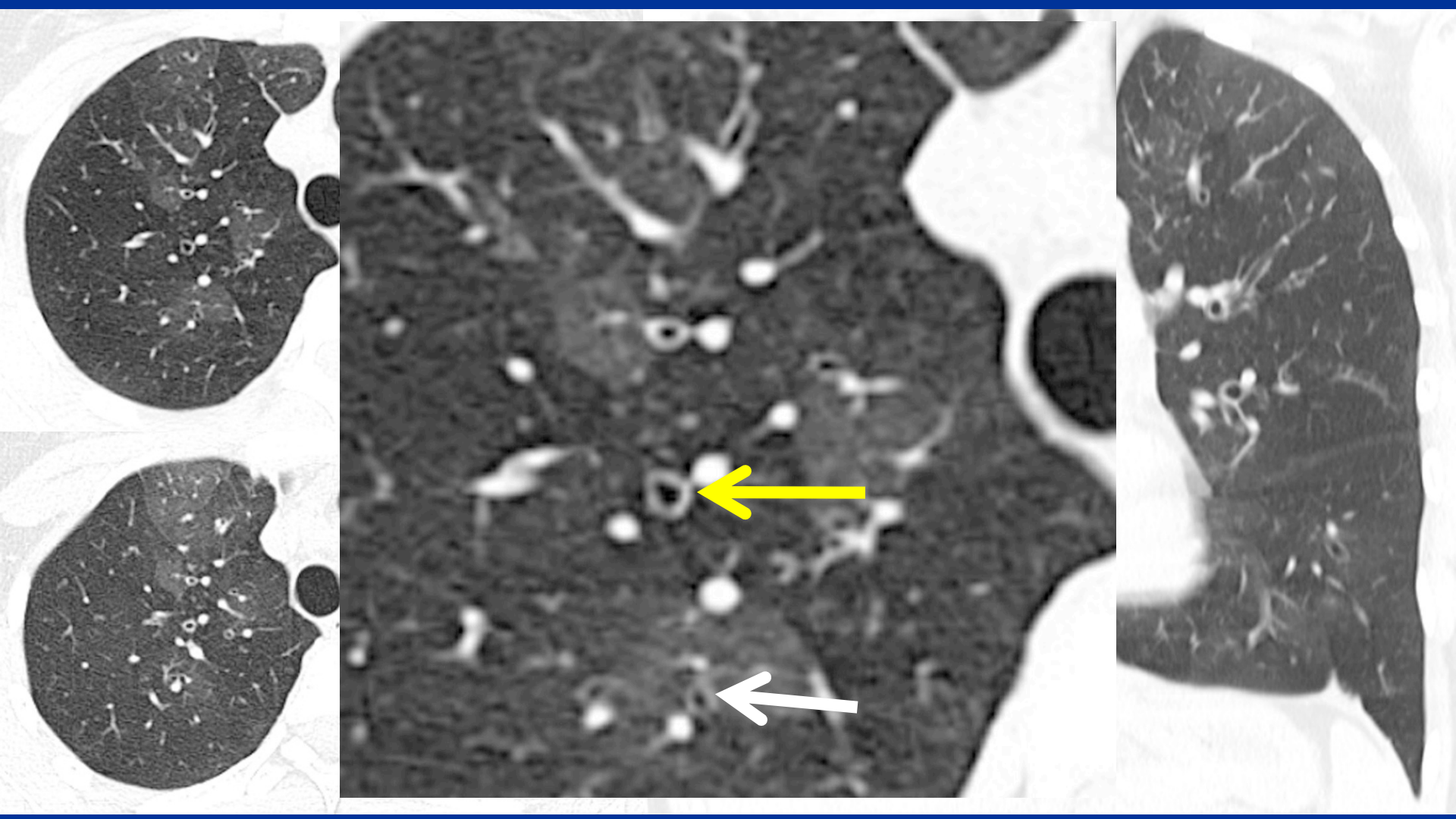
Rheumatoid arthritis

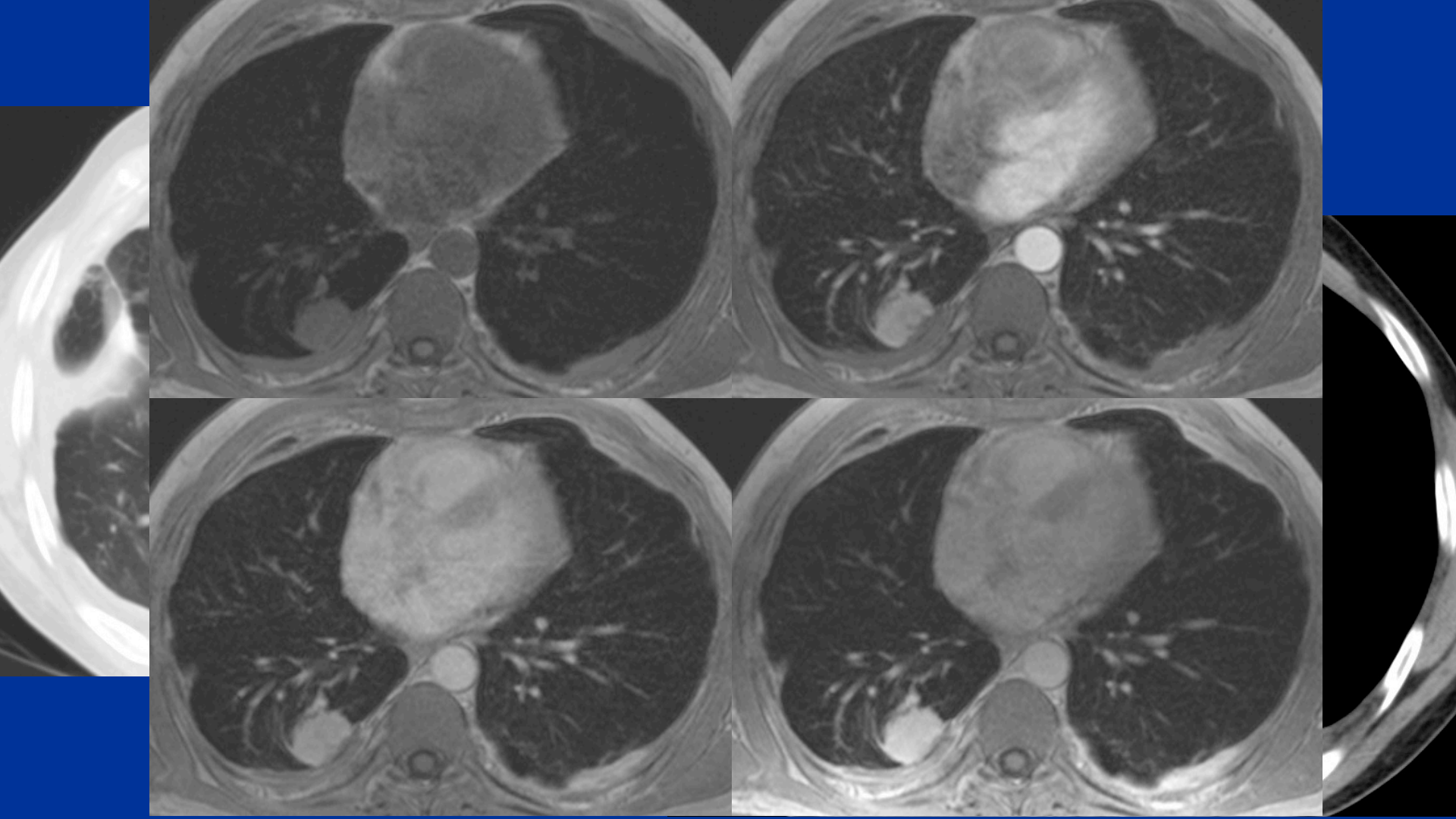
- Interstitial lung disease
  - 50-60% UIP
  - 40% NSIP
  - 10% OP

Rheumatoid arthritis

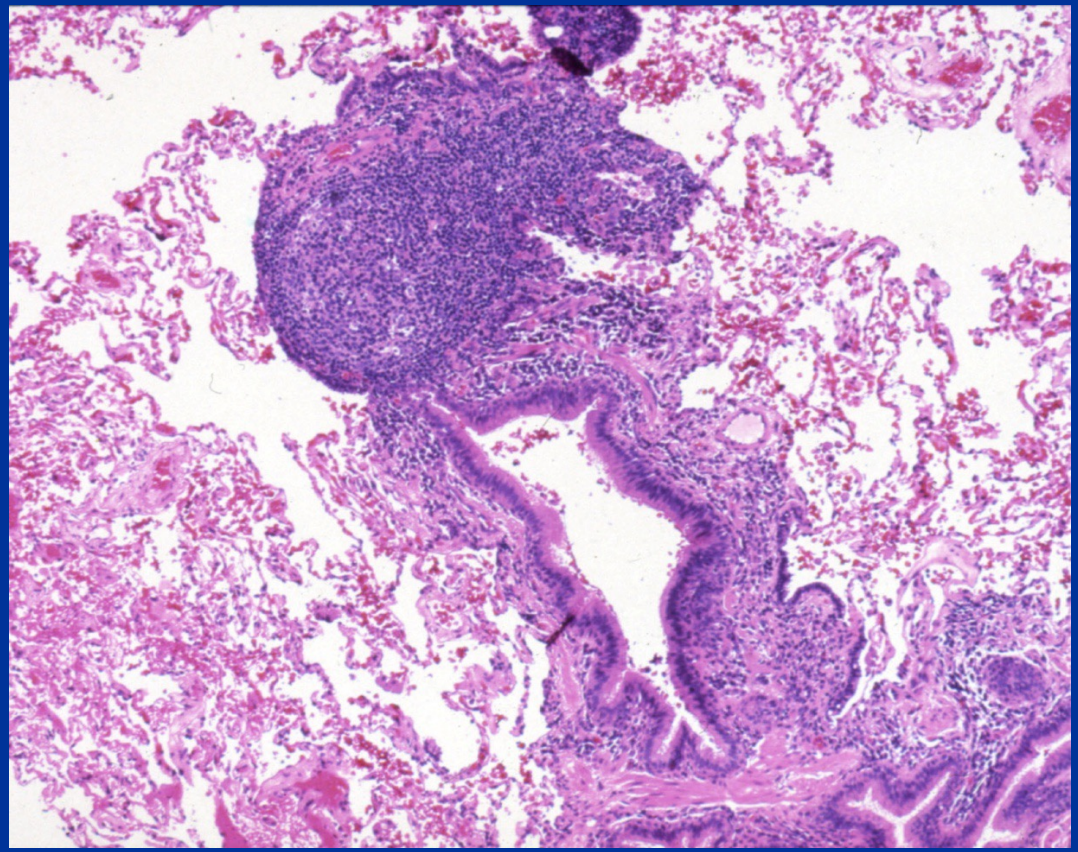


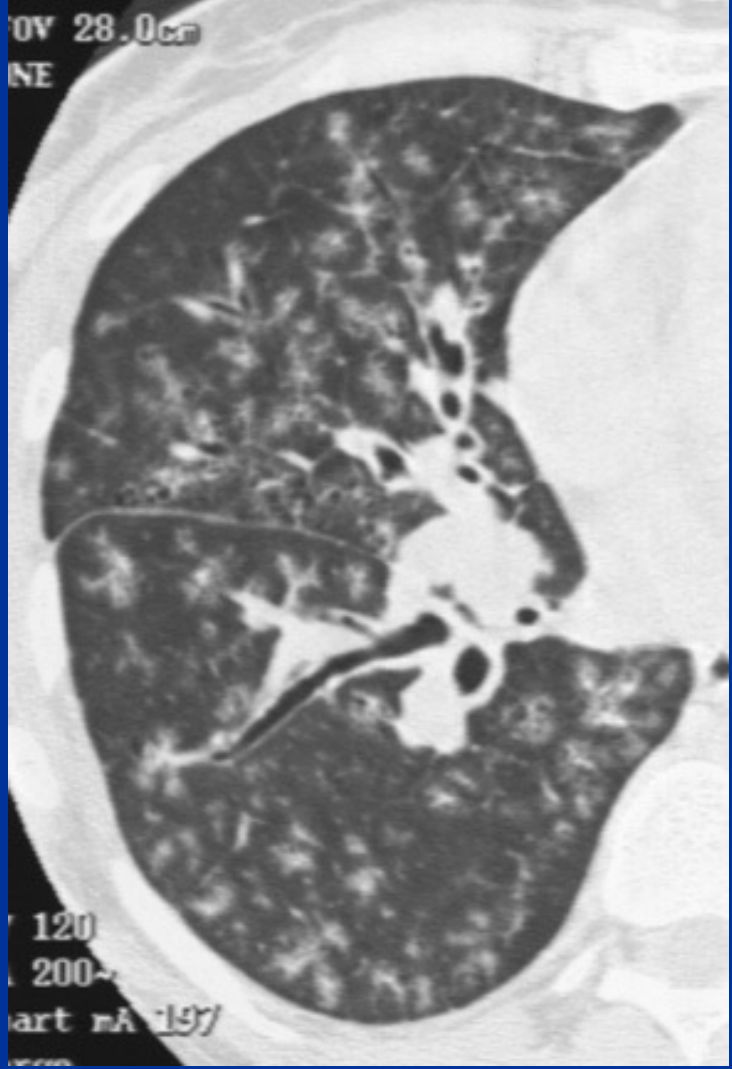






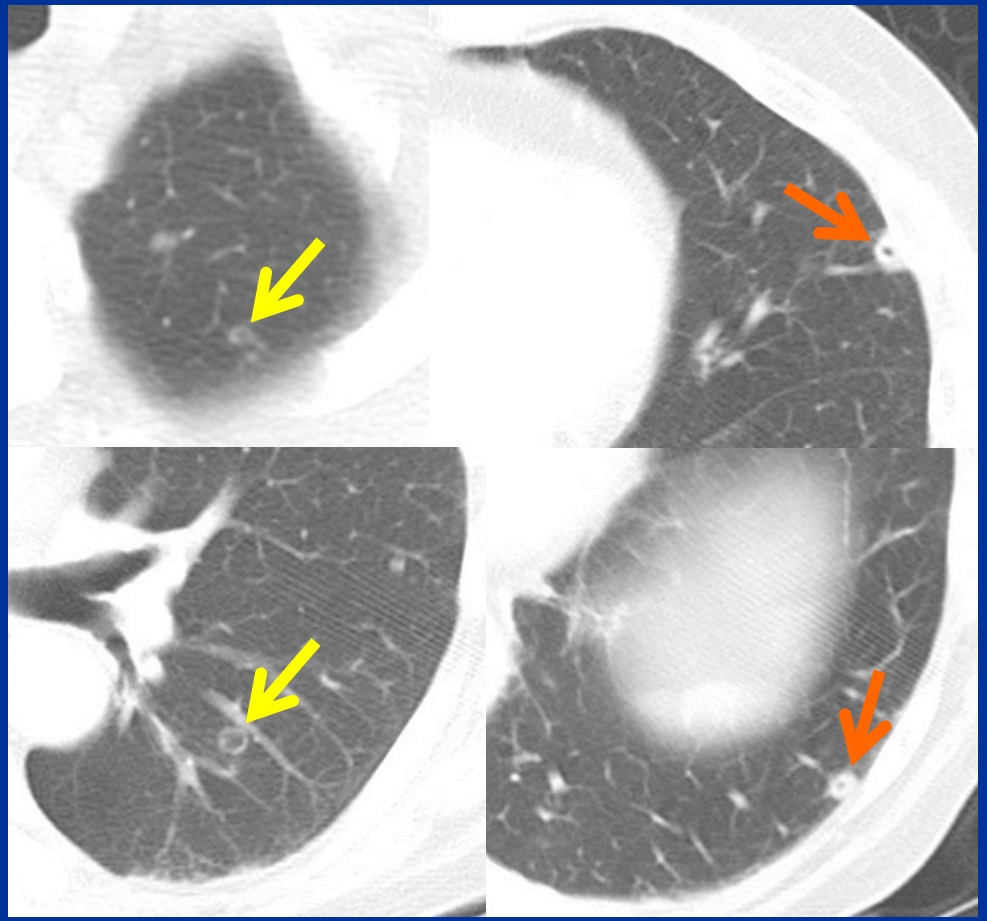
Rheumatoid arthritis





Rheumatoid arthritis nodules

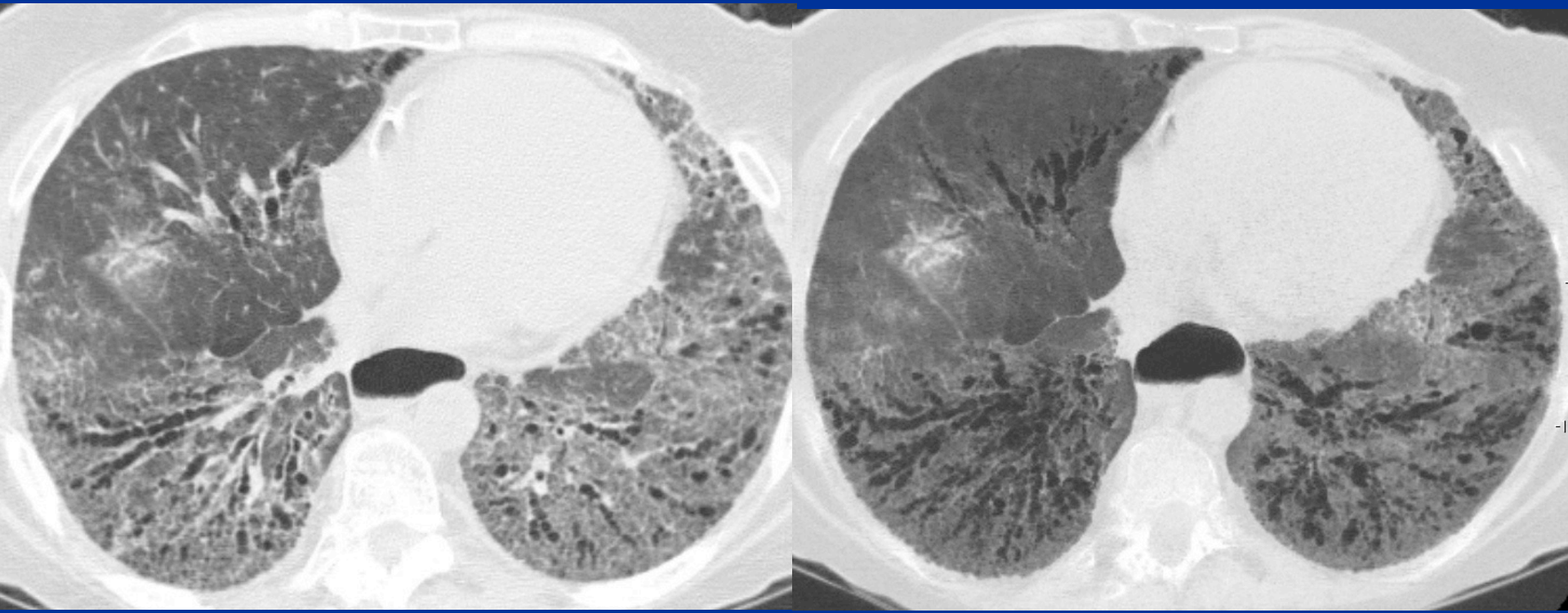
- Often subpleural
- 50% cavitate
- Patholgically identical to skin nodules (necrobiotic)
  - 80% coexistent
- 2x more in men

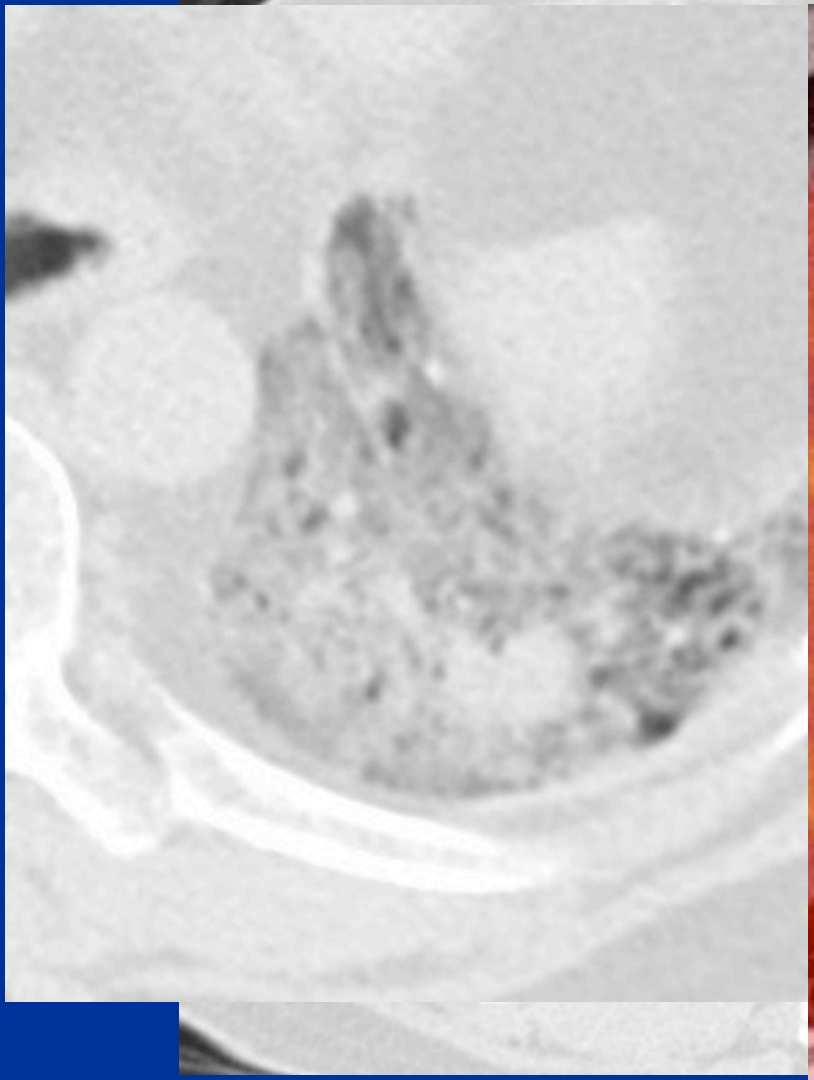


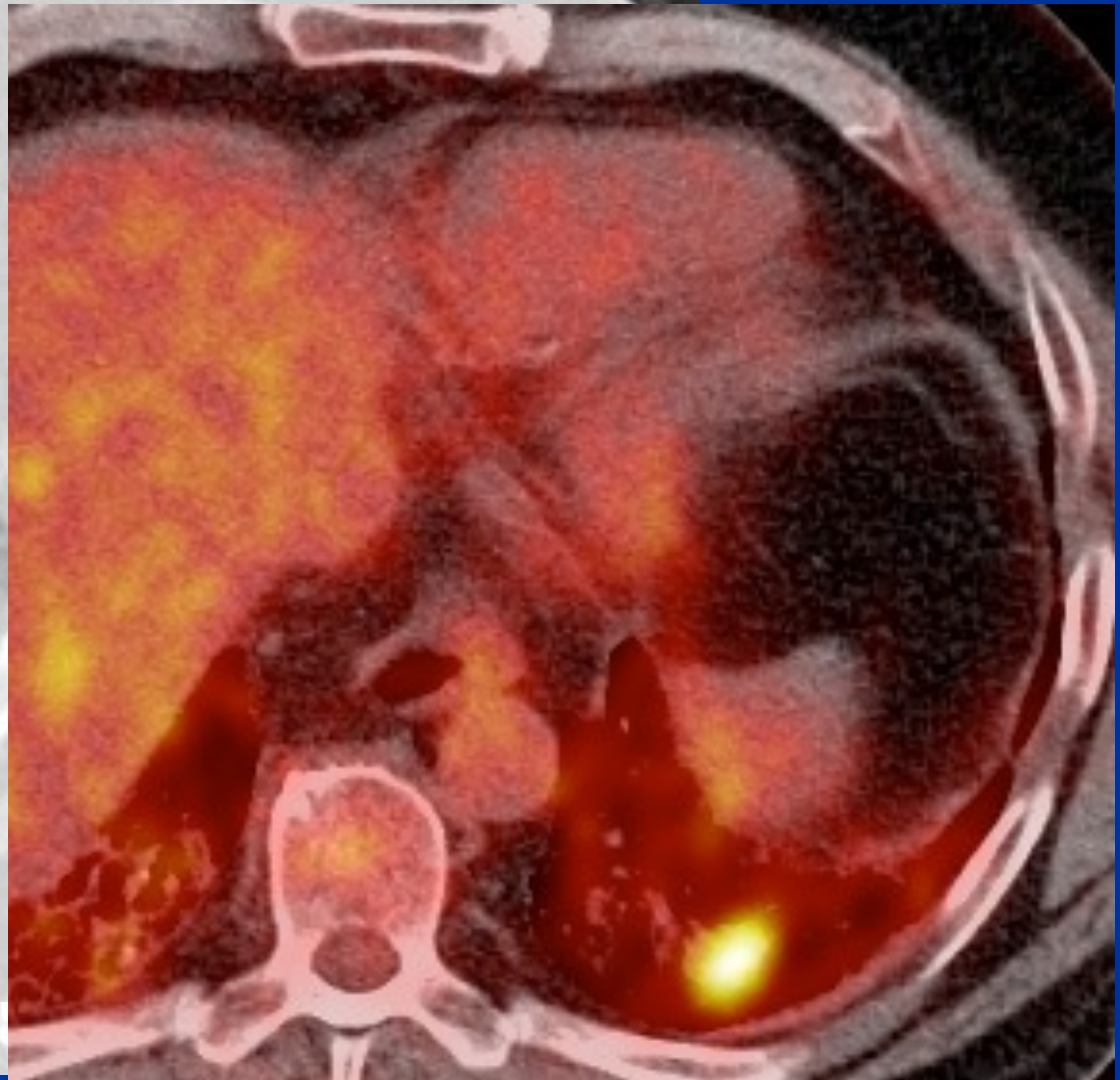
Scleroderma

- Pulmonary fibrosis common (40-90%)
  - NSIP by far most common
- Esophageal dysmotility
- Pulmonary hypertension
- Lung cancer

Scleroderma







Lung Cancer in Chronic Interstitial Pneumonia: Early Manifestation From Serial CT Observations

OBJECTIVE. The purpose of this study was to use serial CT observations to characterize early-stage lung cancer in patients with chronic interstitial pneumonia.

MATERIALS AND METHODS. We found 23 lung cancers in 22 patients during routine follow-up of chronic interstitial pneumonia between 1999 and 2010. Patients with lung cancer found at initial CT were excluded. Two radiologists independently reviewed serial CT scans, determined the earliest scan showing lung cancer, and evaluated the tumor shape, size, density, and location. Delay in diagnosis was measured from the time of the earliest scan showing lung cancer and the subsequent clinical diagnosis.

RESULTS. During the mean follow-up period of 4.1 years, CT scans were obtained eight times on average. The median tumor size at presentation was 11 mm, and at clinical diagnosis was 22 mm. The median delay in diagnosis was 409 days. Fifteen tumors (65.2%) were in t
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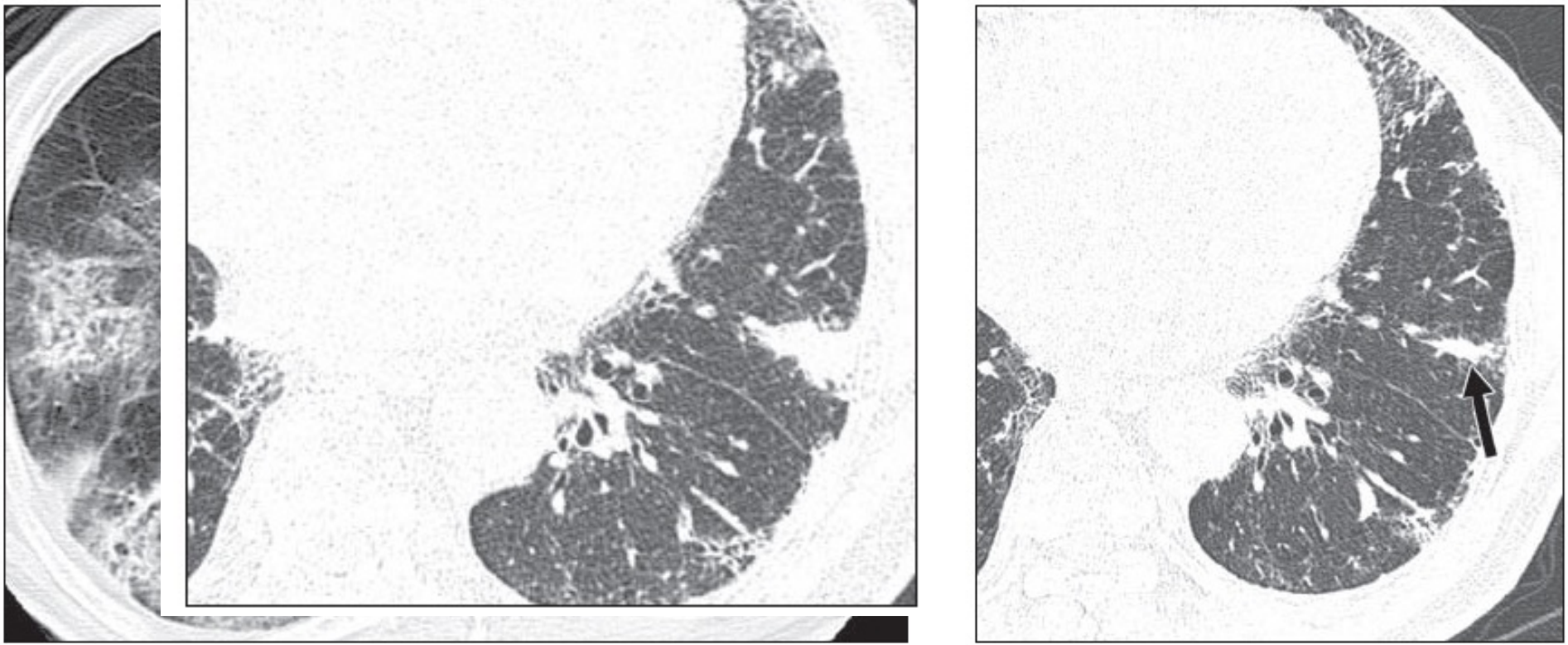
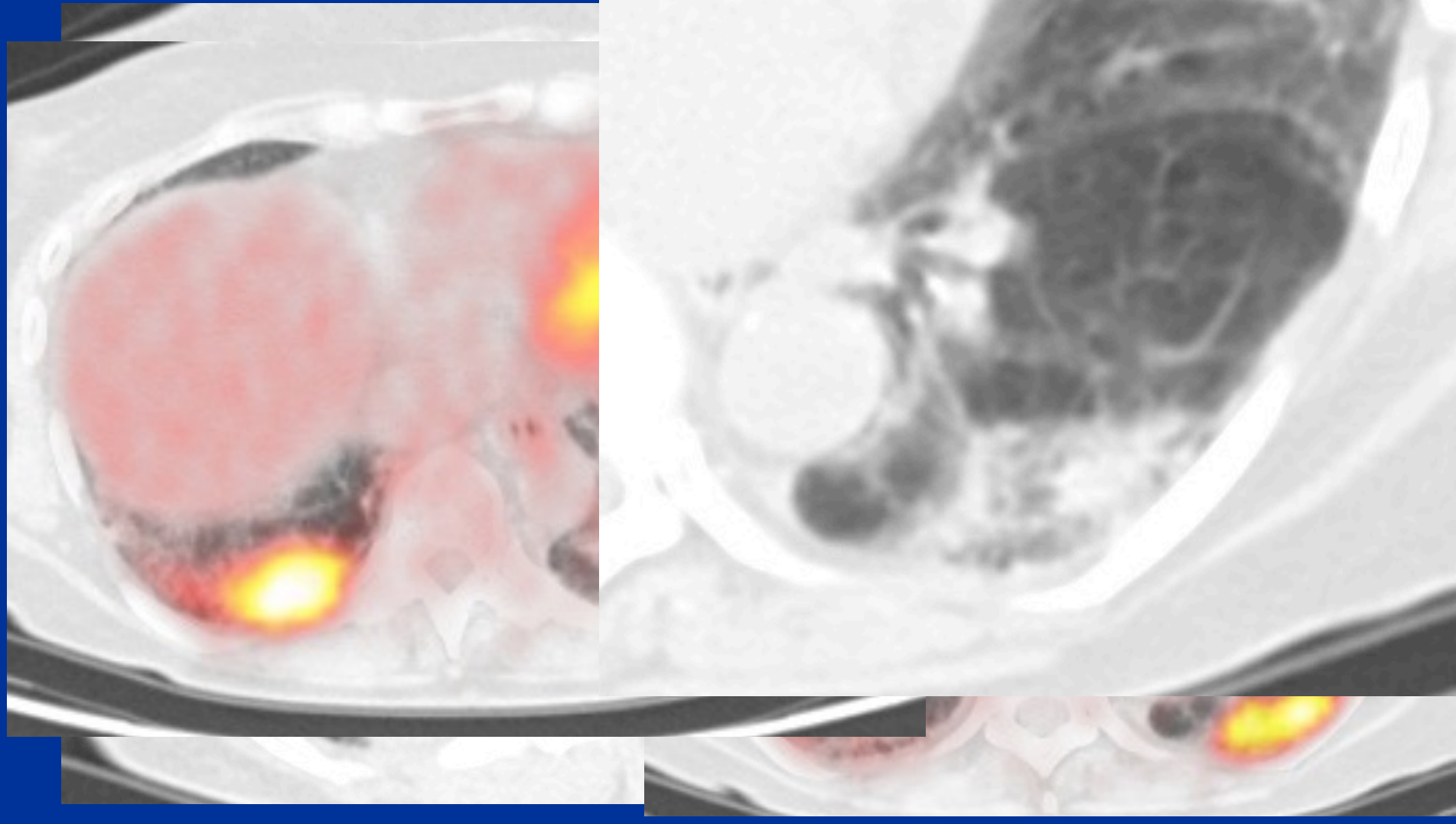
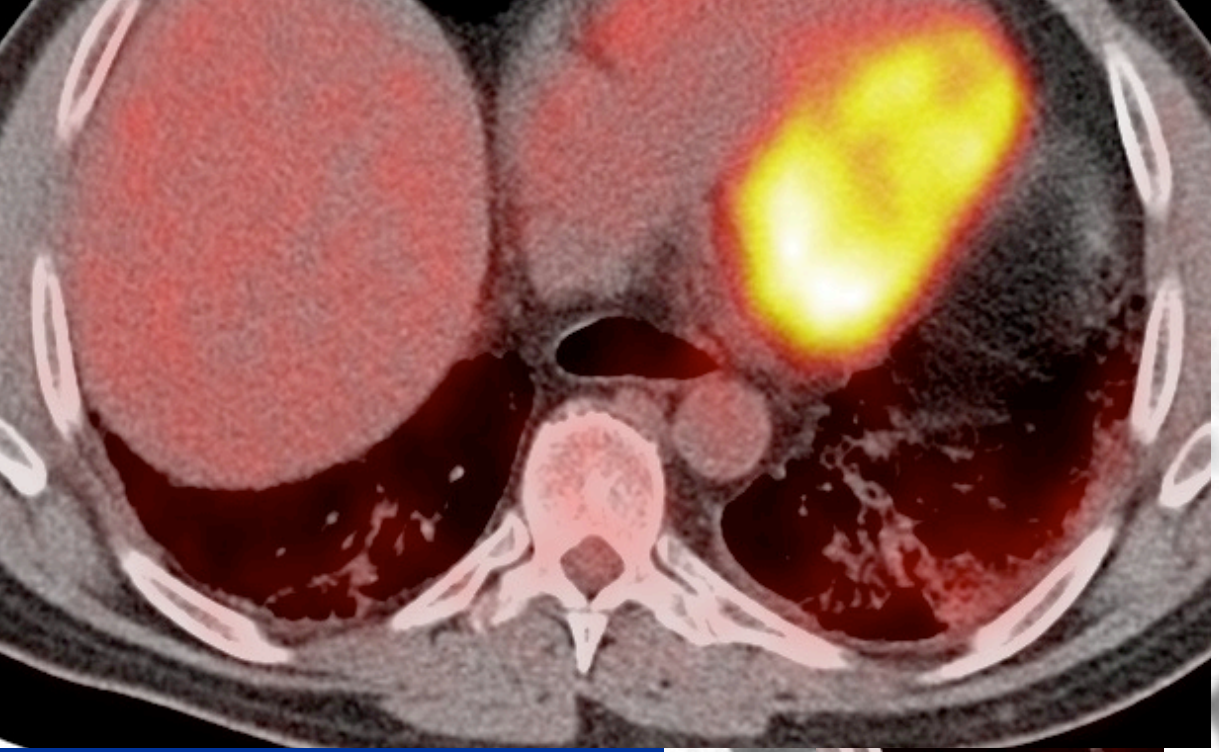
was
ill-defined stellate shape, and two had a bandlike shape. One tumor appeared as an area of ill-defined increased lung attenuation.

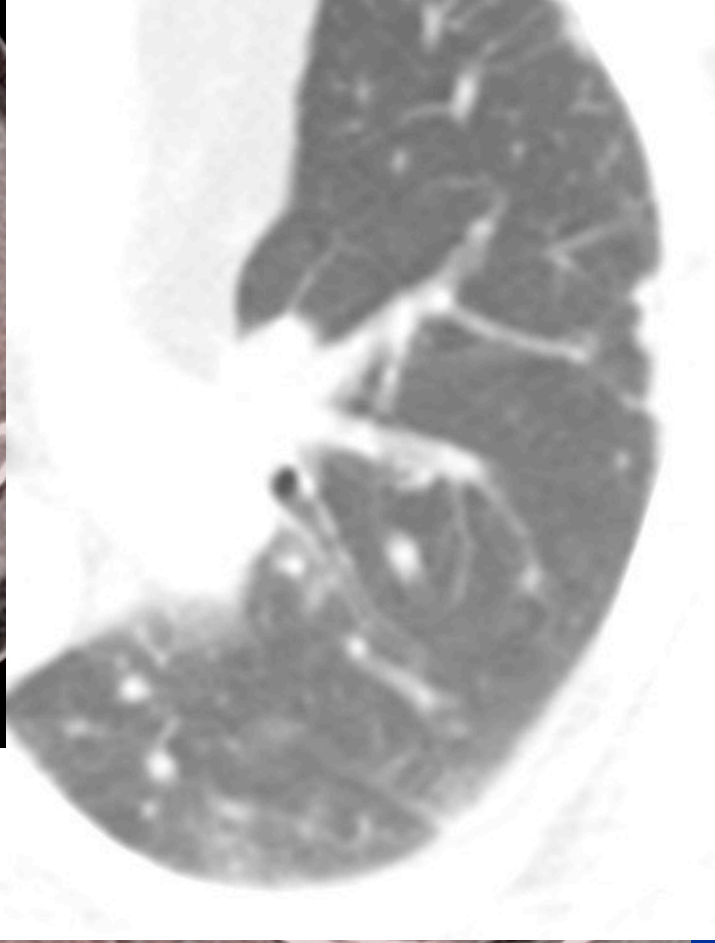
CONCLUSION. Nearly one half of the tumors had a stellate or bandlike shape and were difficult to recognize as tumors initially. Most of the tumors were located at the interface between normal lung and fibrotic cysts; only rarely were tumors located in the midst of honeycomb cysts.

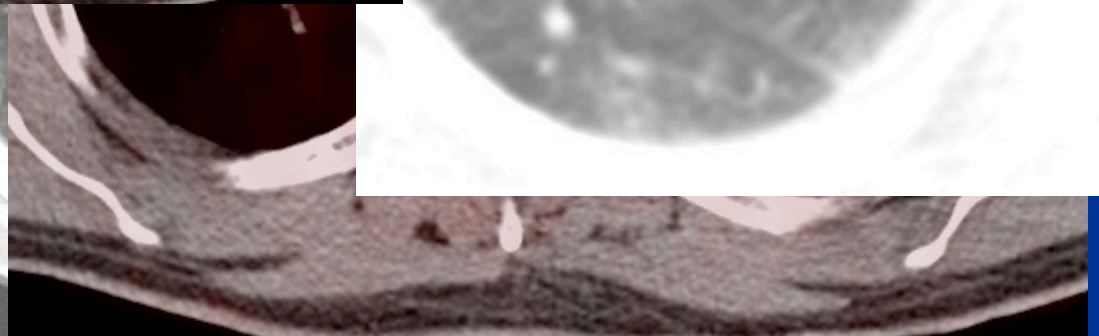
The median delay in diagnosis was 409 days.









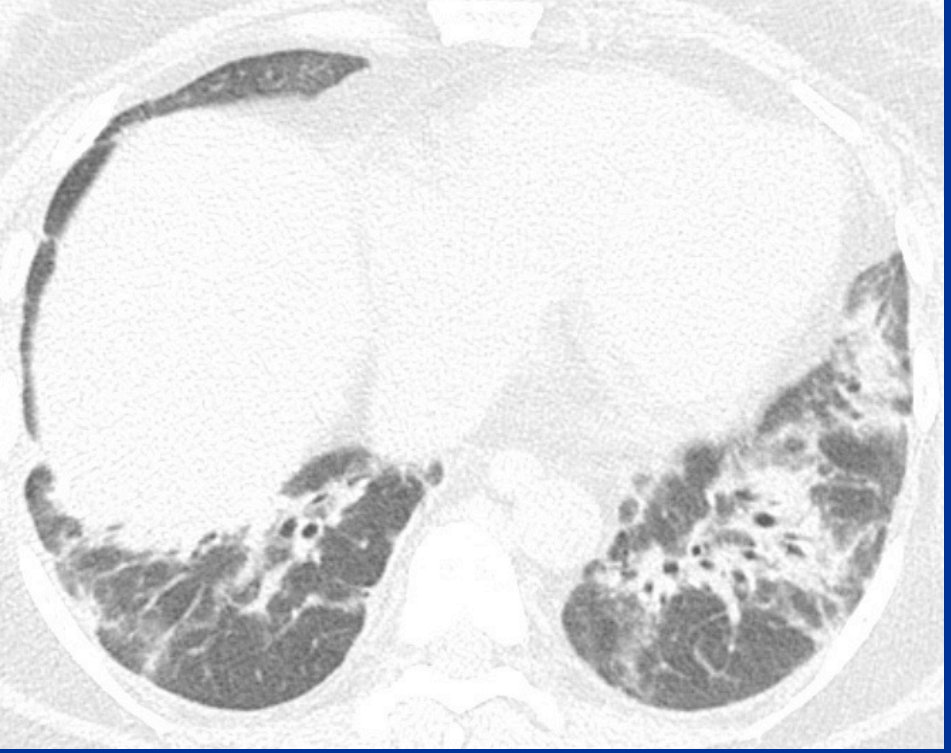


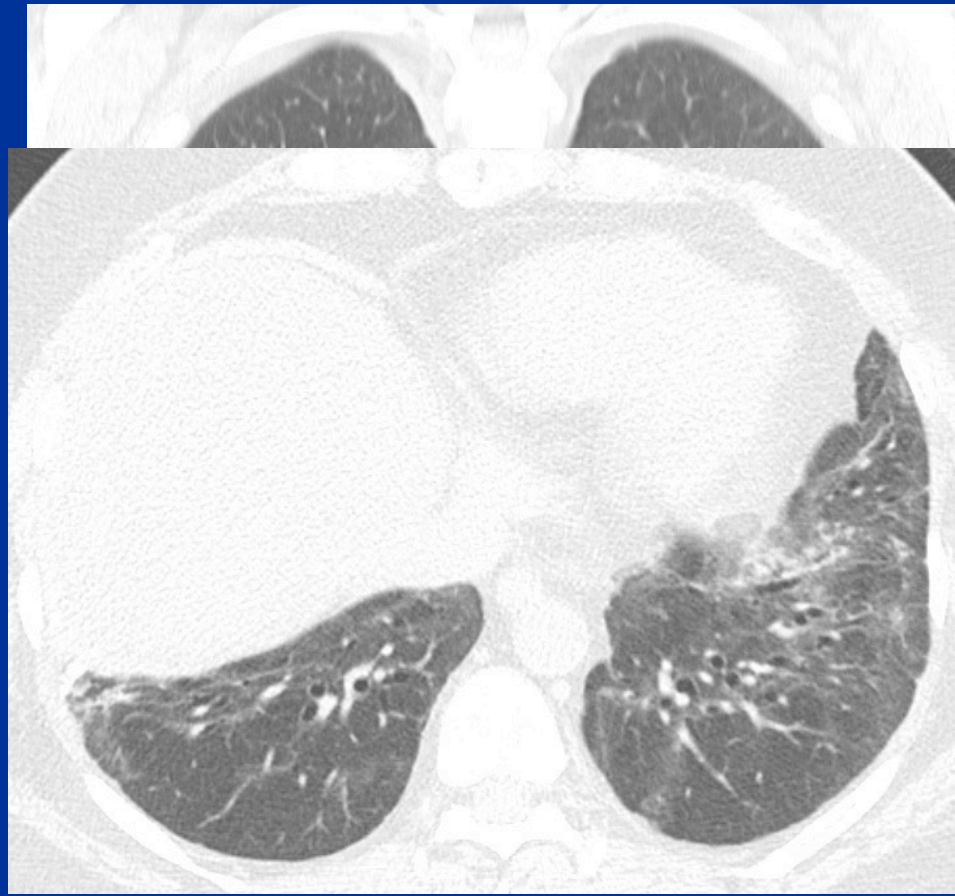
Dermatomyositis and polymyositis

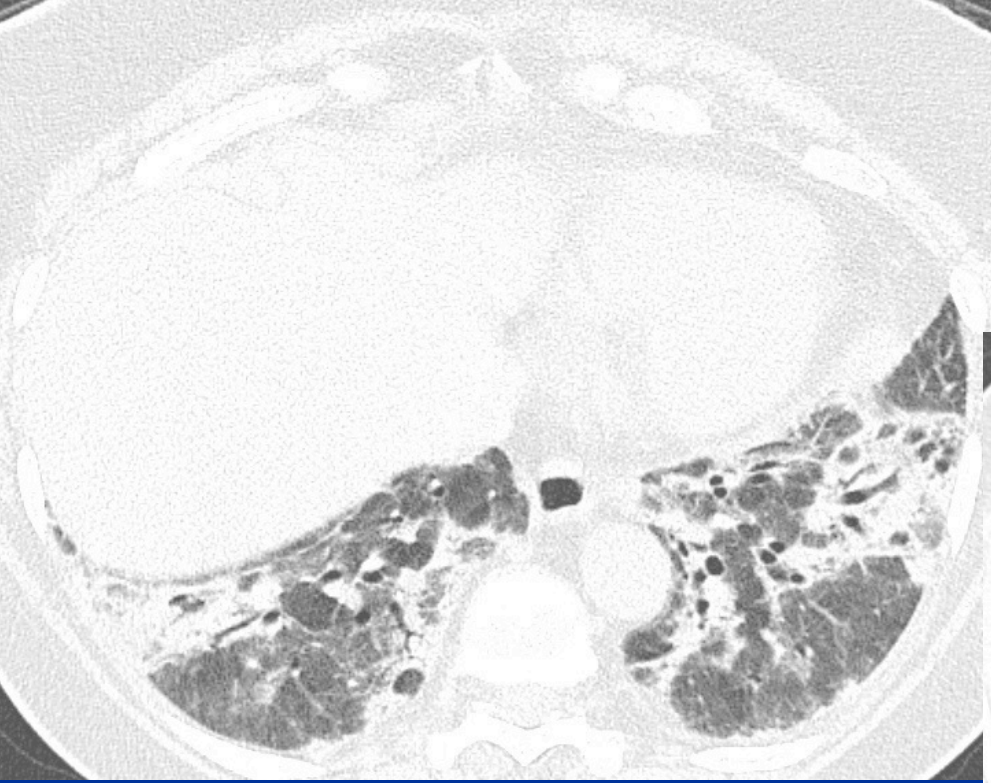
- Inflammatory myopathy
- Interstitial lung disease
  - OP and NSIP characteristic
  - May evolve into NSIP
  - DAD, UIP less common
- Jo-1 antibody specific

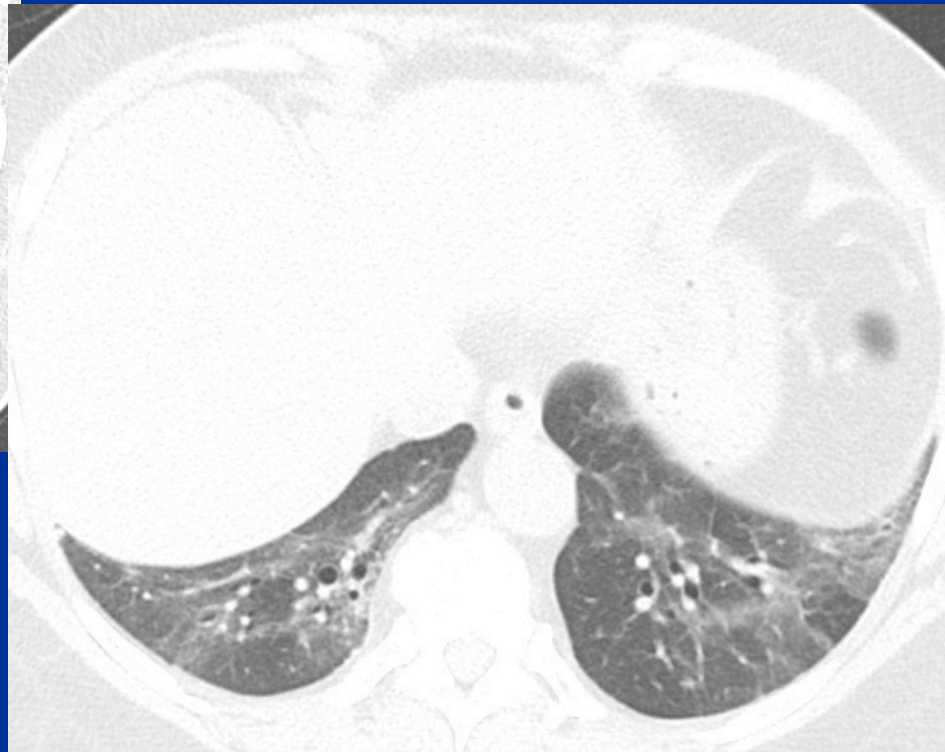
Antisynthetase syndrome

- Antisynthetase syndrome similar to DM/PM
  - Often presents with isolated ILD
  - ILD is almost always basal predominant, OP and/or NSIP patterns
- Jo-1 antibody associated with ILD





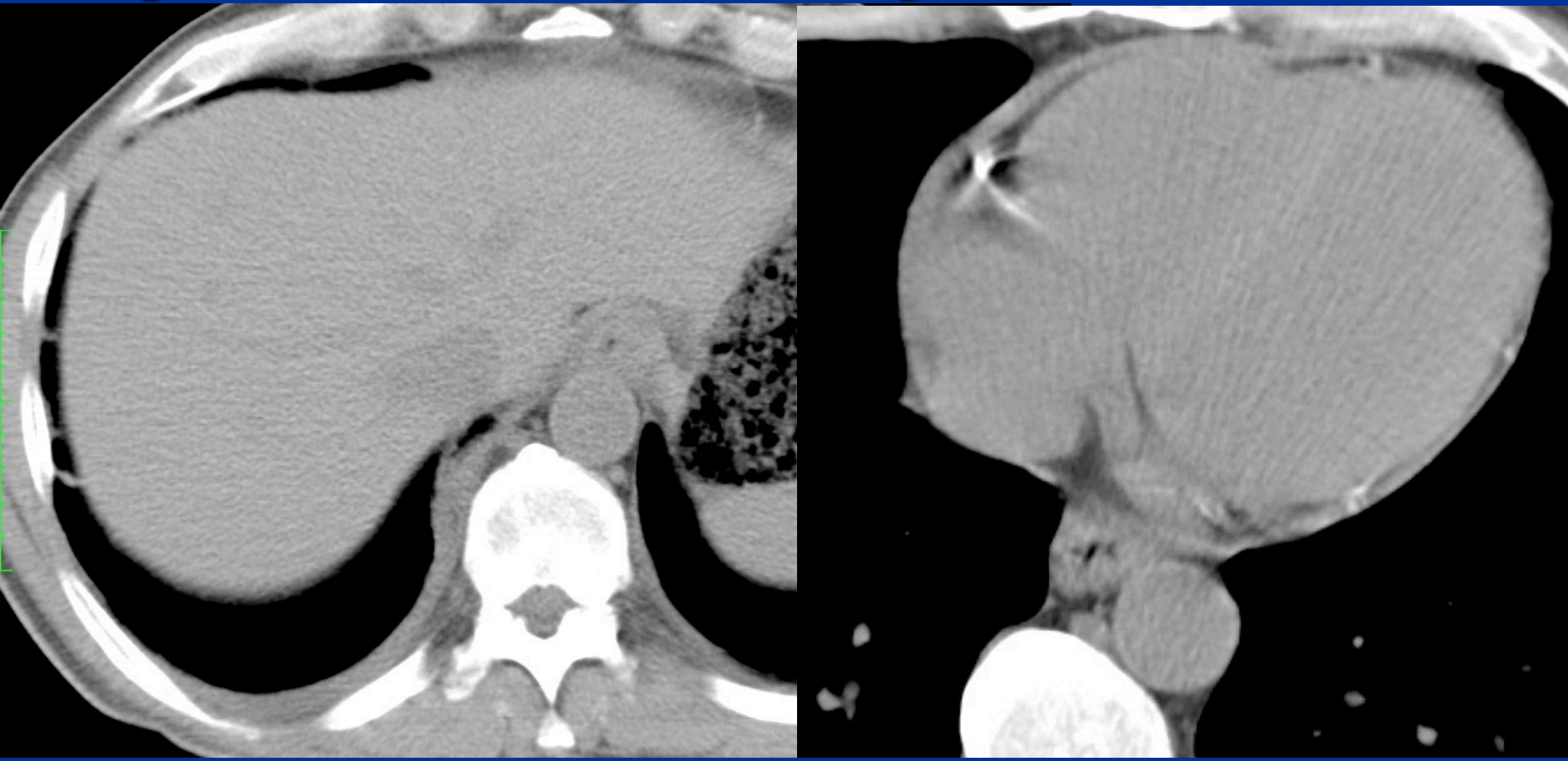




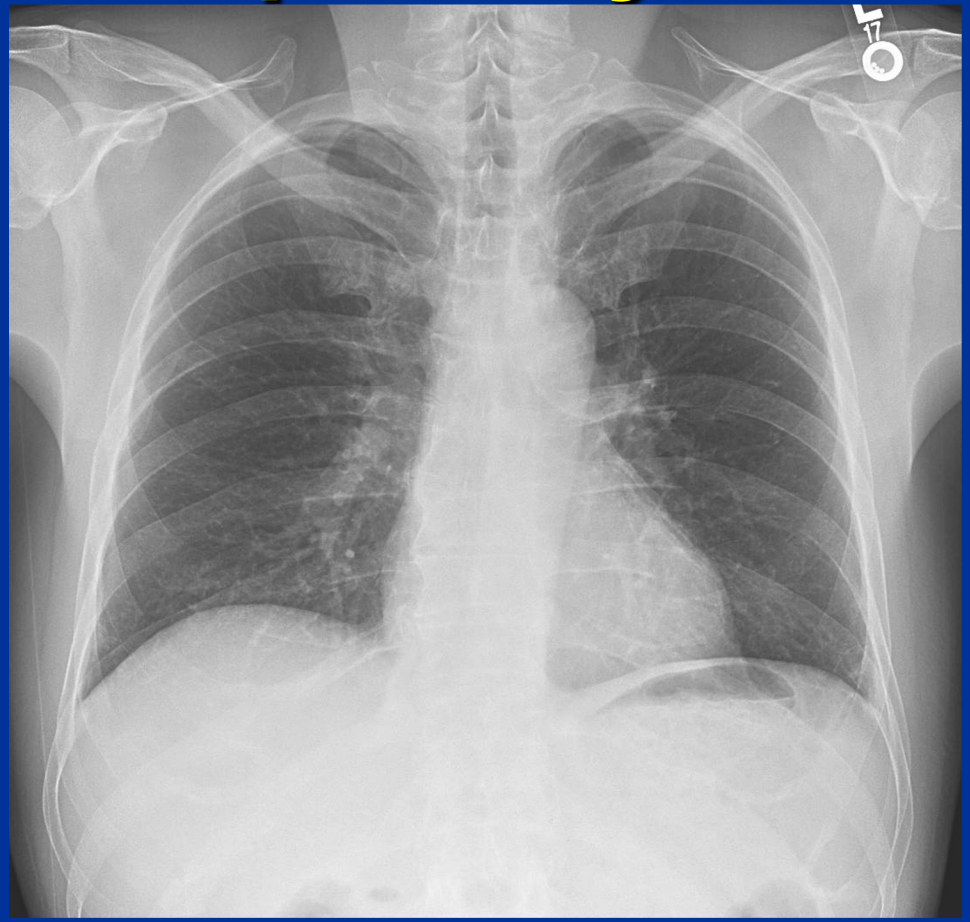
Systemic lupus erythematosus

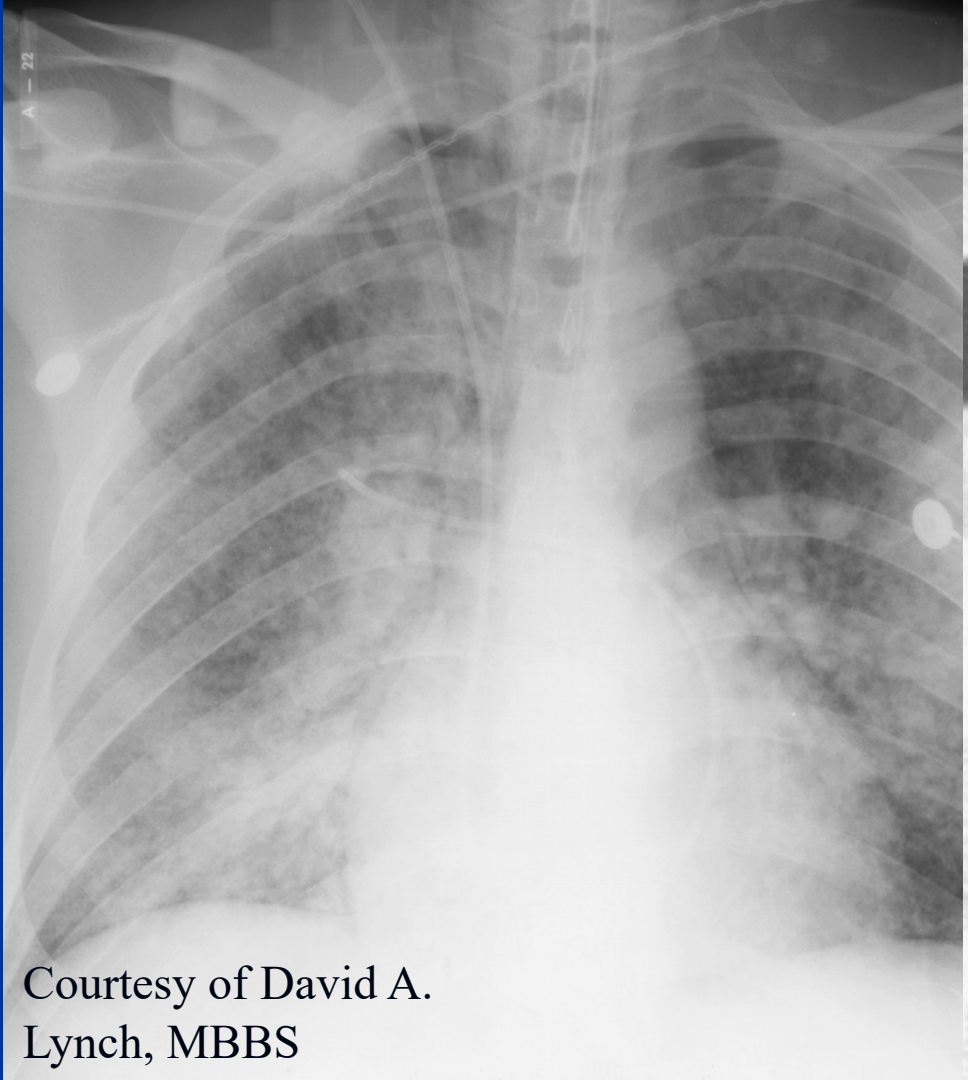
- Serositis more common than ILD
  - Pleural and/or pericardial effusion or thickening
- PNA most common lung dx
- Pulmonary hemorrhage
- Shrinking lung syndrome
- Pulmonary HTN
- Antiphospholipid syndrome (present in 1/3 of SLE)
  - Hypercoagulable
  - Hemorrhage

Systemic lupus erythematosus

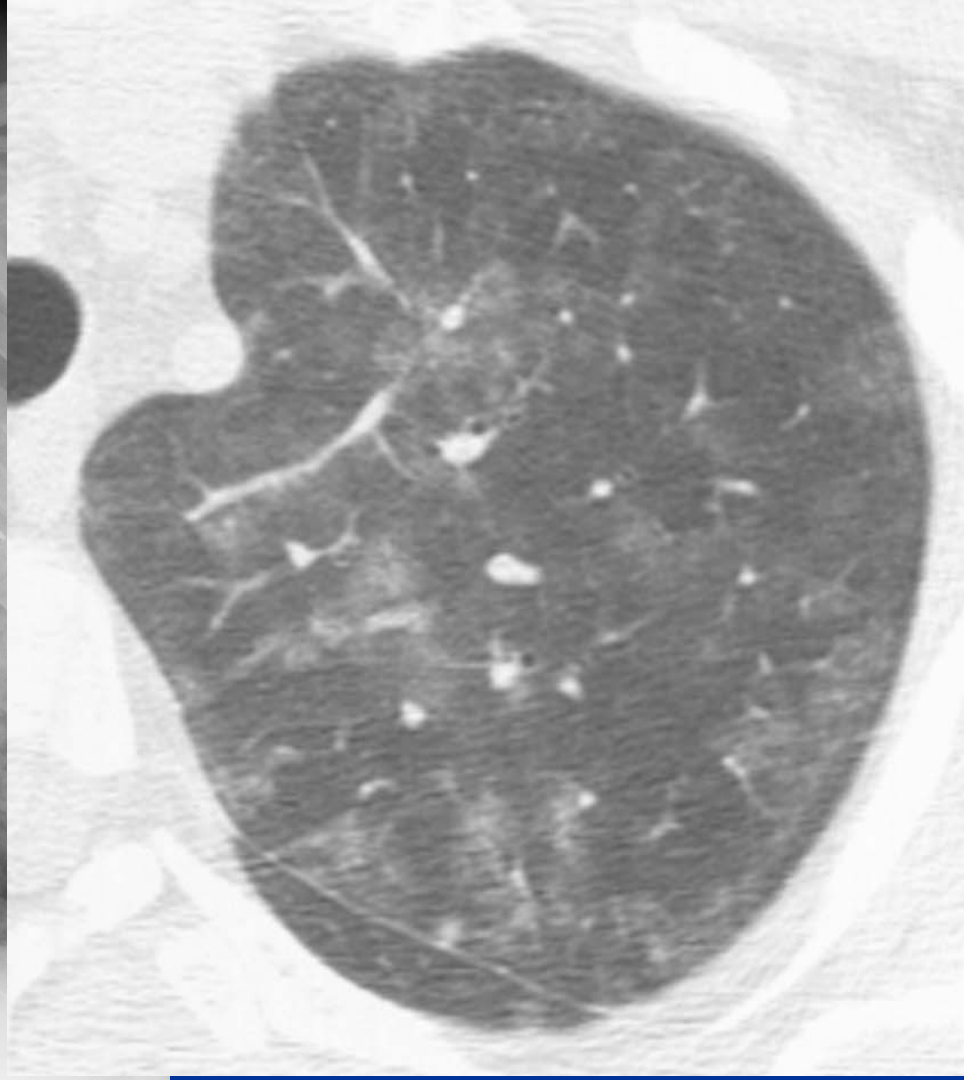


Systemic lupus erythematosus

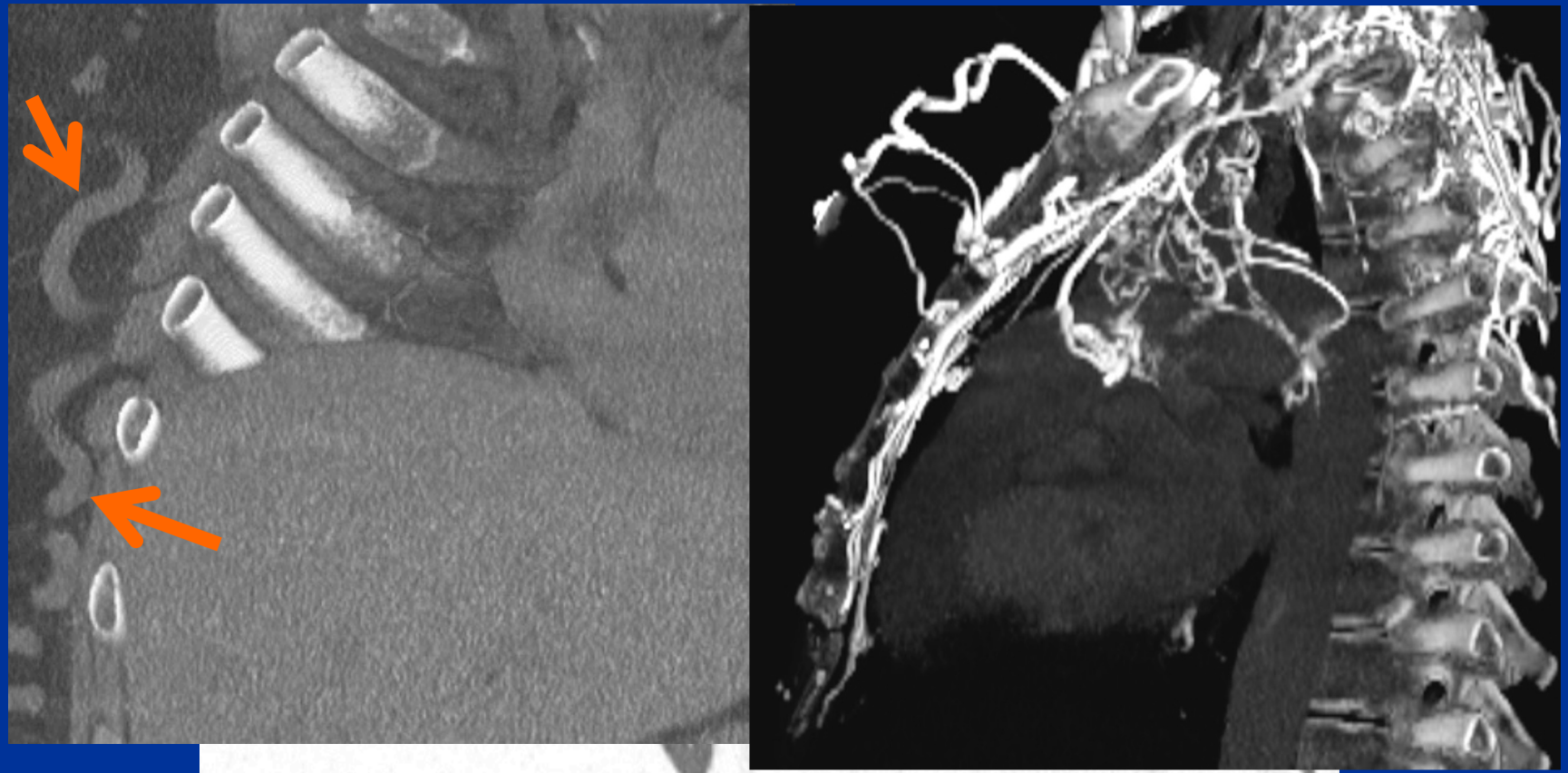




Courtesy of David A. Lynch, MBBS

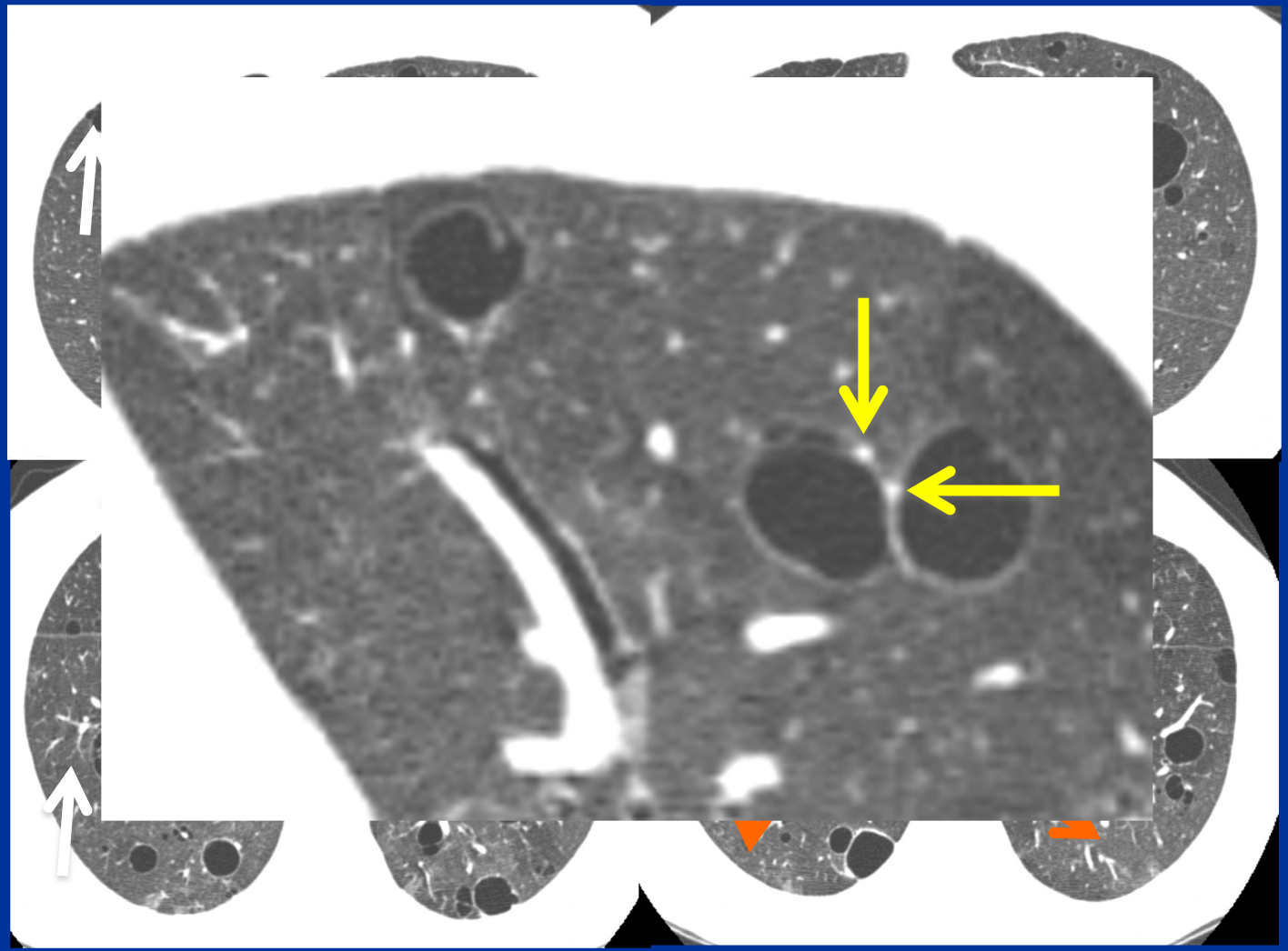


Antiphospholipid syndrome



Sjögren syndrome

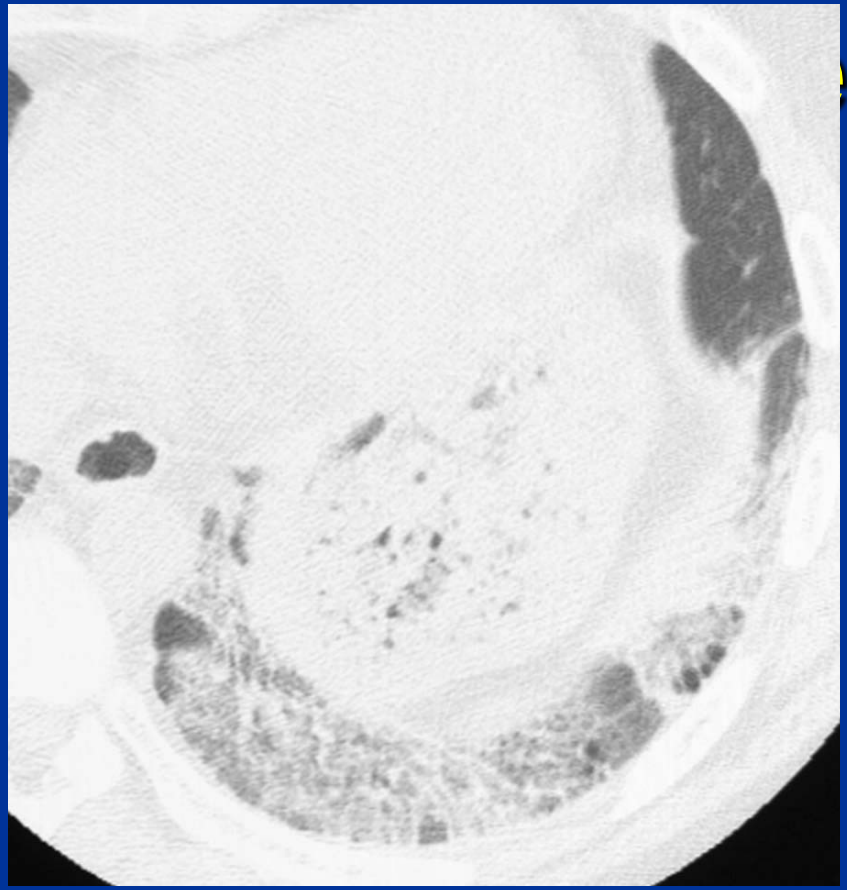
- Interstitial lung disease
  - LIP most likely
  - NSIP and UIP much less likely
- Airways disease (small and large)
- Lymphoma: Rare, usually MALT
  - Large nodules, consolidation



Mixed connective tissue disease

- NOT related to UCTD/IPAF
- Distinct clinical entity with overlap of SLE, scleroderma, and inflammatory myositis
- Anti-ribonucleoprotein (RNP) antibody
- Lung disease common (60%)
  - NSIP most common
- Pulmonary HTN, pleural and pericardial effusions, esophageal dysmotility

Mixed connective tissue



Courtesy of
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UCTD/IPAF (IP with autoimmune features)

- Suspected connective tissue disease not meeting ACR criteria for specific diagnosis
- Nonspecific antibody profile
- Up to 25% of all CTDs
- UIP pattern but NSIP also common
- Many patients previously IPF => IPAF
  - Research designation

Summary

- RA: UIP and OB, pleuritis
- Systemic sclerosis: NSIP, esophageal dysmotility; CA
- IIM: NSIP+OP; Jo-1 antibody
- SLE: Serositis, pulmonary hemorrhage
  - Antiphospholipid syndrome
- Sjögren: LIP
- Mixed: NSIP
- All: serositis and PAH

Thank You

Acknowledgement:
David A. Lynch, MBBS

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