

Interstitial Lung Diseases

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Interstitial Lung Disease

- Idiopathic IP's
 - IPF
 - NSIP
 - COP
 - RBILD
 - AIP
- CTD-ILD
- Sarcoidosis
- HP
- Pneumoconiosis
- Eosinophil ILD
- LCH
- LAM

Clinical Features

- Clinical Signs:
 - Chronic dry, nonproductive cough
 - » Paroxysmal and can be debilitating
 - Unexplained exertional dyspnea
 - Chest pain (rare)
- Infrequent in patients younger than 40 years old

What causes ILD? (200 causes!)

- 1) **Occupational & Environmental** – Silicosis, Asbestosis, Hypersensitivity Pneumonitis
- 2) **Drug Induced** – Amiodarone, Nitrofurantoin, Methotrexate, Cocaine
- 3) **Connective Tissue Diseases** – Lupus, RA, Scleroderma
- 4) **Primary Diseases** – Sarcoidosis, LAM
- 5) **Idiopathic (25%)** – IPF, NSIP
- 6) **Genetics**



Fungal and Bacteria:

Farmer's lung	Moldy hay, grain, silage		
Humidifier lung; air conditioner lung	Contaminated forced-air systems; water reservoirs		
Bagassosis	Moldy sugarcane (ie, bagasse)		
Mushroom worker's lung	Moldy mushroom compost		
Enoki mushroom worker's lung (Japan)	Moldy mushroom compost		
Suberosis	Moldy cork		
Detergent lung; washing powder lung	Detergents (during processing or use)		
Malt worker's lung	Moldy barley		
Sequoiosis	Moldy wood dust		
Maple bark stripper's lung	Moldy maple bark		
Cheese washer's lung	Moldy cheese		
Woodworker's lung	Oak, cedar, and mahogany dust, pine		
Hardwood worker's lung	Kiln-dried wood		
Paprika slicer's lung	Moldy paprika pods		
Sauna taker's lung	Contaminated sauna water		
Familial HP	Contaminated wood dust in walls		
Wood trimmer's lung	Contaminated wood trimmings		
Composter's lung	Compost		
Basement shower HP	Mold on unventilated shower		
Hot tub lung	Hot tub mists; mold on ceiling		

Animal and Insect proteins

Animal Proteins	
Pigeon breeder's or pigeon fancier's disease	Parakeets, budgerigars, pigeons, chickens, turkeys
Pituitary snuff taker's lung	Bovine and porcine pituitary proteins
Fish meal worker's lung	Fish meal dust
Bat lung	Bat droppings
Furrier's lung	Animal pelts
Animal handler's lung; laboratory worker's lung	Urine, serum, pelts, proteins
Insect Proteins	
Miller's lung	Dust-contaminated grain
Lycoperdonosis	Lycoperdon puffballs

Question:

- How do we narrow this list to the actual cause in me?

History & Physical

- A detailed history is very important including previous occupations and exposures.
- Emphasis on an Early and Accurate diagnosis

Silicosis



www.cdc.gov/Spanish/niosh/images/silifig0.gif

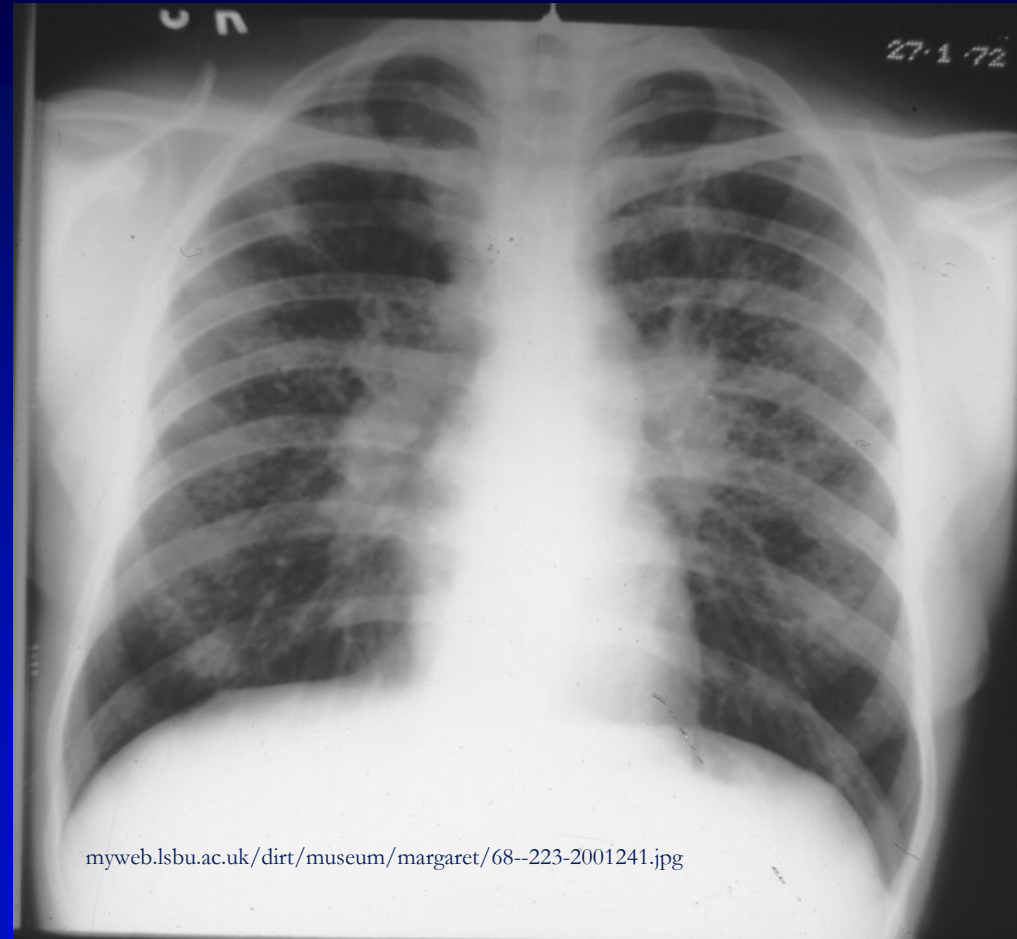


www.cowart.info/.../Parrotman-782740.JPG

Physical Findings

- Physical Examination can be diagnostic.

Sarcoidosis



Scleroderma



newsimg.bbc.co.uk/media/images/39397000/jpg/

Serologic Tests Can Help Exclude Other Conditions

Connective tissue diseases

ESR, ANCA
ANA
CCP (for RA)
CK
Aldolase
Anti-myositis panel with Jo-1 antibody
ENA panel

- Scl-70
- Ro (SSA)
- La (SSB)
- Smith
- RNP

Hypersensitivity pneumonitis

Hypersensitivity panel
(if exposure history)

Table 1. Pulmonary Function Test Results

Test	Predicted	Measured	% Predicted
FVC (L)	2.31	2.17	94
FEV ₁ (L)	1.89	1.61	85
FEV ₁ /FVC	0.78	0.70	90
FRC (L)	2.47	1.85	75
RV (L)	1.91	1.24	65
TLC (L)	4.41	3.26	74
D _{LCO} (mL/mm Hg/s)	16.74	3.85	23
D _L /V _A	4.33	1.17	27

Predicted = mean predicted values as per Crapo et al.¹

FVC = forced vital capacity

FEV₁ = forced expiratory volume in the first second.

FEV₁/FVC = ratio of FEV₁ to FVC

FRC = functional residual capacity.

RV = residual volume.

TLC = total lung capacity by helium dilution

D_{LCO} = Diffusing capacity for carbon monoxide.

D_L/V_A = ratio of diffusing capacity to alveolar volume.

Focus on the TLC and DLCO, both low

6-Minute Walk Test



Bronchoscopy

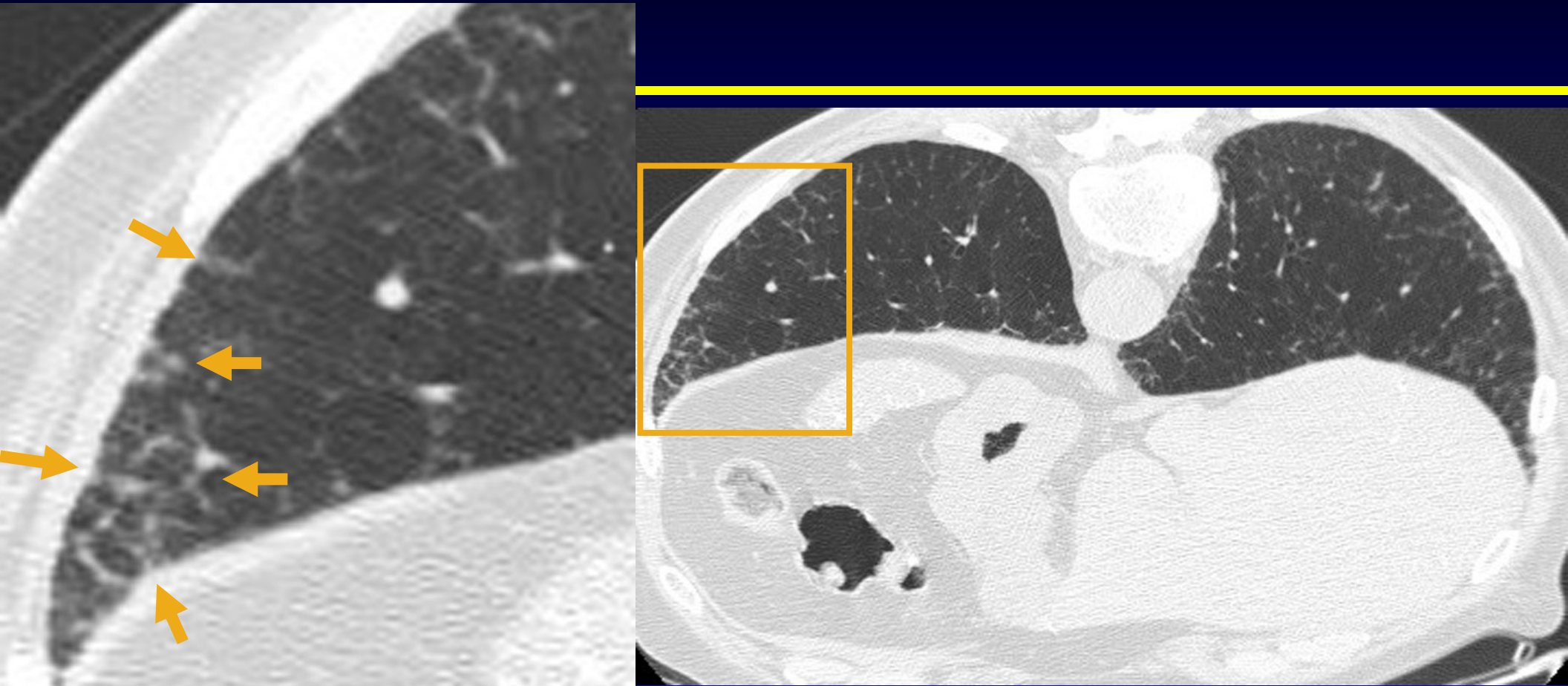


www.pcca.net/images/BronchoscopyNose.jpg

One Example of PF-IPF

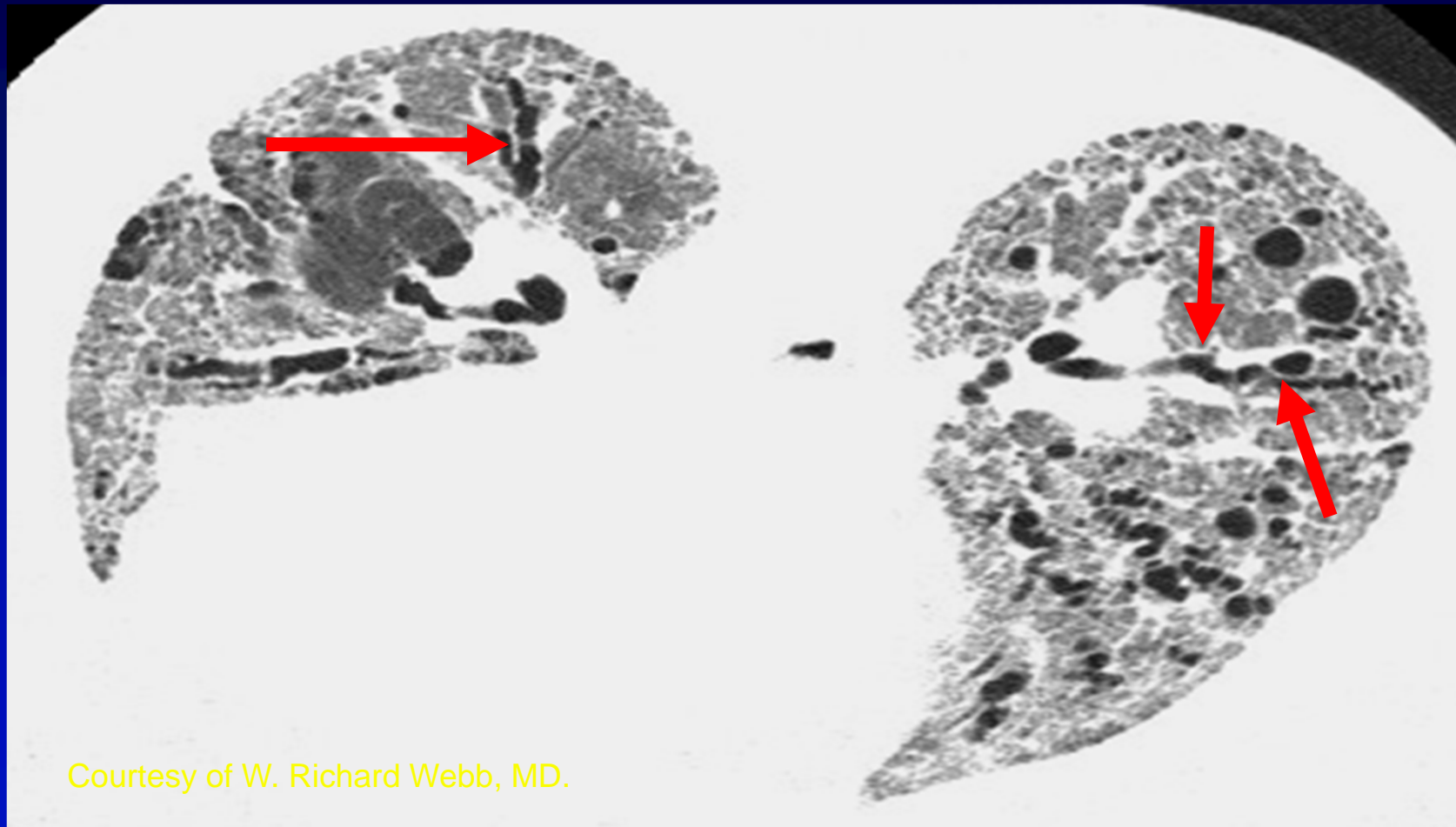
- IPF is the commonest idiopathic ILD

Early HRCT Findings in IPF



Courtesy of David A. Lynch, MD.

UIP: Traction Bronchiectasis



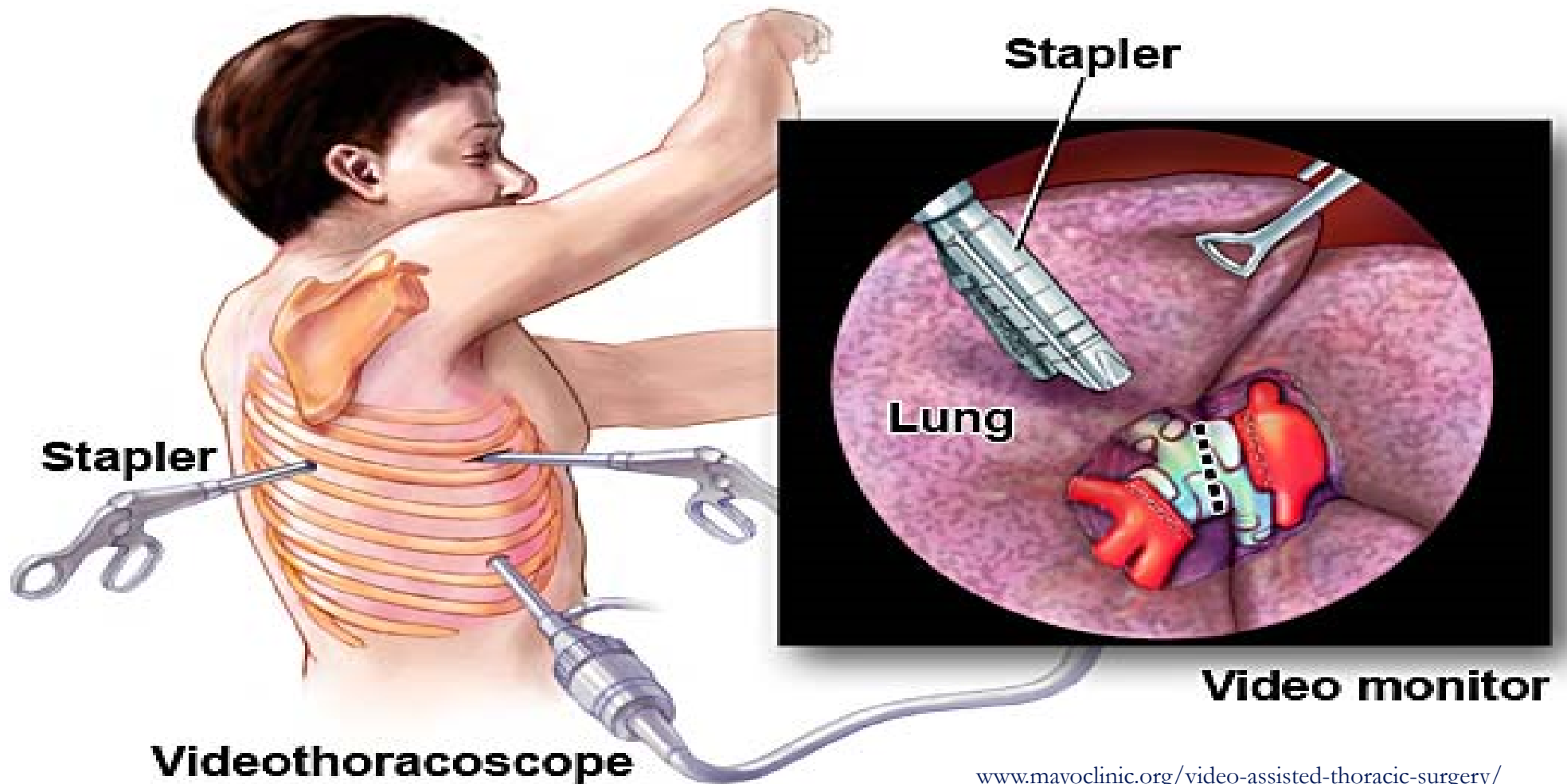
International Consensus Statement on IPF: Histology

- Surgical lung biopsy (VATS) recommended in patients with suspected IPF, especially those with **atypical** clinical or radiographic features

Video-Assisted Thoracic Surgery (VATS)

- High diagnostic accuracy
- Less morbidity and mortality than open lung biopsy
- Ideal biopsy
 - Two or more surgical wedge biopsies with areas of normal lung taken from different areas of the lung
 - Samples should measure 3–5 cm in length and 2–3 cm in depth
- Outpatient thoracoscopic lung biopsy in patients with interstitial or focal lung disease
 - Diagnosis obtained in 61/62 patients
 - 72.5% discharged home within 8 hours
 - 22.5% discharged home within 23 hours

Video-Assisted Thoracoscopic Surgery (VATS)



Approach to the Diagnosis of IPF

Clinical

- History
- Physical
- Laboratory
- PFTs

Radiology

- Chest X-ray
- HRCT

Pathology

- Surgical lung biopsy

Primary care
physicians

Pulmonologists

Radiologists

Pathologists

Multidisciplinary

Pulmonary Function Tests

- Restrictive Lung Disease
 - Low FVC or TLC ($< 80\%$)
 - Normal FEV1/FVC ratio ($> 70\%$)
 - Low Dlco ($< 80\%$)
- Some conditions can have obstructive components as well

Interstitial Lung Disease

High resolution CT scanning:

- 1-2 mm thin sections
- No contrast required
- Pattern may be pathognomonic

ILD: Role of HRCT

HRCT Scans:

- Confirm or exclude diagnosis
- Assess pattern and extent
- **Assess prognosis**



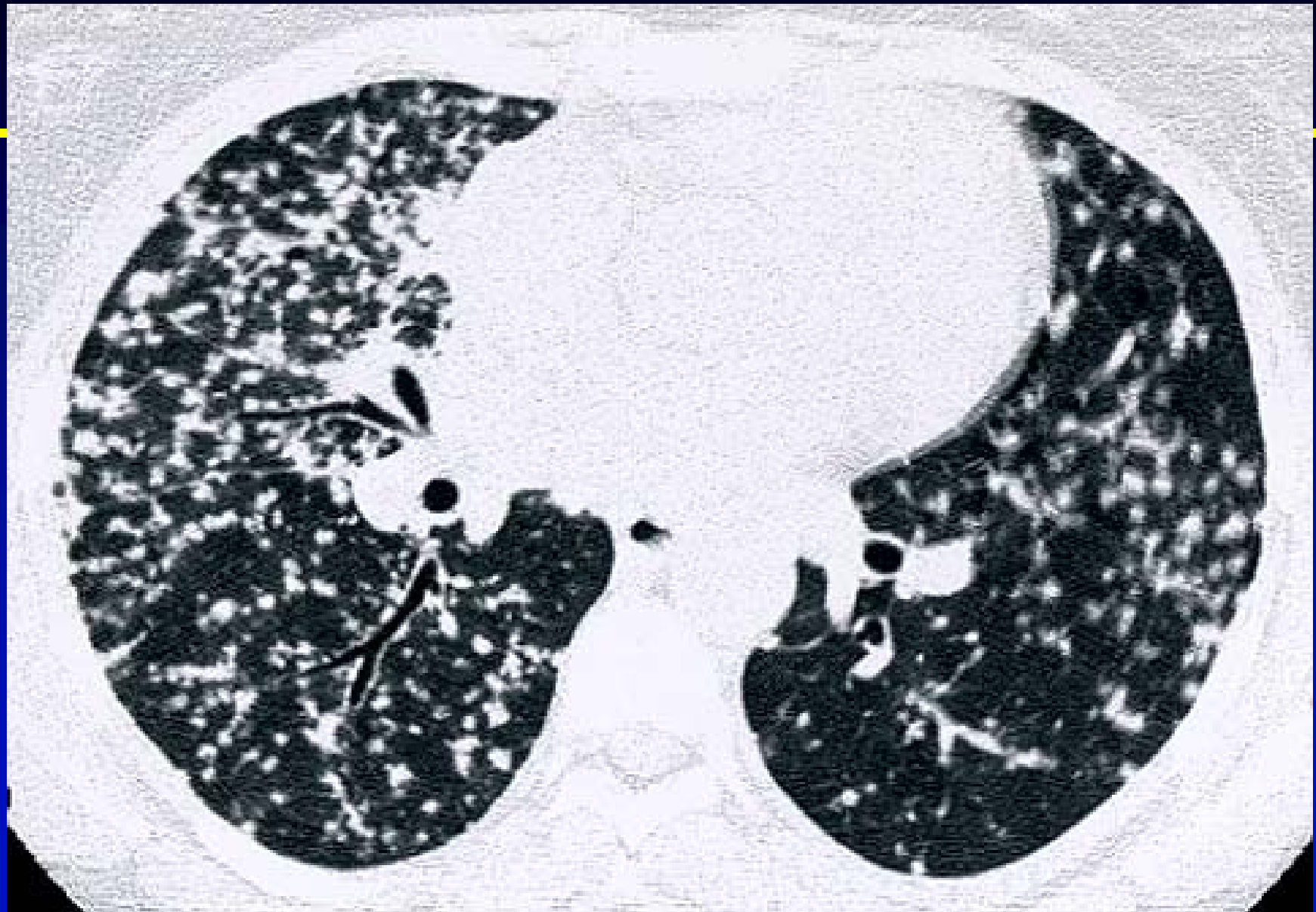




Case Presentation

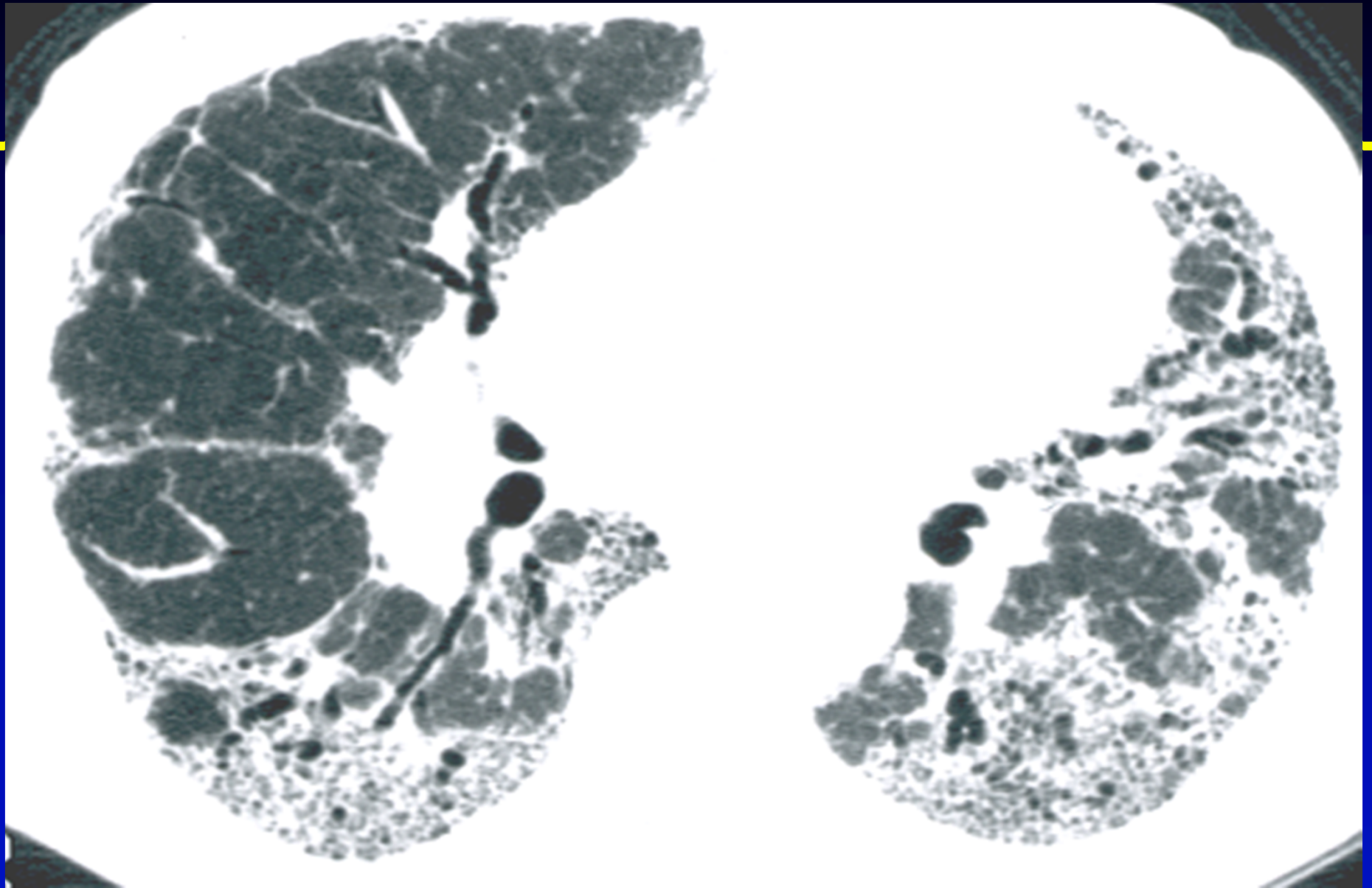
- 43yo F with progressive dry cough x 4 months. No other constitutional symptoms
- Dyspnea on heavy exertion such as climbing stairs or lifting objects
- No prior PMHx
- Life-long non-smoker, no drugs or occupational exposures

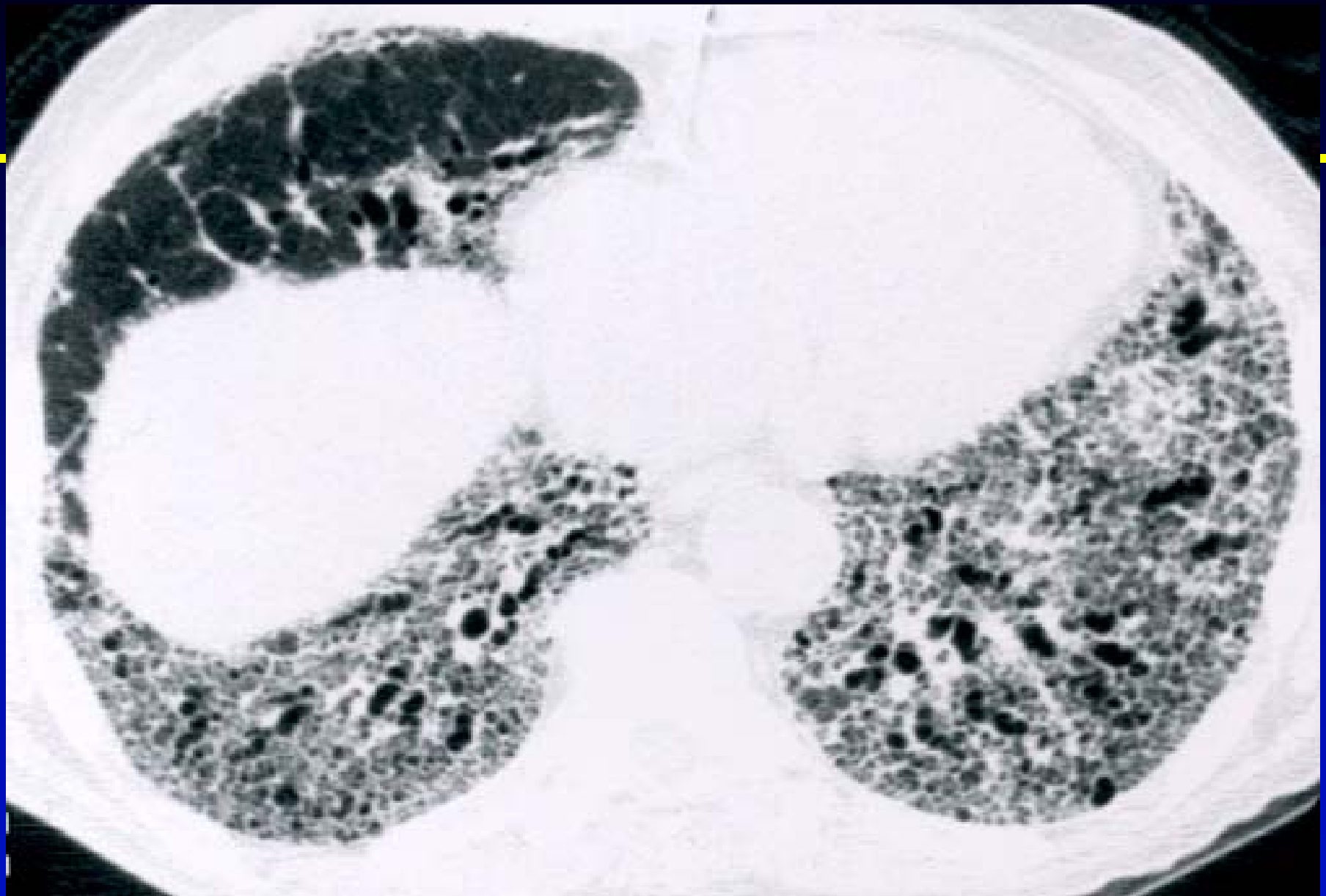




Case Presentation 2

- 65yo M with progressive shortness of breath with activity for 1 year
- Oxygen dependent x 6 months 2L n/c
- No significant smoking history
- No occupational exposures or allergies
- PMHx: Hypertension, Diabetes





Pulmonary Function Testing

- FVC: 1.89 (70) (normal $\geq 80\%$)
- FEV1 1.53 (75) (normal $\geq 80\%$)
- FEV1/FVC 81 (normal $\geq 70\%$)
- DLco 4.6 (21) (normal $\geq 80\%$)

HRCT Patterns in ILDs

- **Upper lobe** (Sarcoid; TB, LCG)
- **Lower lobe** (IPF, CVD-PF)
- **Diffuse; no predominance** (LAM)

HRCT: Patterns

- Cystic lesions
- Ground glass (alveolar)

HRCT: Patterns

Cystic lesions

- IPF (honeycomb cysts)
- Emphysema
- Sarcoidosis
- LCG and LAM

Interstitial Lung Diseases (ILDs)

Alveolar disorders:

- DIP; AIP; LIP; HP
- Cryptogenic organizing pneumonia
- Pulmonary alveolar proteinosis (PAP)
- Chronic eosinophilic pneumonia

Idiopathic Interstitial Pneumonias

Histopathological subtypes:

- Usual (UIP)
- Desquamative (DIP/RBILD)
- Acute (AIP)
- Nonspecific (NSIP)

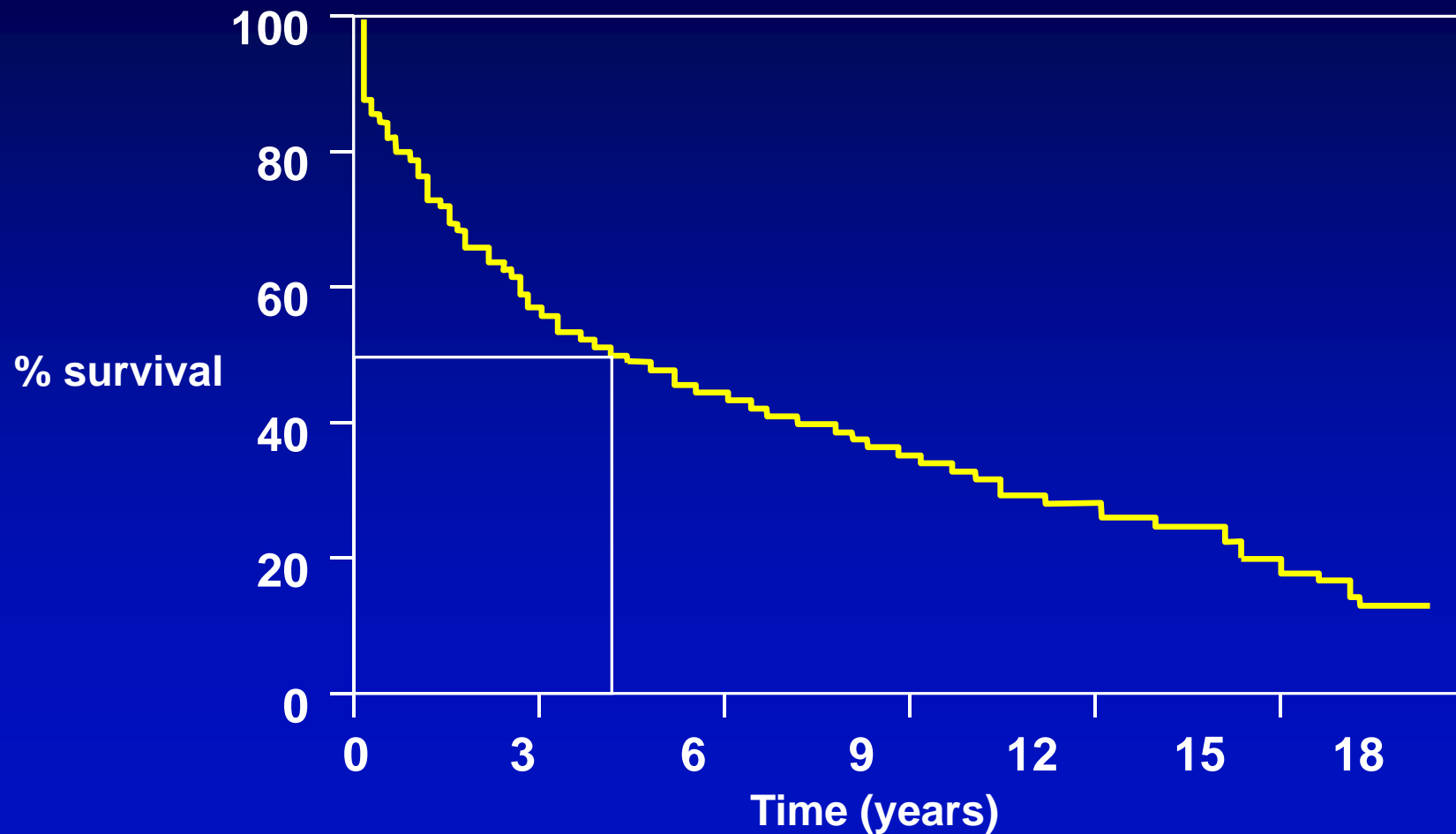
Katzenstein and Myers, *AJRCCM* 1998;157;1301

Idiopathic Pulmonary Fibrosis (IPF)

IPF is synonymous with:

- **Cryptogenic Fibrosing Alveolitis**
- **Usual Interstitial Pneumonia (UIP)**

Idiopathic Pulmonary Fibrosis: Overall Survival



M Turner-Warwick. *Thorax*. 1980;35:171.

Normal Lung



IPF Lung



Idiopathic Pulmonary Fibrosis (IPF)

- Age > 50
- M:F - 2:1
- Progressive breathlessness
- Bibasilar crackles, clubbing
- PERIPHERAL Interstitial pattern
- Subpleural honeycombing



What symptoms can I expect?

- Breathlessness (worse with exercise)
- Hacking dry cough
- Fatigue and weakness
- Appetite and weight loss
- Enlargement of fingertips (clubbing)

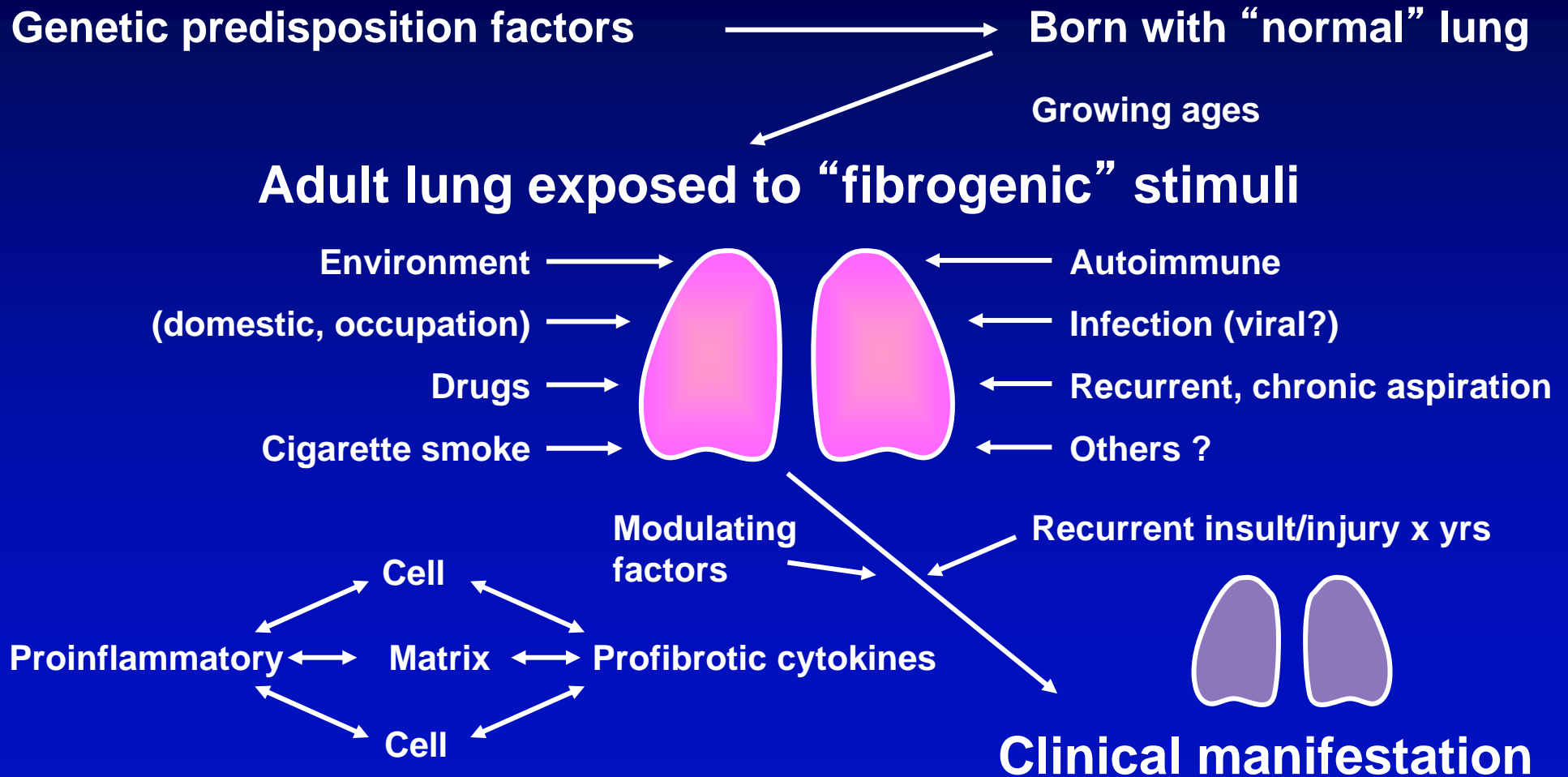


Am I just unfortunate?

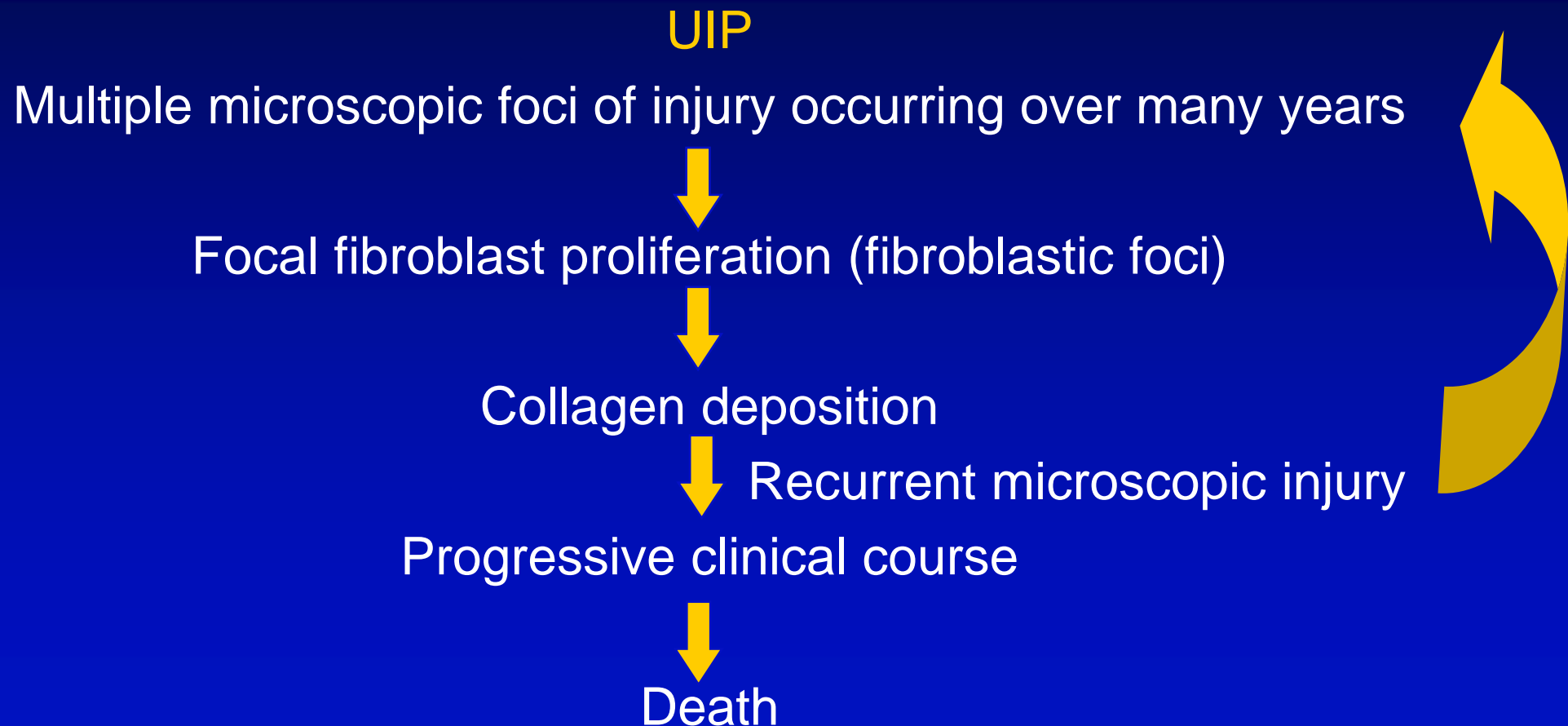
- Over 5 million people worldwide have fibrosis
- Over 300,000 in the US (likely more)
 - 100,000 new cases every year
 - 40,000 die each year (same as breast cancer) perhaps more (51/1,000 000 people)
- UK- one in 3,000 has PF.
 - 3,000 die annually from PF

Pulmonary Fibrosis of Unknown Etiology

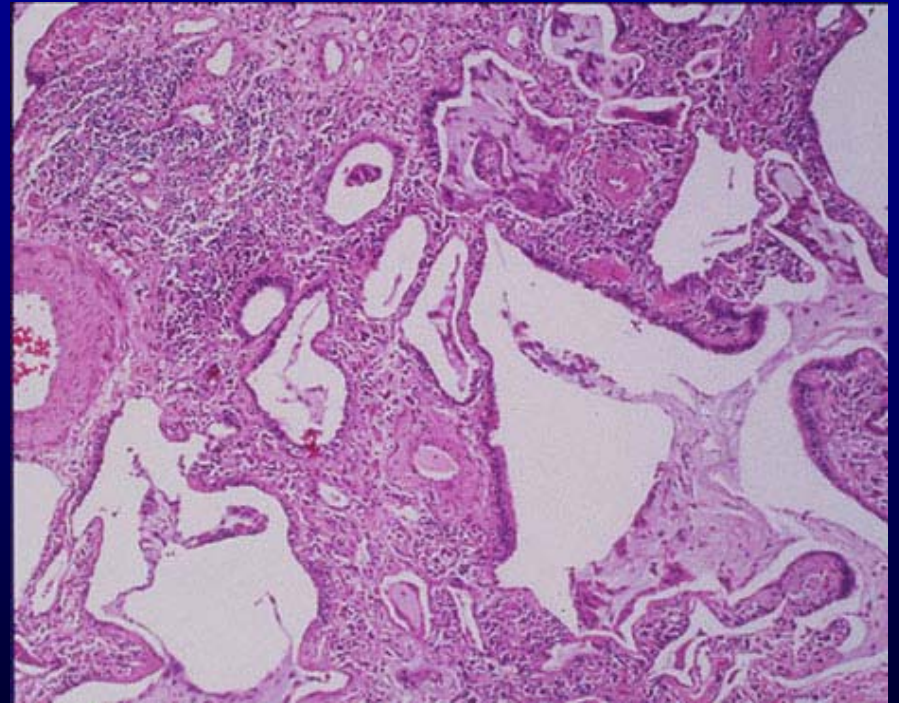
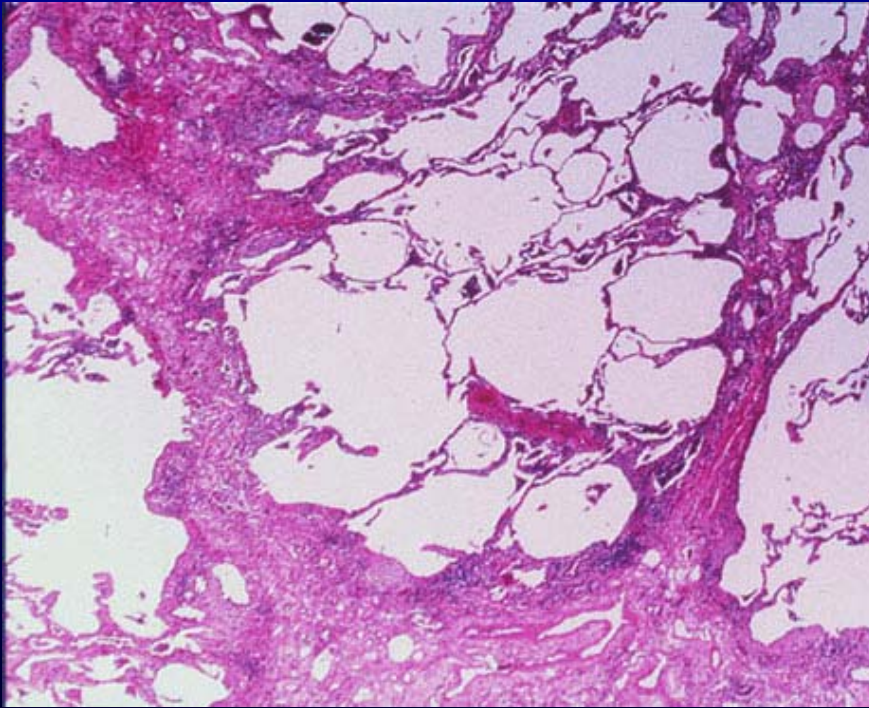
Conceptual pathogenesis of unusual interstitial pneumonia (UIP)

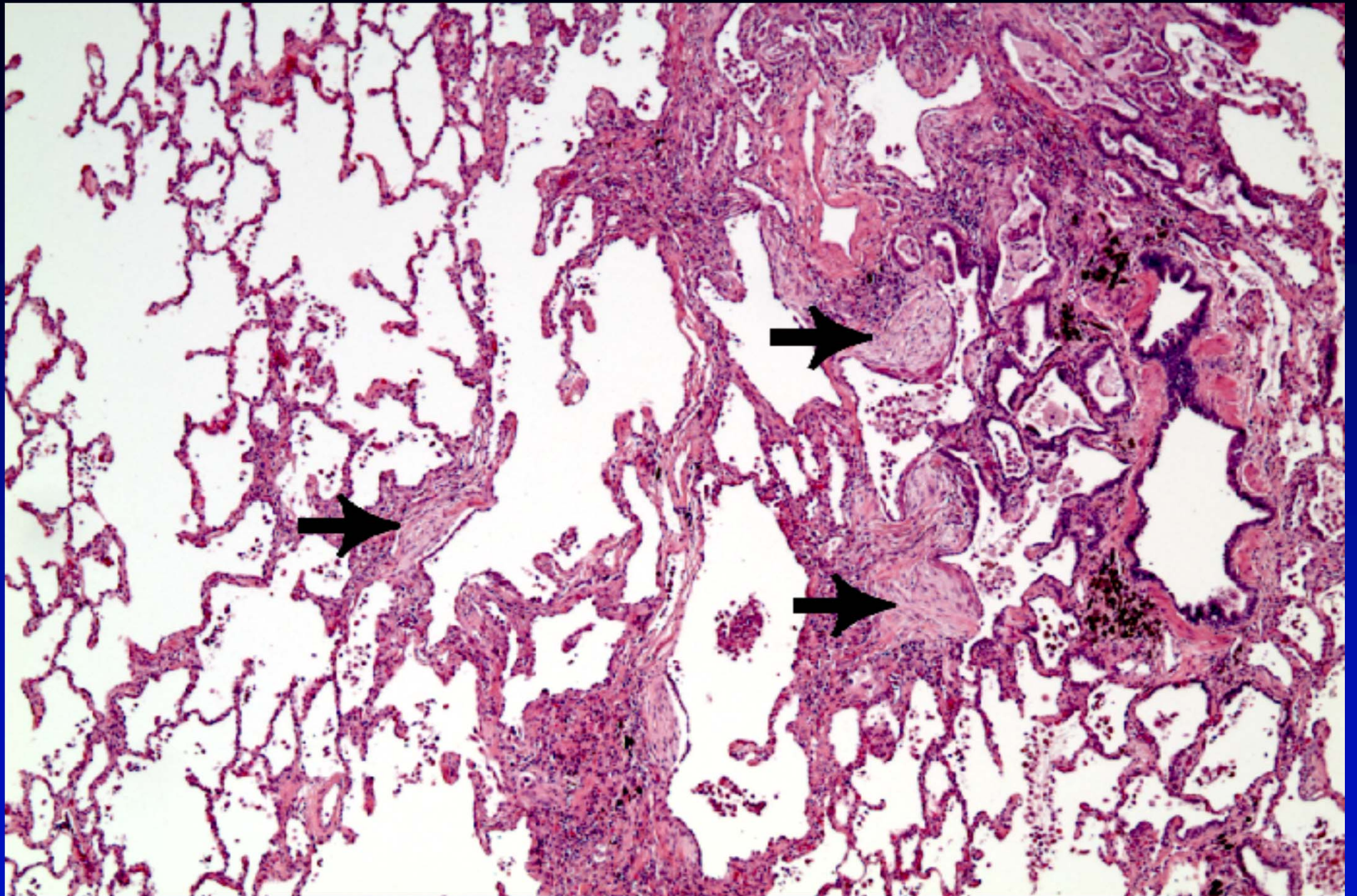


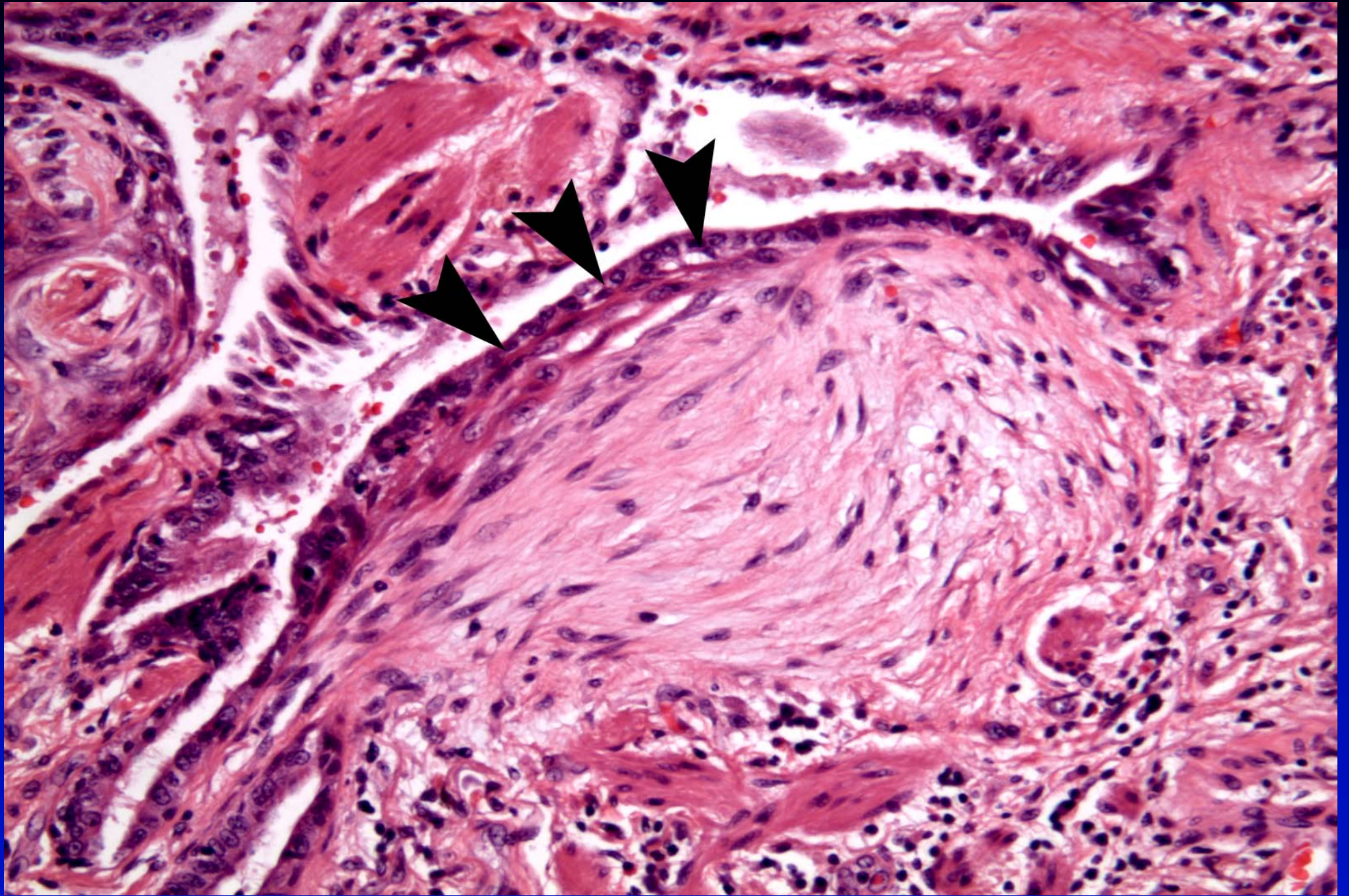
Pathogenesis and course of UIP

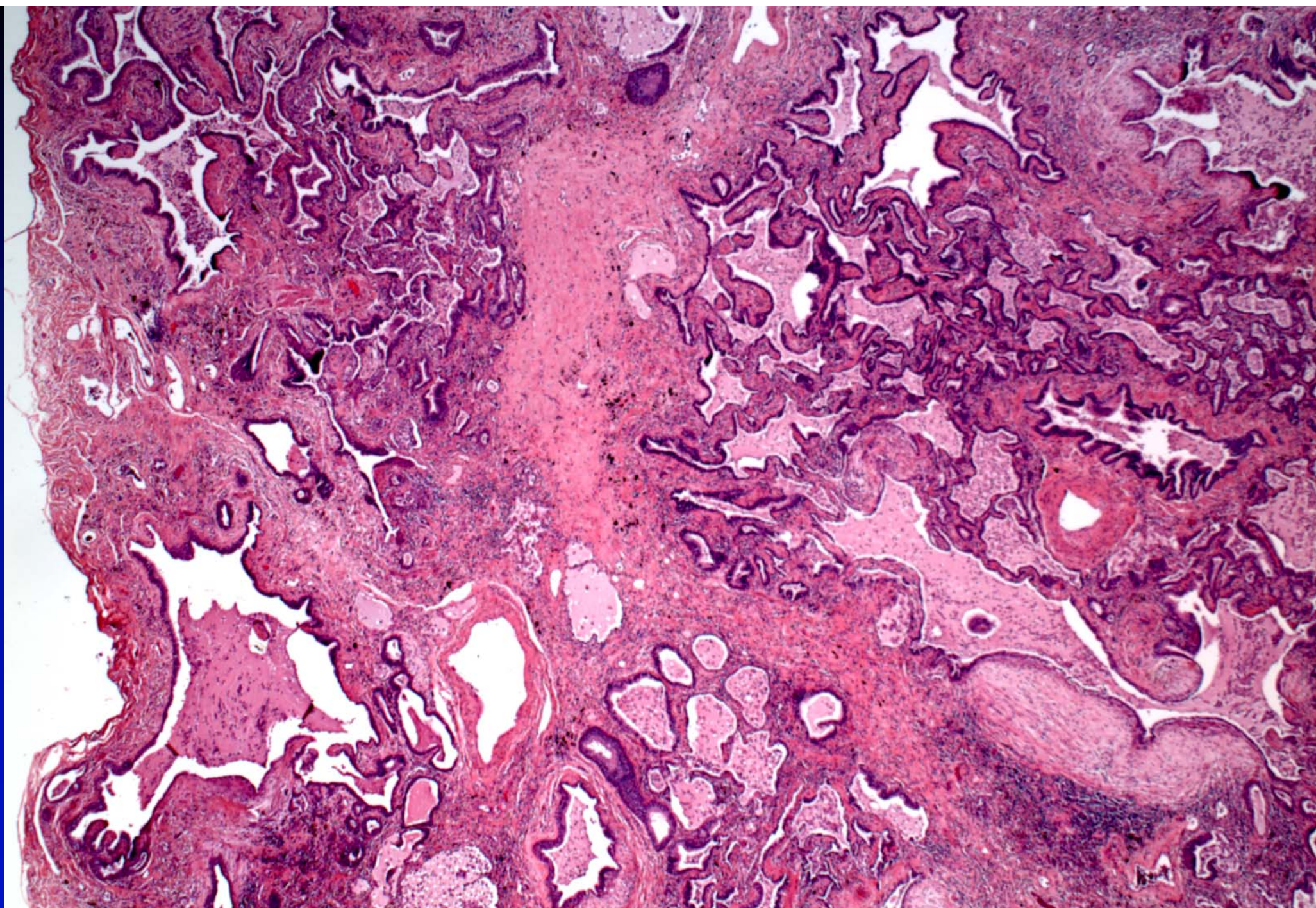


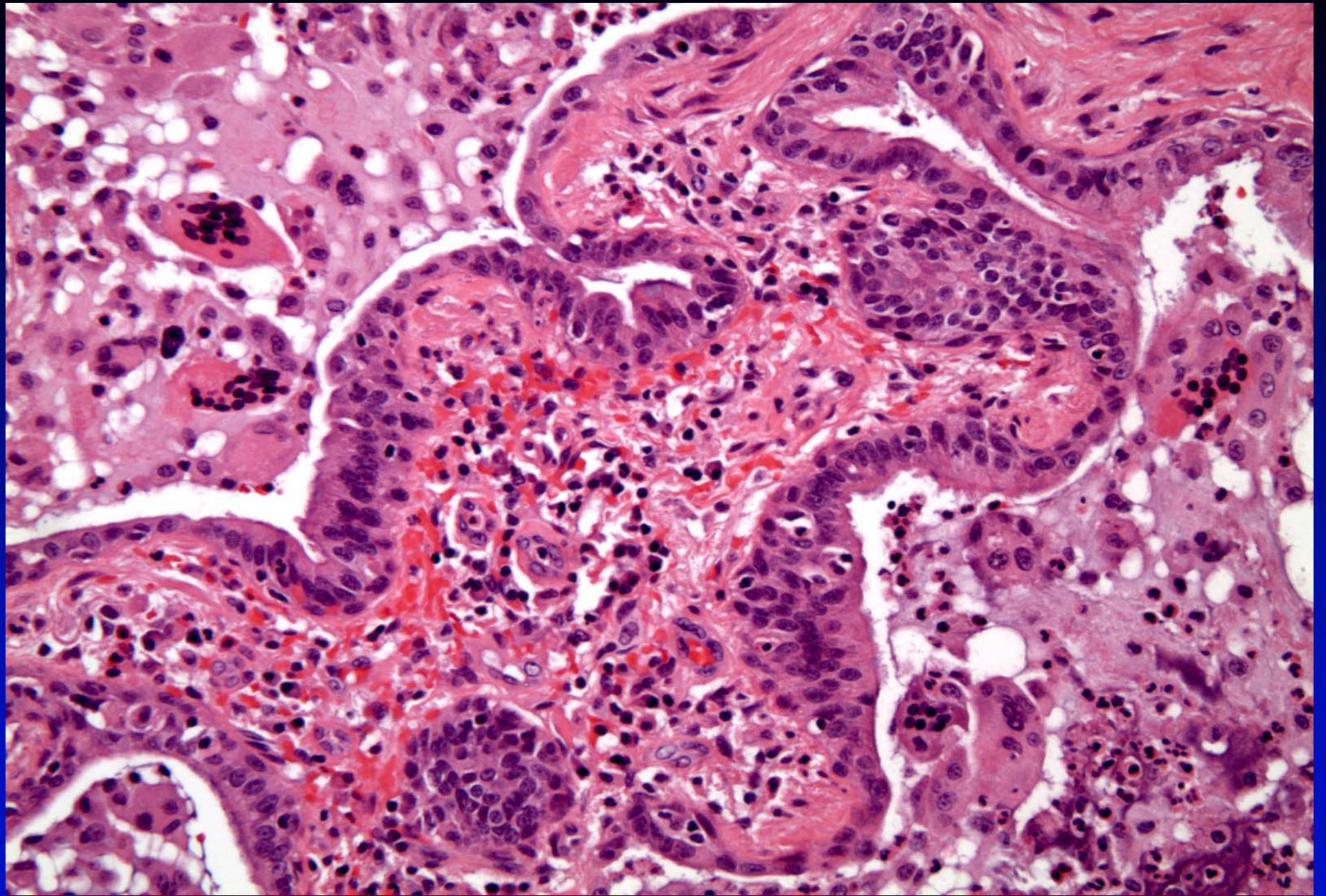
Temporal heterogeneity of UIP

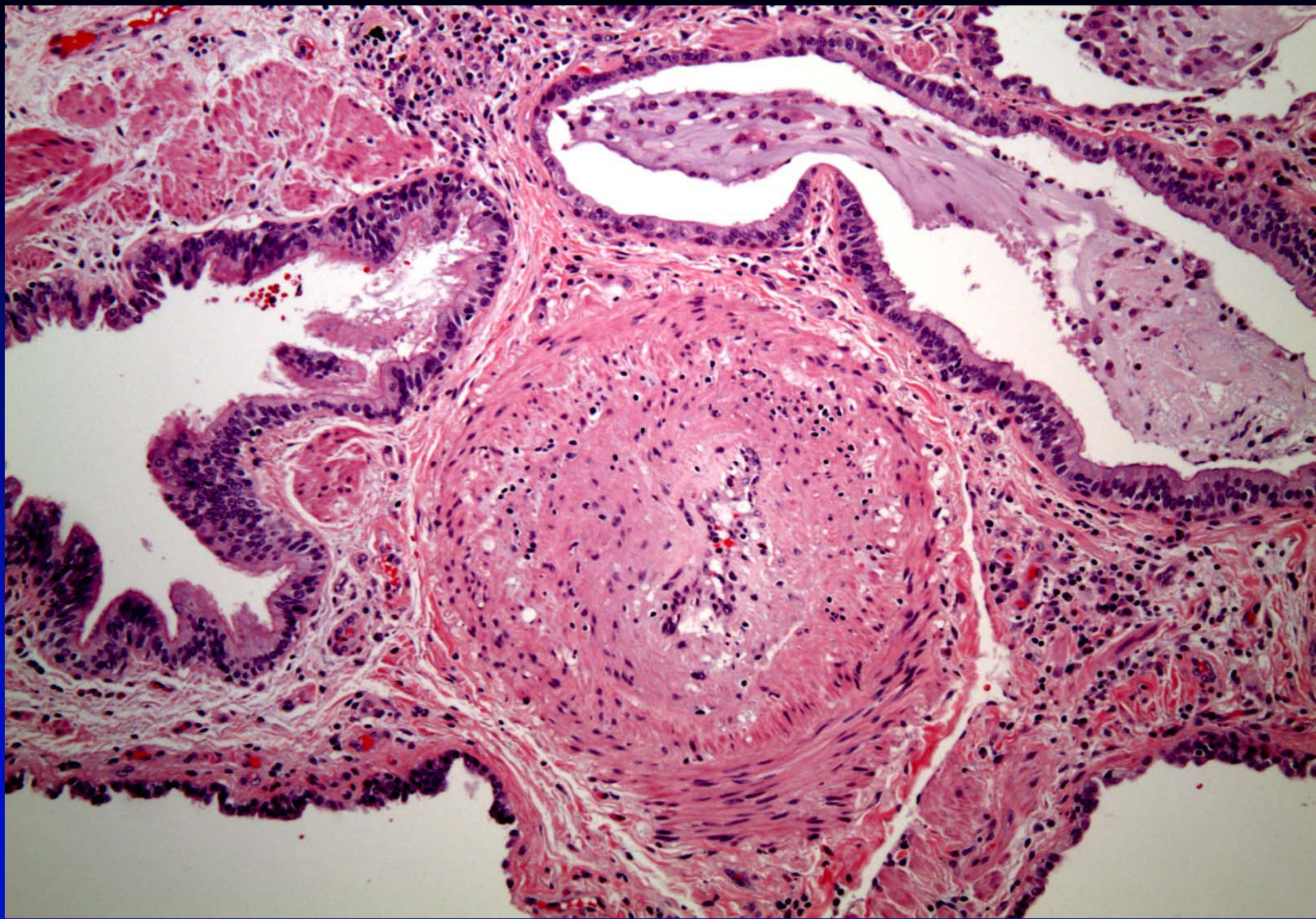


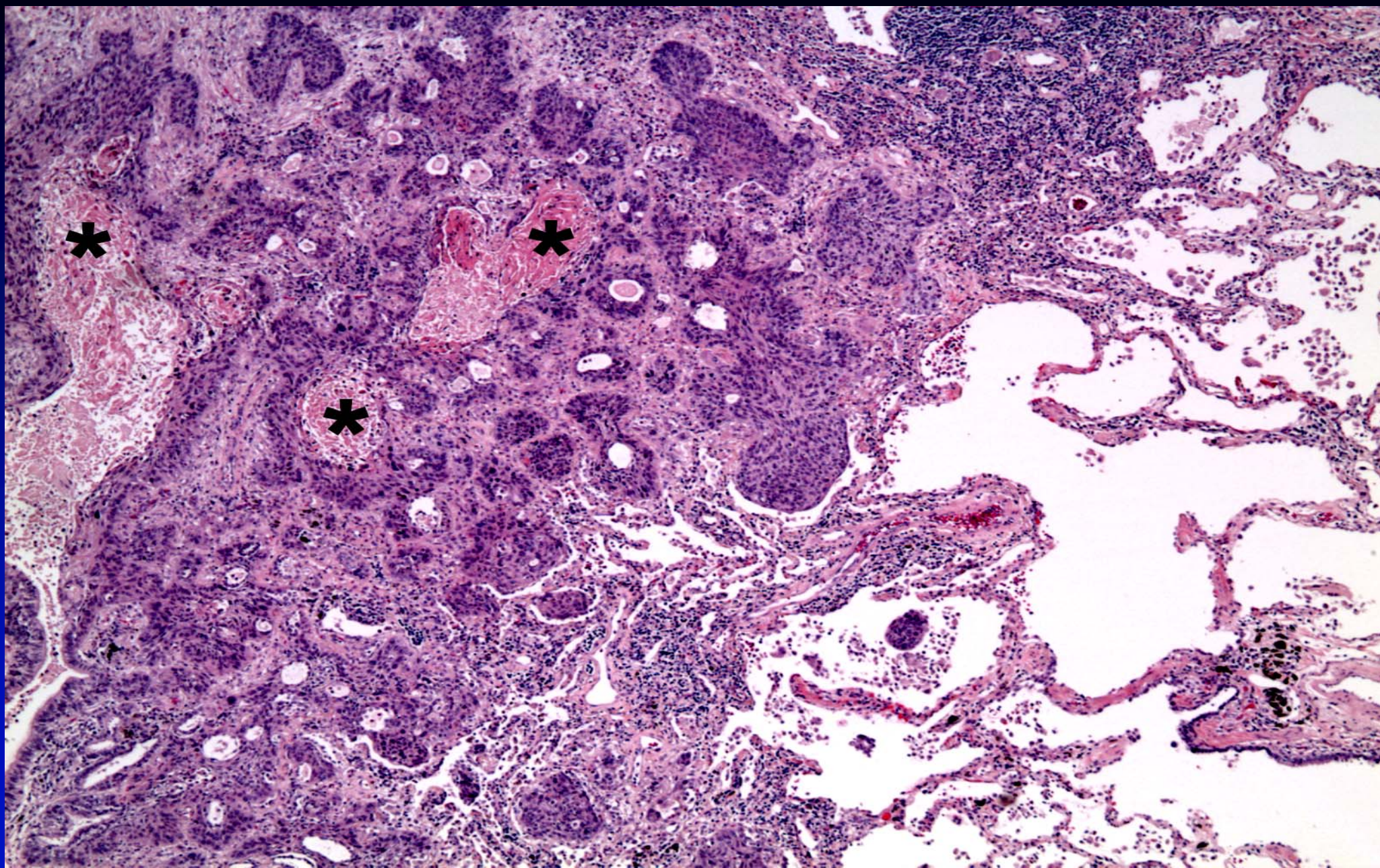














CASE# 507-10302 SPECIMEN DATE



CASE# 507-10302 SPECIMEN DATE

CANCER DIAGNOSIS

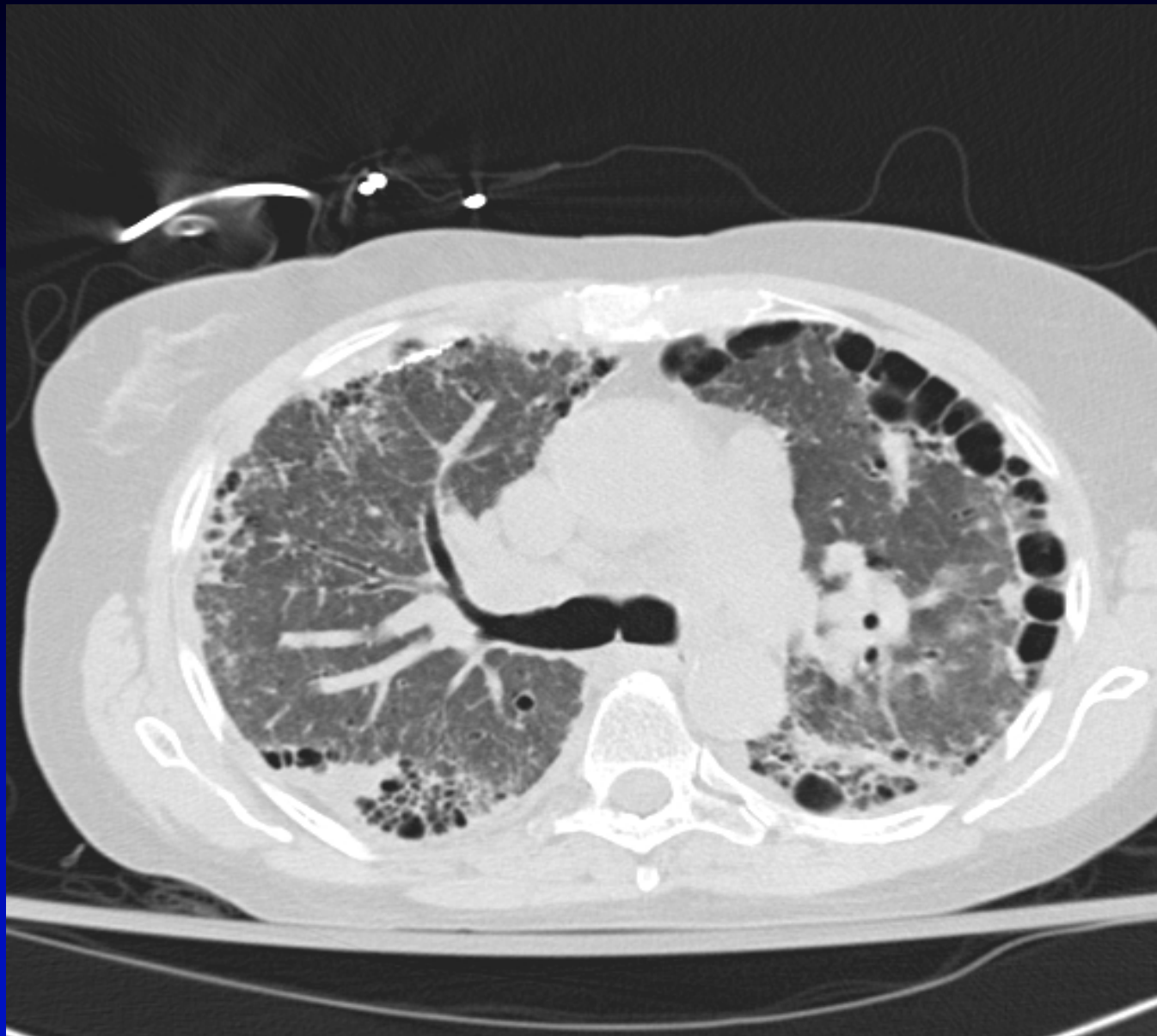
UIP: HRCT Features

- Patchy, heterogeneous
- Lower lobes, subpleural
- Reticular (linear) lines
- Honeycomb cysts
- Ground glass minimal or absent

UIP: subpleural, patchy







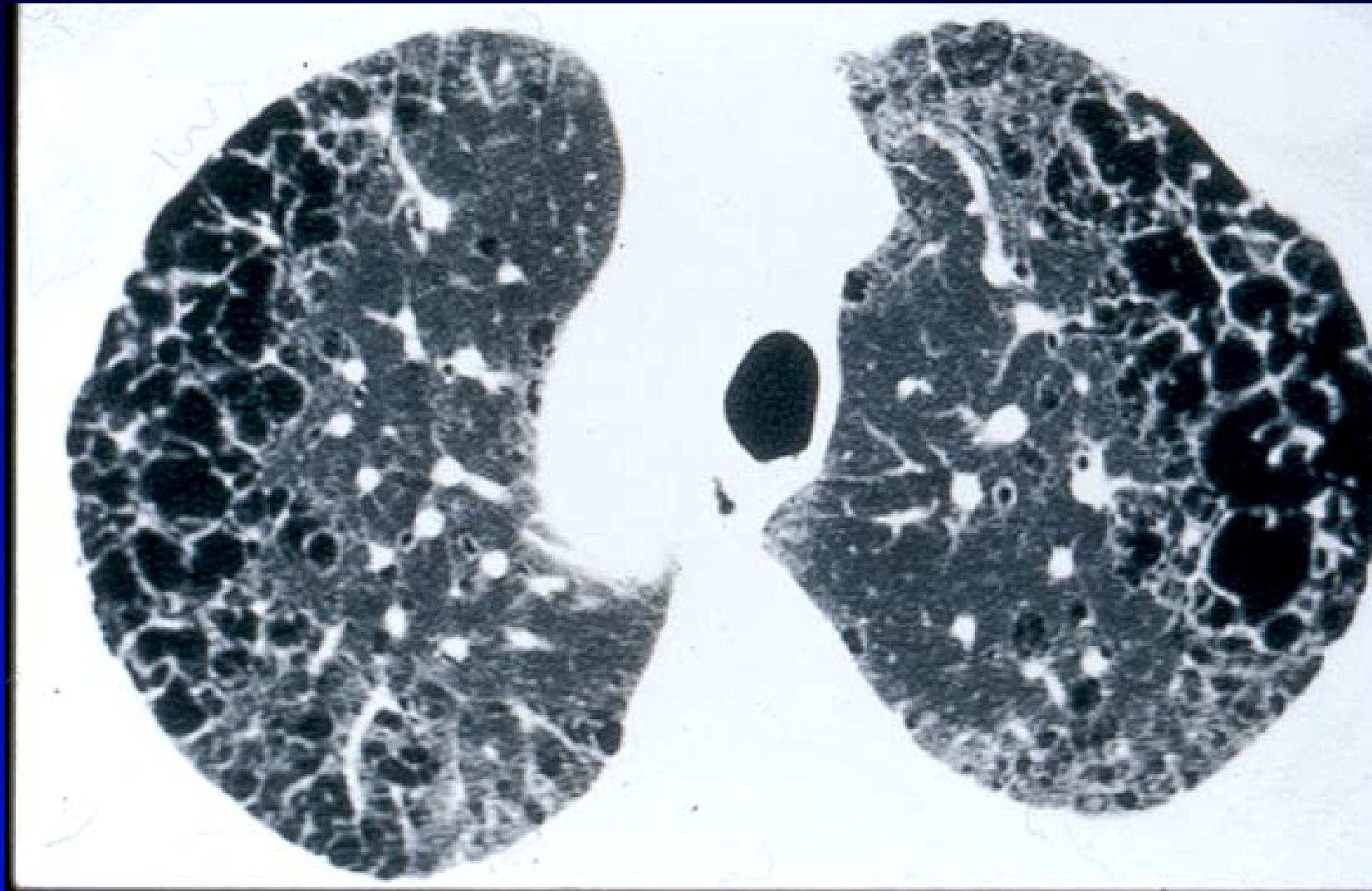




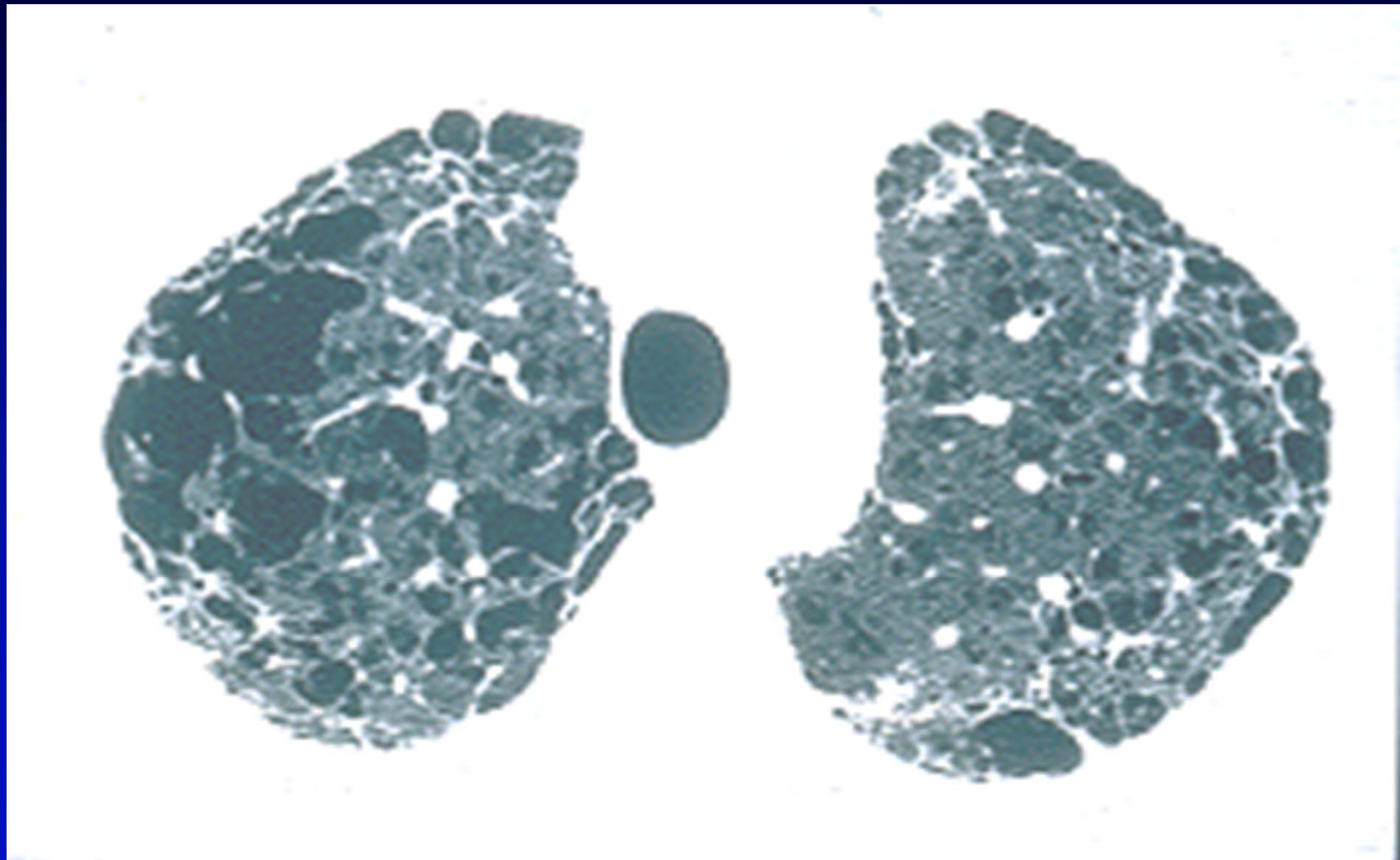
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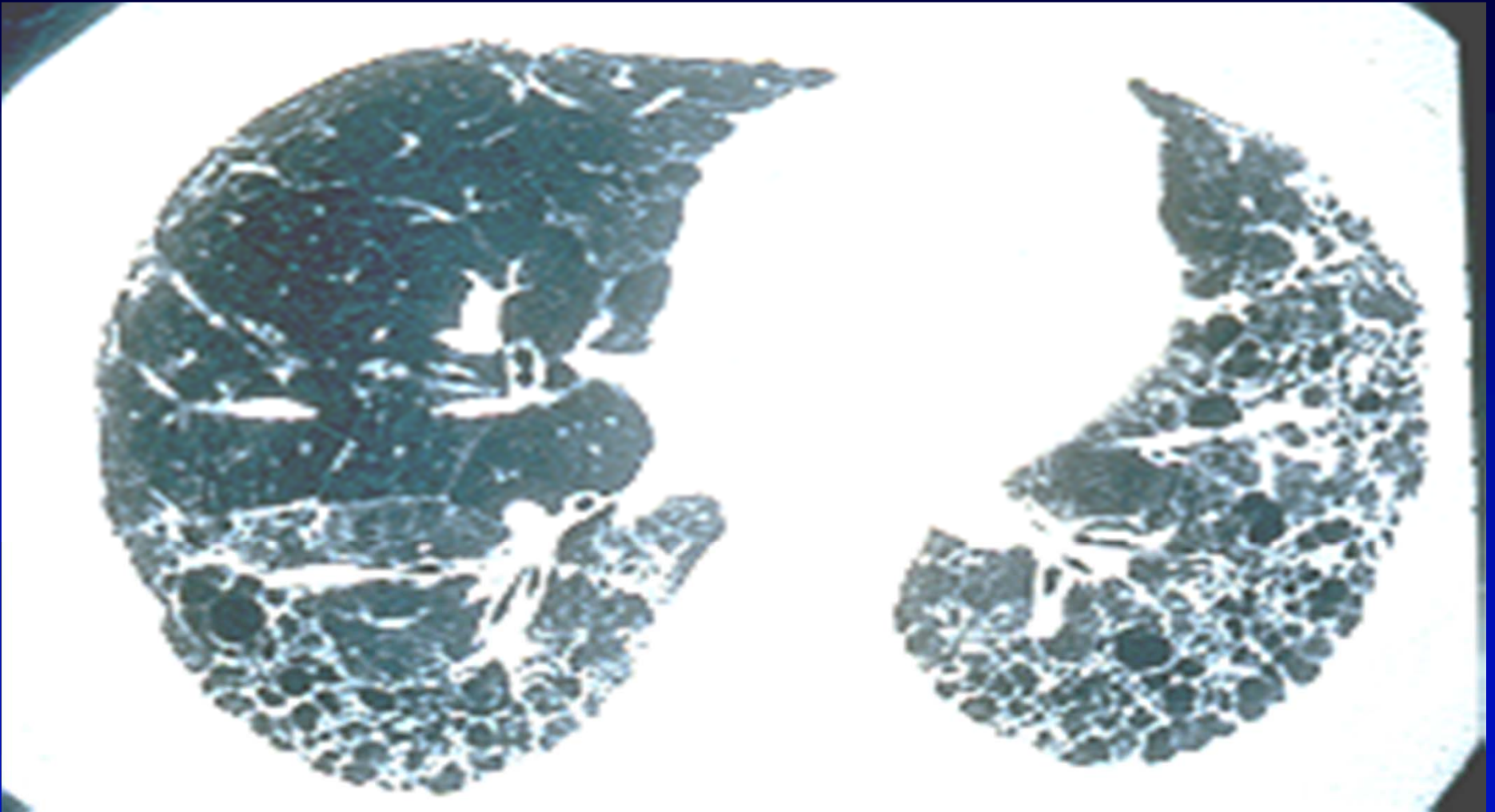
Honeycomb cysts (UIP)



Honeycomb cysts (UIP)



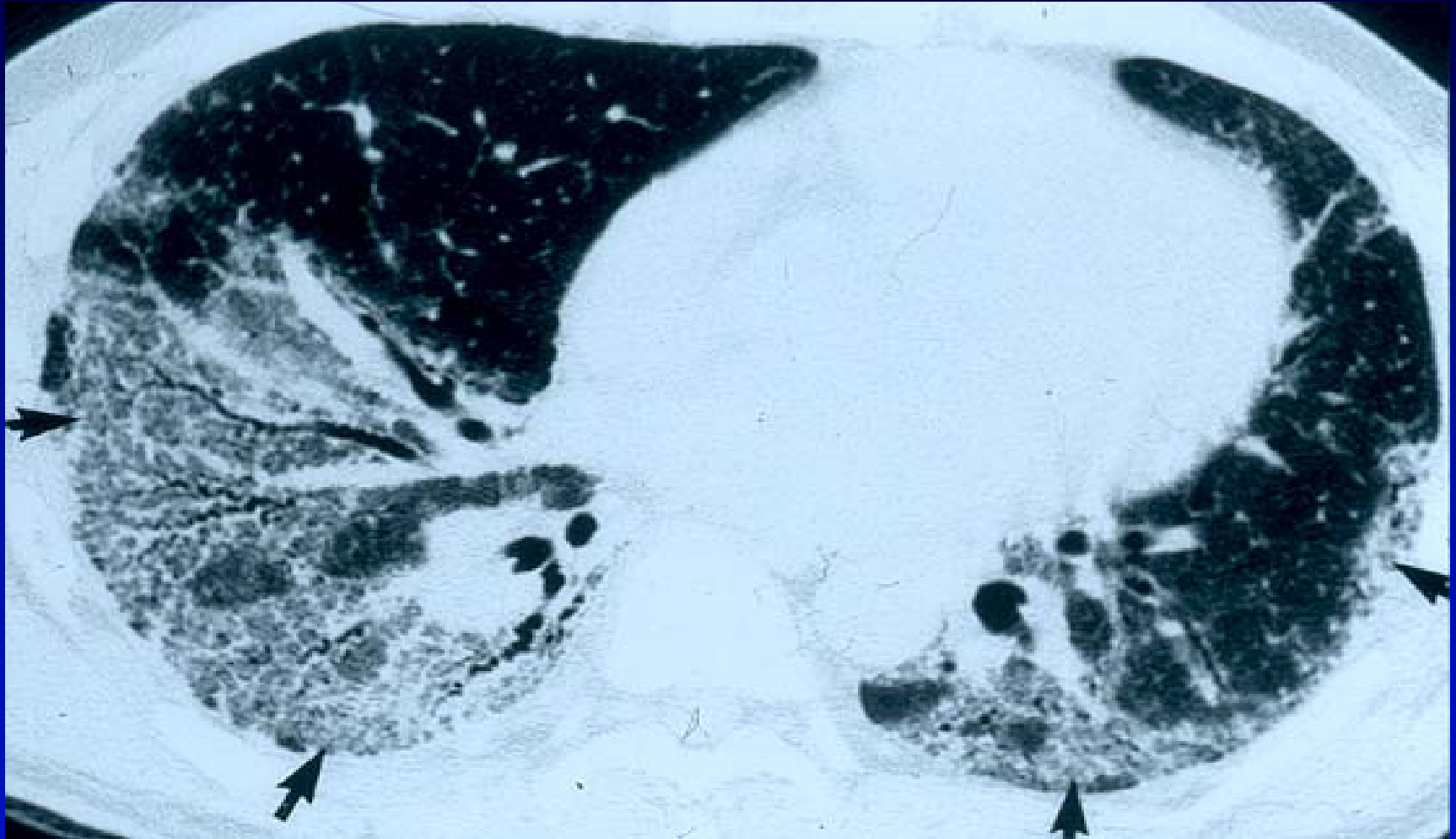
UIP: Honeycomb change; patchy; subpleural

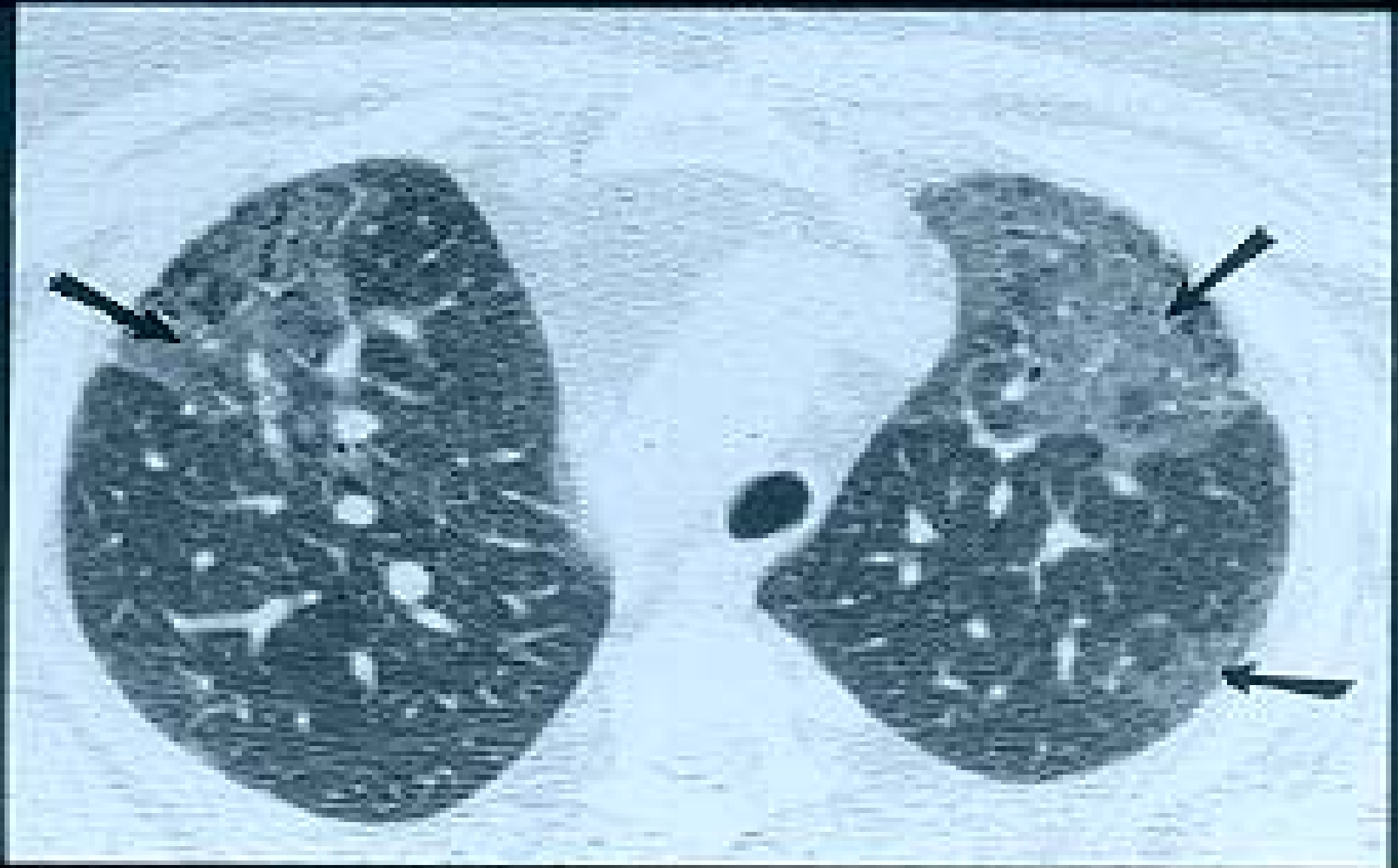


UIP: HRCT Features

- Patchy, heterogeneous
- Lower lobes, subpleural
- Reticular (linear) lines
- Honeycomb cysts
- Ground glass minimal or absent

Coarse GGO: UIP



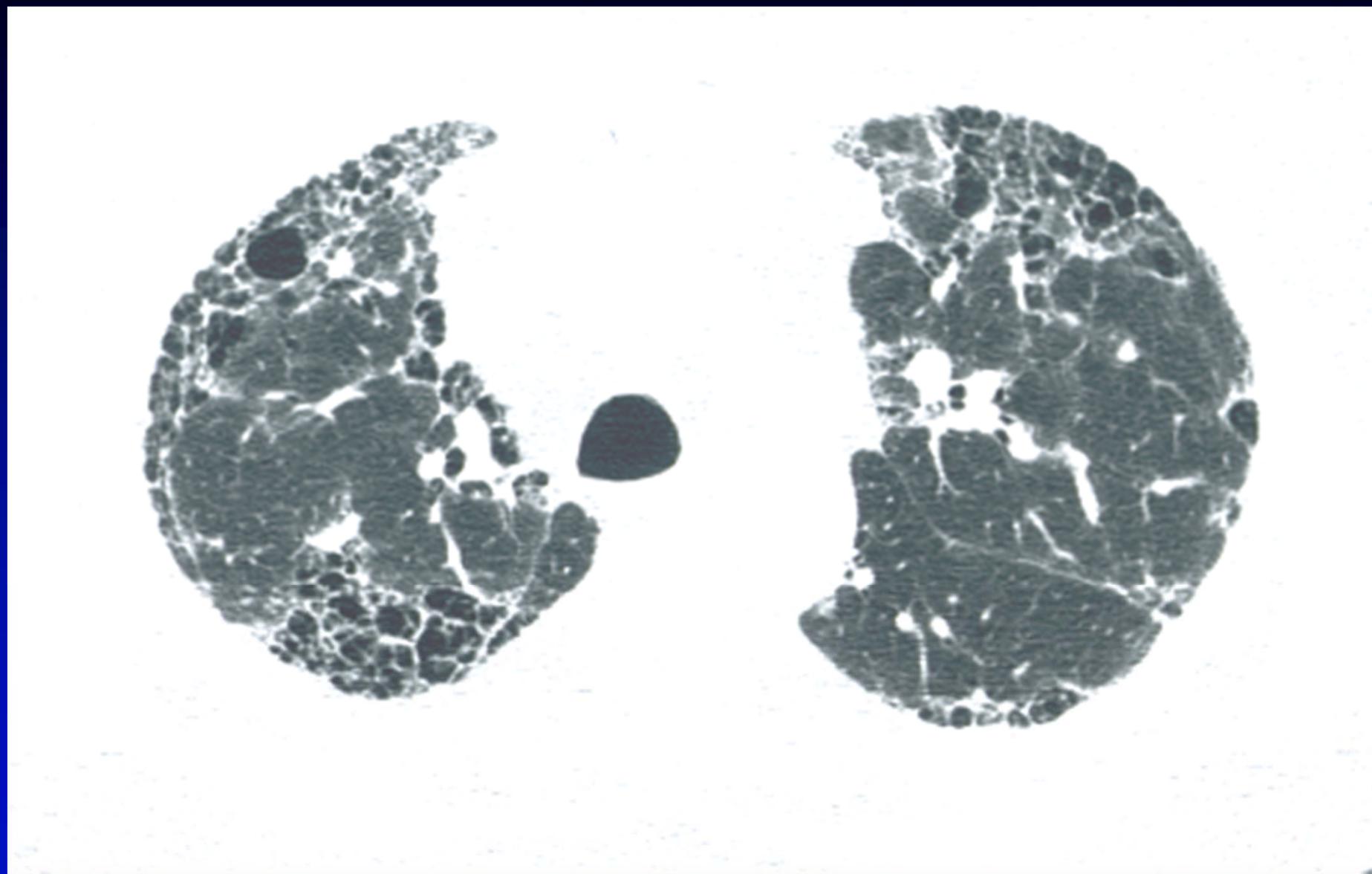


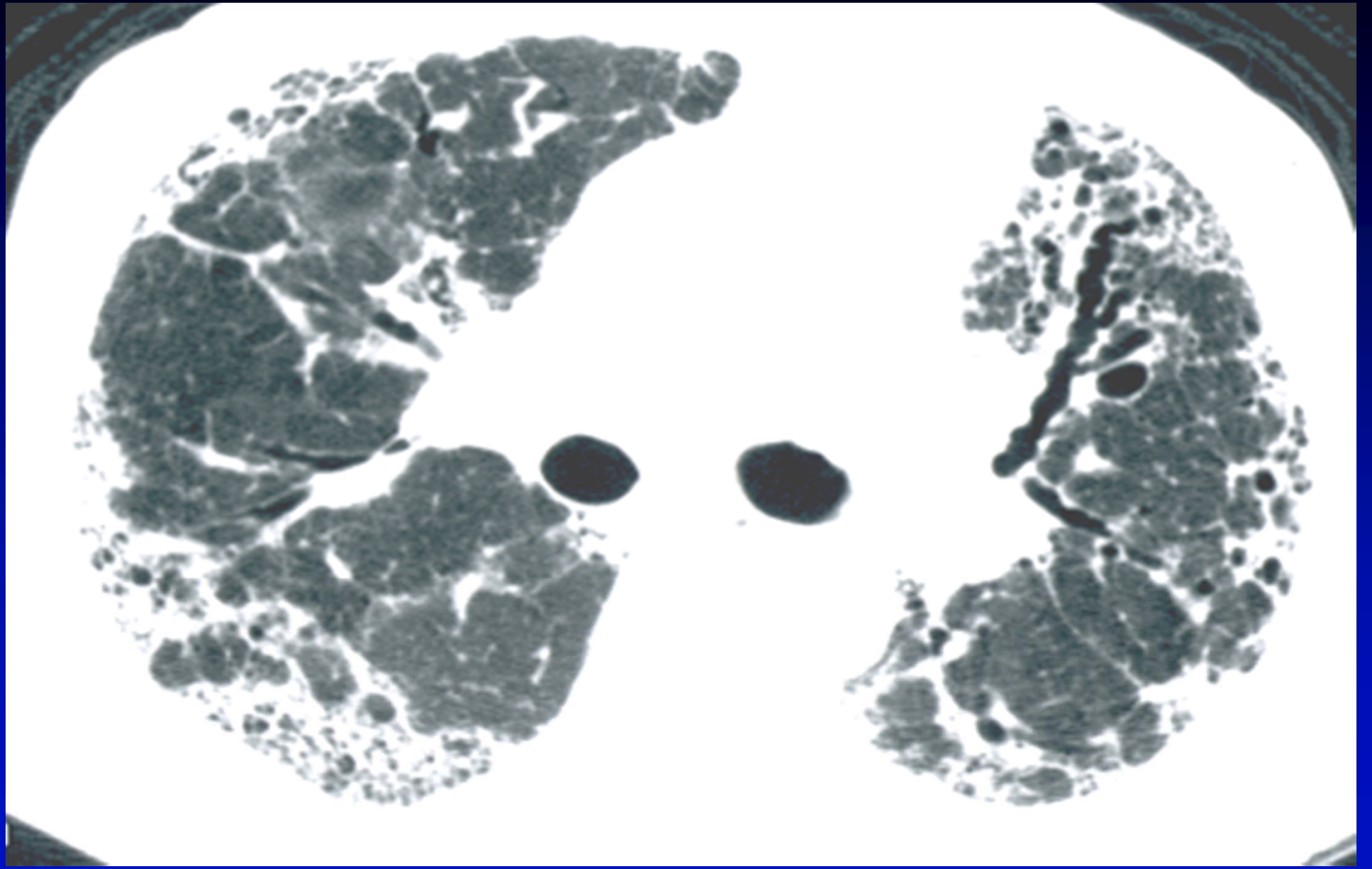


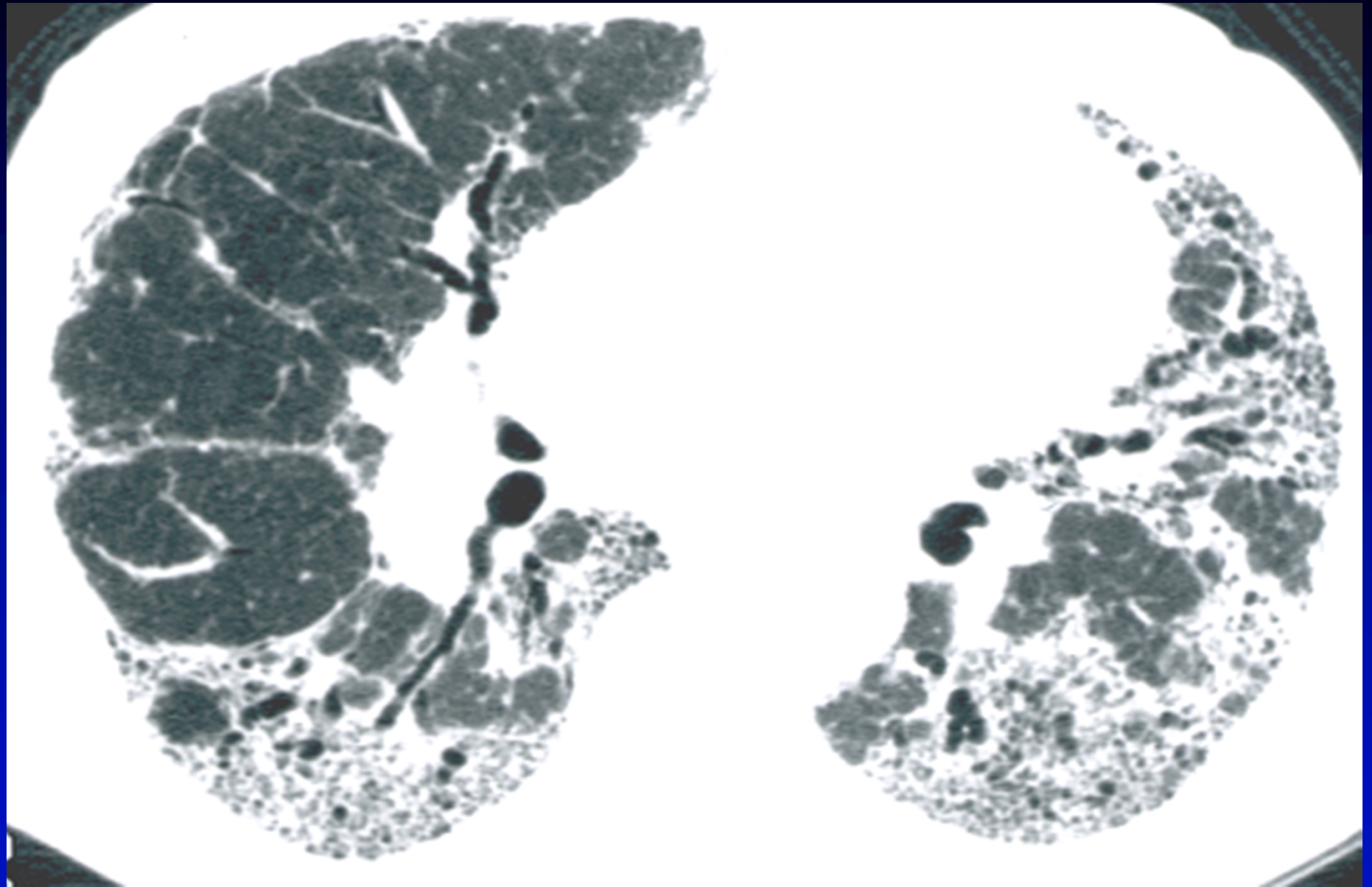
"And now there go the Wilsons! . . . Seems like everyone's evolving except us!"

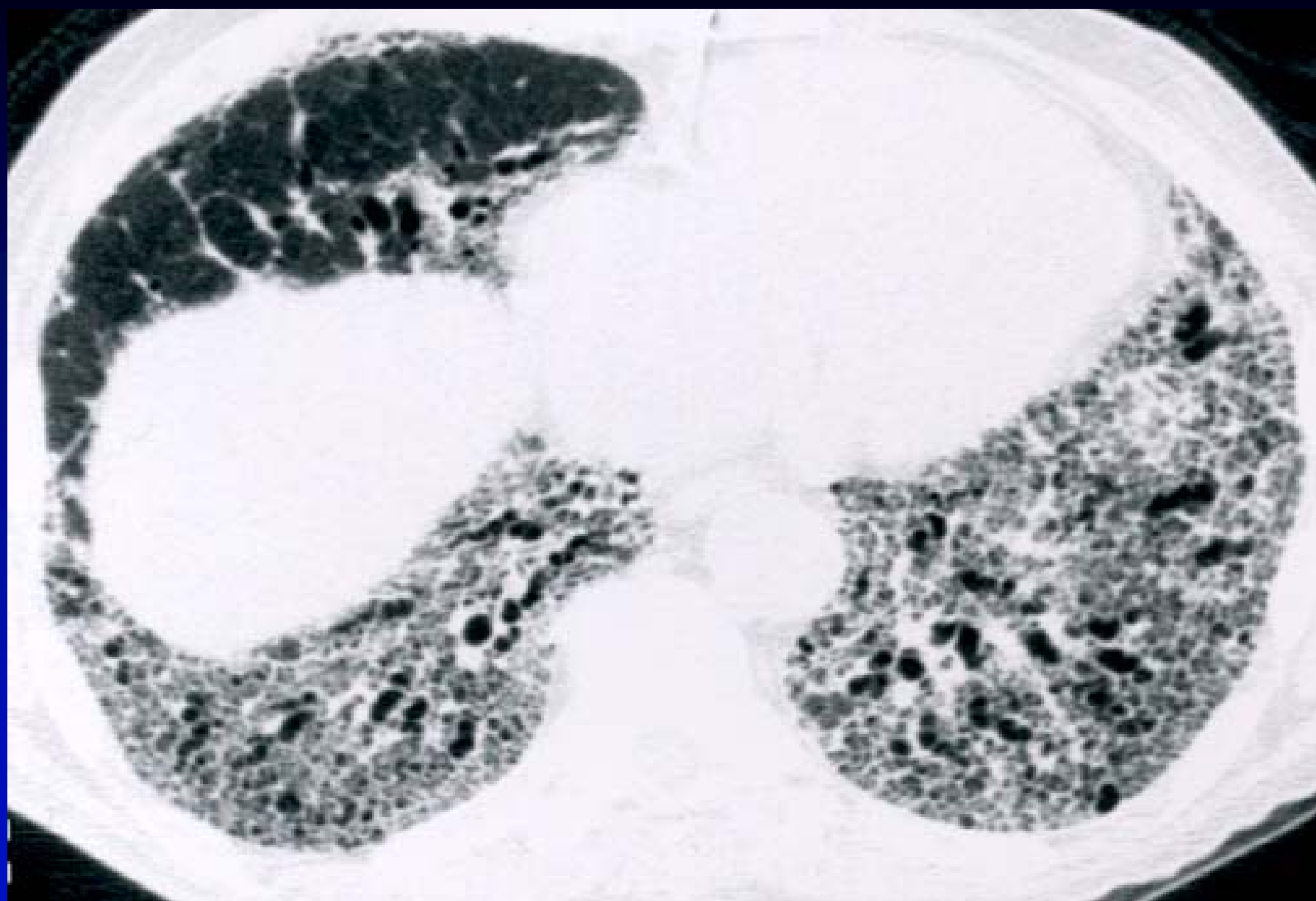
UIP: HRCT Features

- **Proclivity for basilar regions:**
 - **Worsens as descend from upper to lower lobes**









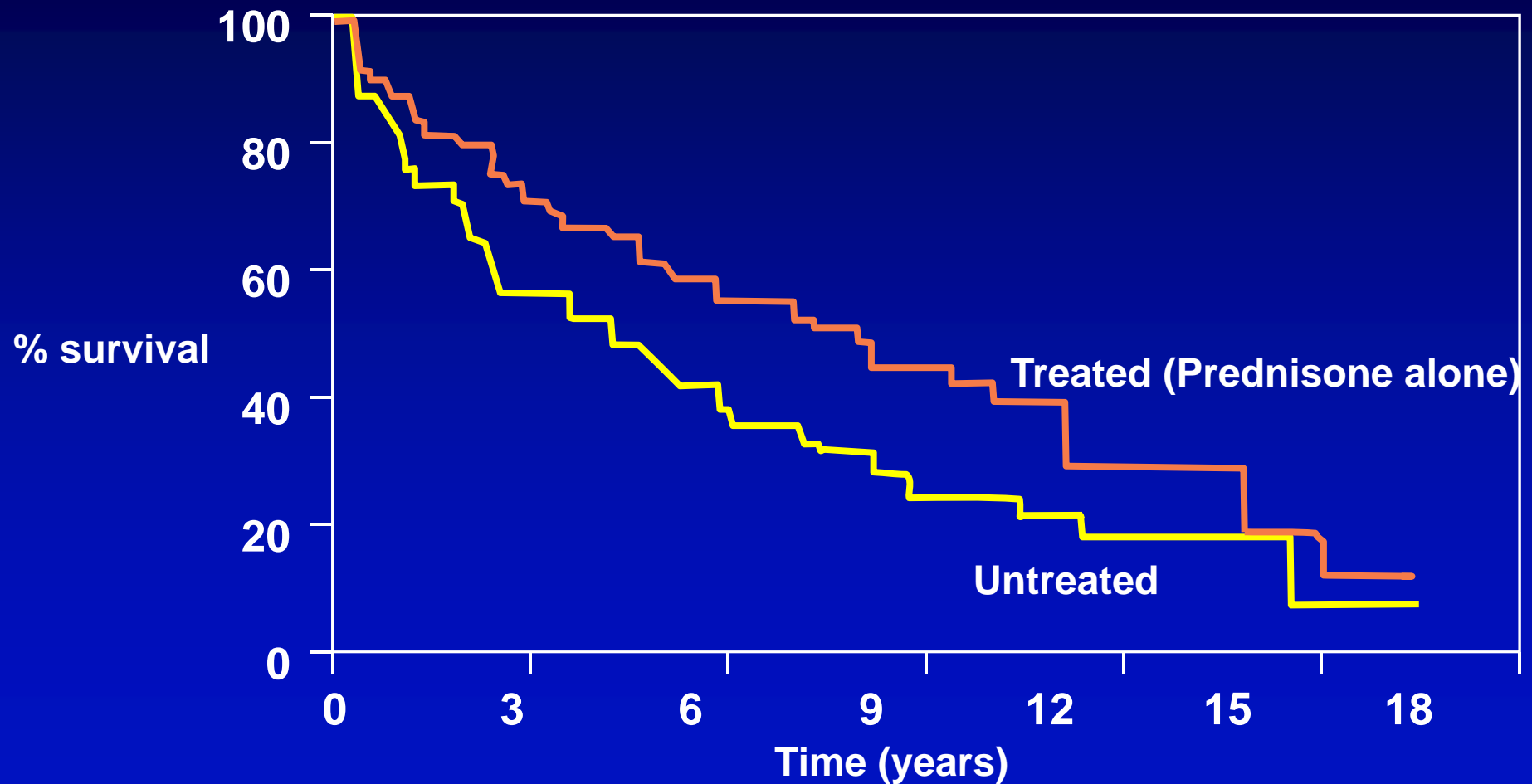
Treatment Modalities

- Non-Pharmaceutical Treatment
- Pharmaceutical Treatment

Pulmonary Rehabilitation

- PR shown to:
 - increase exercise endurance
 - decrease dyspnea
 - improve health related QoL
 - Reduce health care costs?
- Typical course is 8 to 12 weeks, 2-3x per week

Idiopathic Pulmonary Fibrosis: Survival



M. Turner-Warwick, et al. *Thorax*. 1980;35:593-599.

Prednisone:

- 10-15% response
- Optimal Duration and Dose unknown
- Side Effects:
 - Weight gain/body changes
 - Risk of infection
 - Skin breakdown and bruisability
 - Adrenal insufficiency

Failed antifibrotic therapies for IPF

- N-Acetylcysteine (NAC)
- Anti TGF- β therapies
- Lovastatin
- Relaxin
- ACE Inhibitors
- PGE₂
- Leukotriene receptor antagonist
- Endothelin receptor antagonist
- Anti TNF- α Therapies
- Others

Treatment:

The NEW ENGLAND
JOURNAL *of* MEDICINE

ESTABLISHED IN 1812

MAY 29, 2014

VOL. 370 NO. 22

Efficacy and Safety of Nintedanib in Idiopathic
Pulmonary Fibrosis

The NEW ENGLAND JOURNAL *of* MEDICINE

ORIGINAL ARTICLE

A Phase 3 Trial of Pirfenidone in Patients
with Idiopathic Pulmonary Fibrosis

Antifibrotic therapies for IPF

Pirfenidone

- Decreases fibroblast proliferation
- Decreases ECM production
- Inhibits TGF- β collagen synthesis
- Inhibits mitogenic effects of PDGF

Ameliorated fibrosis in a hamster model of bleomycin lung

Lung Transplant

Double lung transplant performed through bilateral anterior thoracotomies.



Native lung with end-stage emphysema



Donor lung after implantation

Lung transplantation

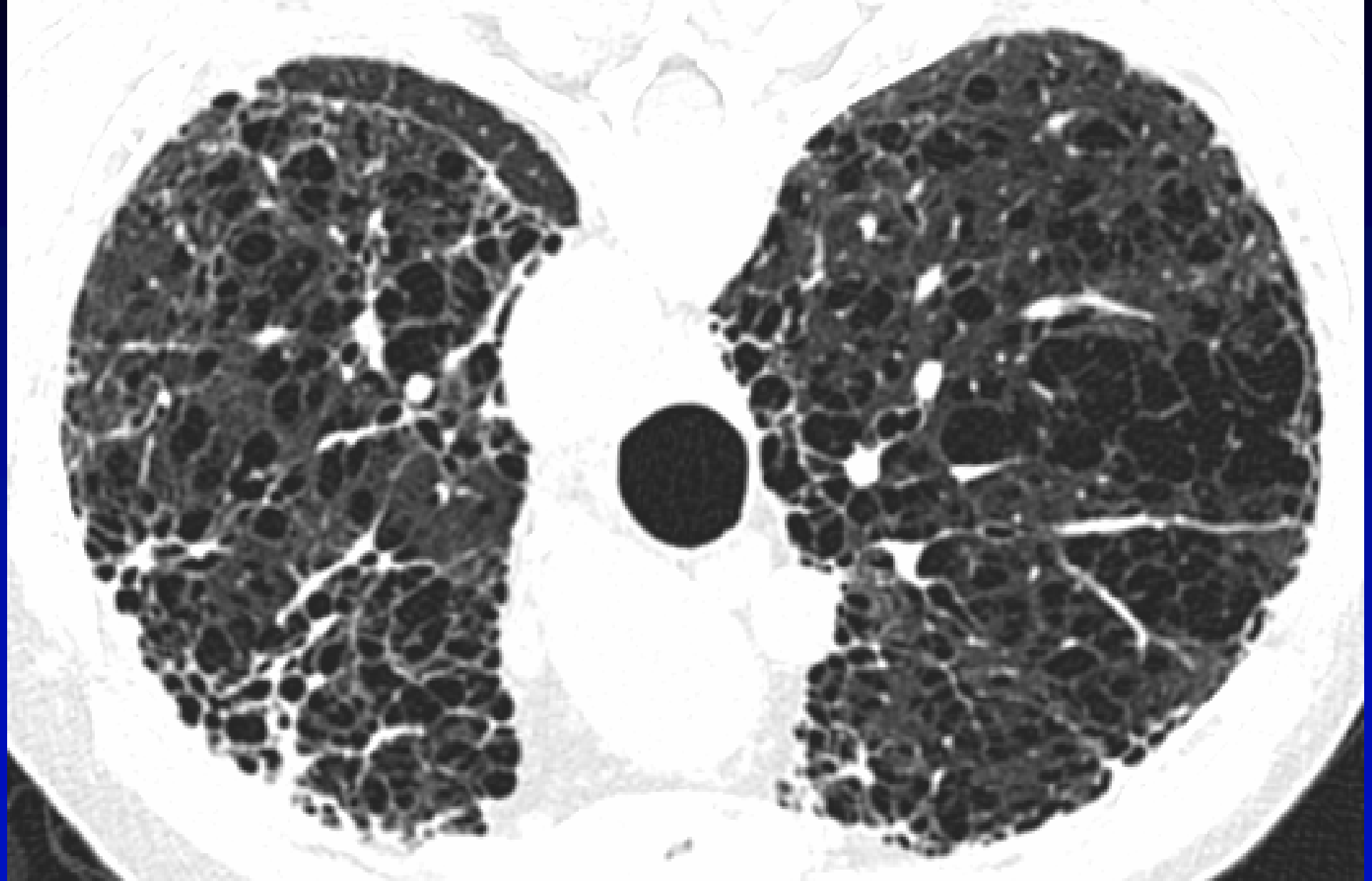
Criteria for listing:

- **Limited life expectancy (< 3 y)**
- **Failure of medical therapy**
- **Age \leq 65 yrs (moving to 75yo)**
- **No extrapulmonary organ failure**

Pulmonary Fibrosis Transplant Listing

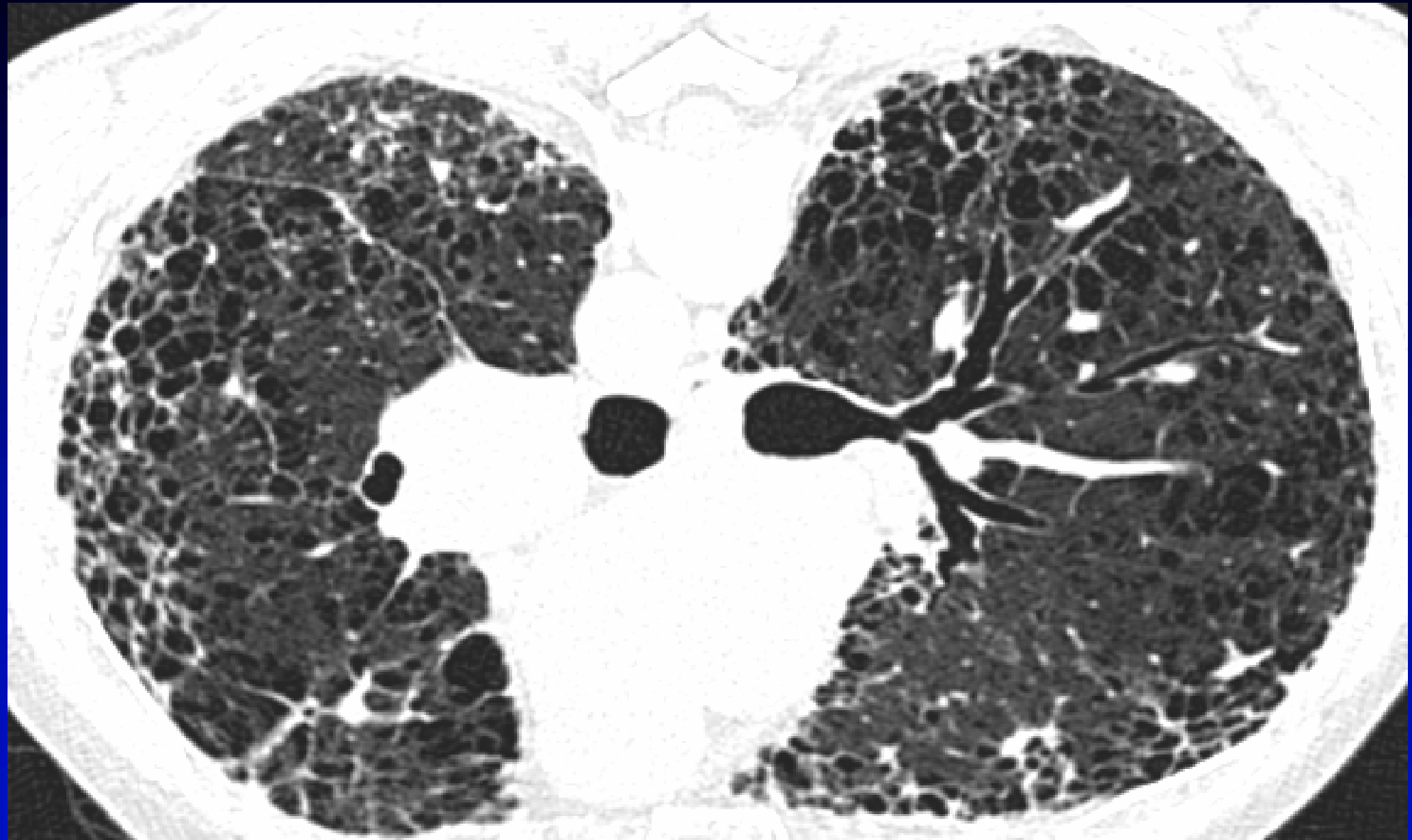
- Oxygen Dependent
- Failure of medical therapy
- FVC < 60% and/or Dlco < 40
- Pulmonary Hypertension

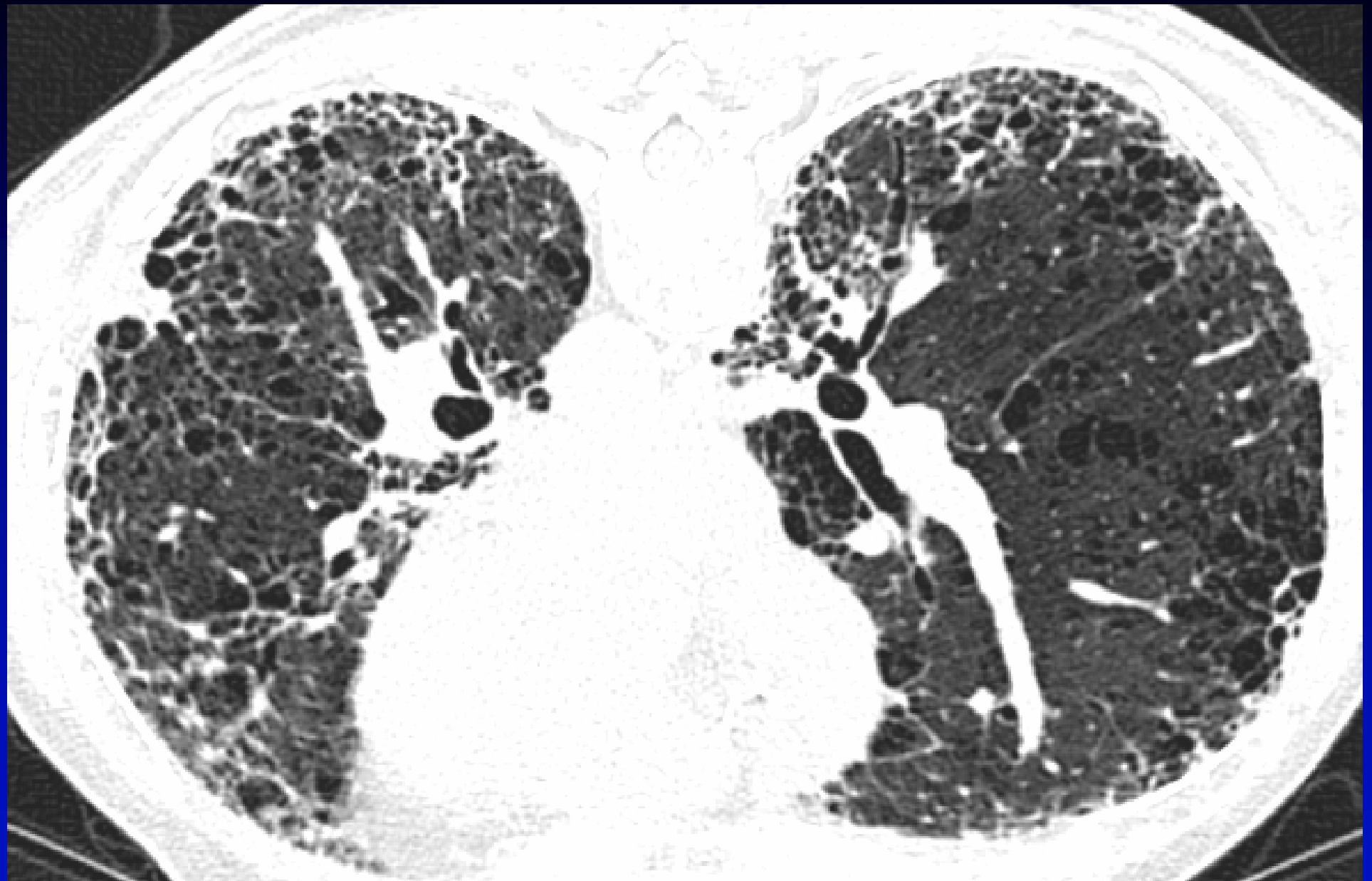
- **Coexistent Emphysema and IPF**
- **Combined PF and Emphysema
(CPFE)**

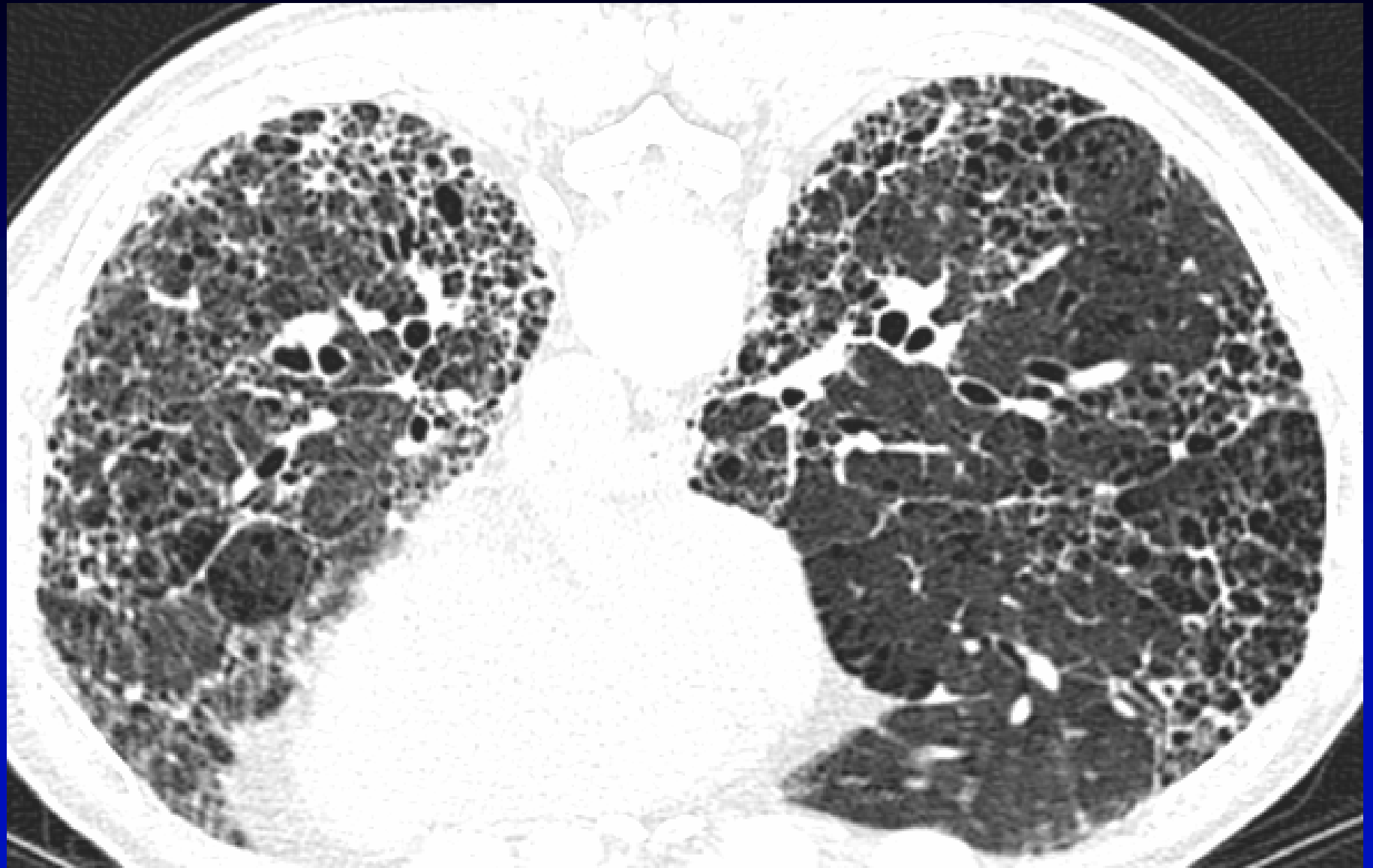




[A]







Idiopathic Interstitial Pneumonias

Syndromes distinct from IPF/UIP:

- Desquamative Interstitial Pneumonia (DIP)
- Respiratory Bronchiolitis ILD (RBILD)
- Nonspecific Interstitial Pneumonia (NSIP)

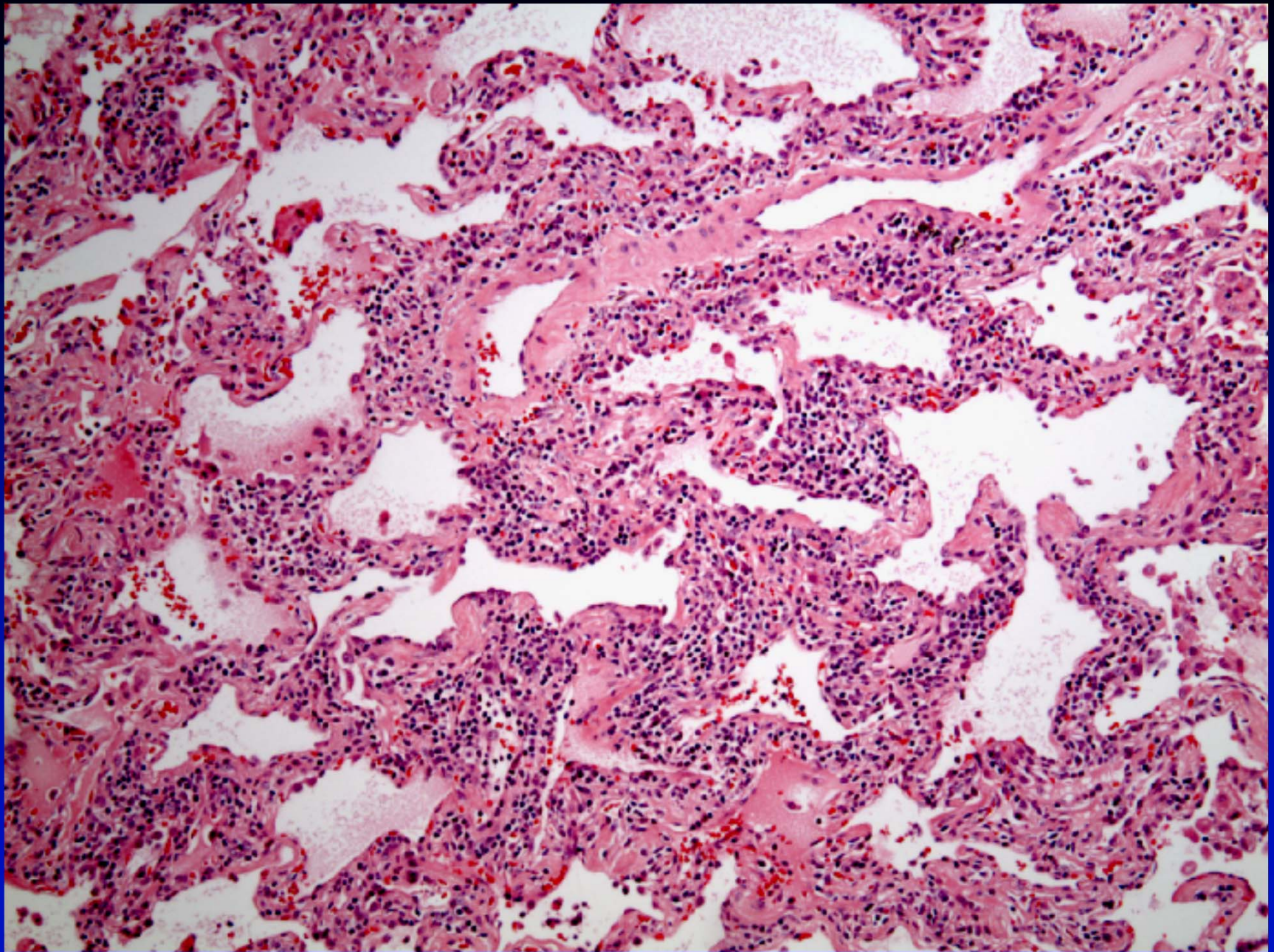
Acute interstitial pneumonia (A.I.P.)

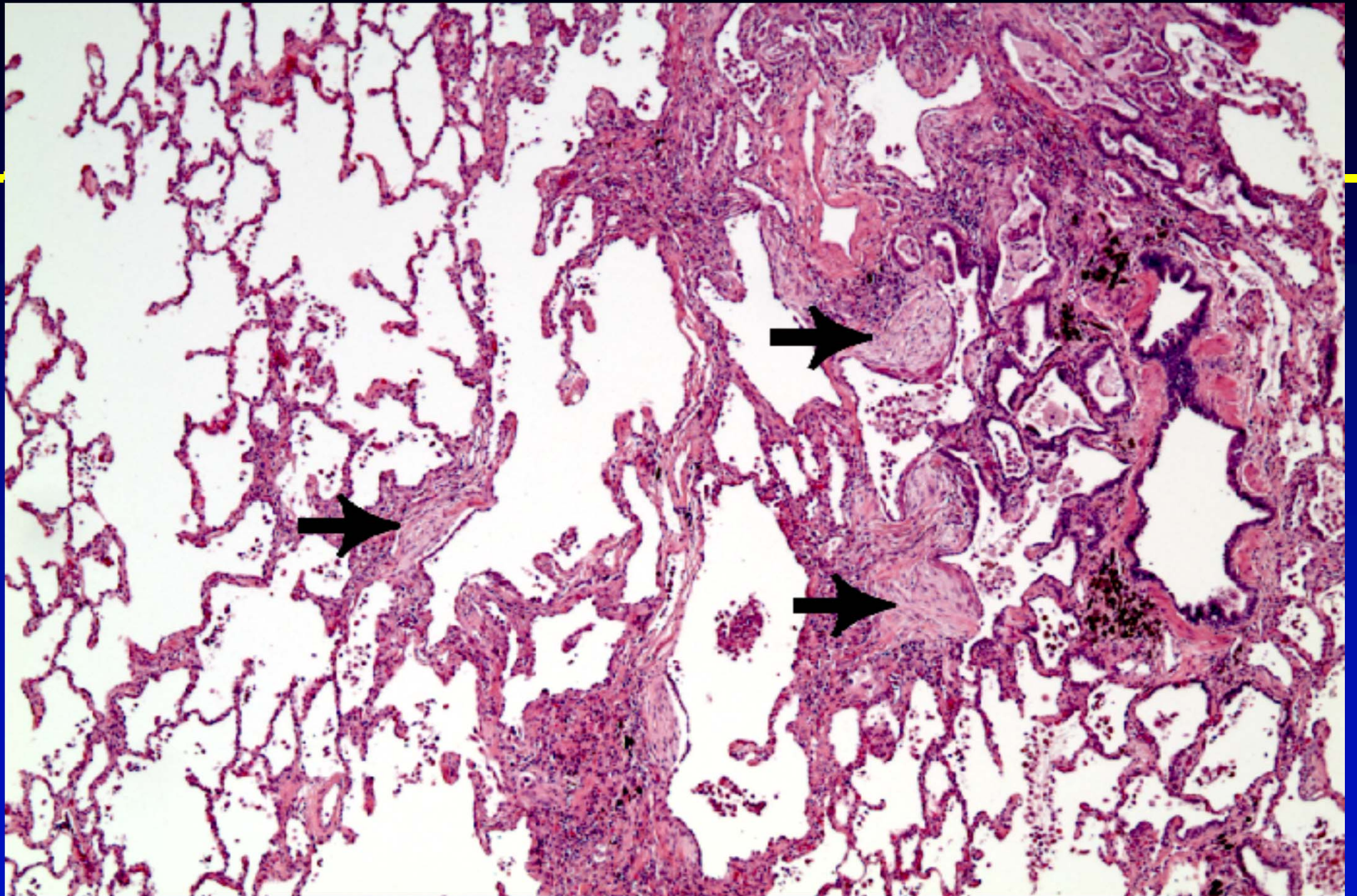
- Subacute, progresses to ARDS
- Diffuse lung damage (DAD)
- **Ground glass opacities on HRCT**
- May respond to IV pulse steroids

Nonspecific interstitial pneumonia

Histological criteria (NSIP):

- Temporal homogeneity
(lesions of same age)
- Lacks features of other CILDs
(UIP, DIP/RBILD, AIP)





Nonspecific interstitial pneumonia

- **Comprises 13-36% of idiopathic interstitial pneumonias from retrospective studies (1998-2001)**

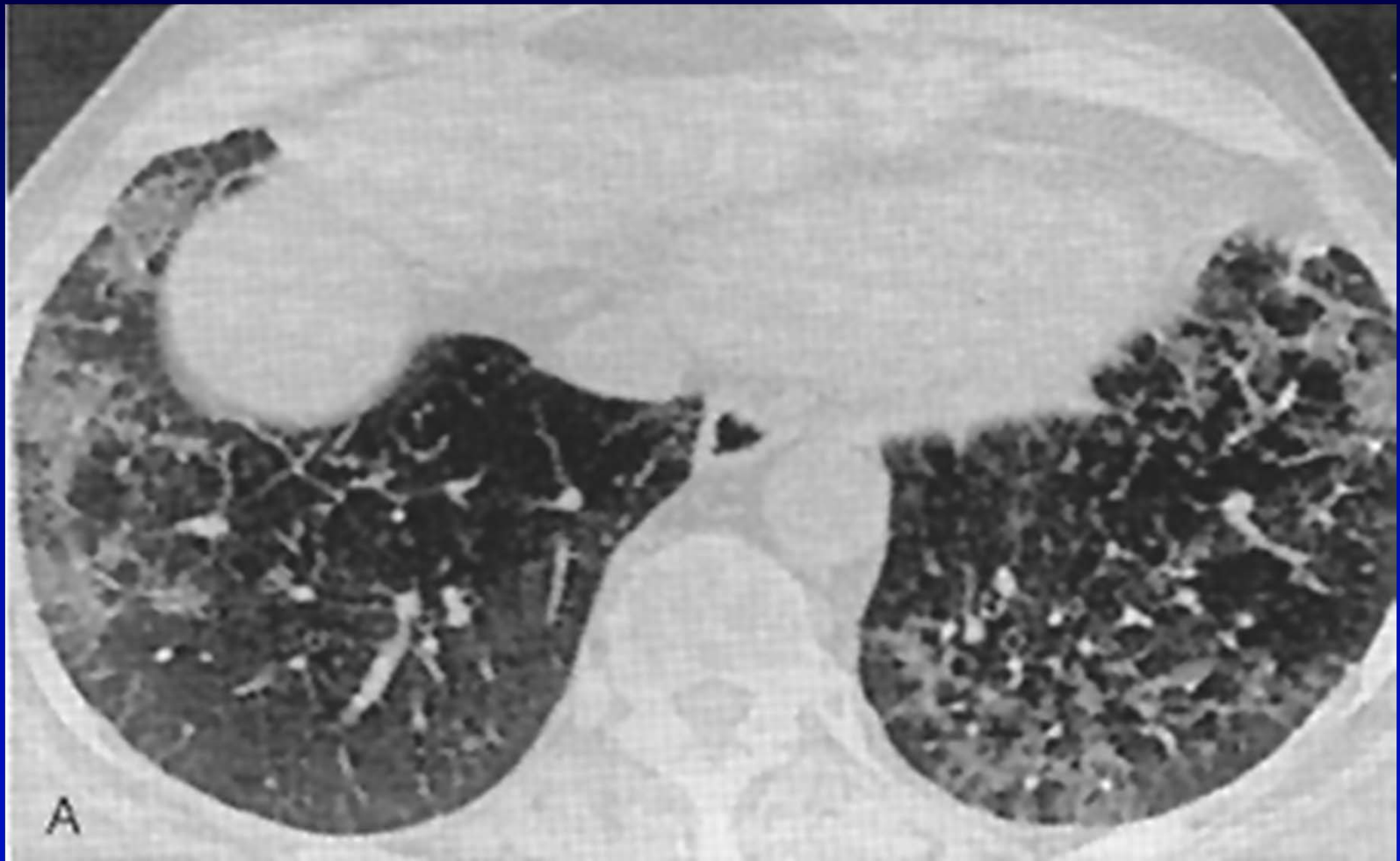
Nonspecific interstitial pneumonia

HRCT features (n=23):

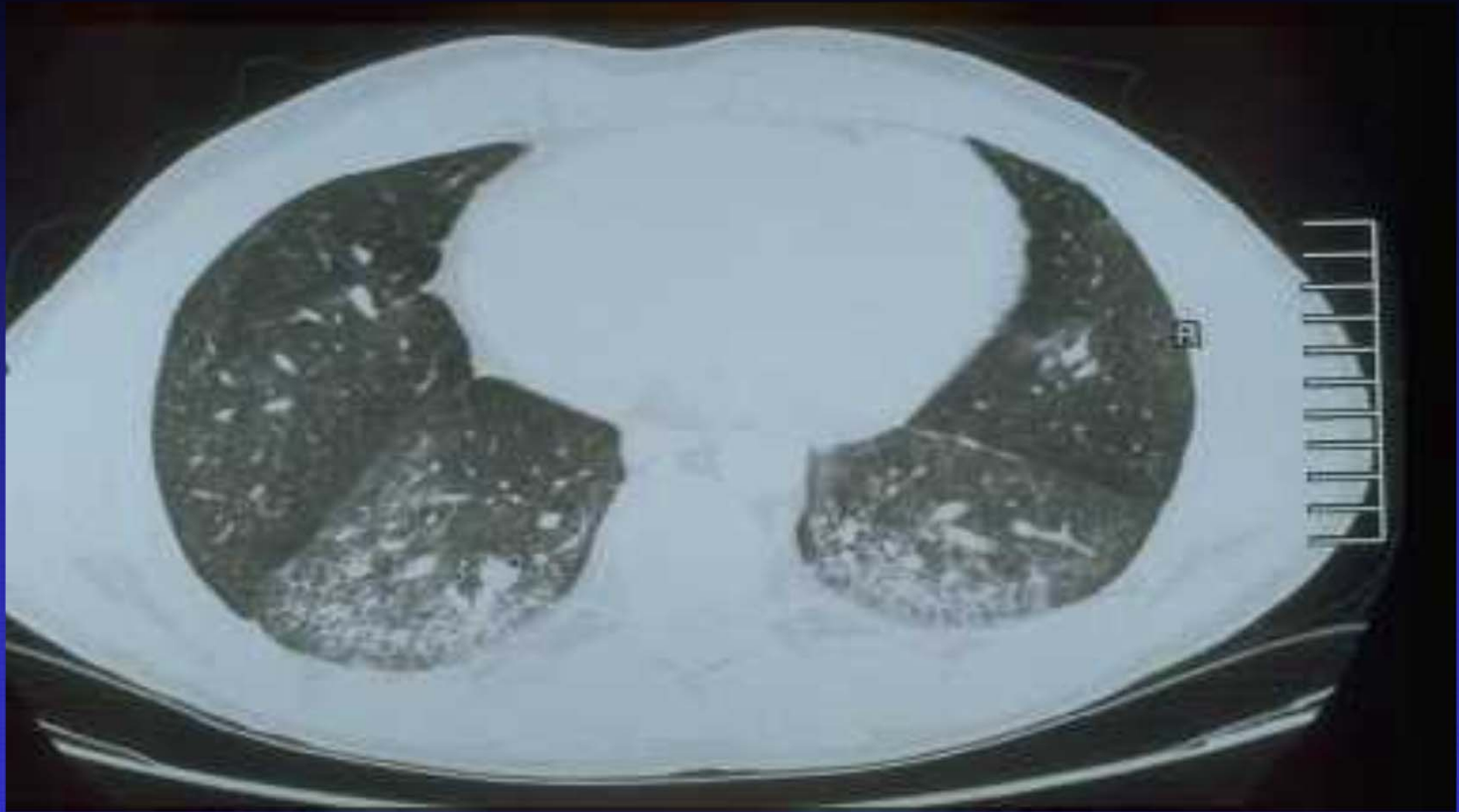
- Honeycombing not found
 - **Bilateral patchy ground glass**
 - Irregular linear opacities
-

Kim, *AJR* 1998;171:1645

Nonspecific interstitial pneumonia



Nonspecific interstitial pneumonia











[4]

Idiopathic interstitial pneumonias

Compared to UIP, other idiopathic interstitial pneumonias show:

- Better response to steroids
- Improved survival

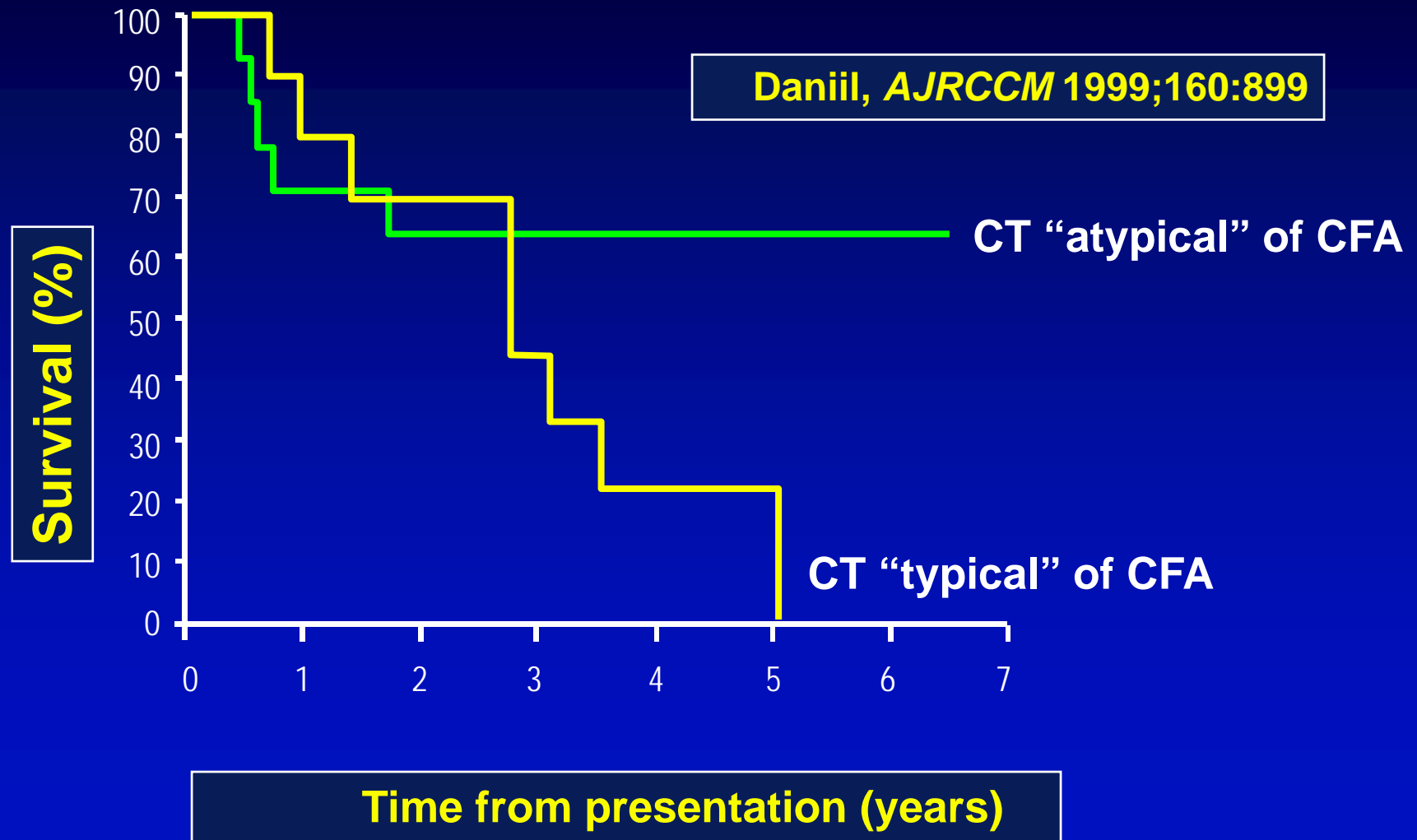
Idiopathic pulmonary fibrosis

Survival worse if:

- HRCT “typical” of IPF
 - UIP on surgical lung biopsy
-

Daniil, *AJRCCM* 1999;160;899

HRCT appearance vs survival



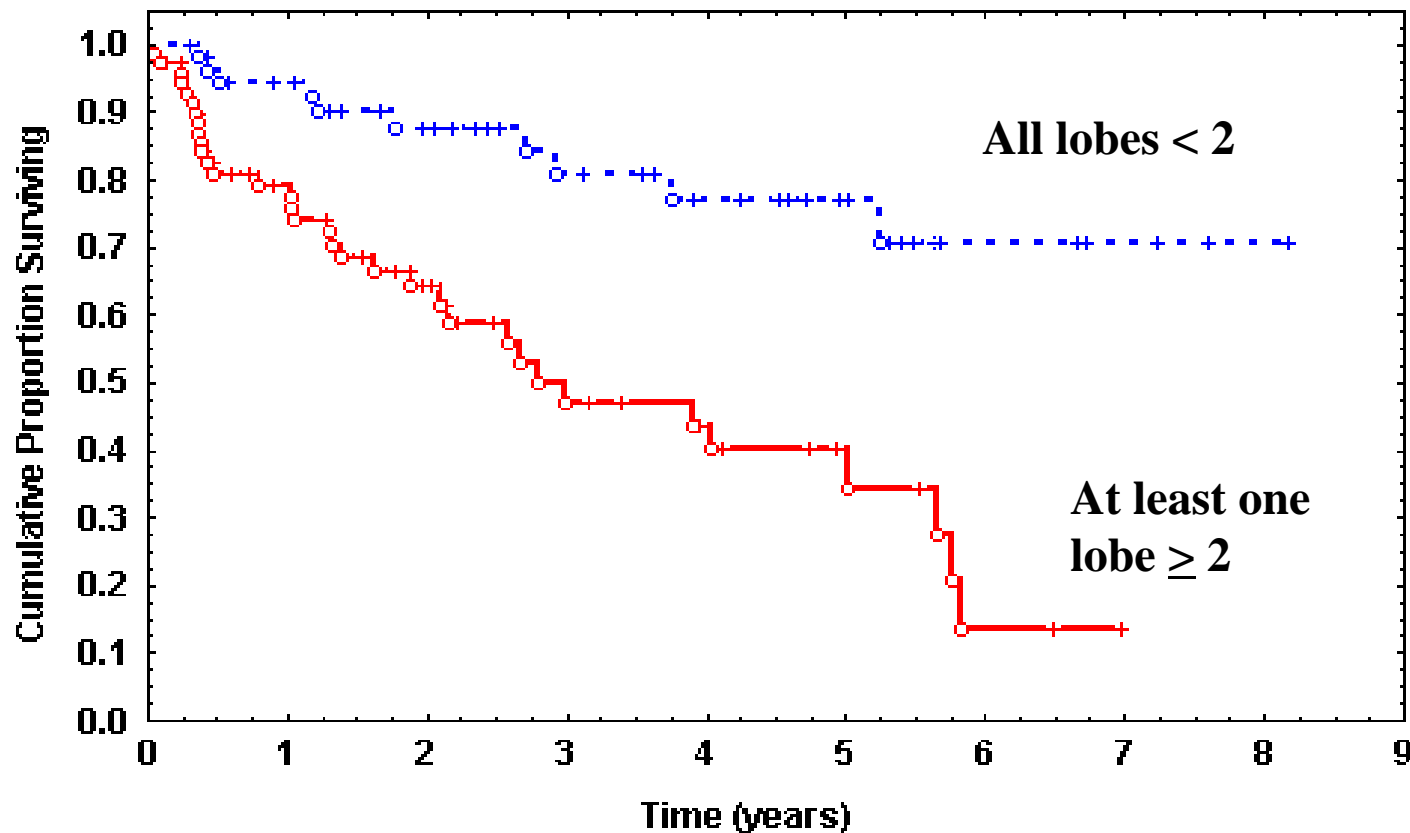
Idiopathic pulmonary fibrosis

**“Classical” CT scan for “IPF”
correlates with diagnosis of
UIP and high mortality**

HRCT-fib ≥ 2 predicted worse survival

I.I.P.s

Flaherty *Eur Respir J* 2002;19;275



Significance of CT pattern

- Extensive ground glass opacities suggests diagnosis *other than UIP*
 - Diagnosis of UIP can be assumed if *classical* CT features are present
-

Ground Glass Patterns (HRCT)

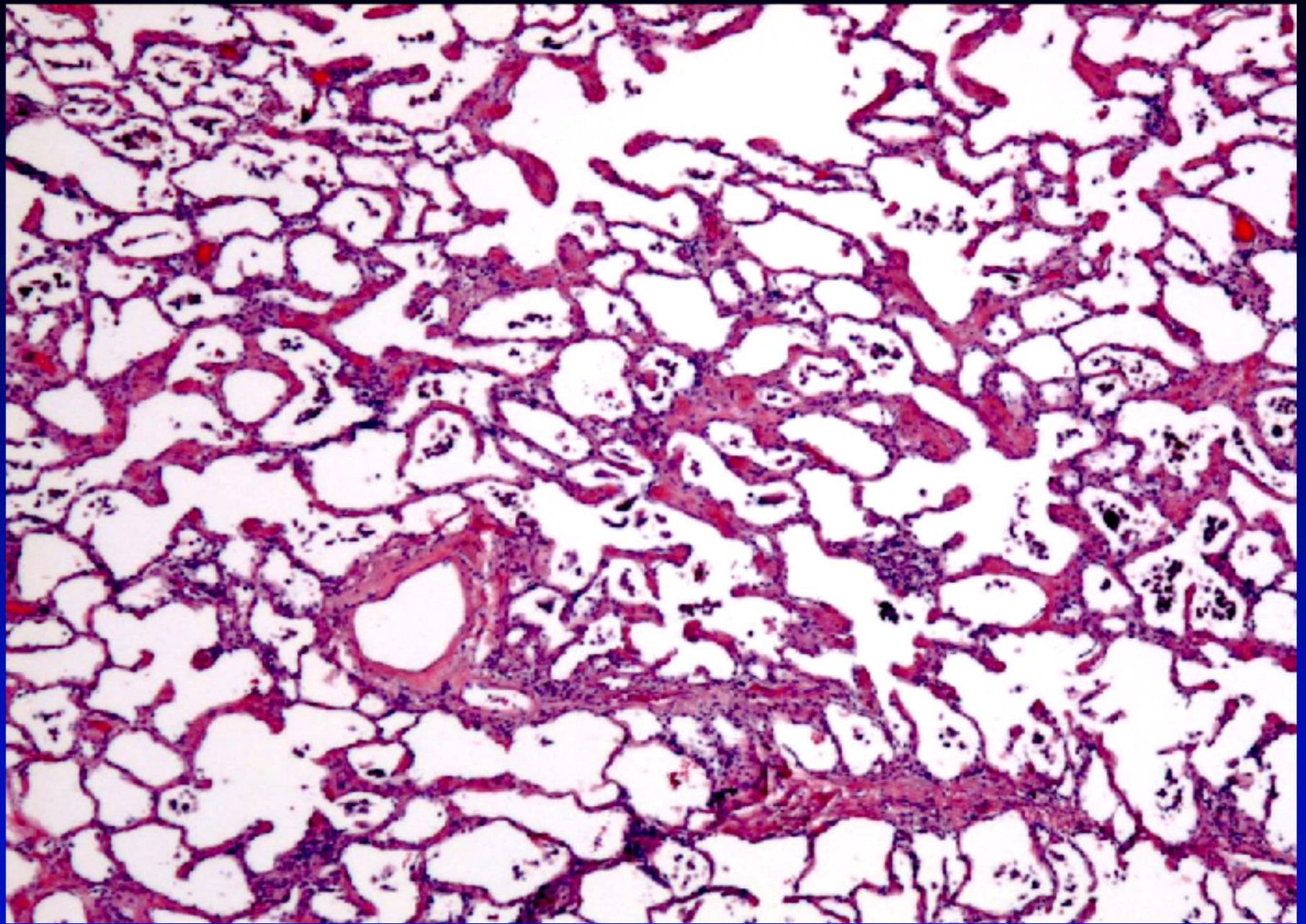
- Idiopathic interstitial pneumonias *other than UIP* (e.g., NSIP, DIP, AIP, LIP)
- Hypersensitivity pneumonia
- Cryptogenic organizing pneumonia
- Pulmonary Alveolar Proteinosis

Collagen vascular diseases

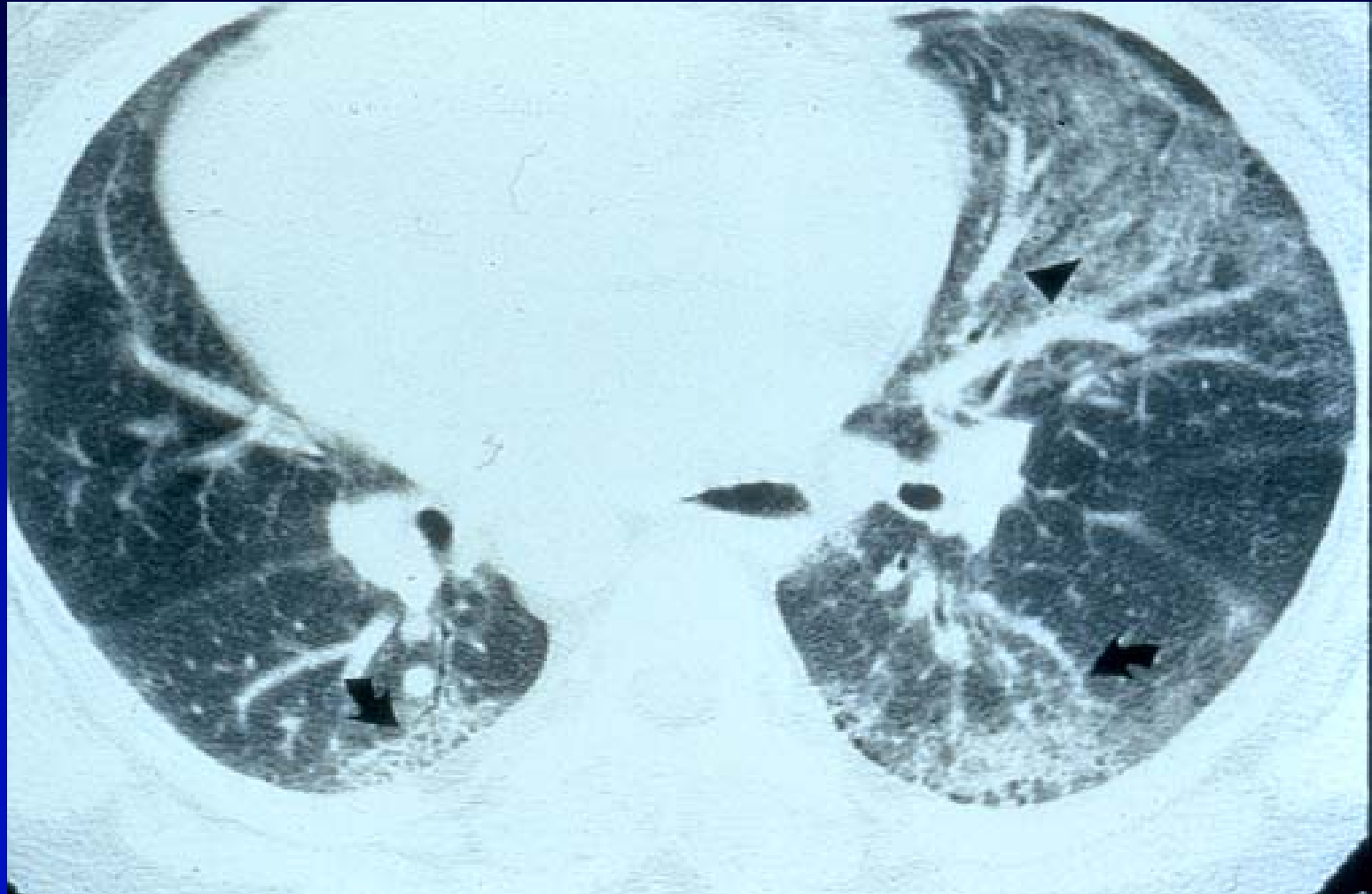
- Pulmonary fibrosis can be different or resembles IPF
- Course more indolent in CVD

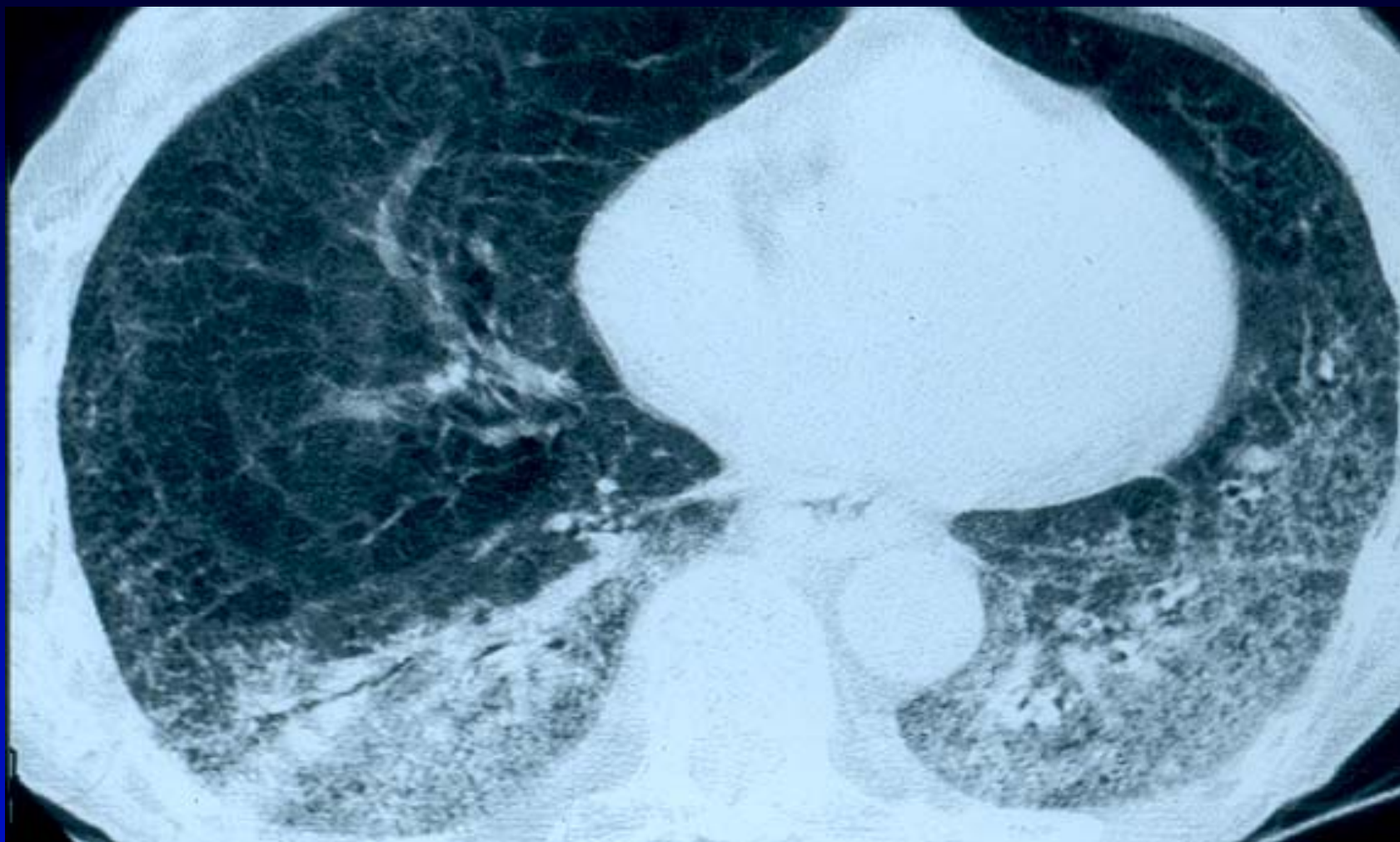
Collagen vascular diseases

- **NSIP most common pattern in CVD-associated ILD**
- **Other patterns: LIP; BOOP; UIP; constrictive bronchiolitis**

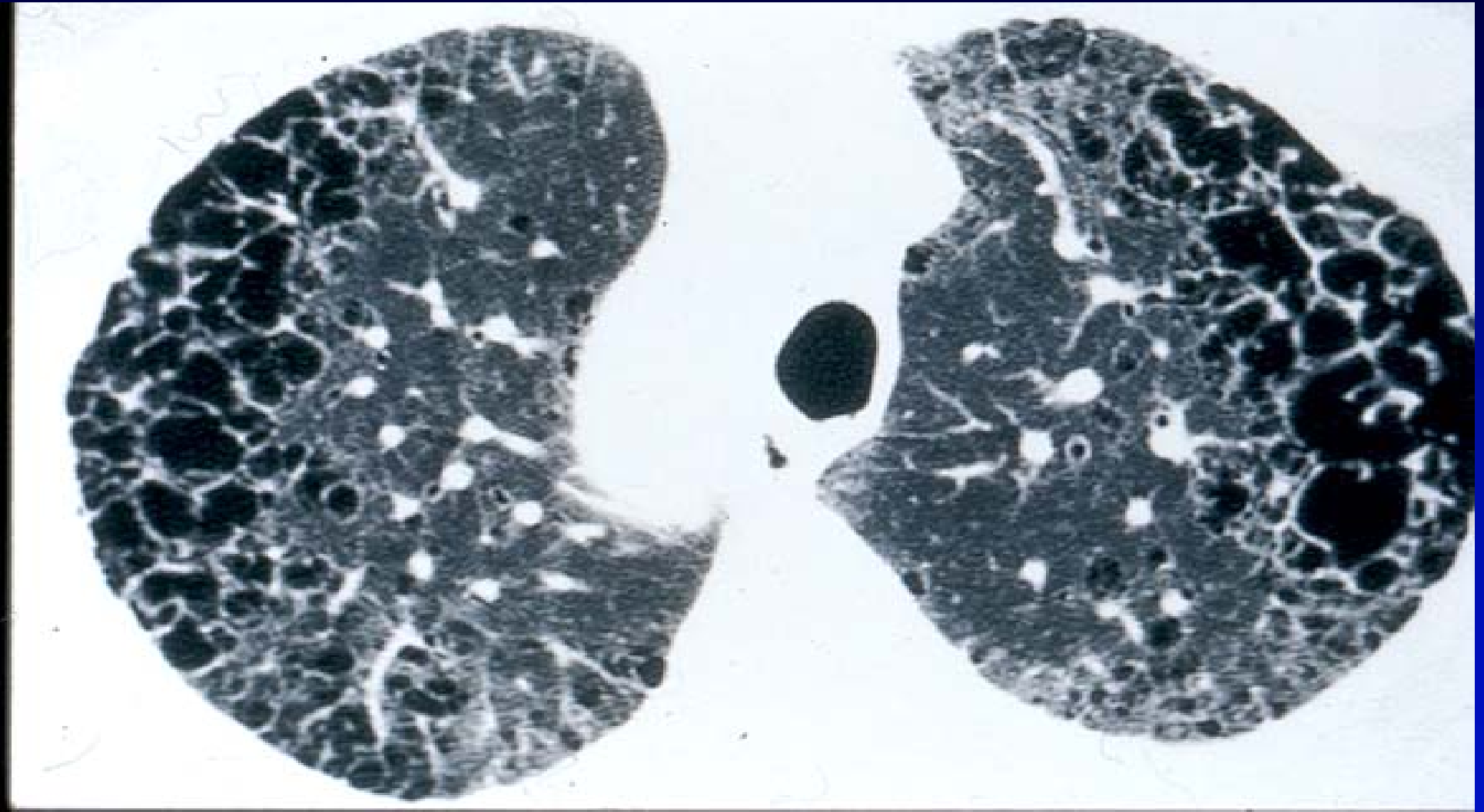


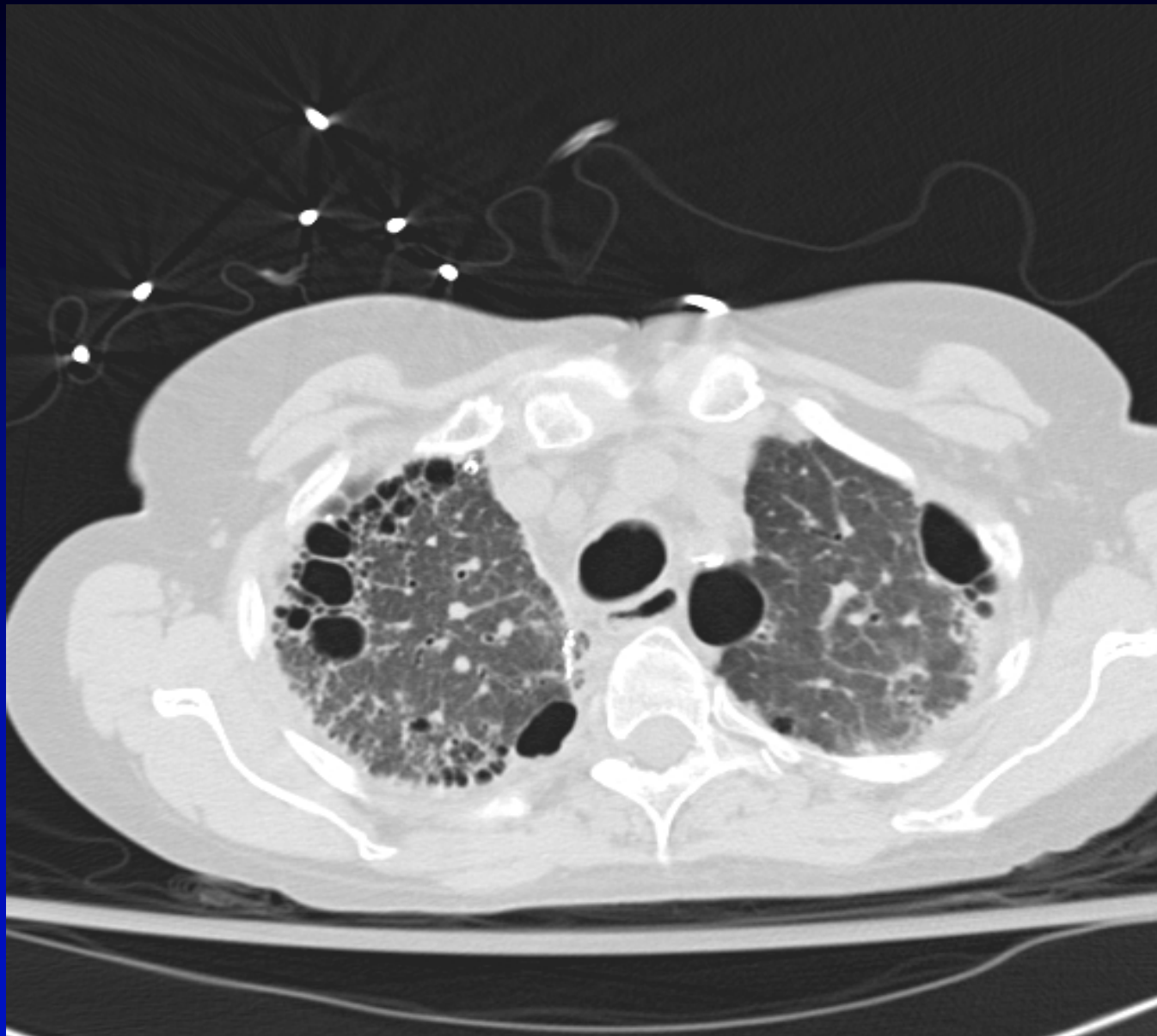
Scleroderma (PSS)

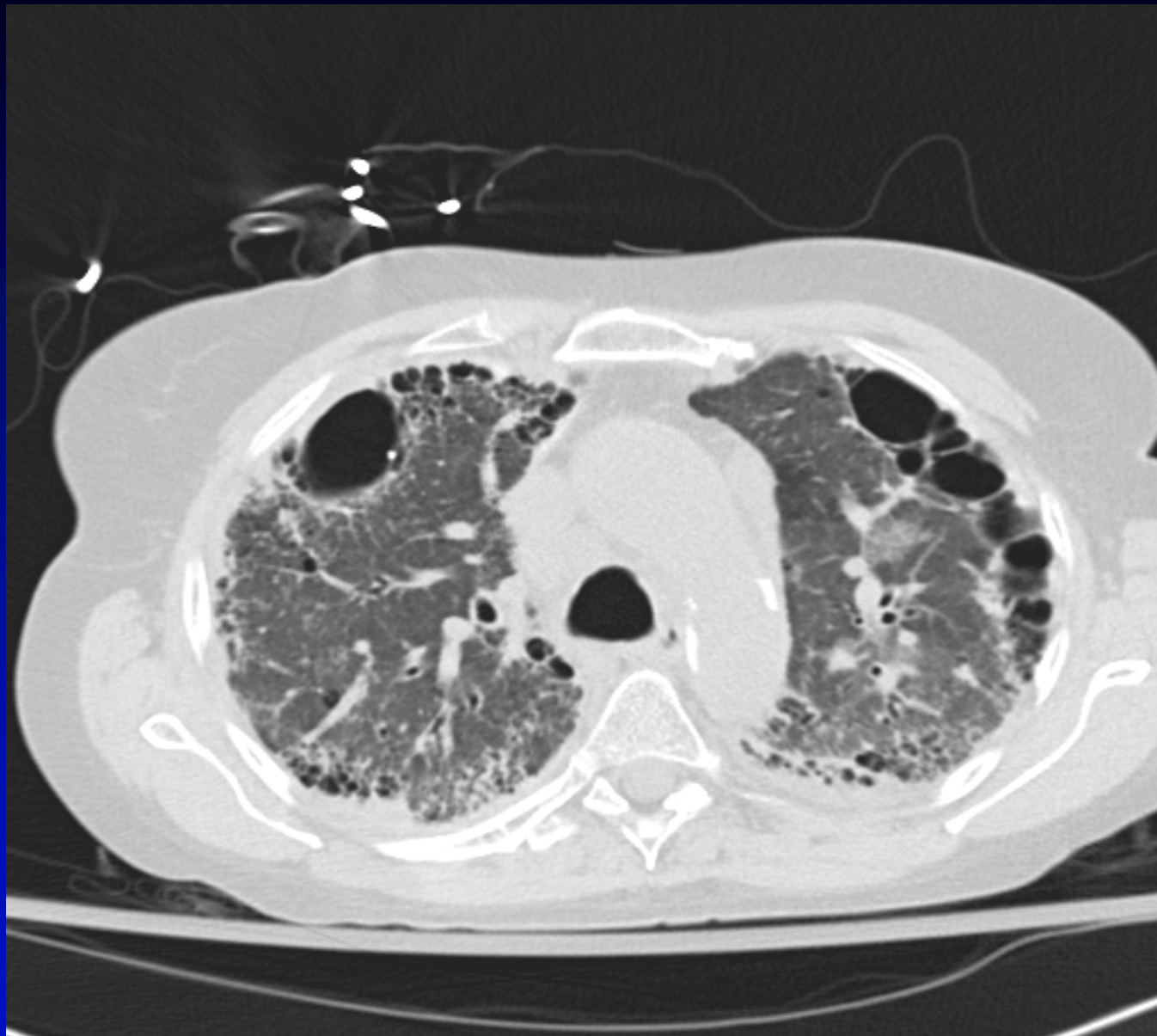


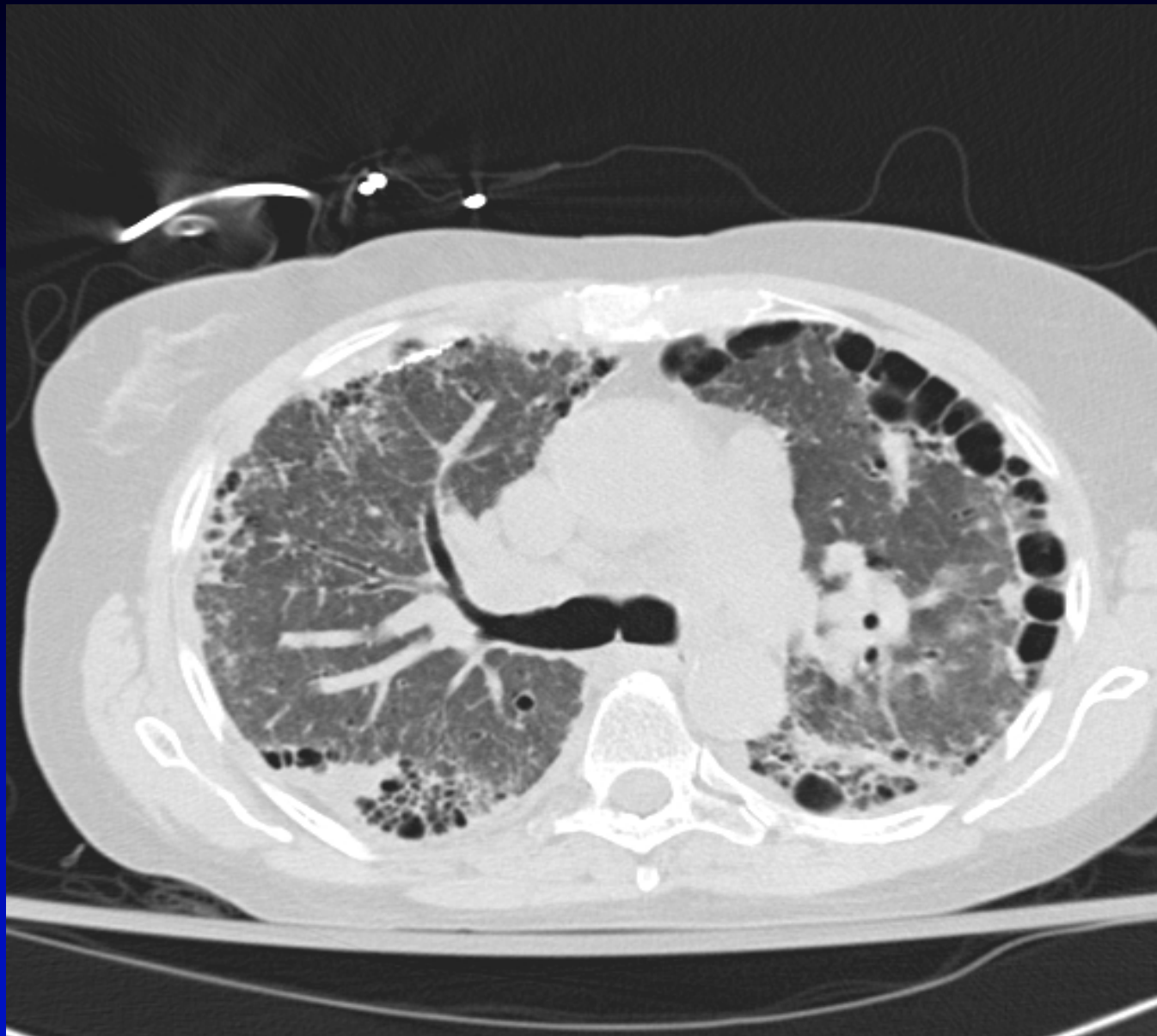


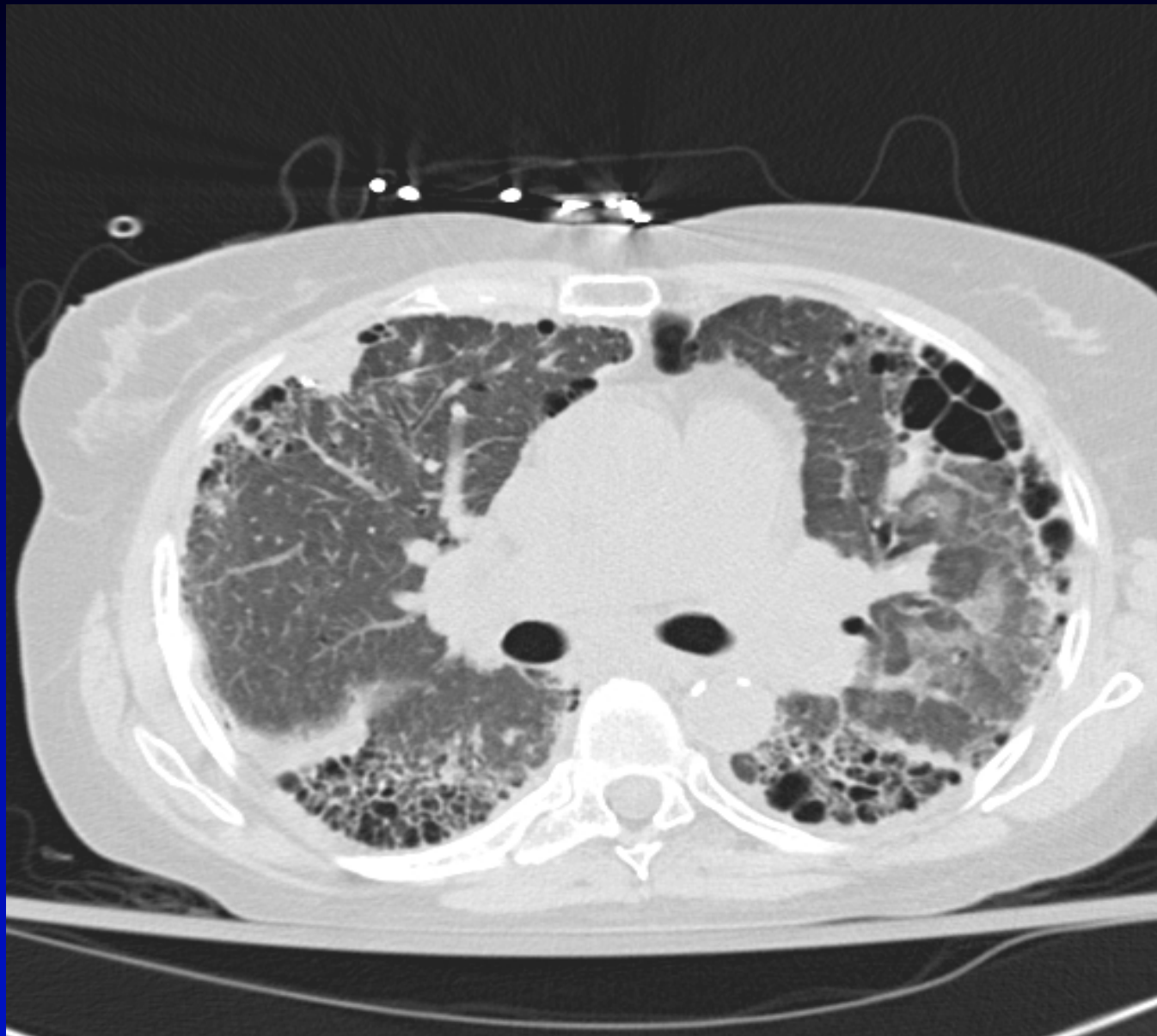
Honeycomb cysts (MCTD)

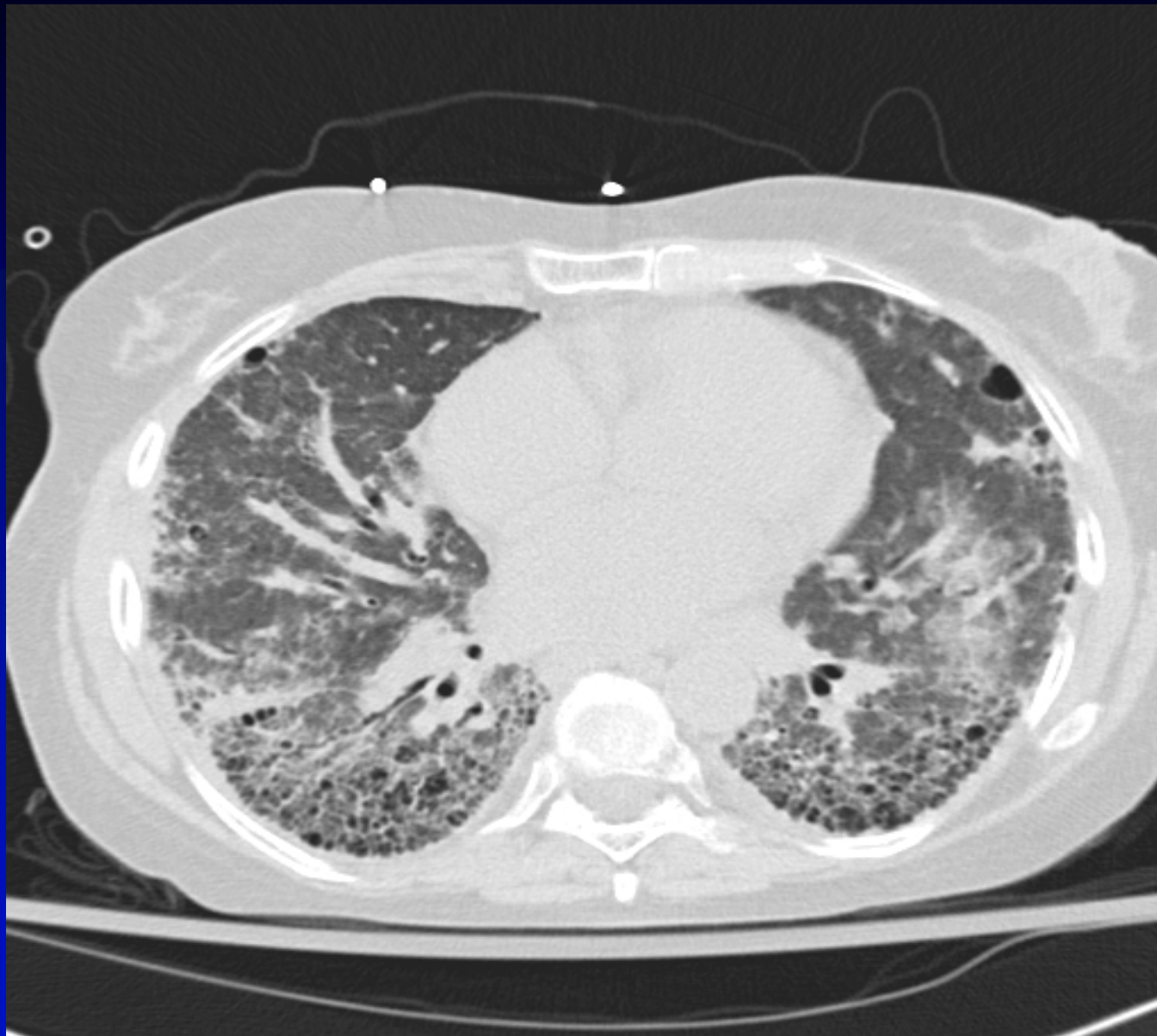


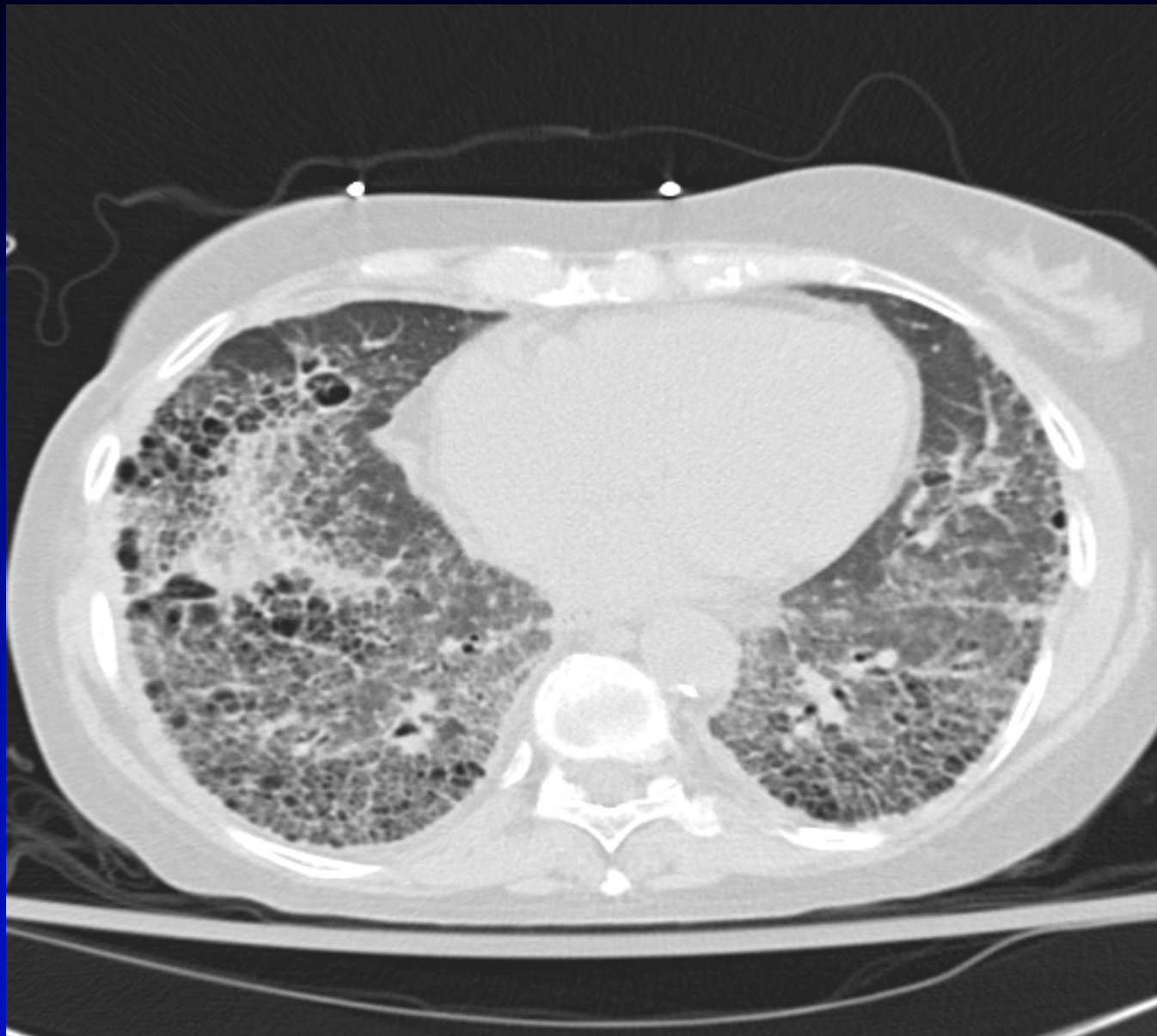




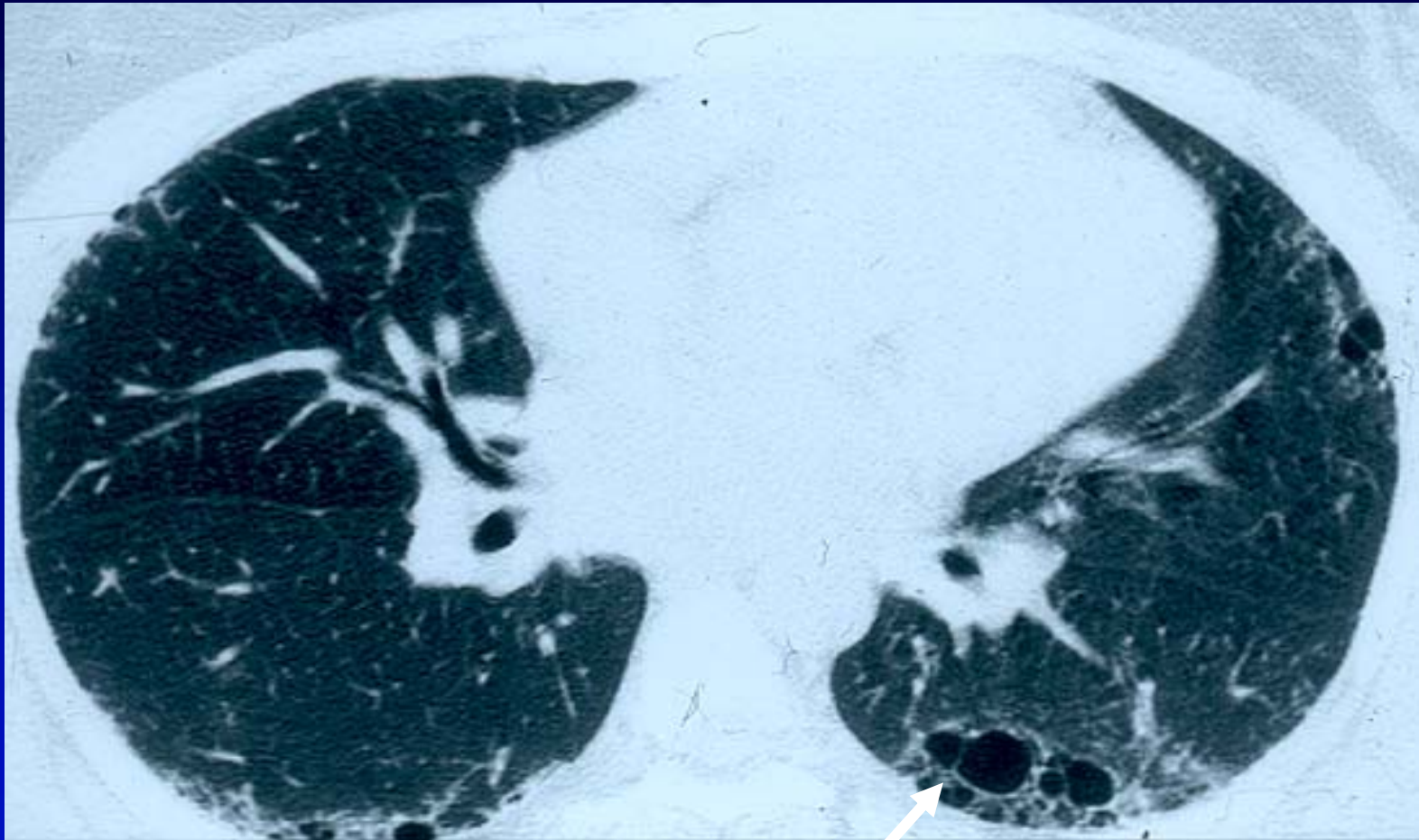


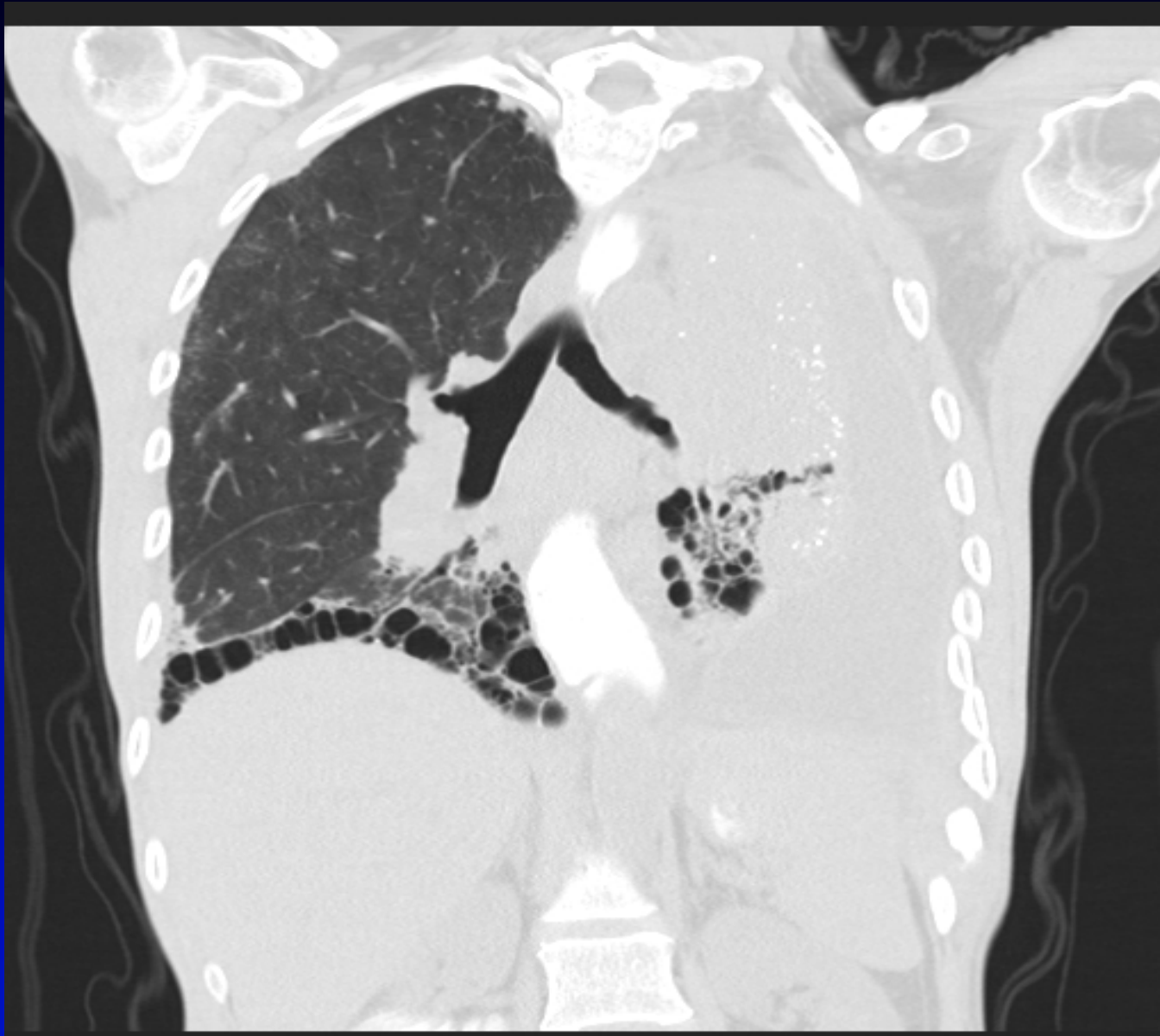




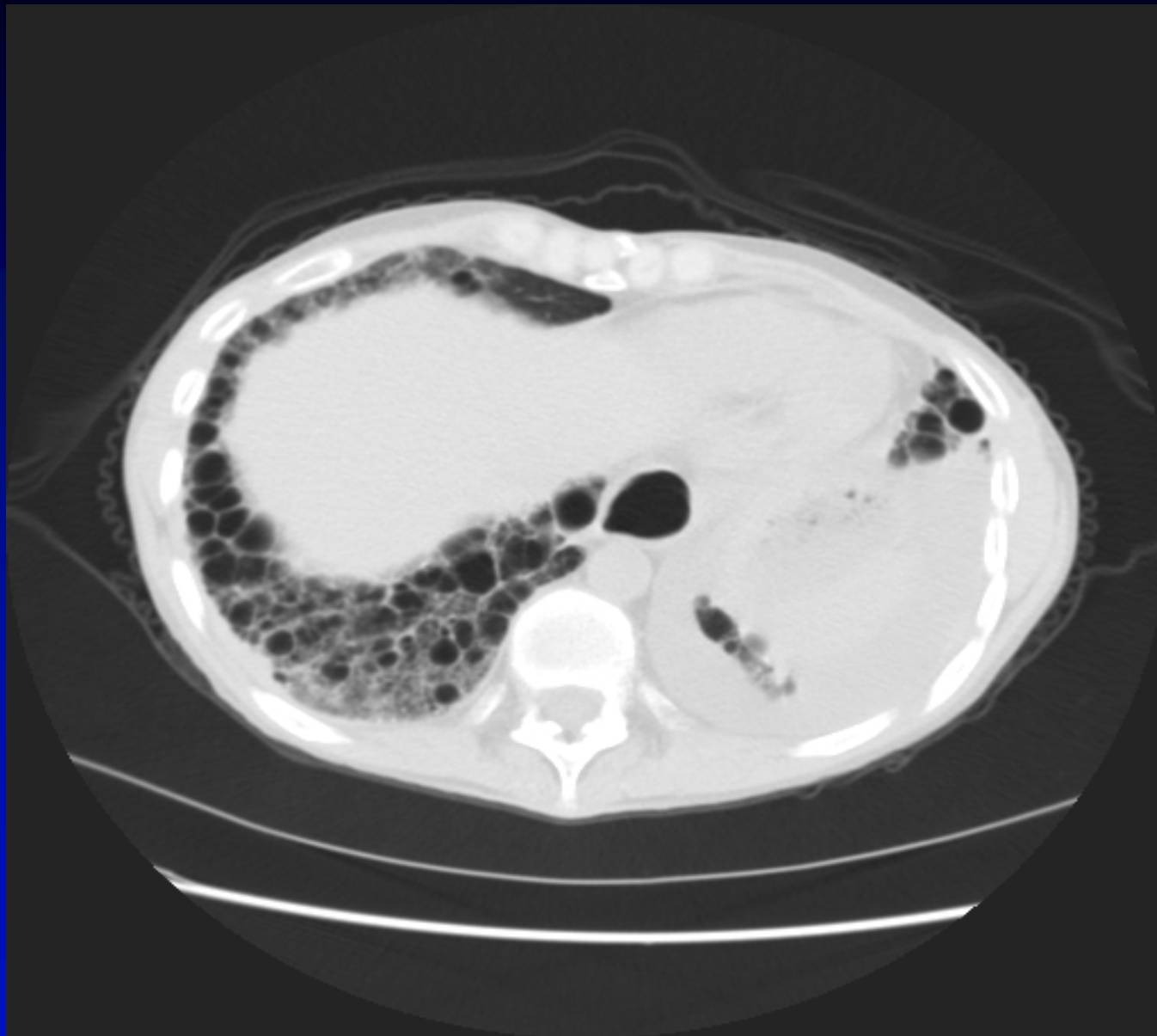


Honeycomb cysts: Scleroderma (PSS)



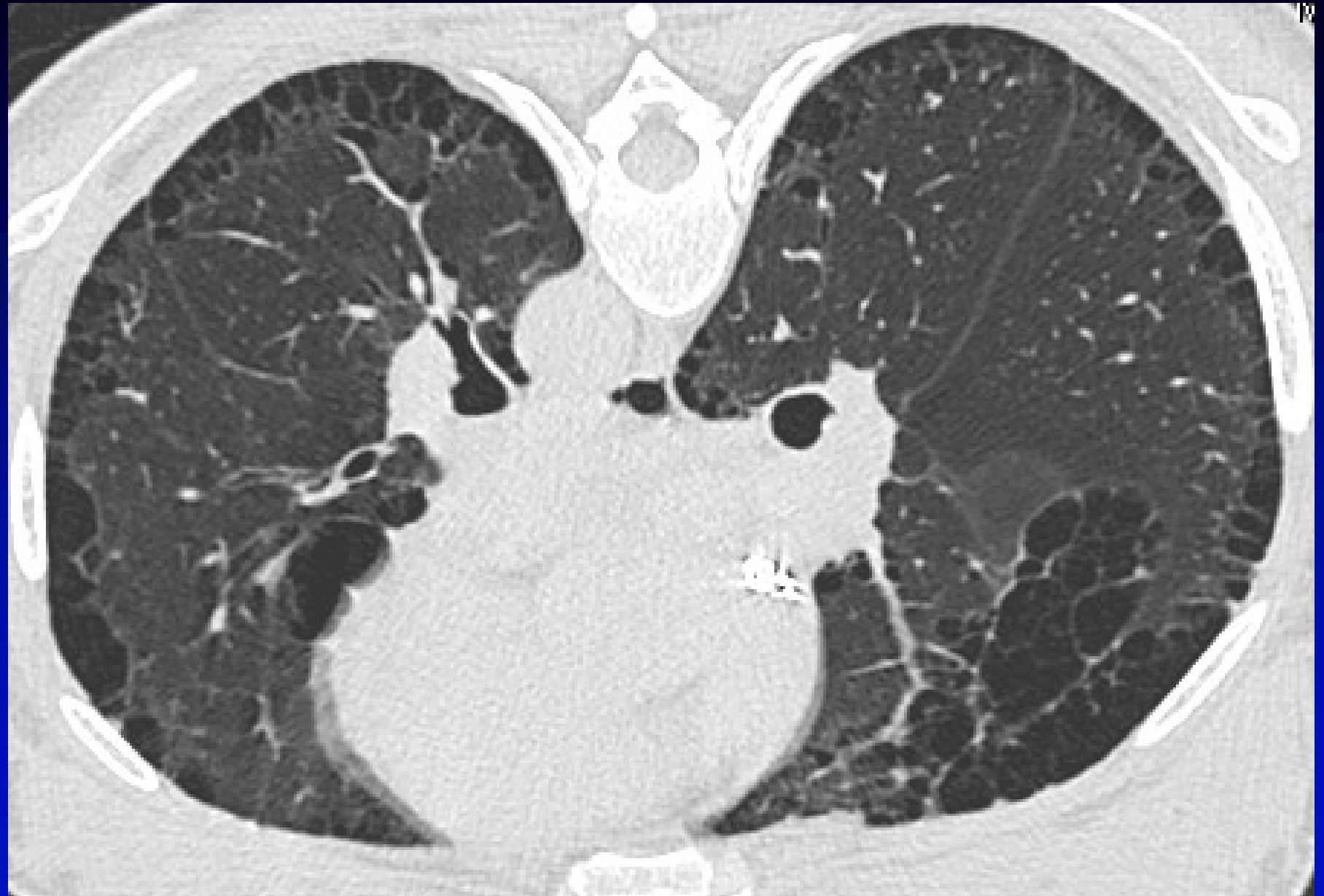






- **Scleroderma-associated ILD**



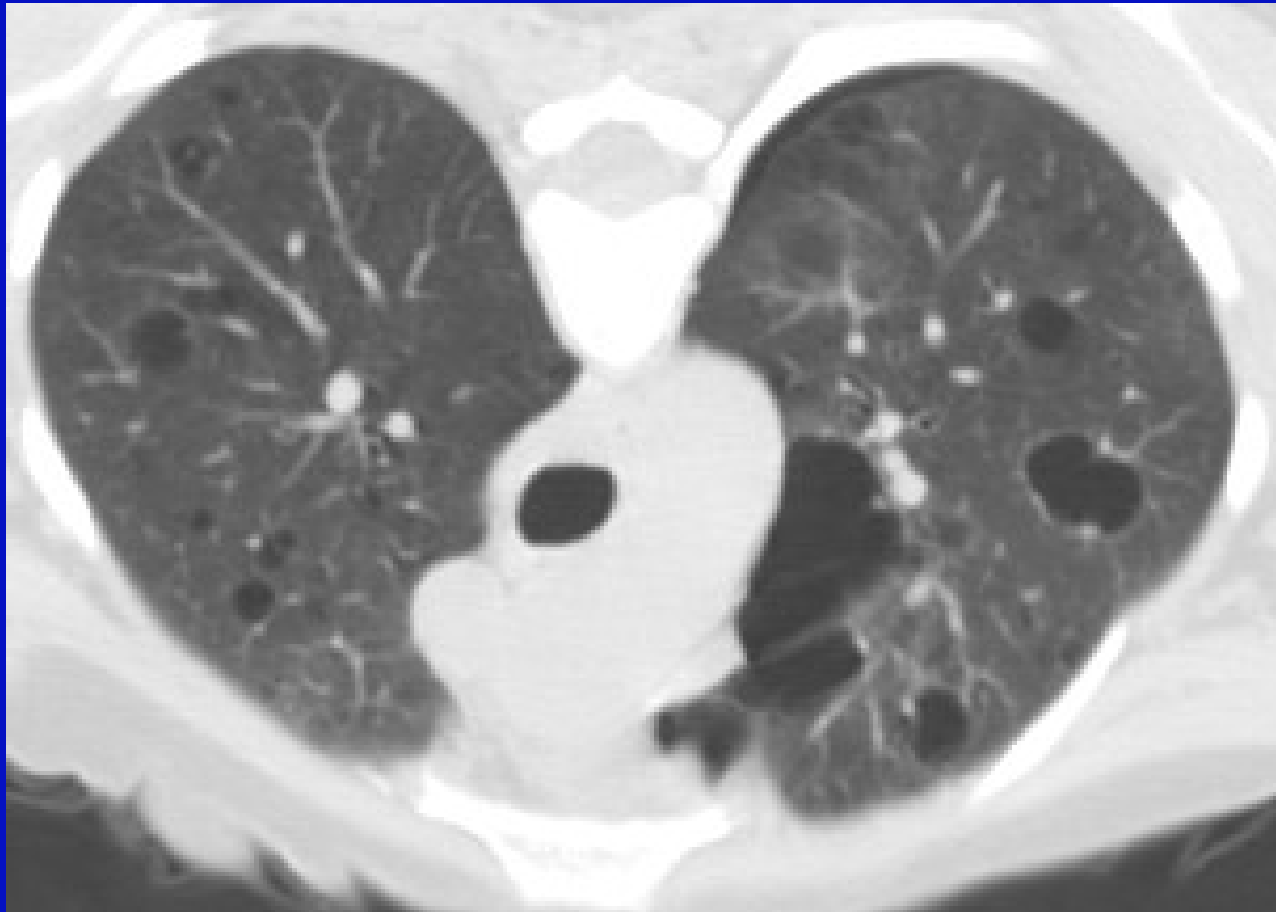




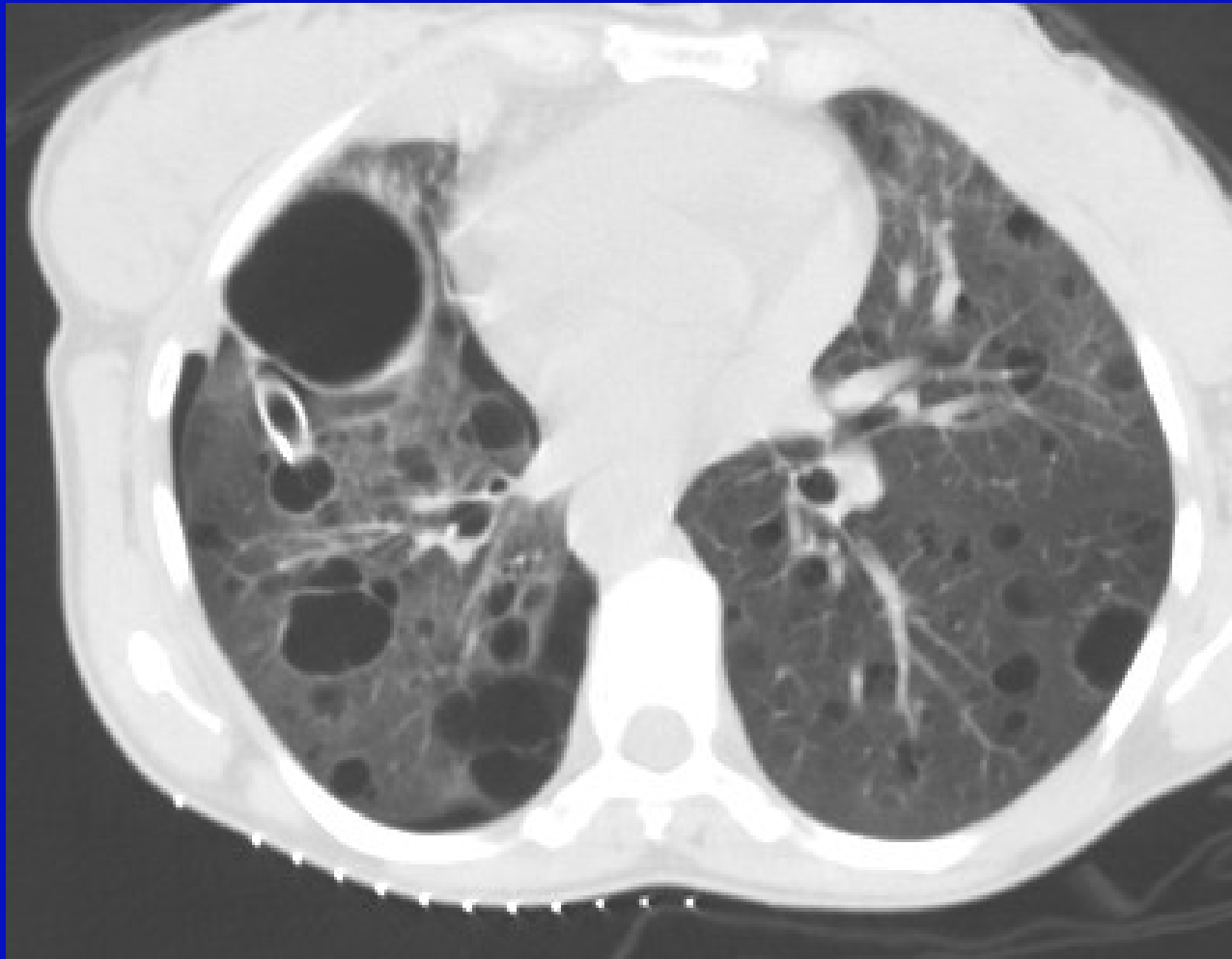




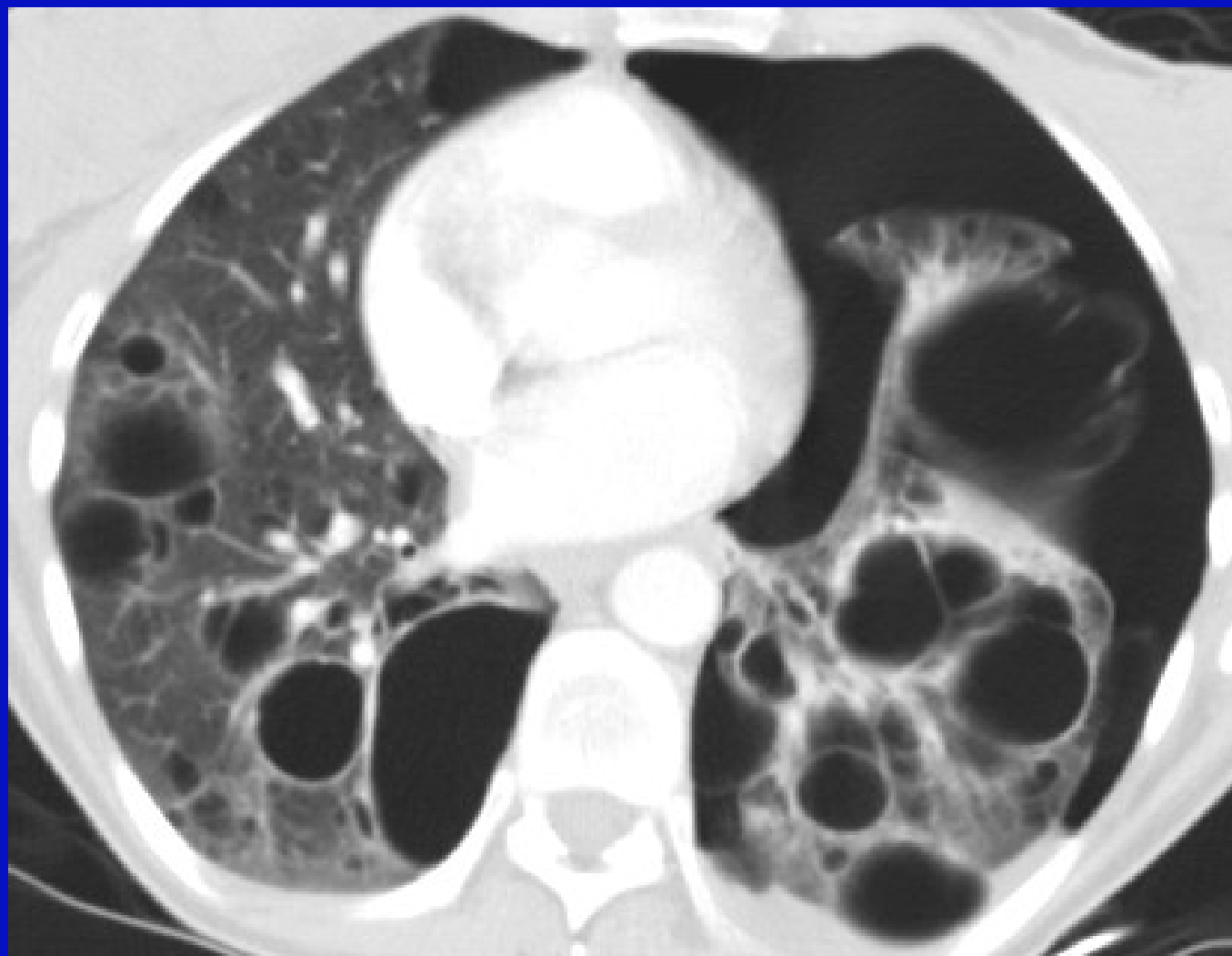
Sjogren's Syndrome



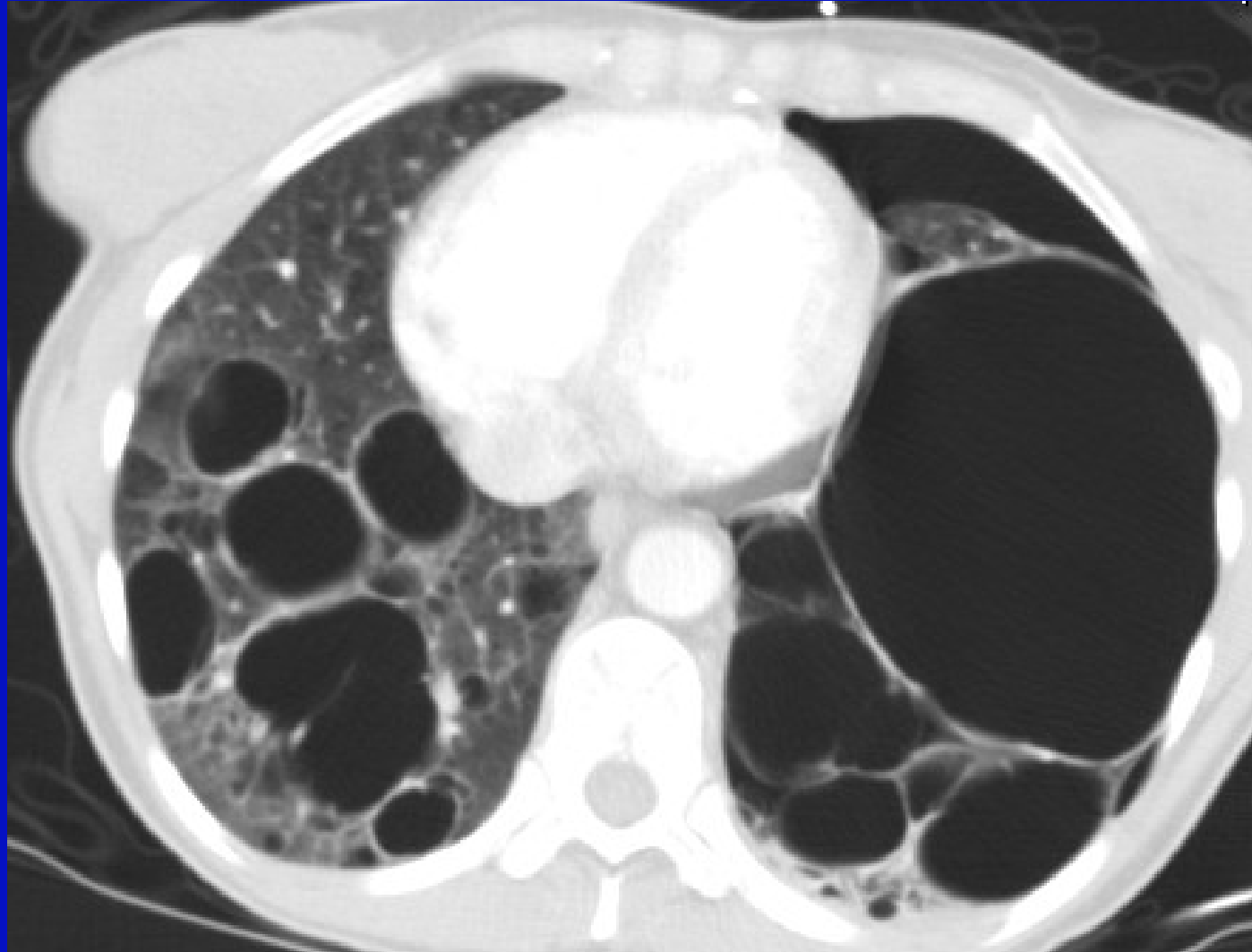
Sjogren's Syndrome



Sjogren's Syndrome



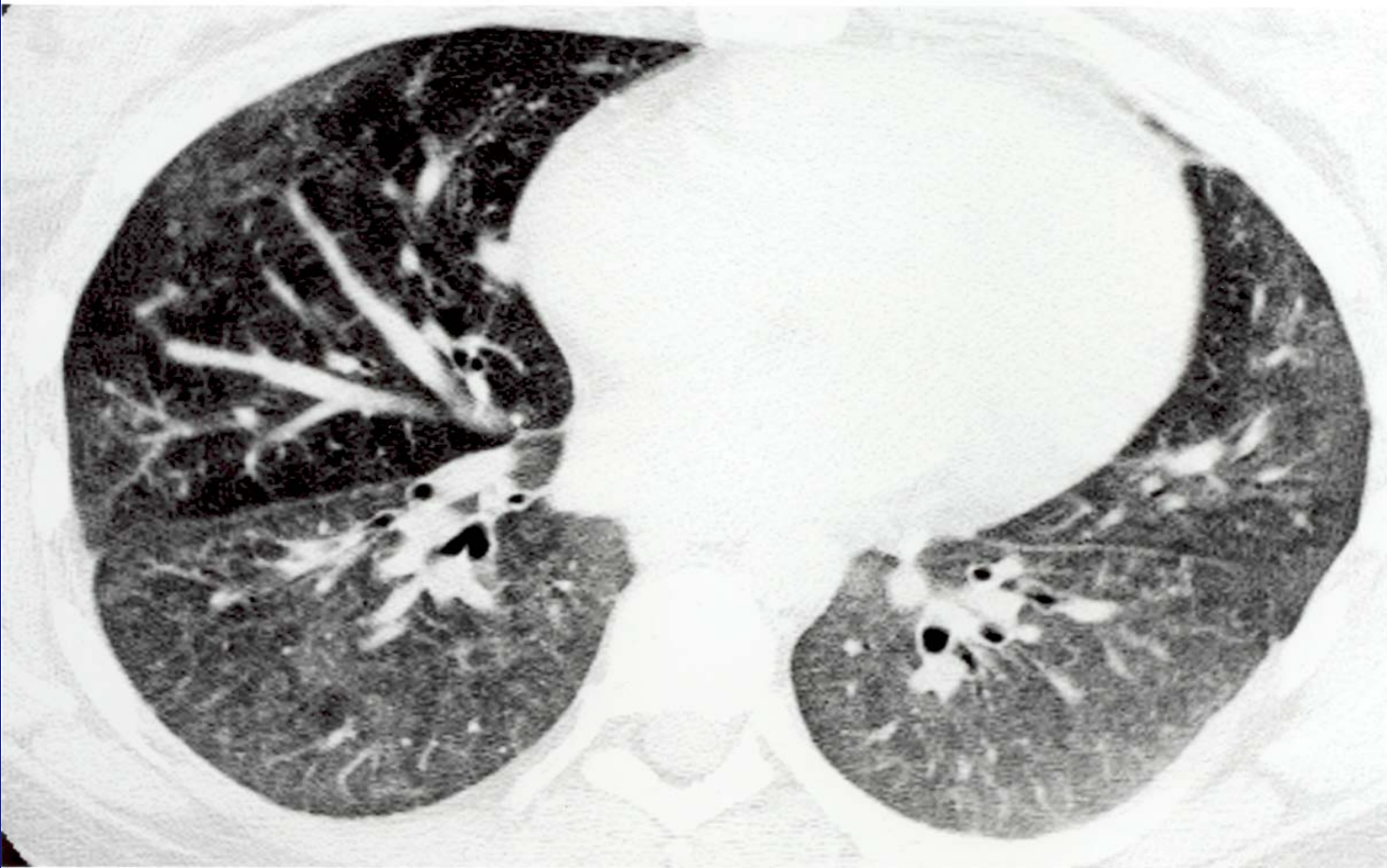
Sjogren's Syndrome



Collagen vascular diseases

-
- Ground glass opacities on CT
surrogate marker of inflammation
-

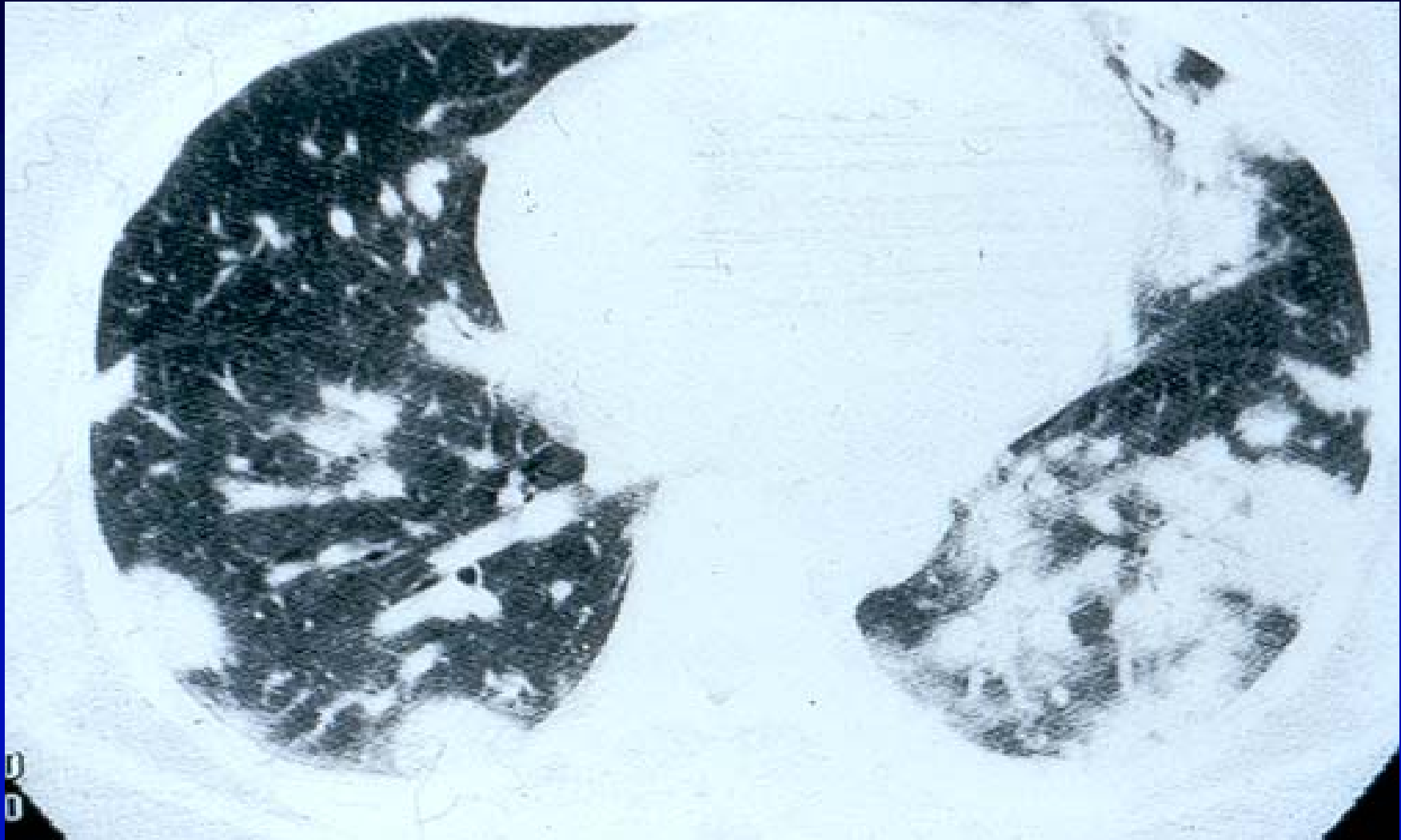
BOOP complicating RA: Diffuse GGO

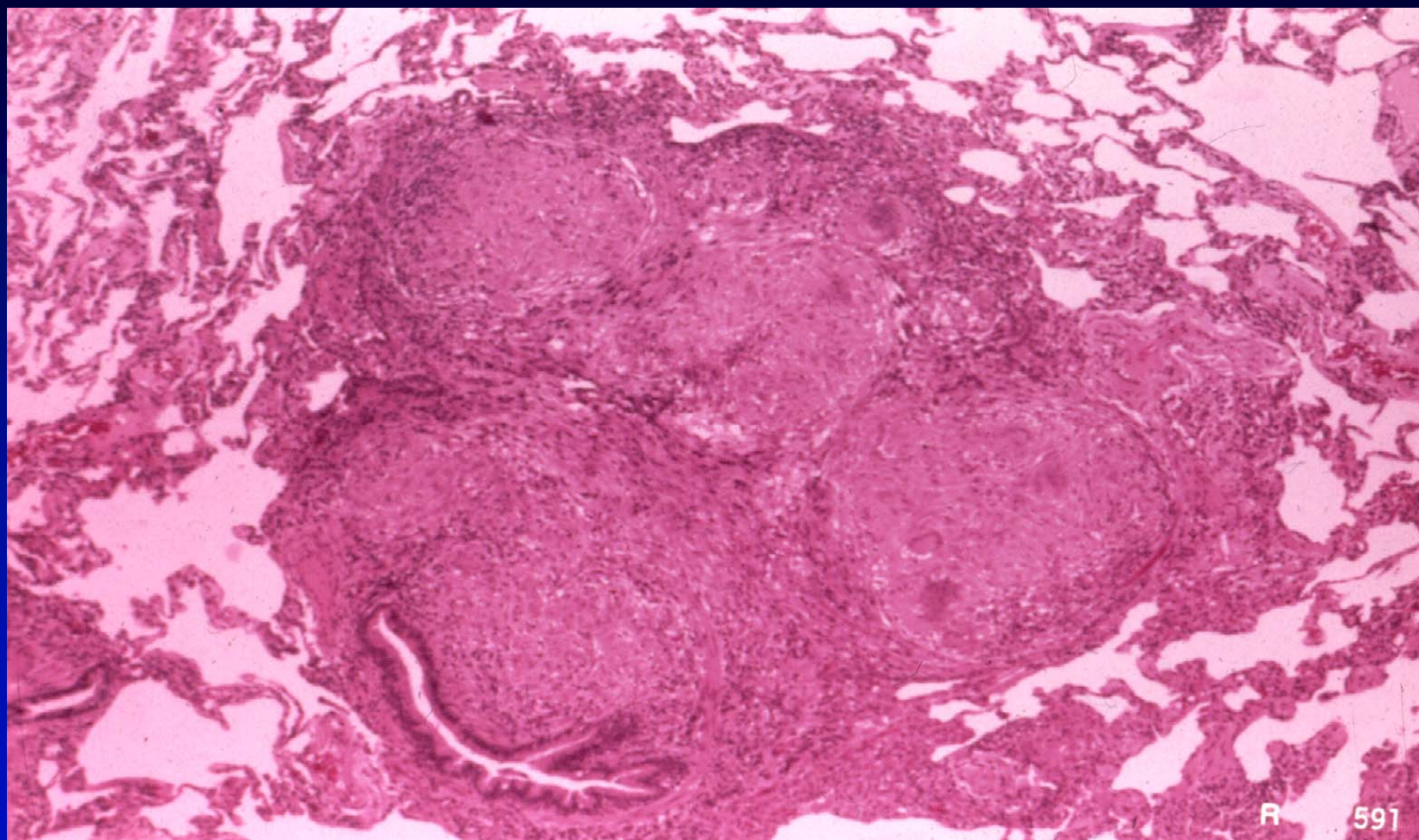


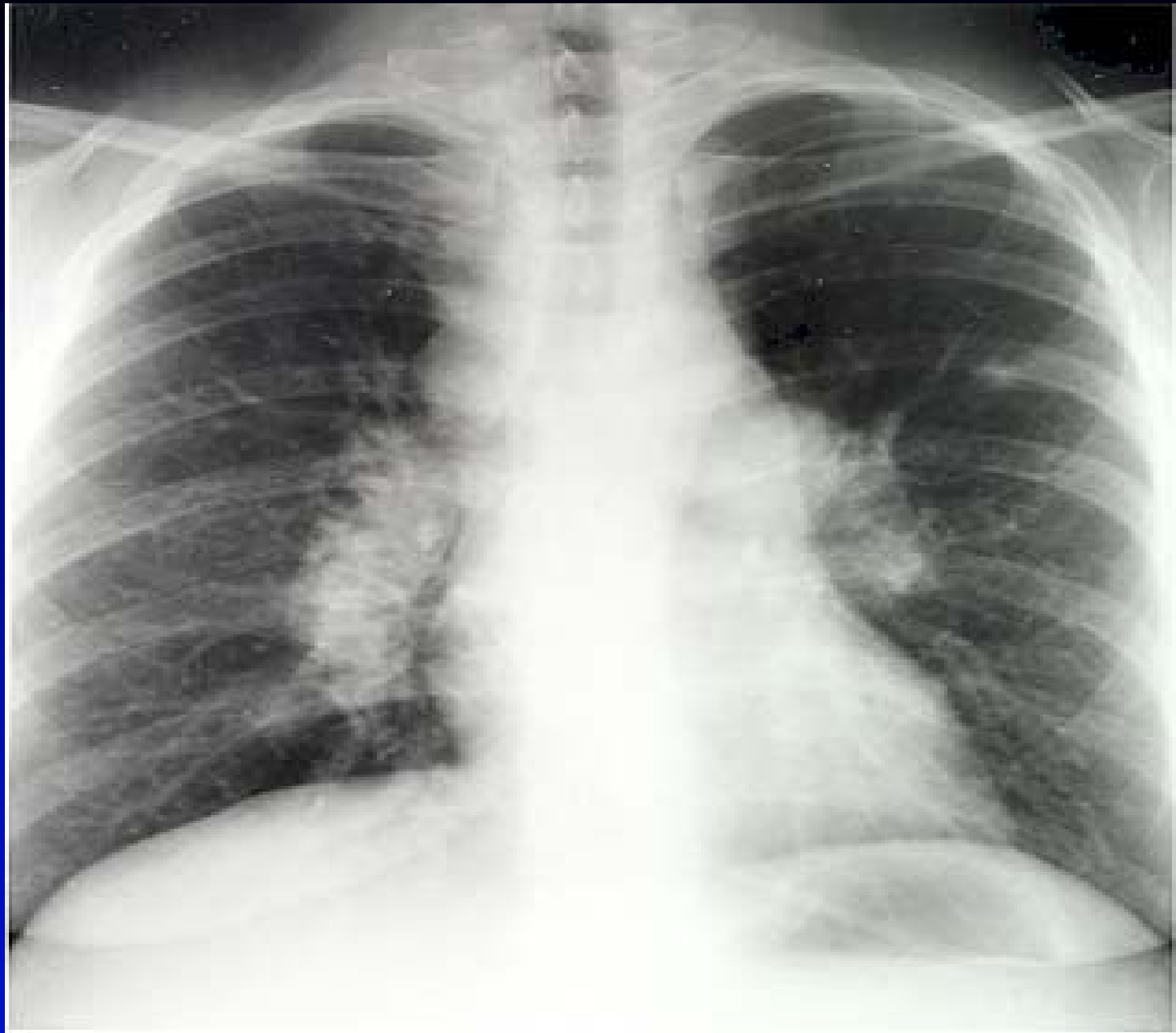
Relapsing BOOP complicating RA

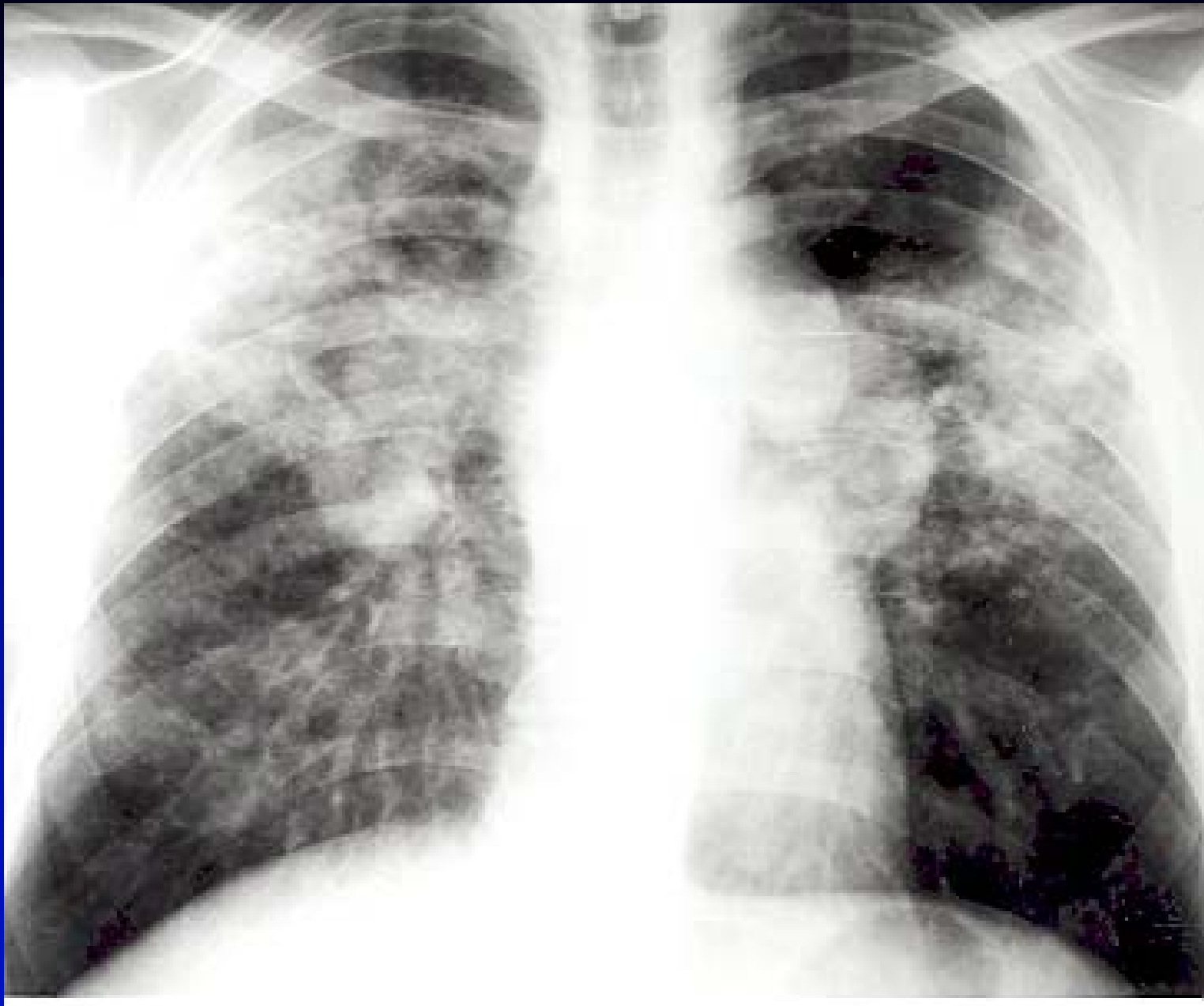


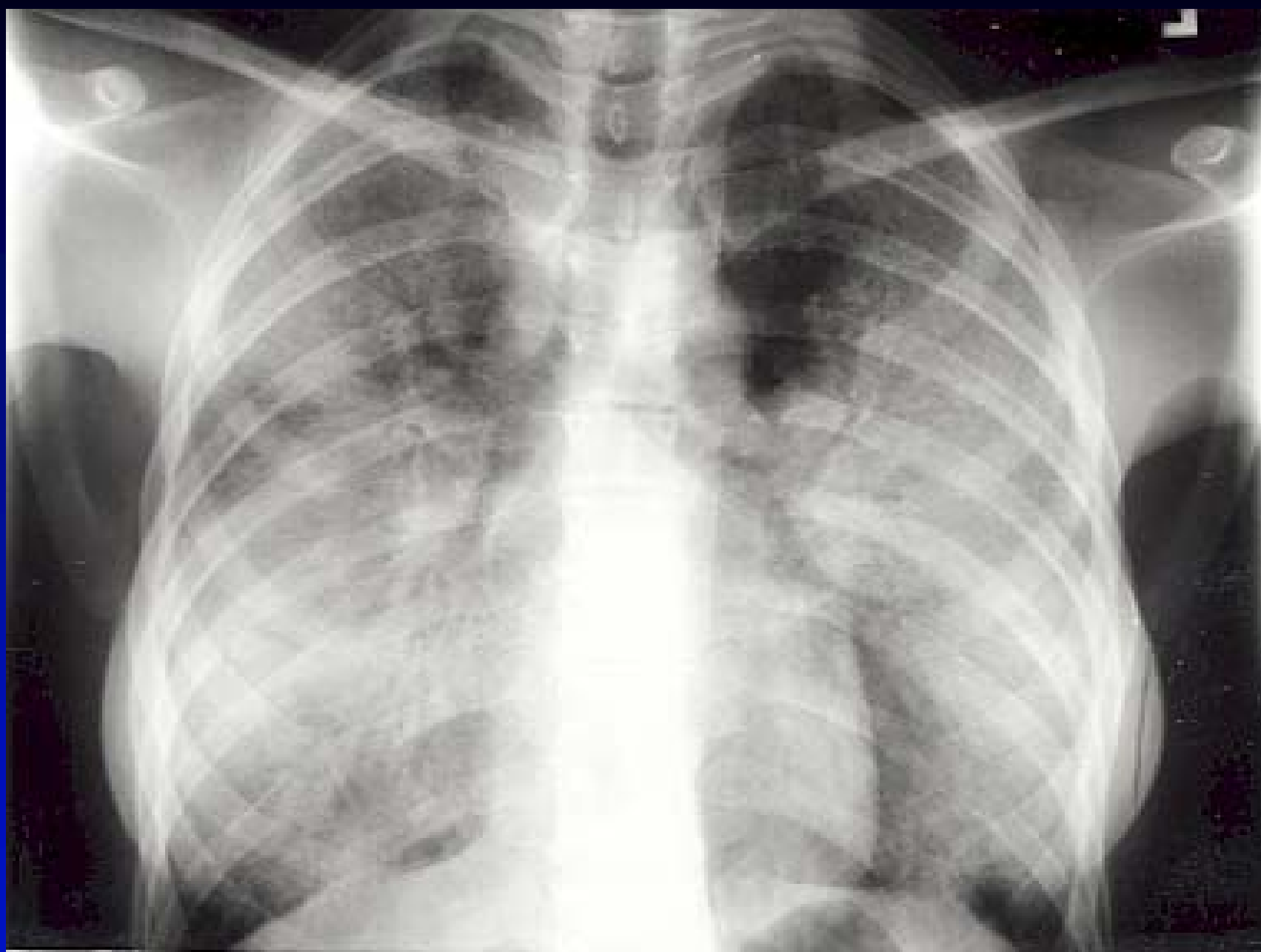
Ground glass opacities (MCTD)

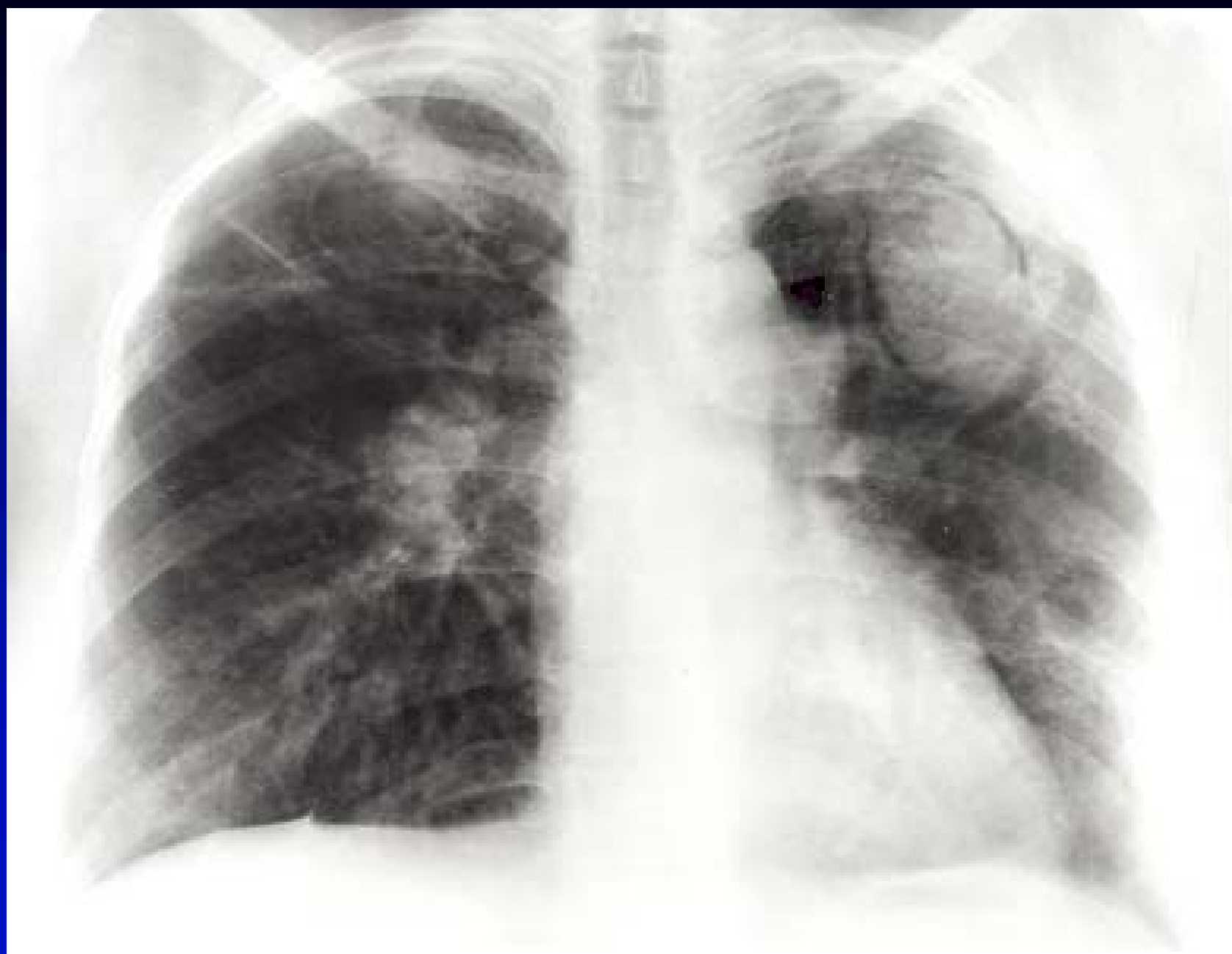






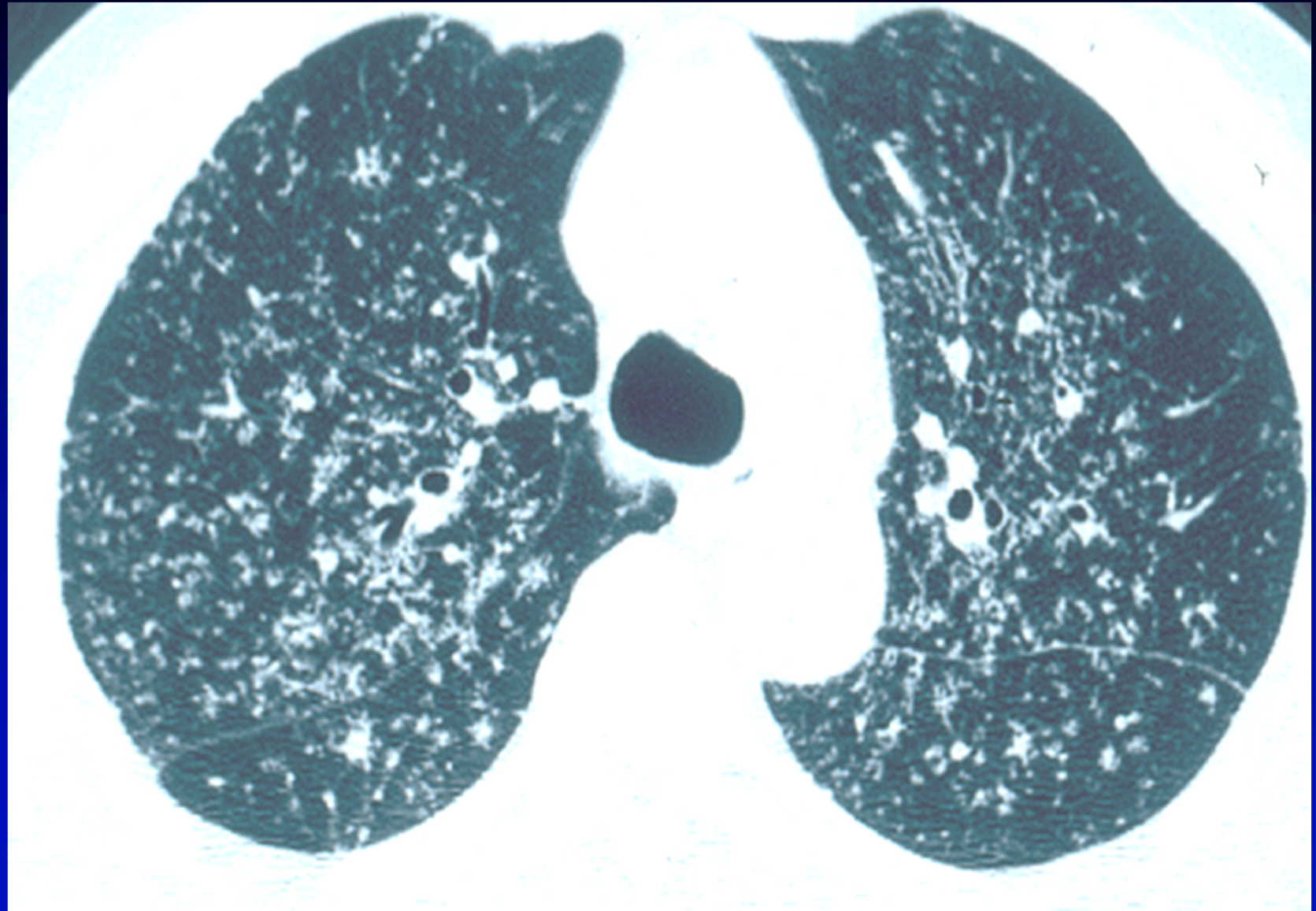


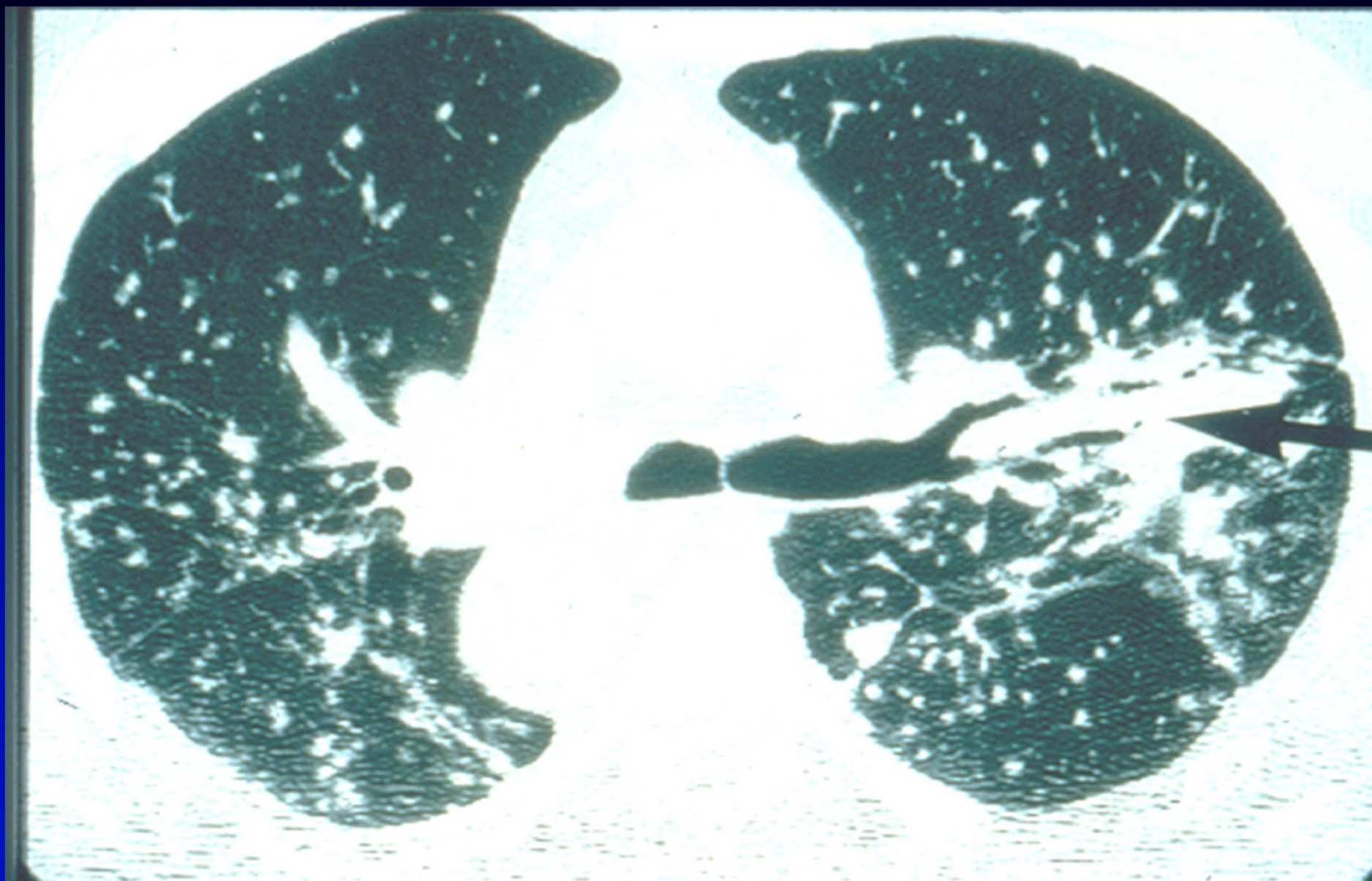




HRCT scans in sarcoidosis

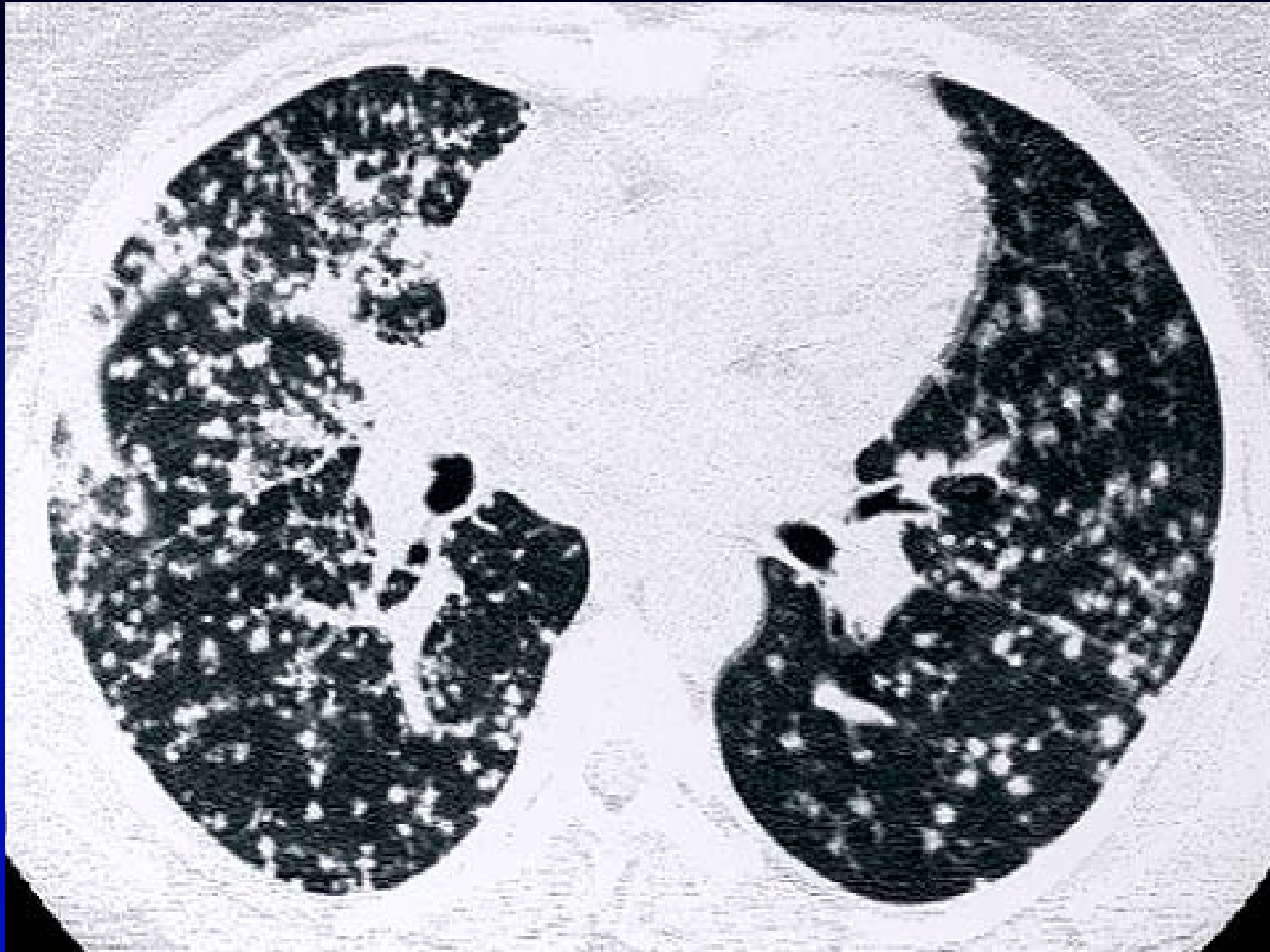
- Upper lobe predominance
- Central bronchovascular bundles
- Micronodules (<3 mm)
- Confluent alveolar opacities
- Distortion, fibrosis, cysts





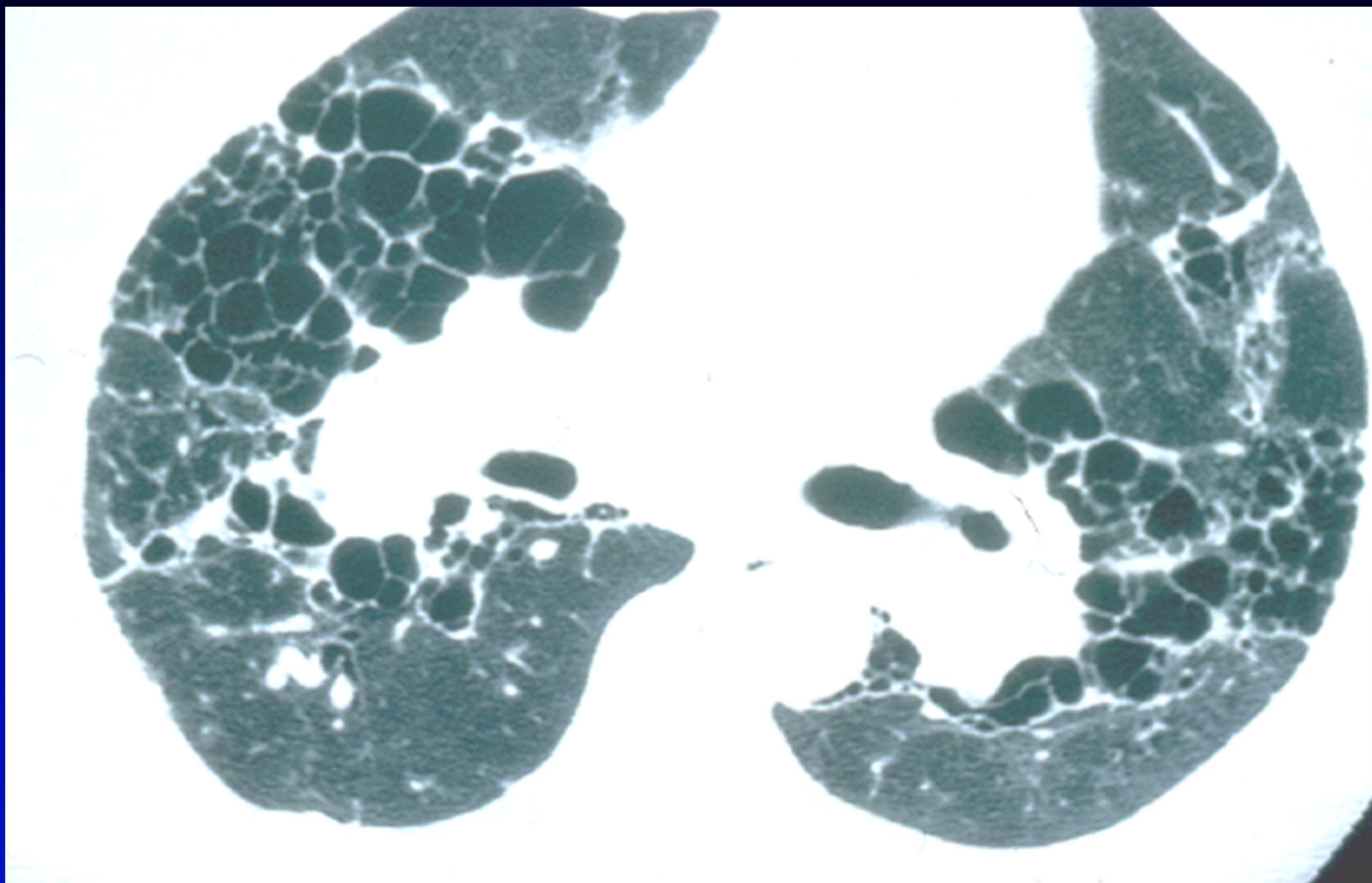


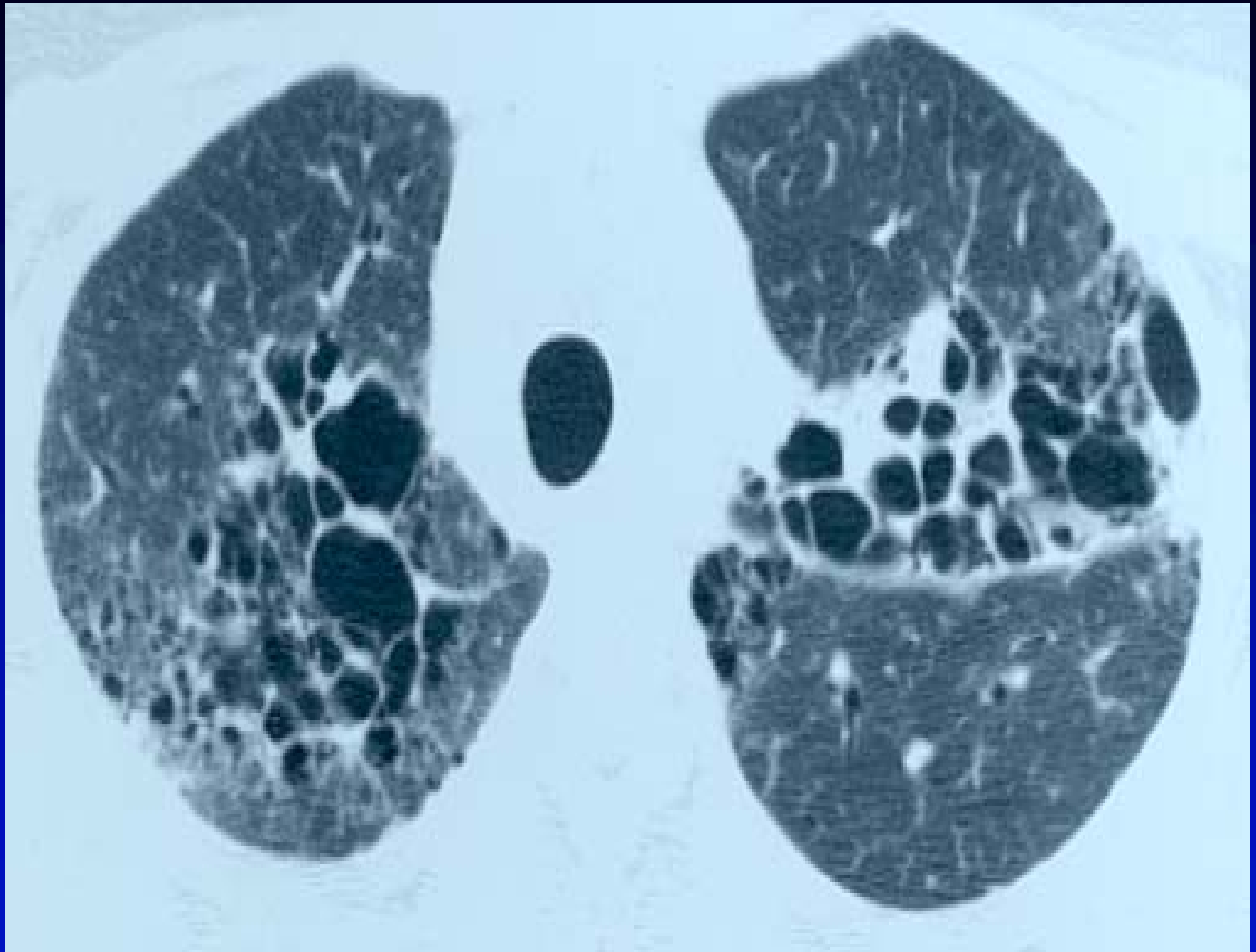


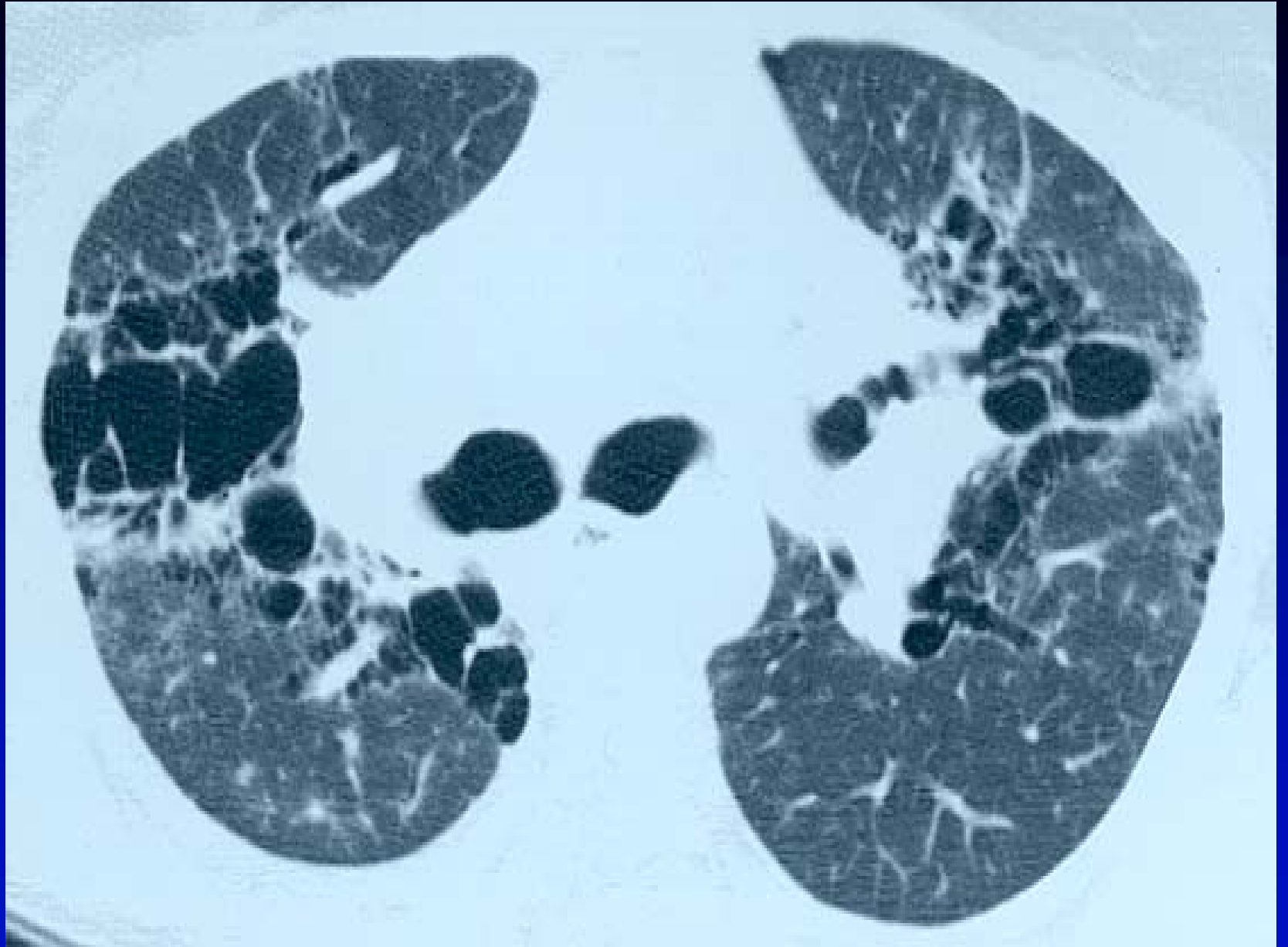


HRCT scans in sarcoidosis

- Upper lobe predominance
- **Central bronchovascular bundles**
- Micronodules (<3 mm)
- Confluent alveolar opacities
- Distortion, fibrosis, cysts

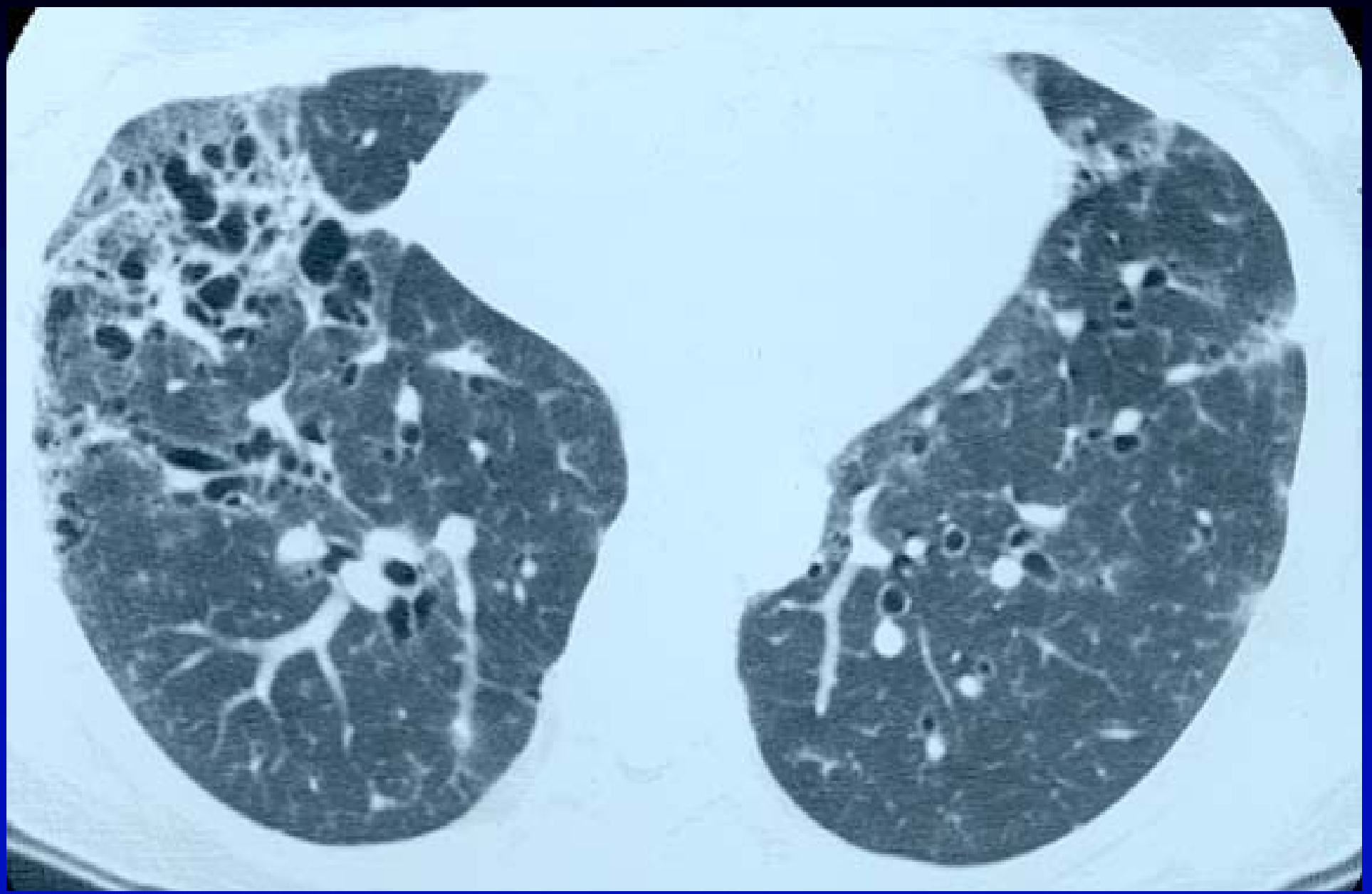




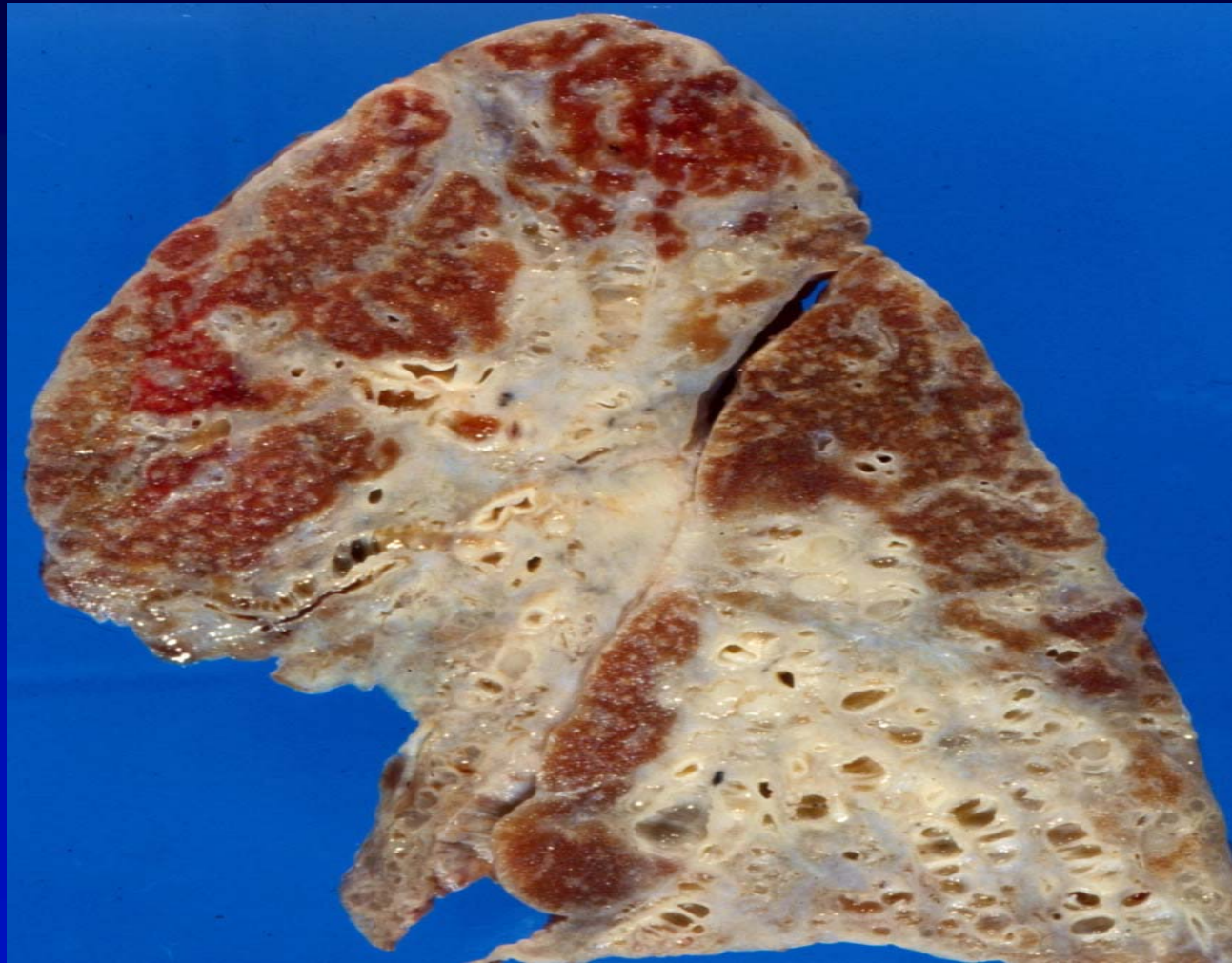






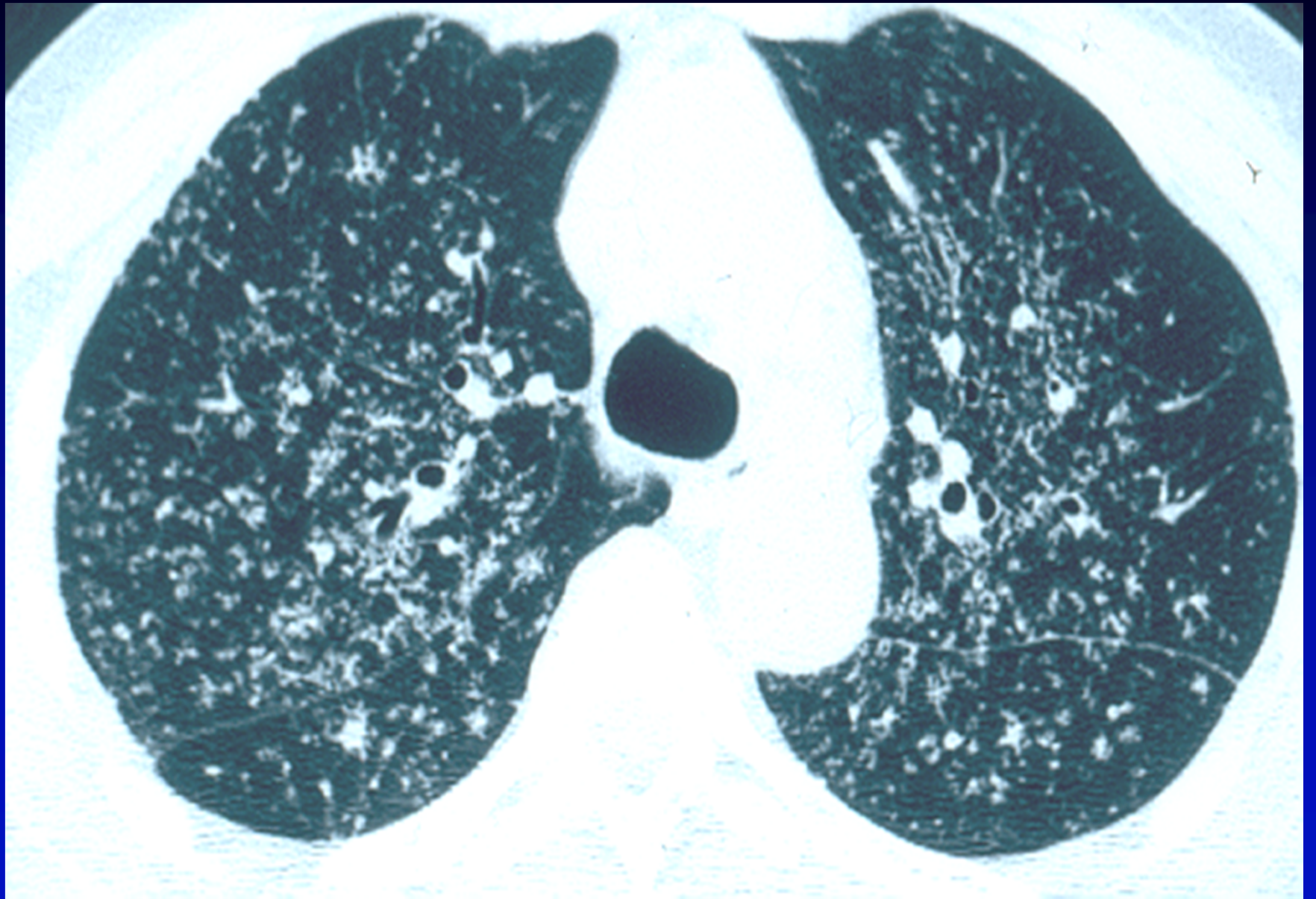


Sarcoidosis

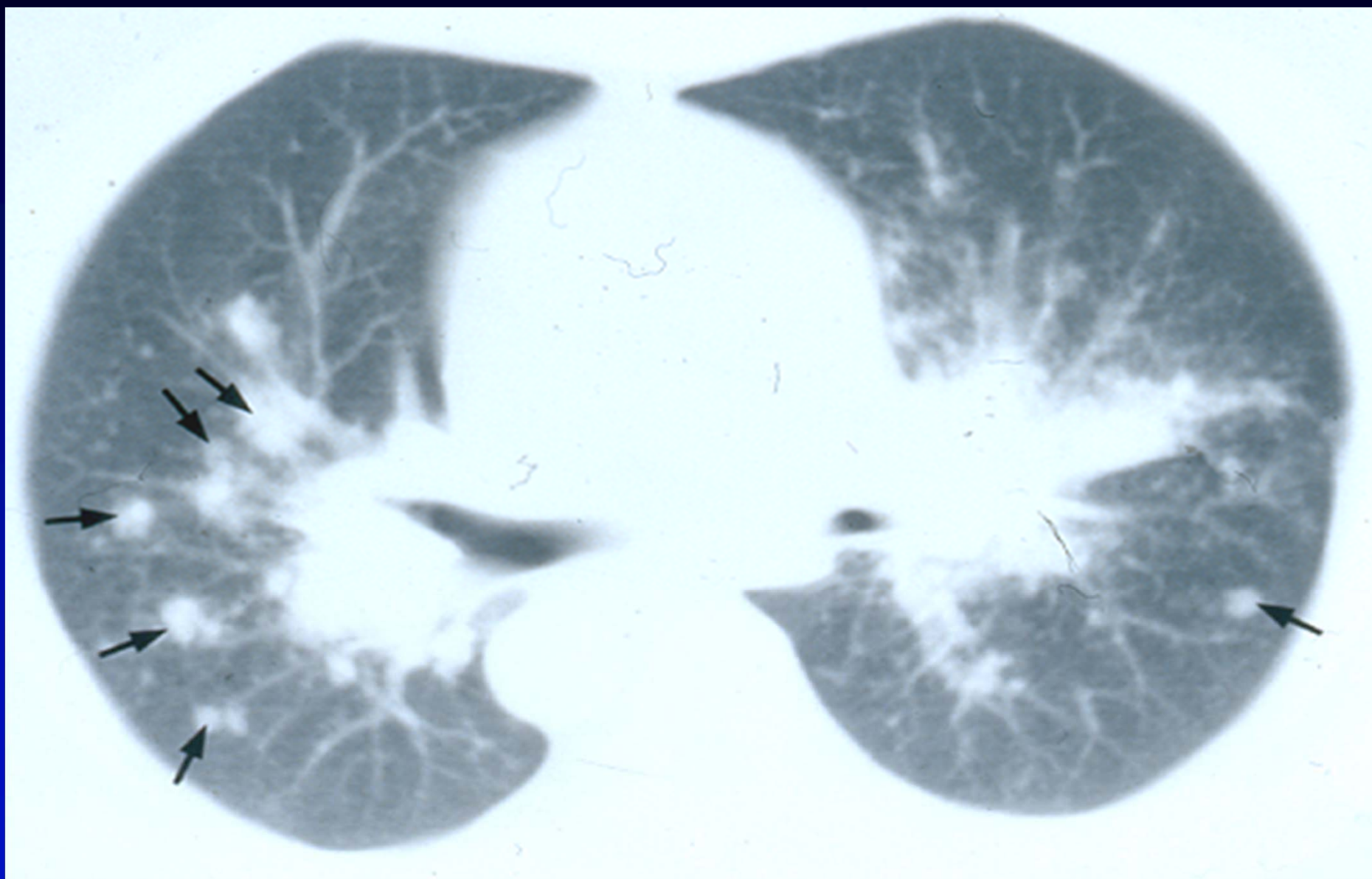


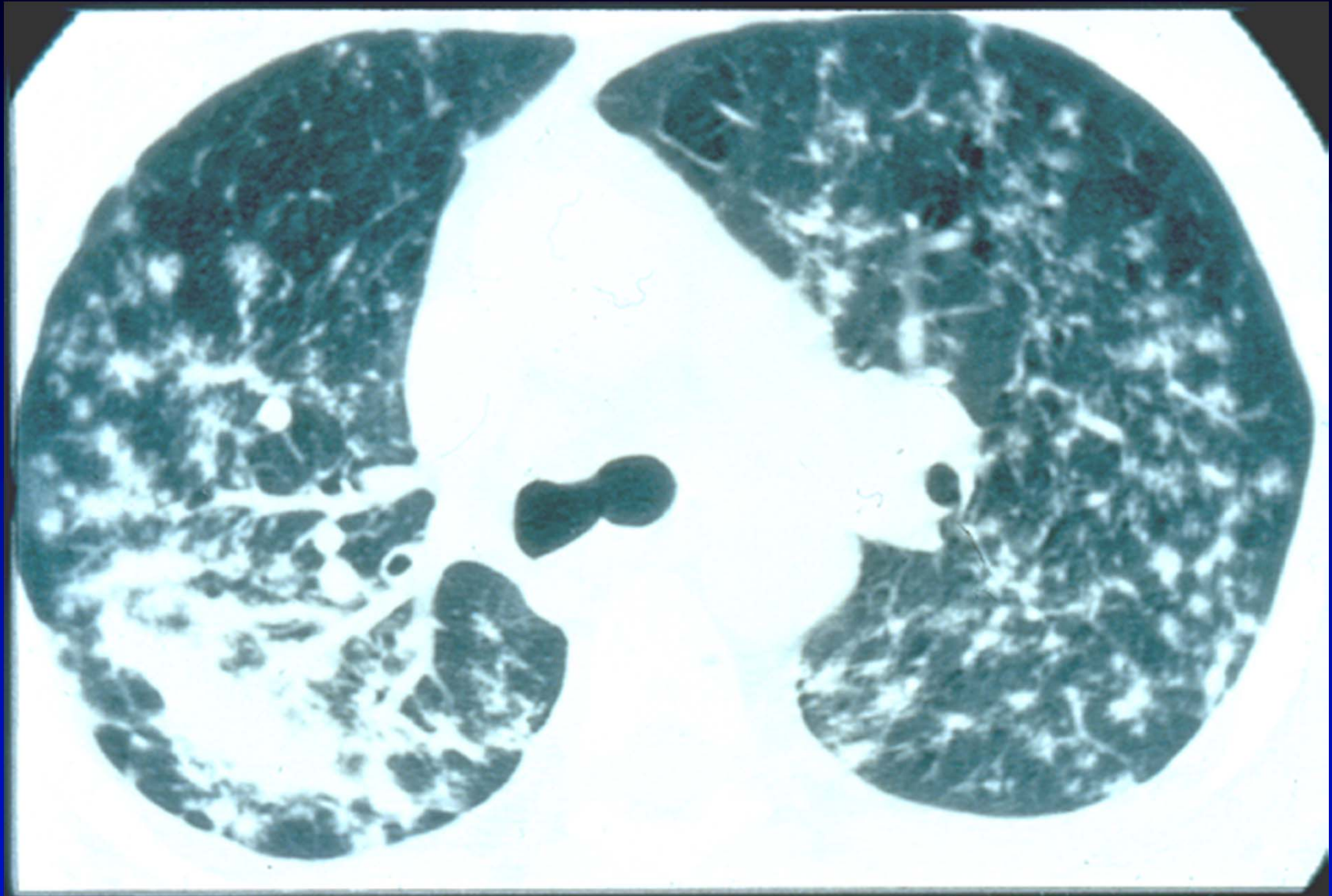
HRCT scans in sarcoidosis

- Upper lobe predominance
- Central bronchovascular bundles
- **Micronodules (<3 mm)**
- Confluent alveolar opacities
- Distortion, fibrosis, cysts



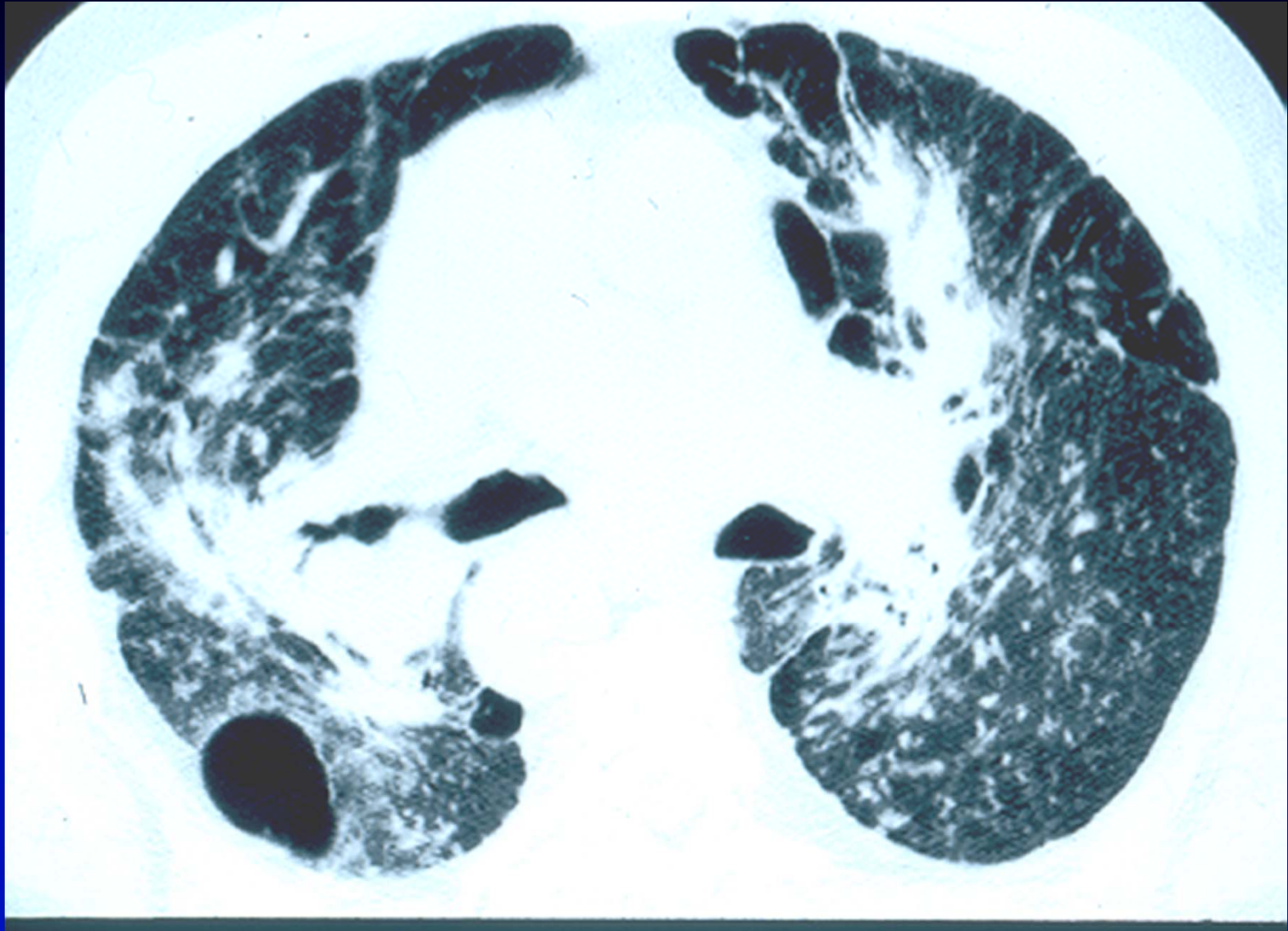


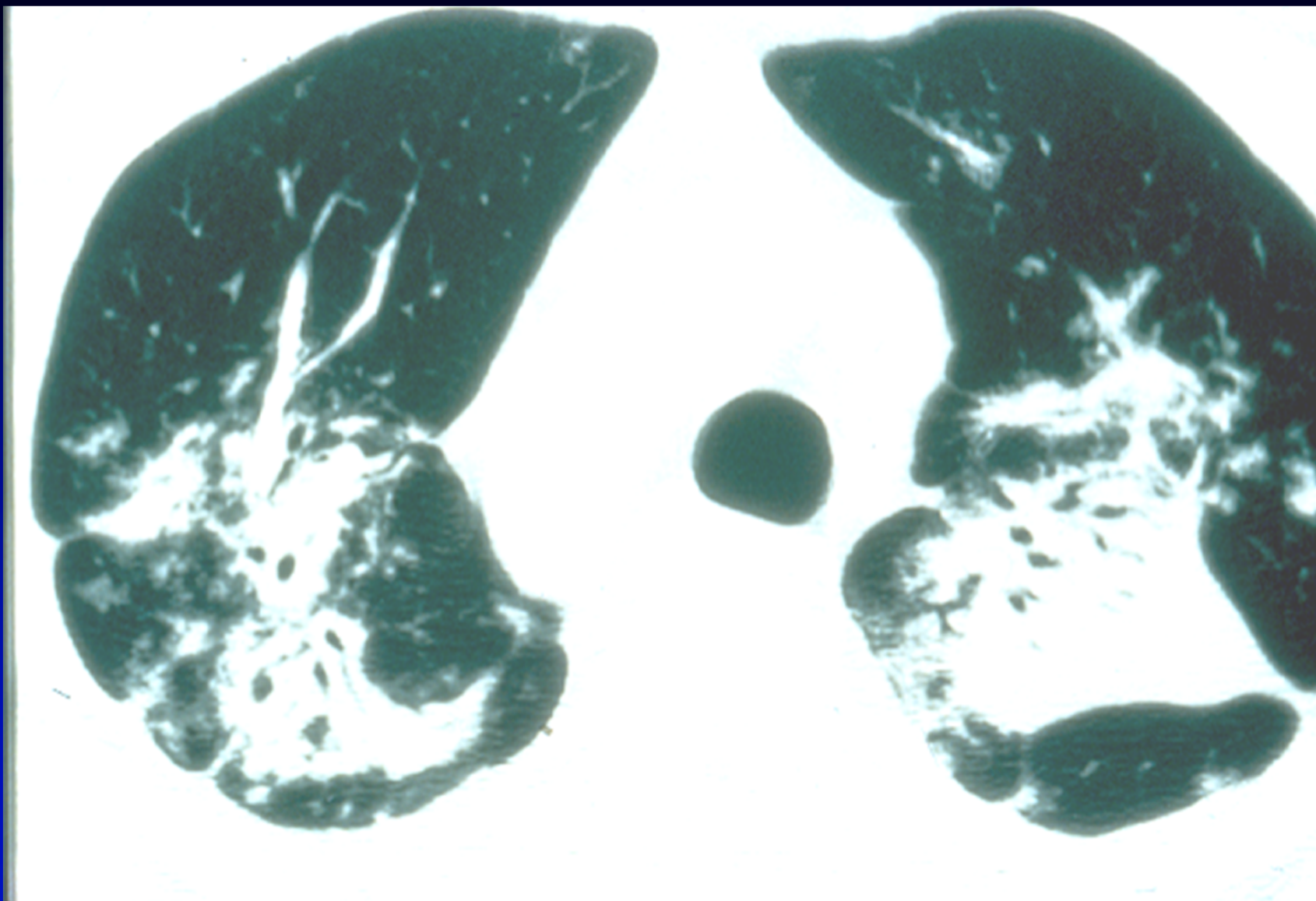


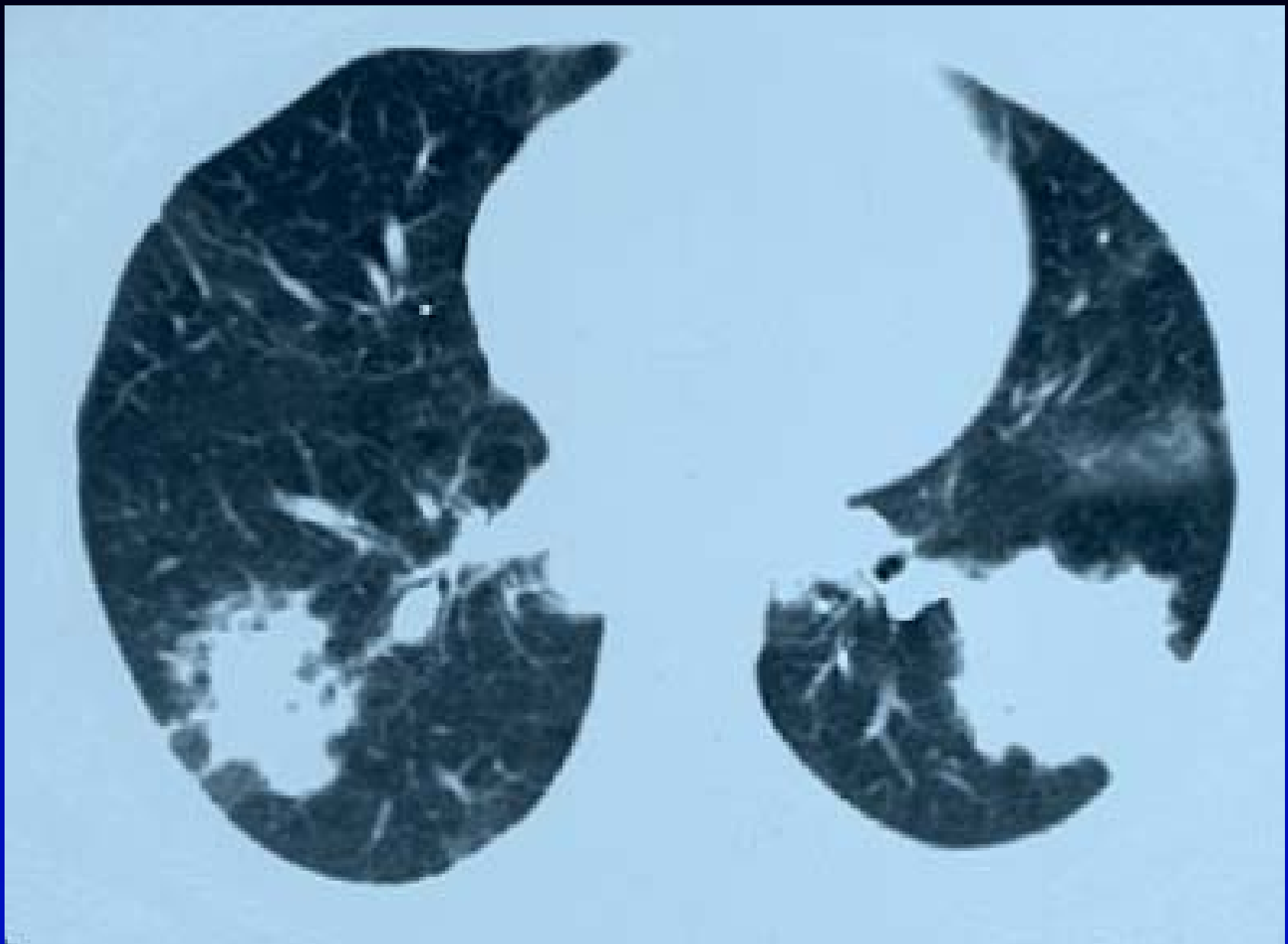


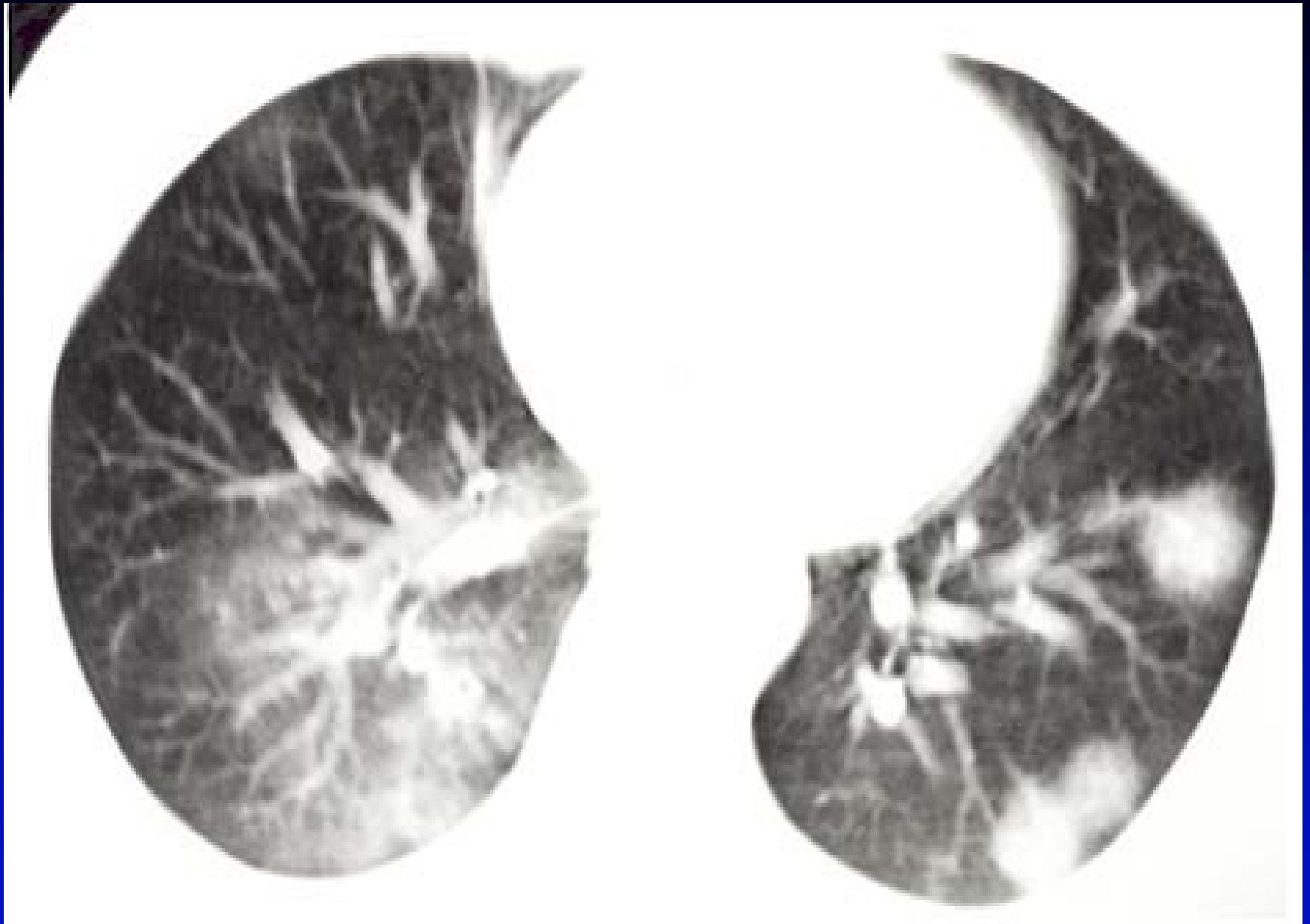
HRCT scans in sarcoidosis

- Upper lobe predominance
- Central bronchovascular bundles
- Micronodules (<3 mm)
- **Confluent alveolar opacities**
- Distortion, fibrosis, cysts





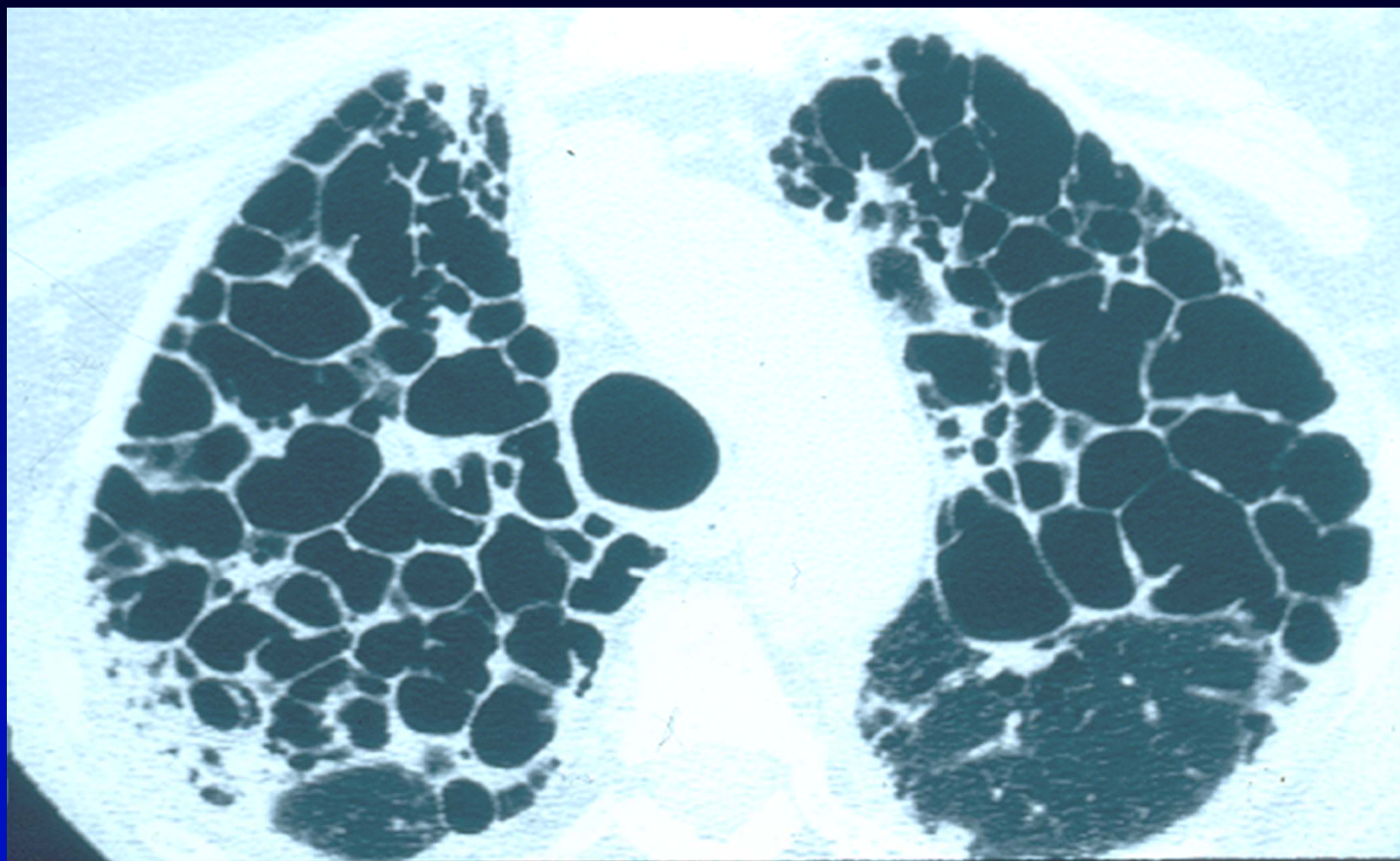


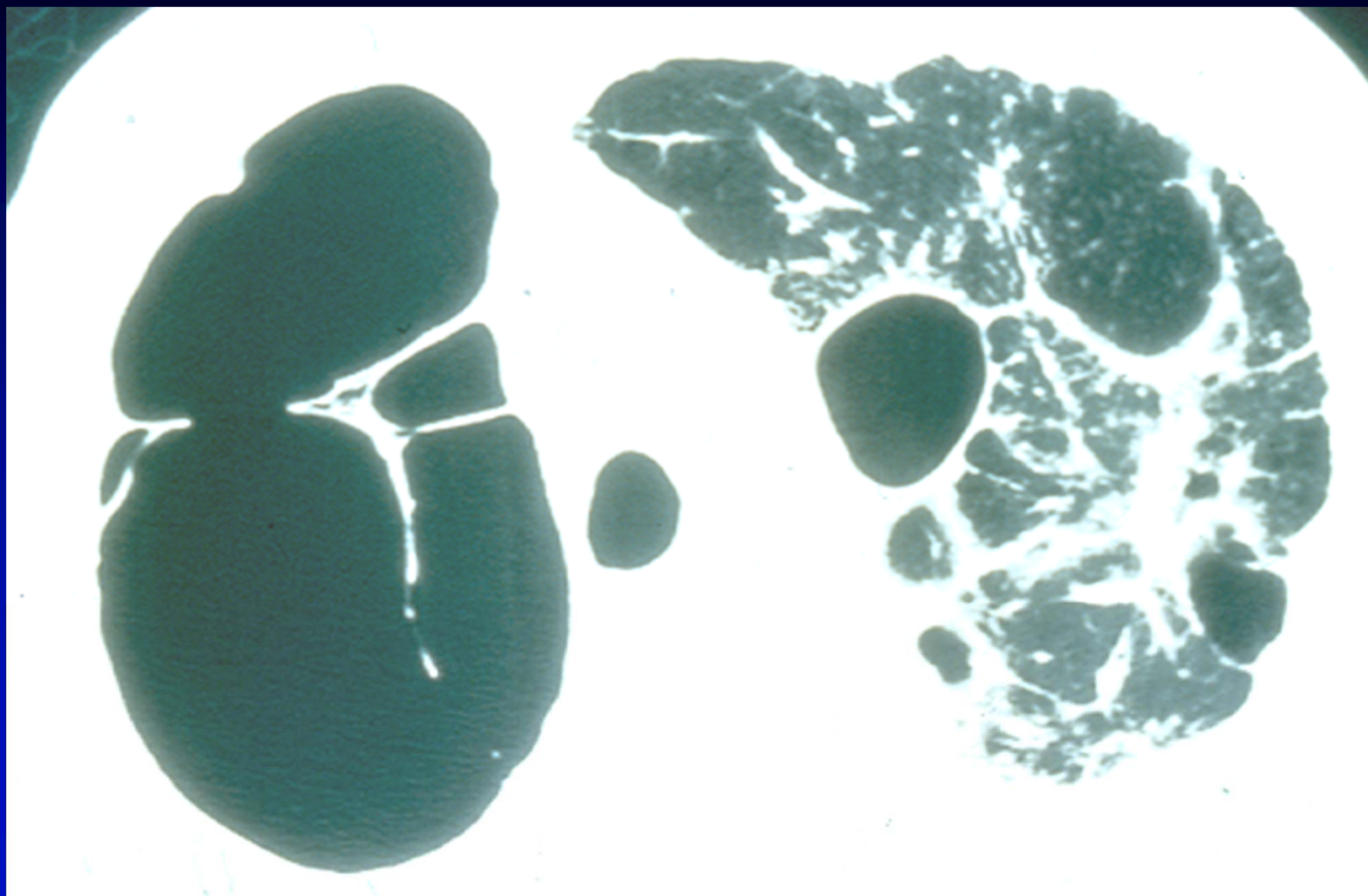


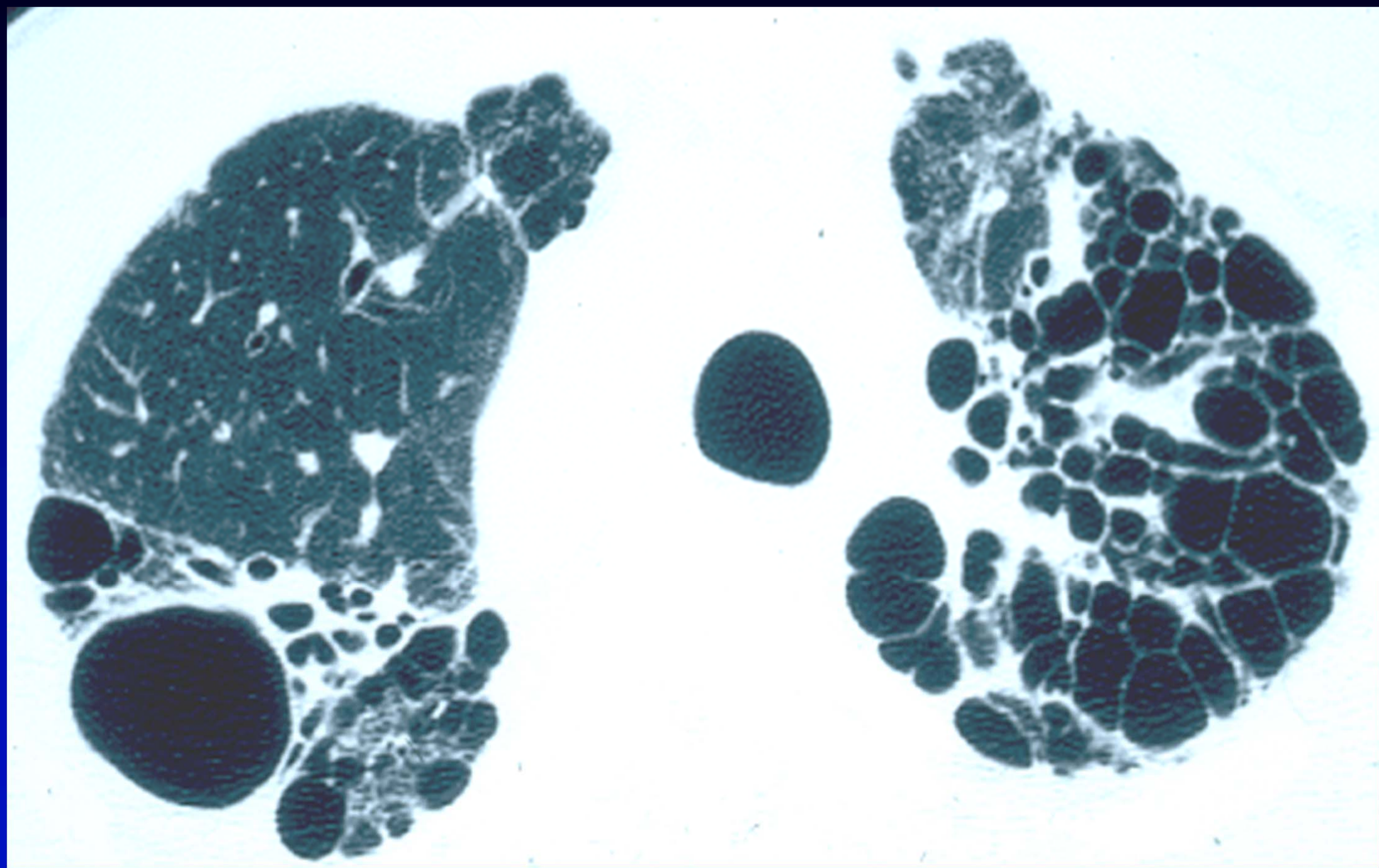
HRCT scans in sarcoidosis

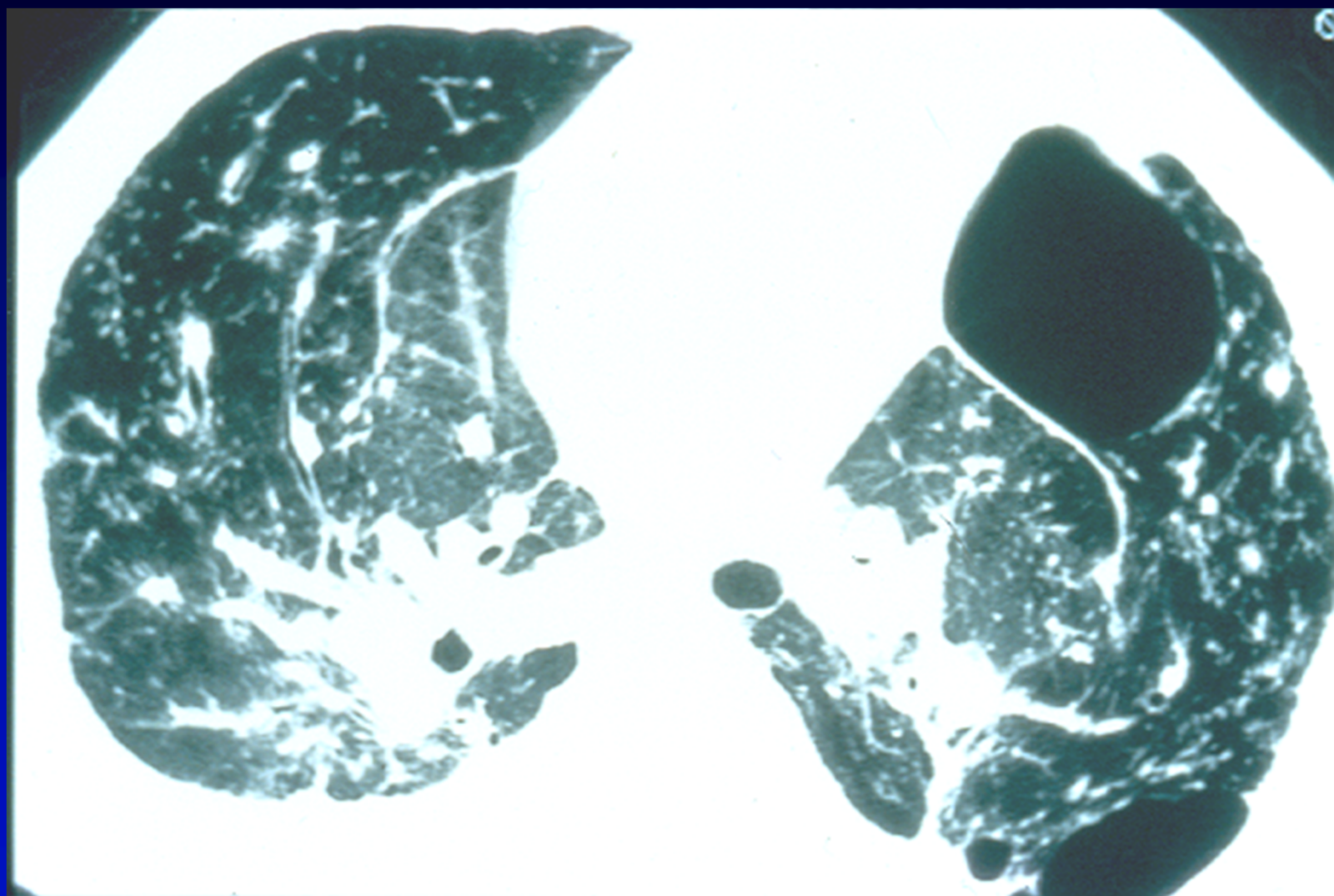
- Upper lobe predominance
- Central bronchovascular bundles
- Micronodules (<3 mm)
- Confluent alveolar opacities
- Distortion, fibrosis, cysts

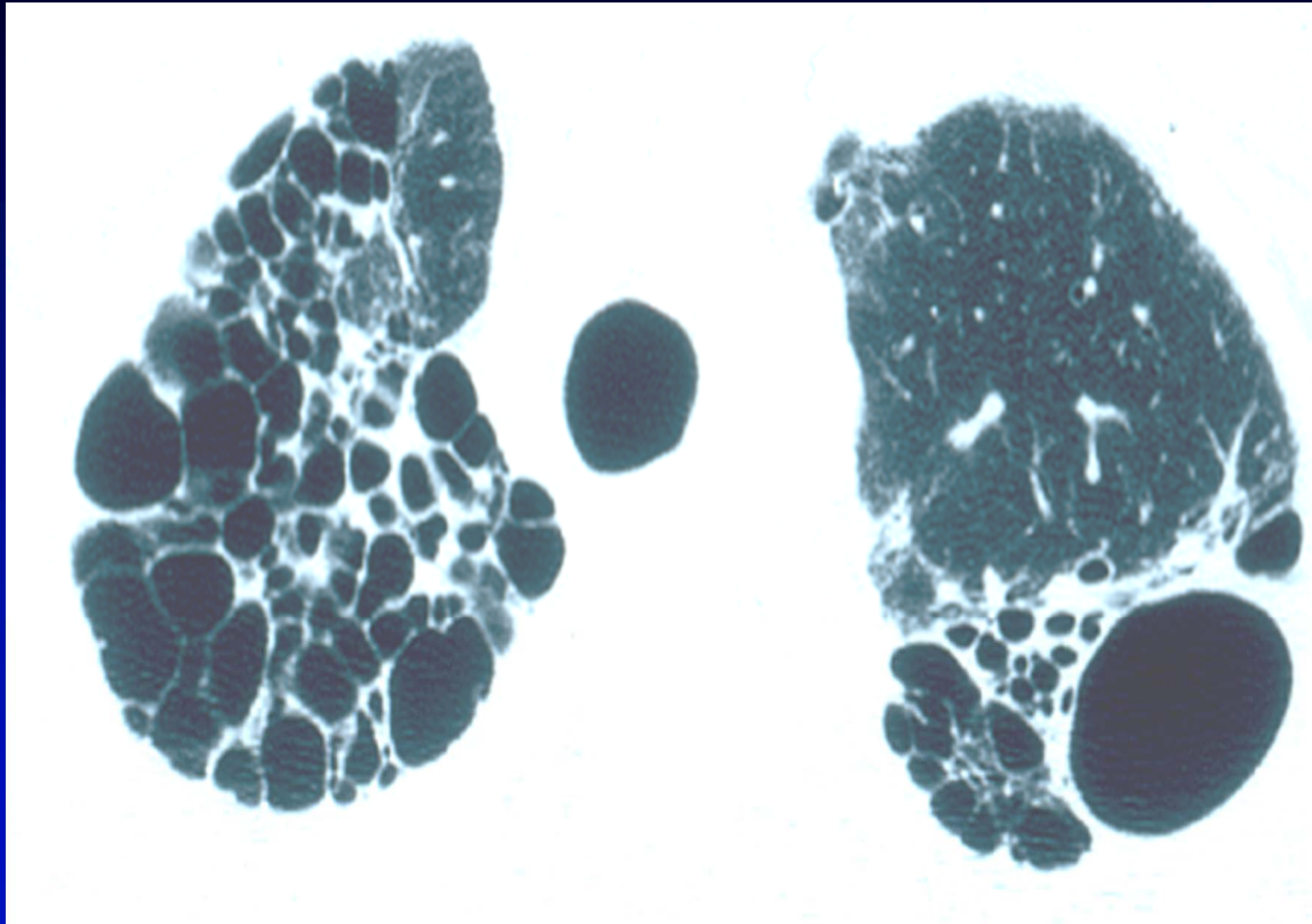


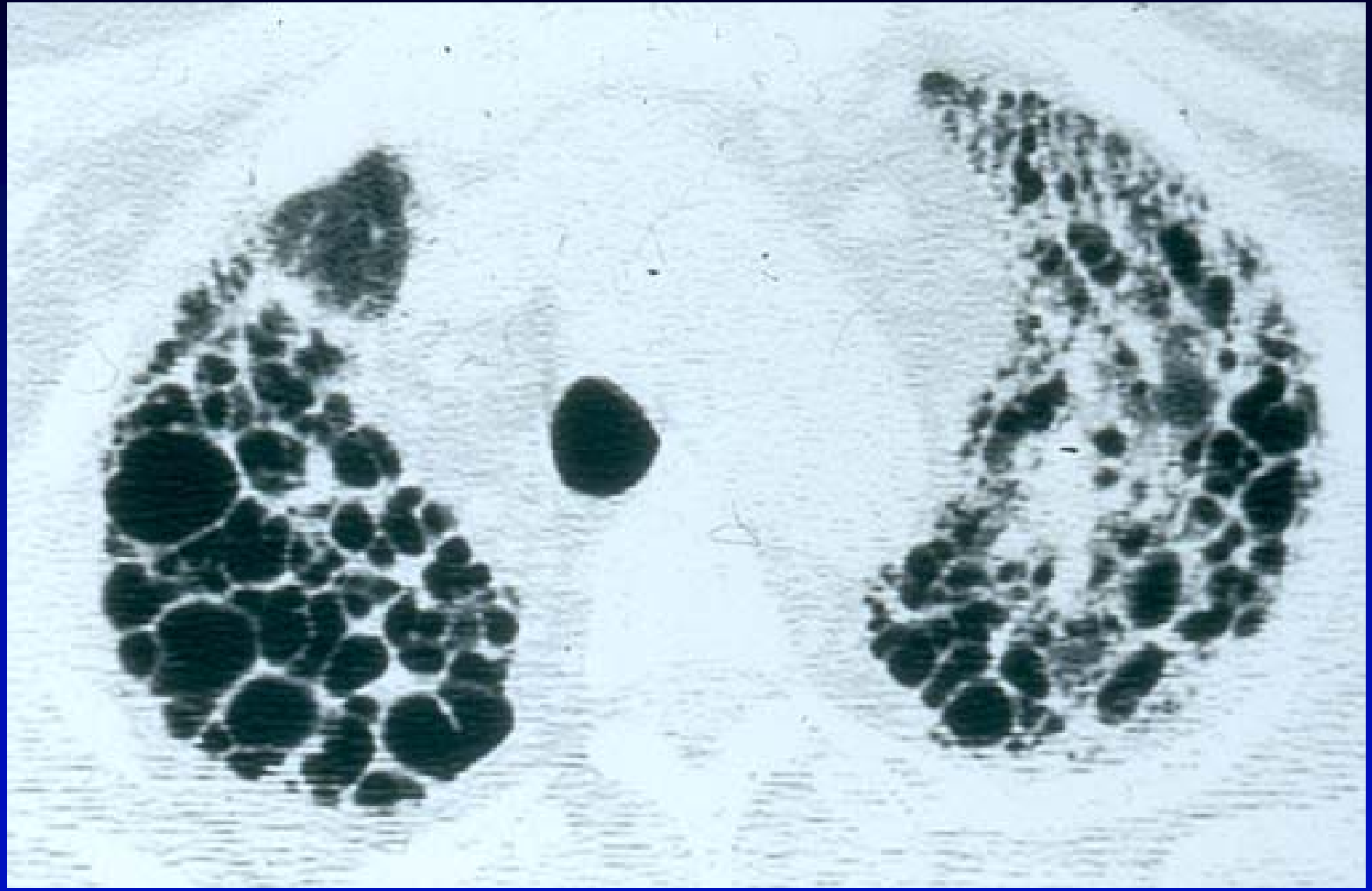


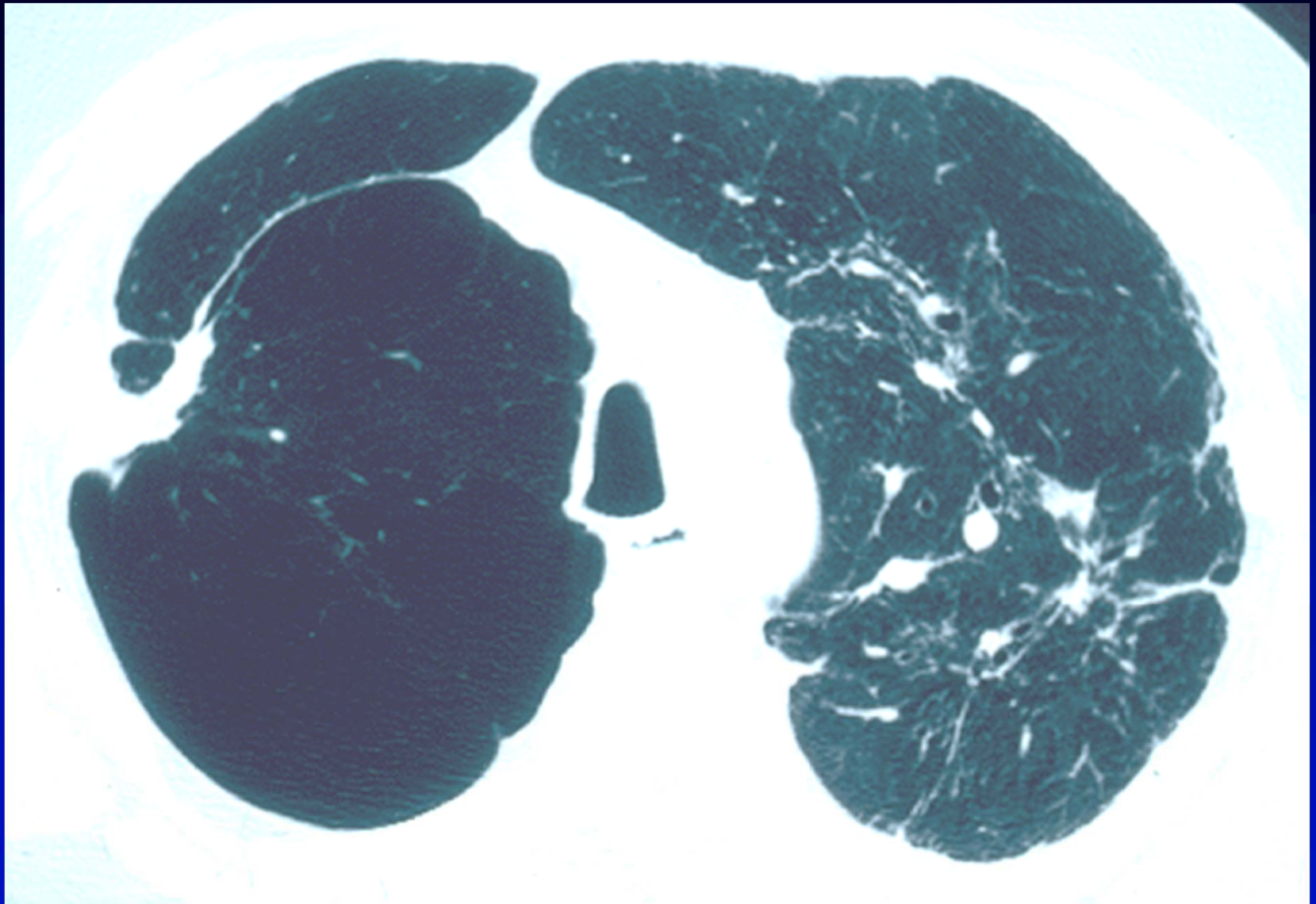


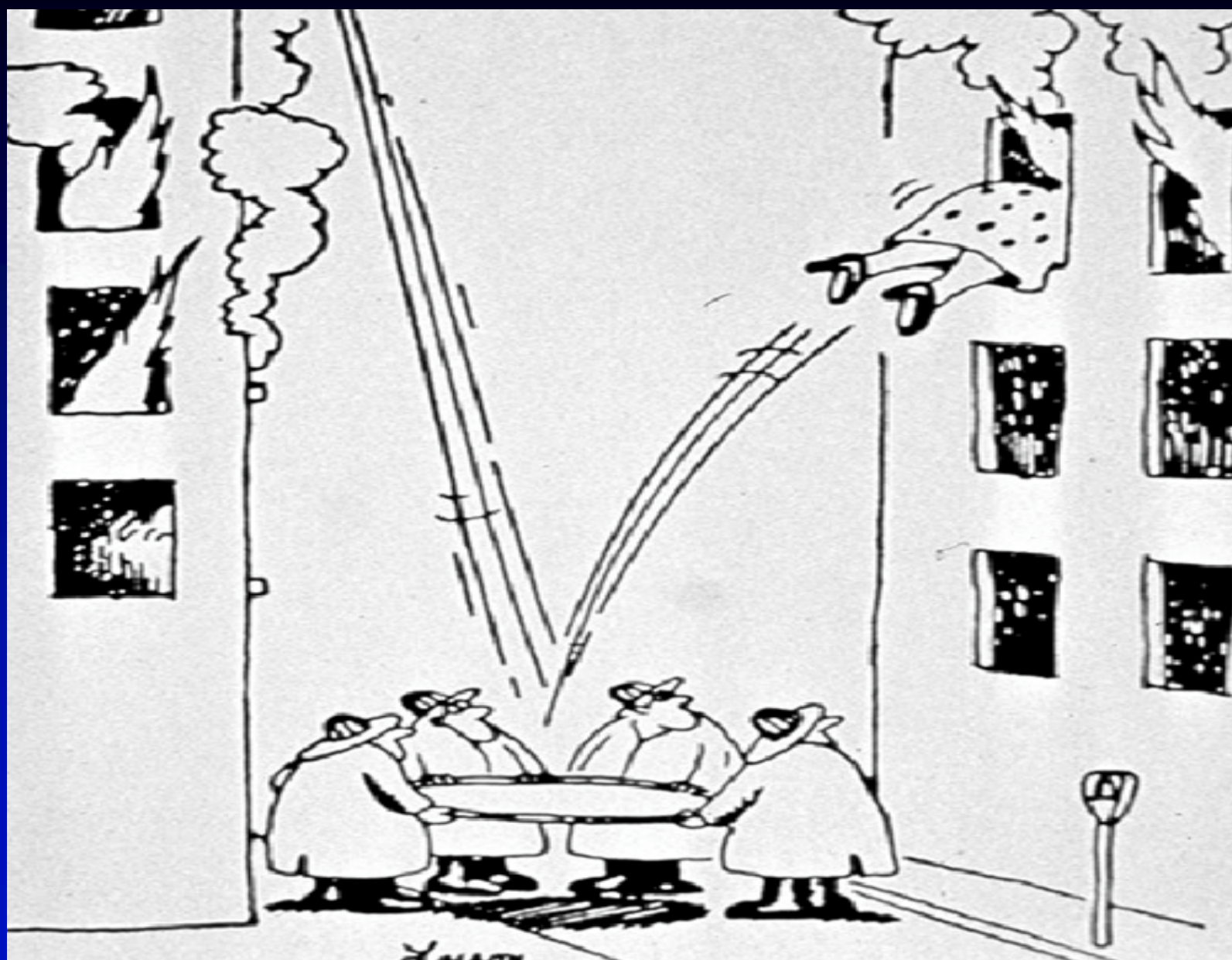












Sarcoidosis: HRCT

Potentially reversible if:

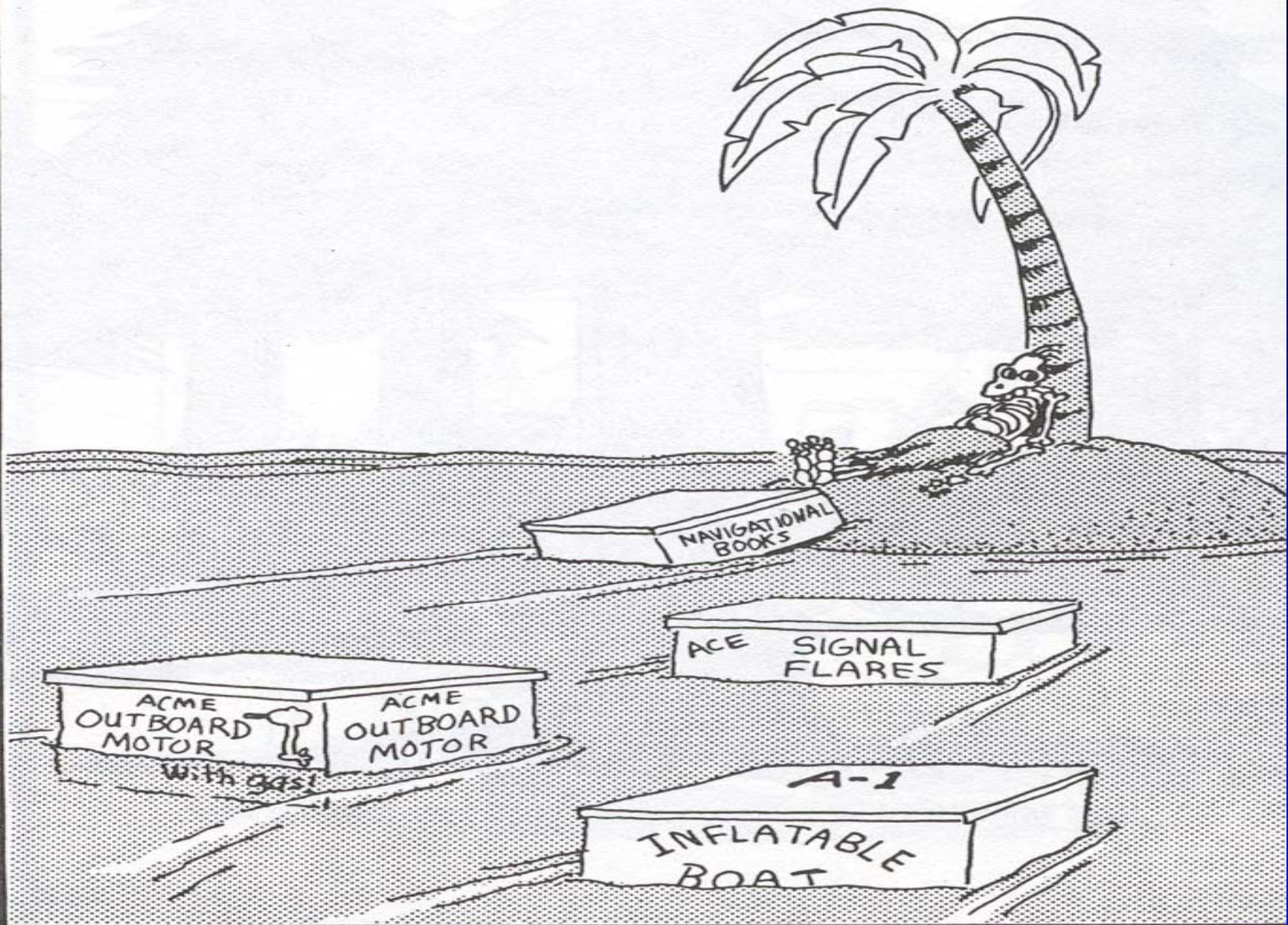
- **Ground glass opacities**
- **Nodules**
- **Consolidation**

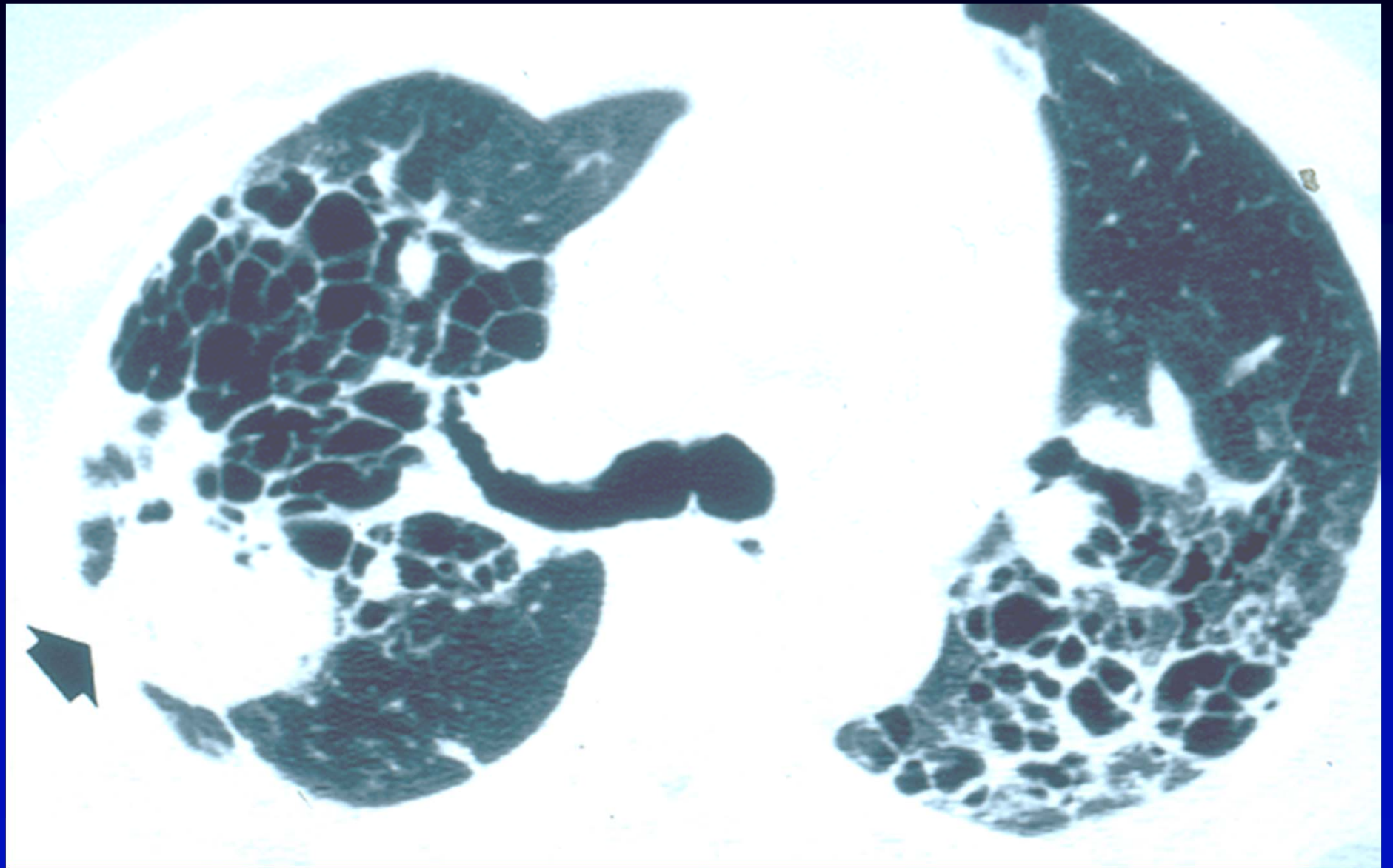
Sarcoidosis: HRCT

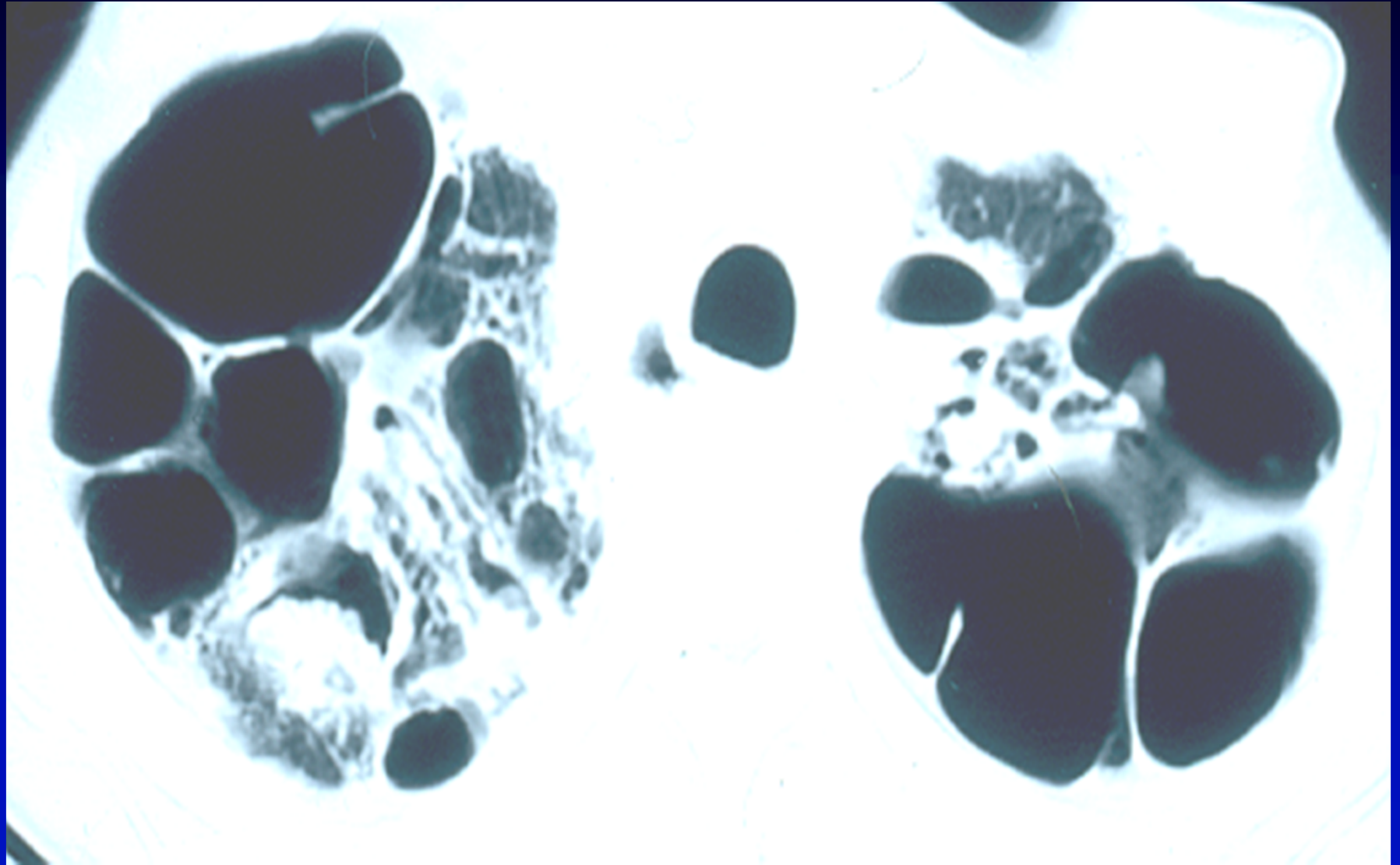
Adverse prognostic factors:

- Honeycomb cysts
- Traction bronchiectasis
- Broad septal bands
- Distortion

Larson

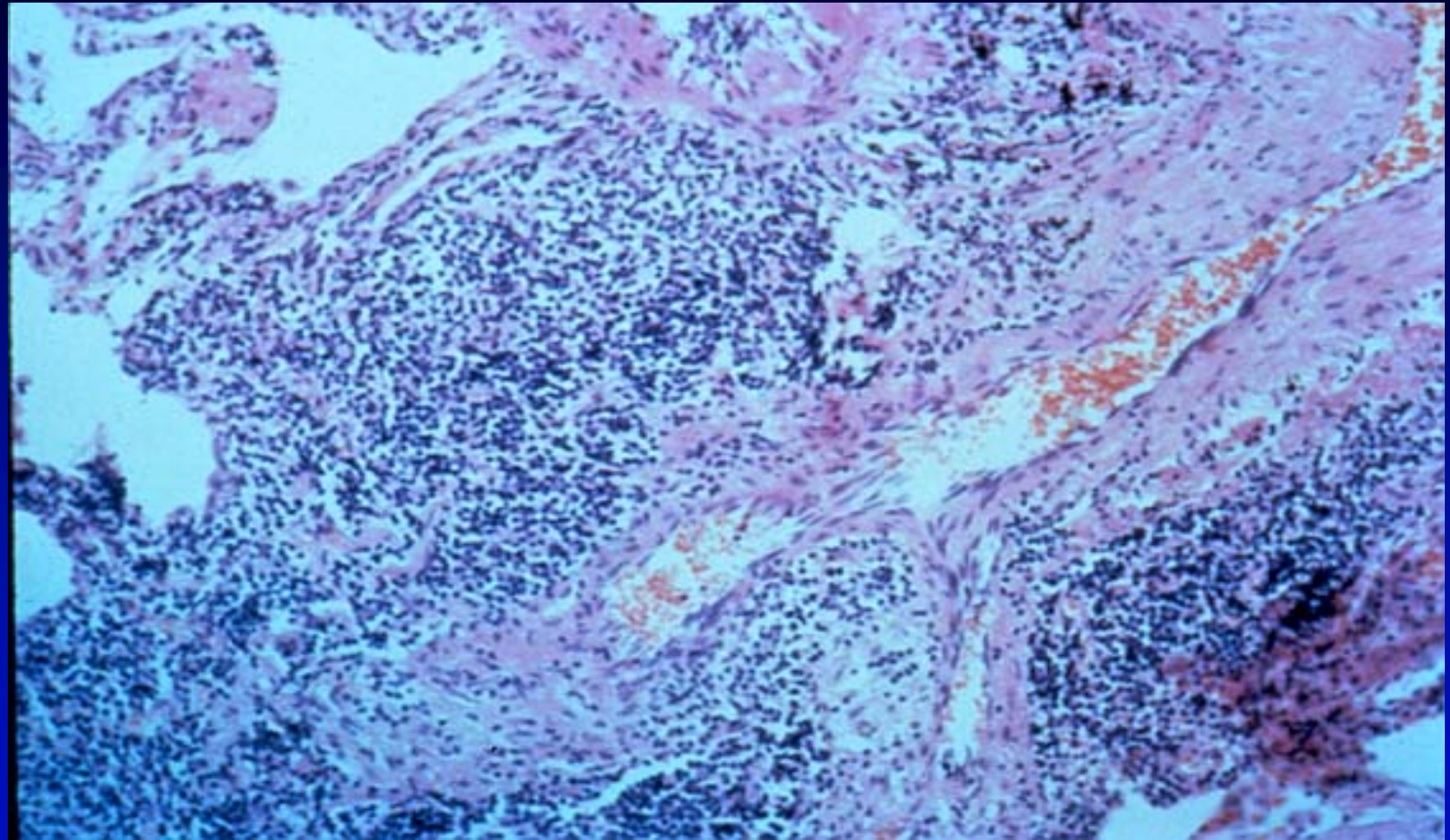


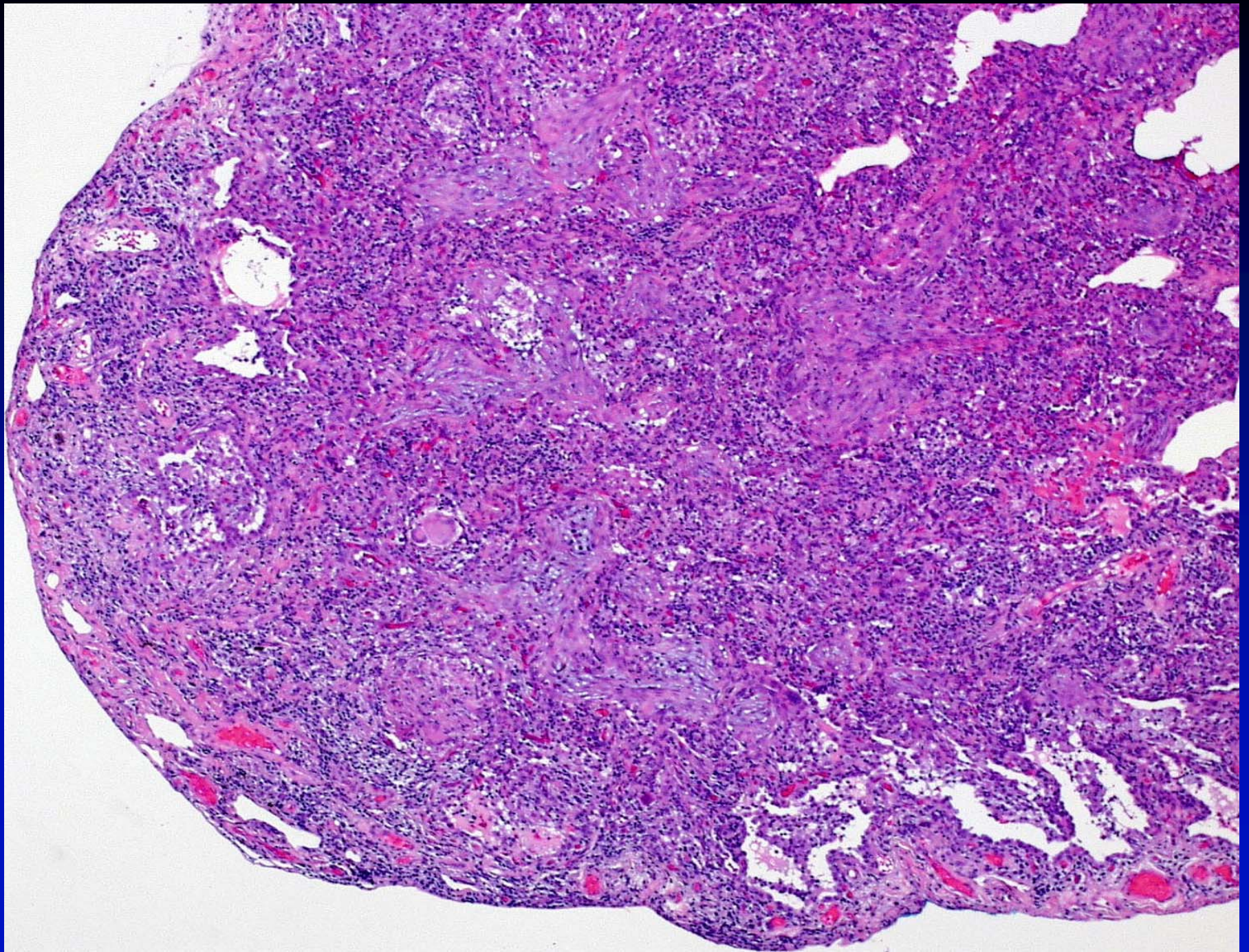


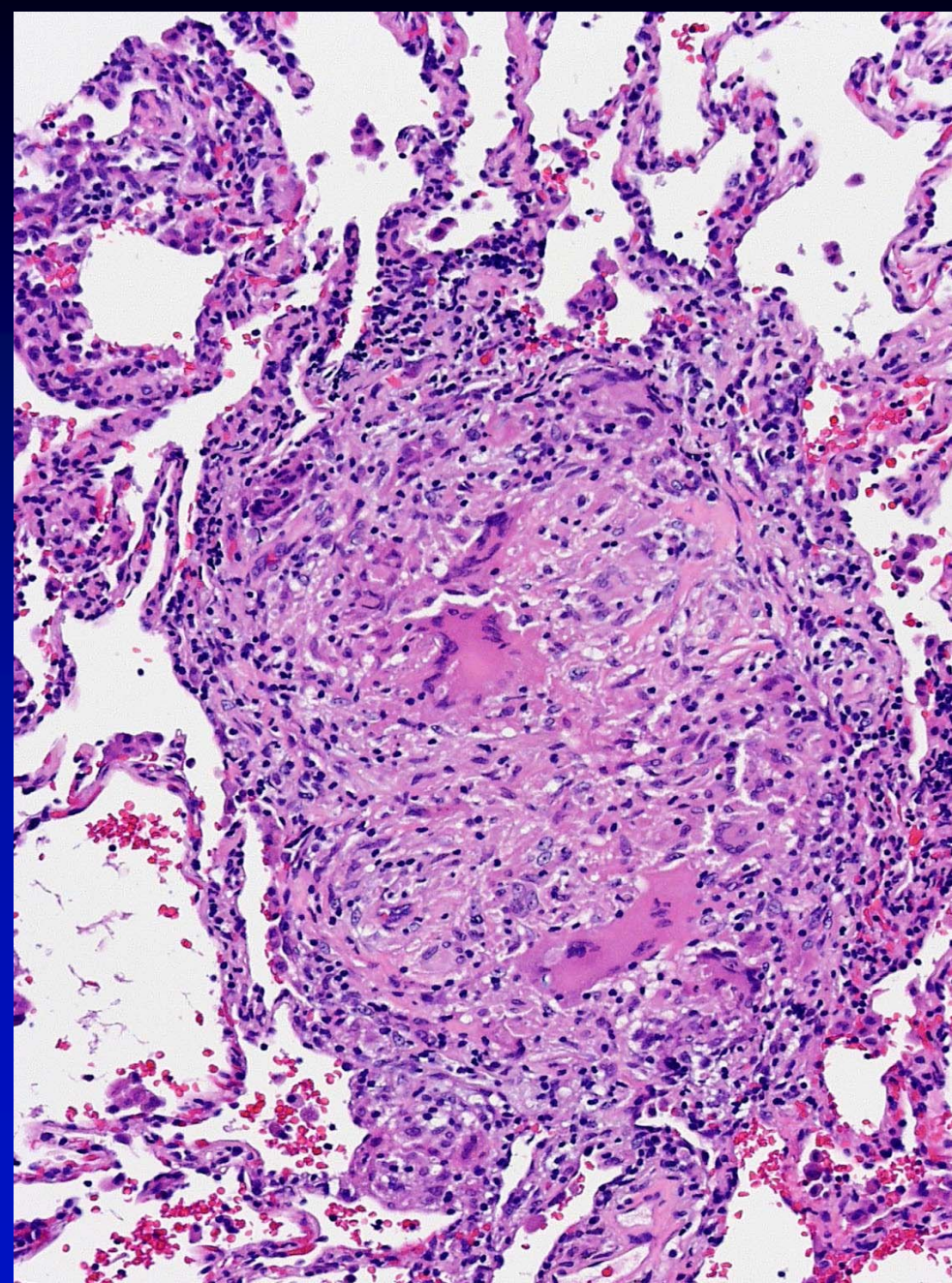
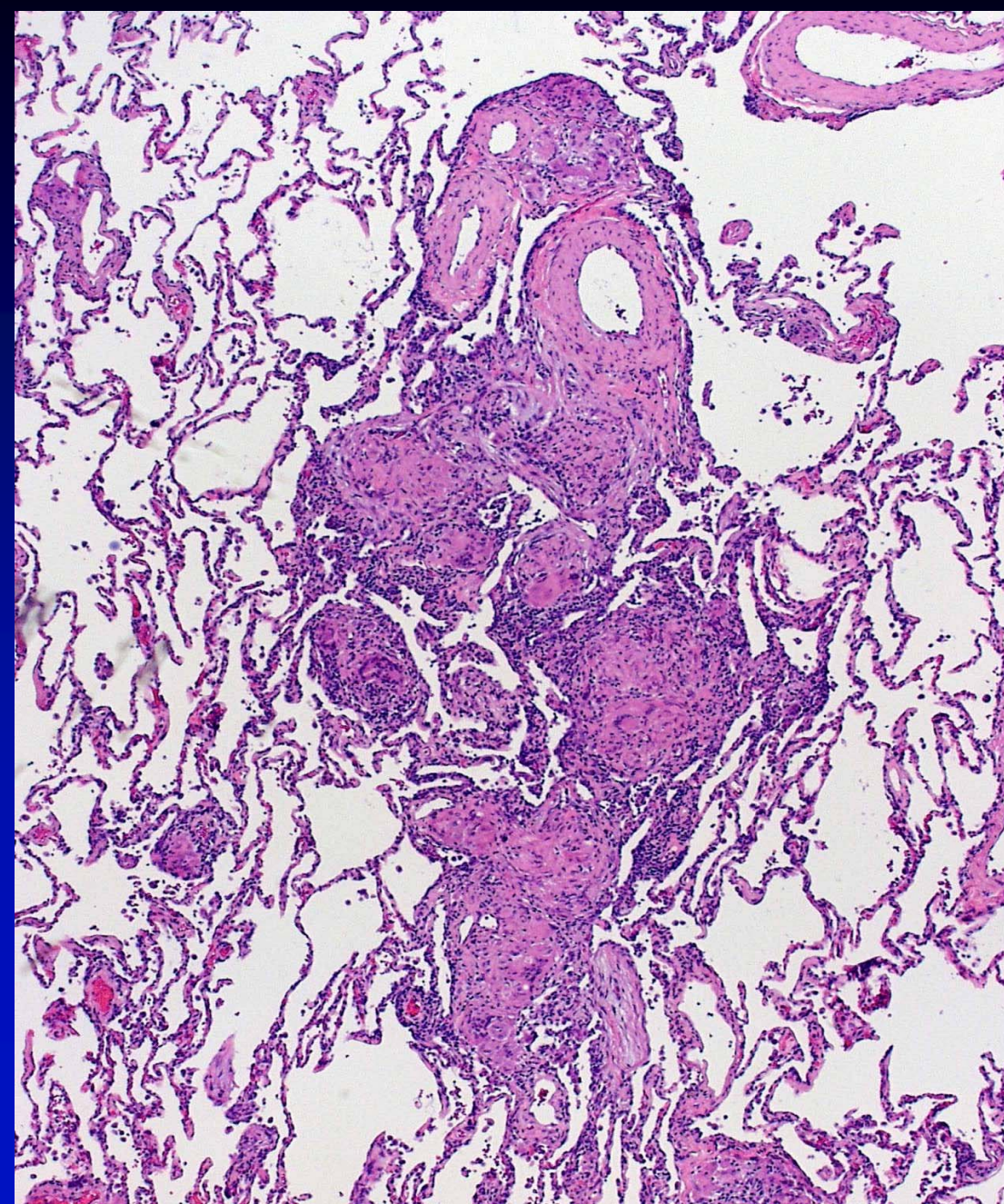




**"I feel a lot better since I ran
out of those pills you gave me."**



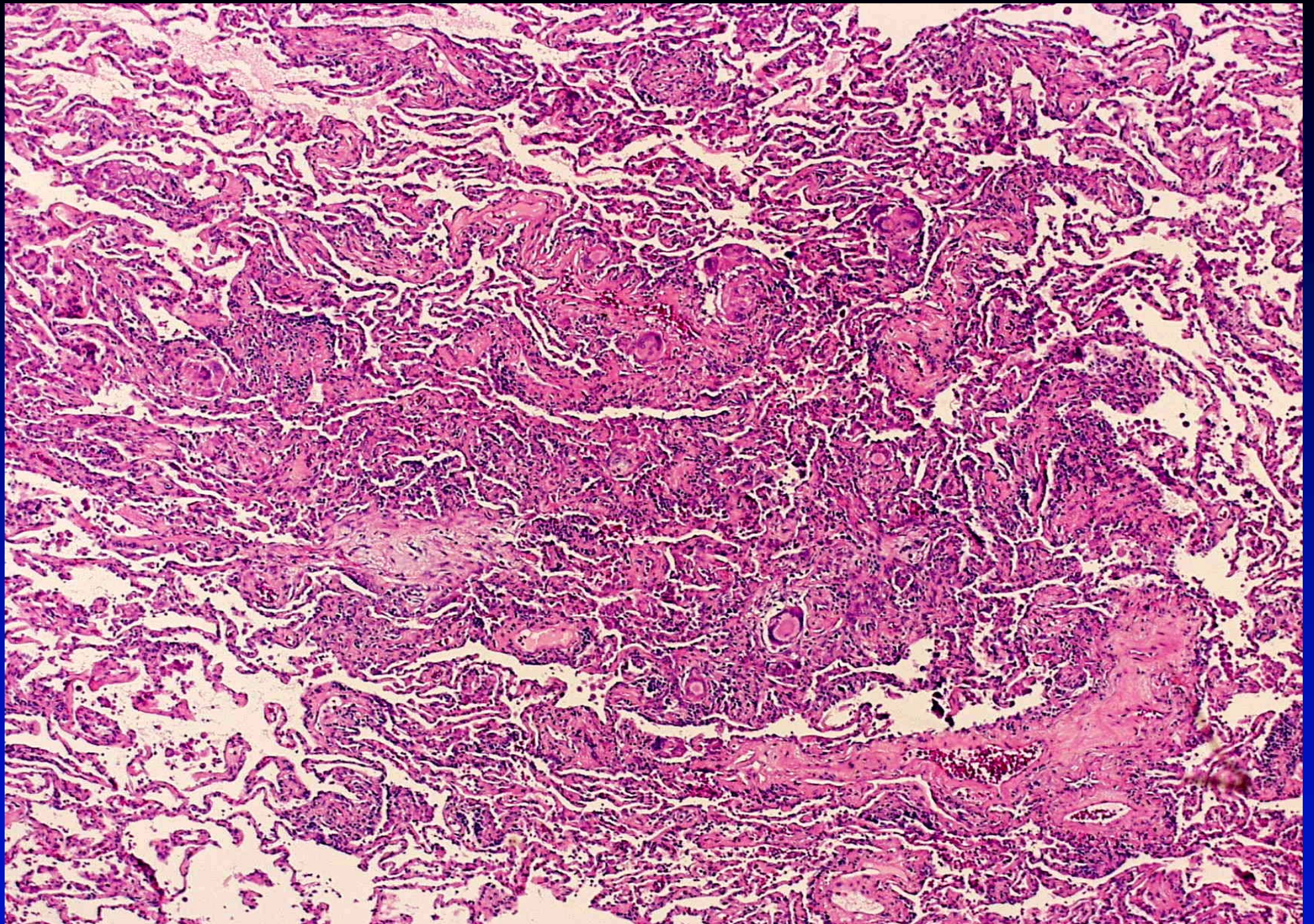


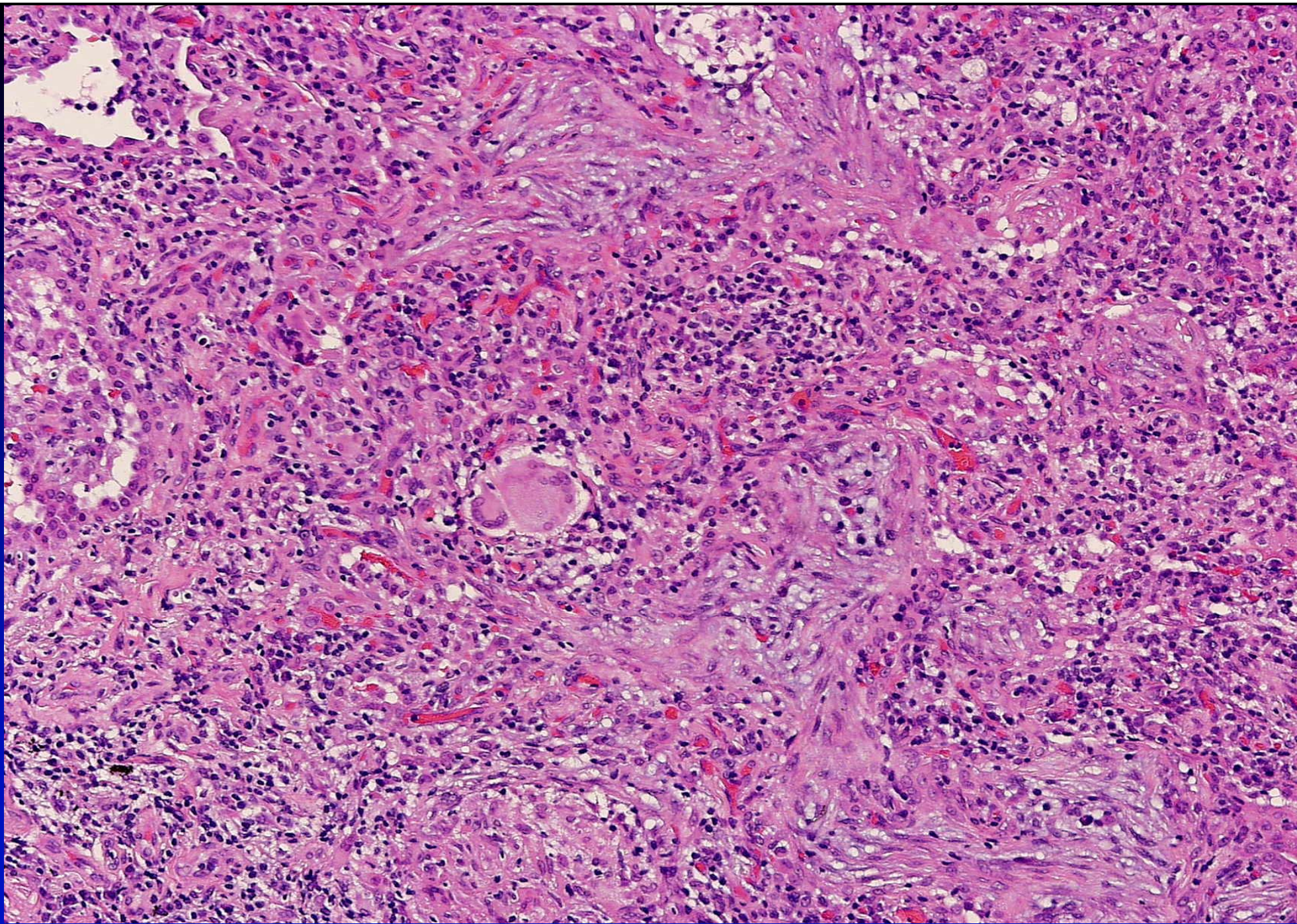


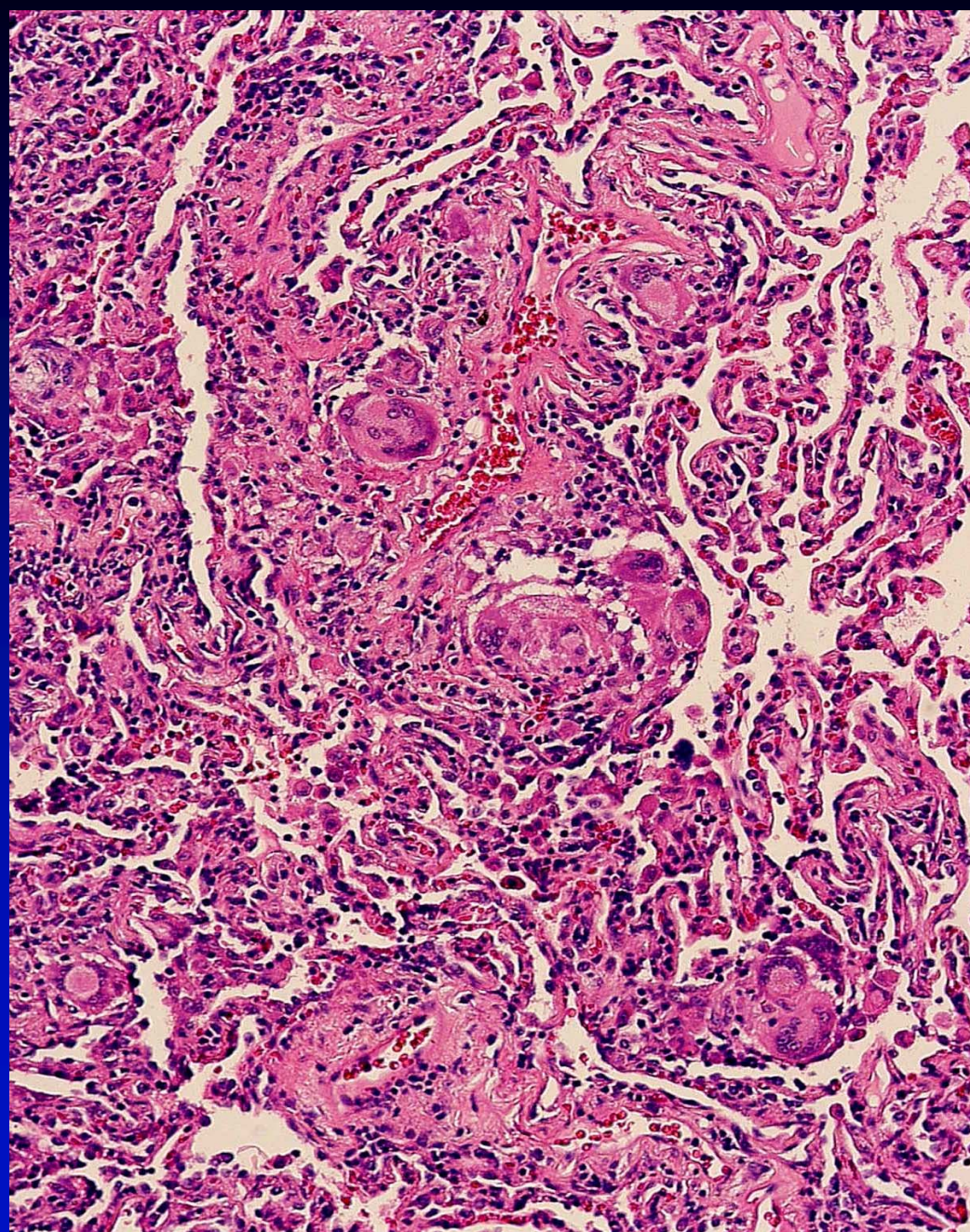
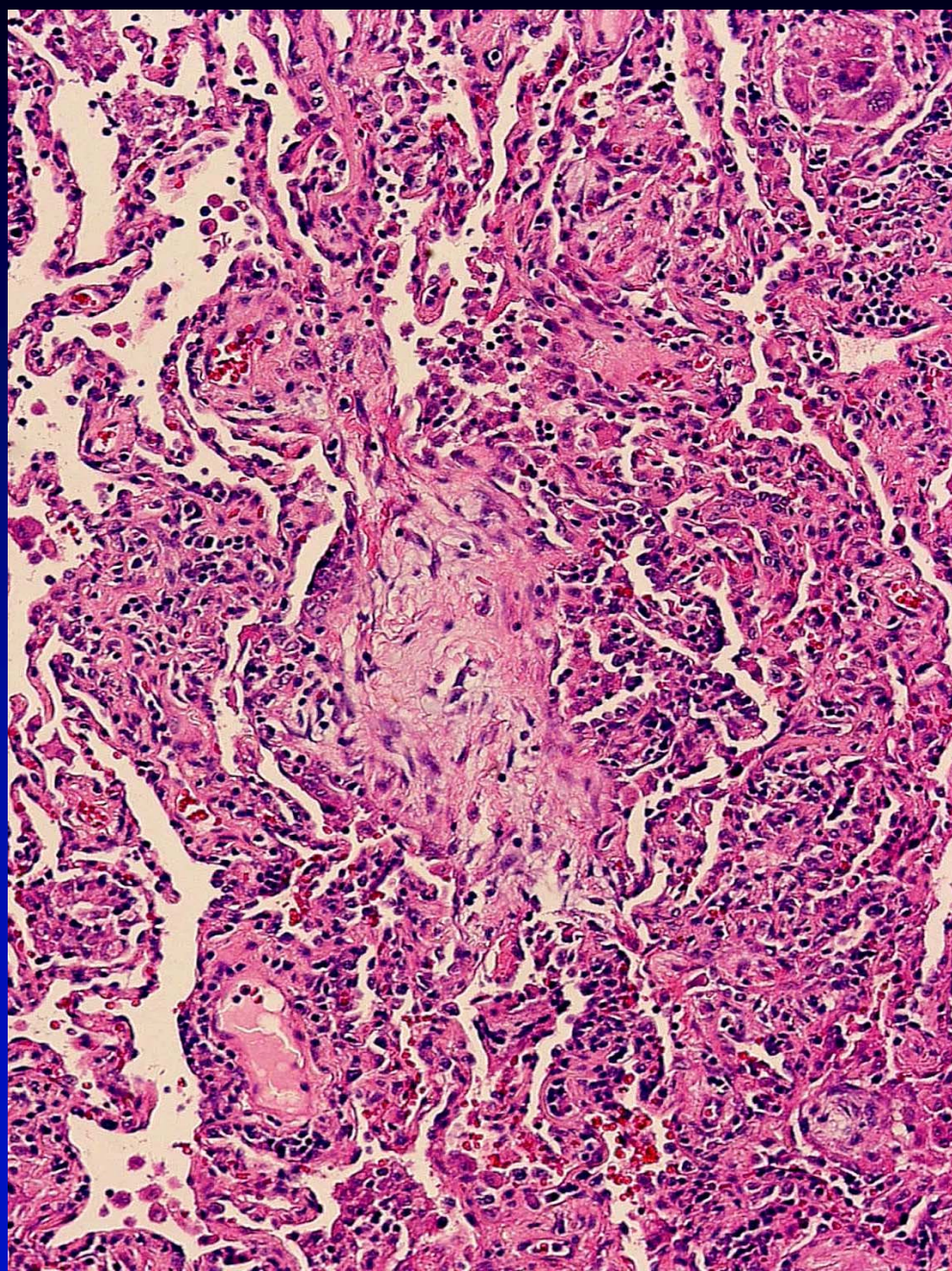
Hypersensitivity pneumonia

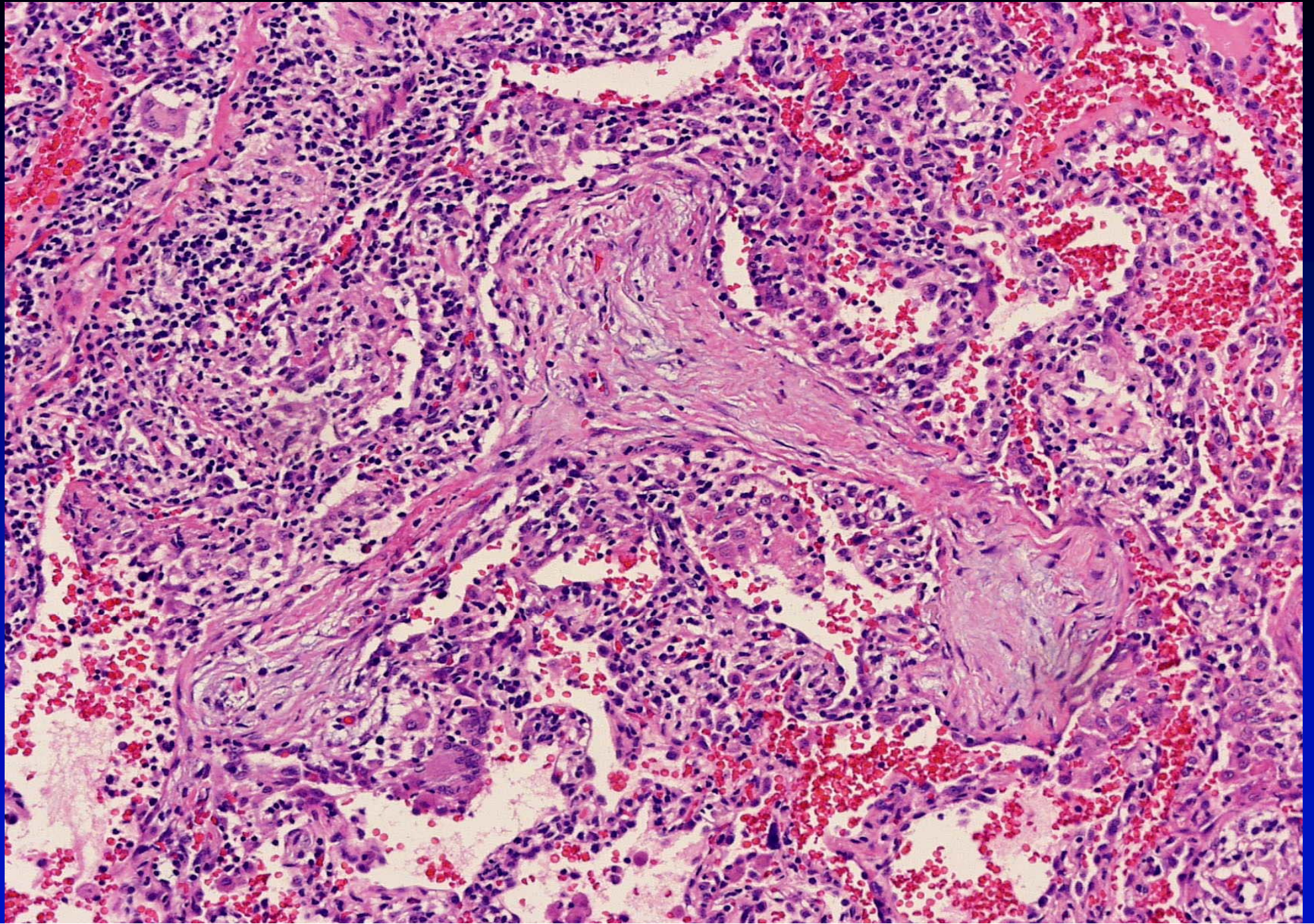
Synonymous with:

- **Extrinsic allergic alveolitis**









Hypersensitivity pneumonia

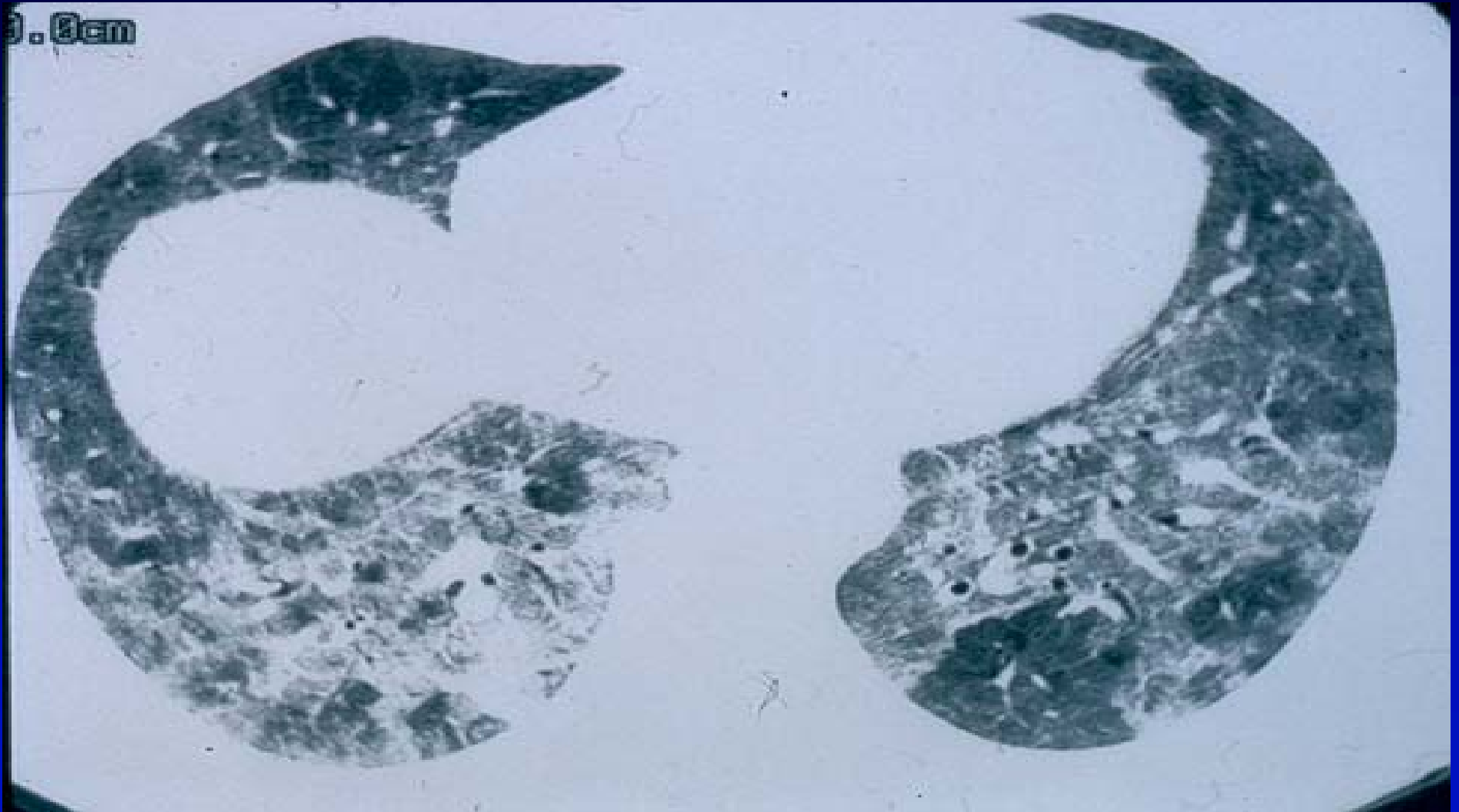
- Environmental exposure
- Serum precipitating antibodies
- BAL lymphocytosis (often > 40%)
- Resolves after cessation exposure

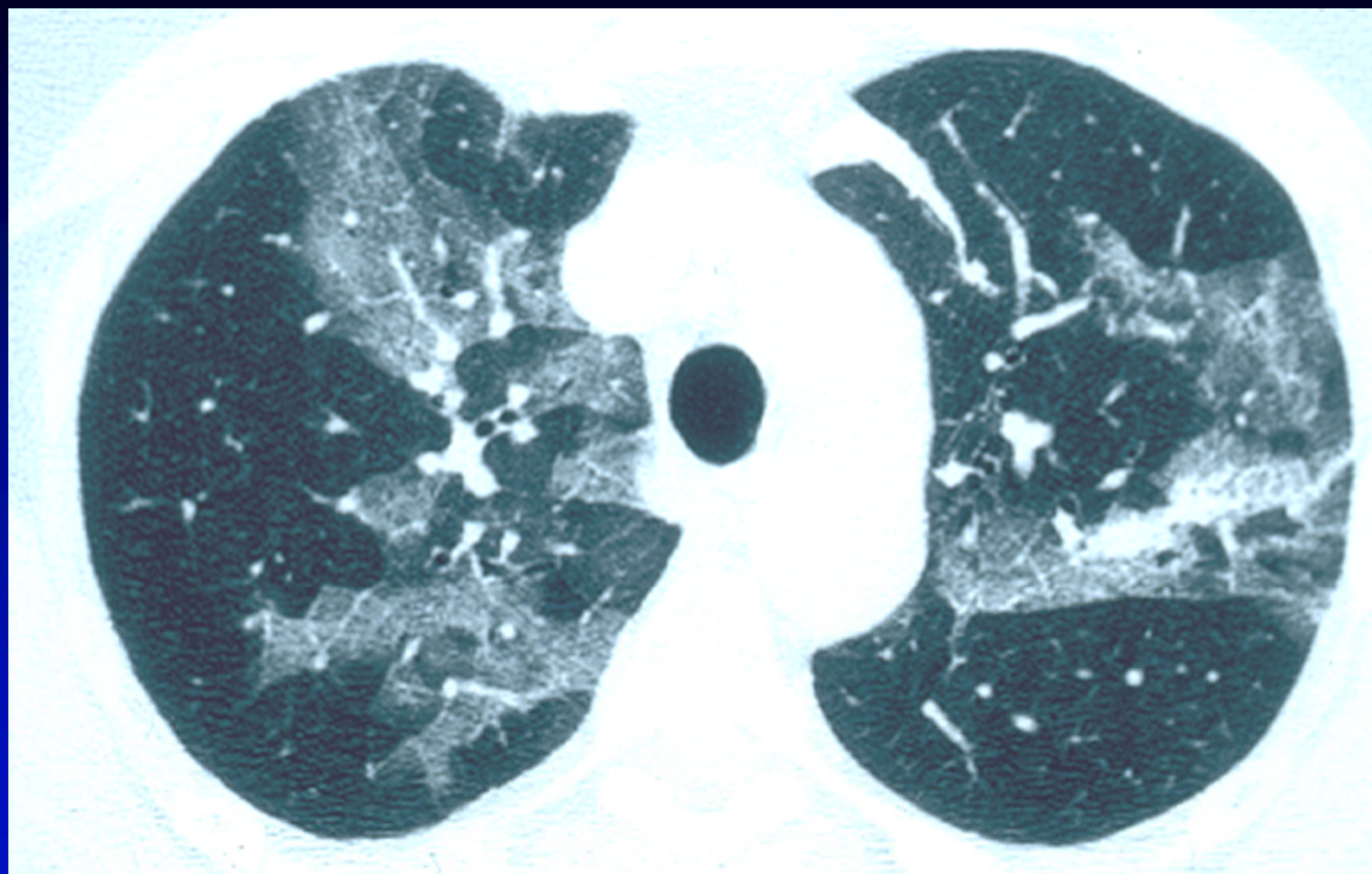
Acute Hypersensitivity Pneumonia

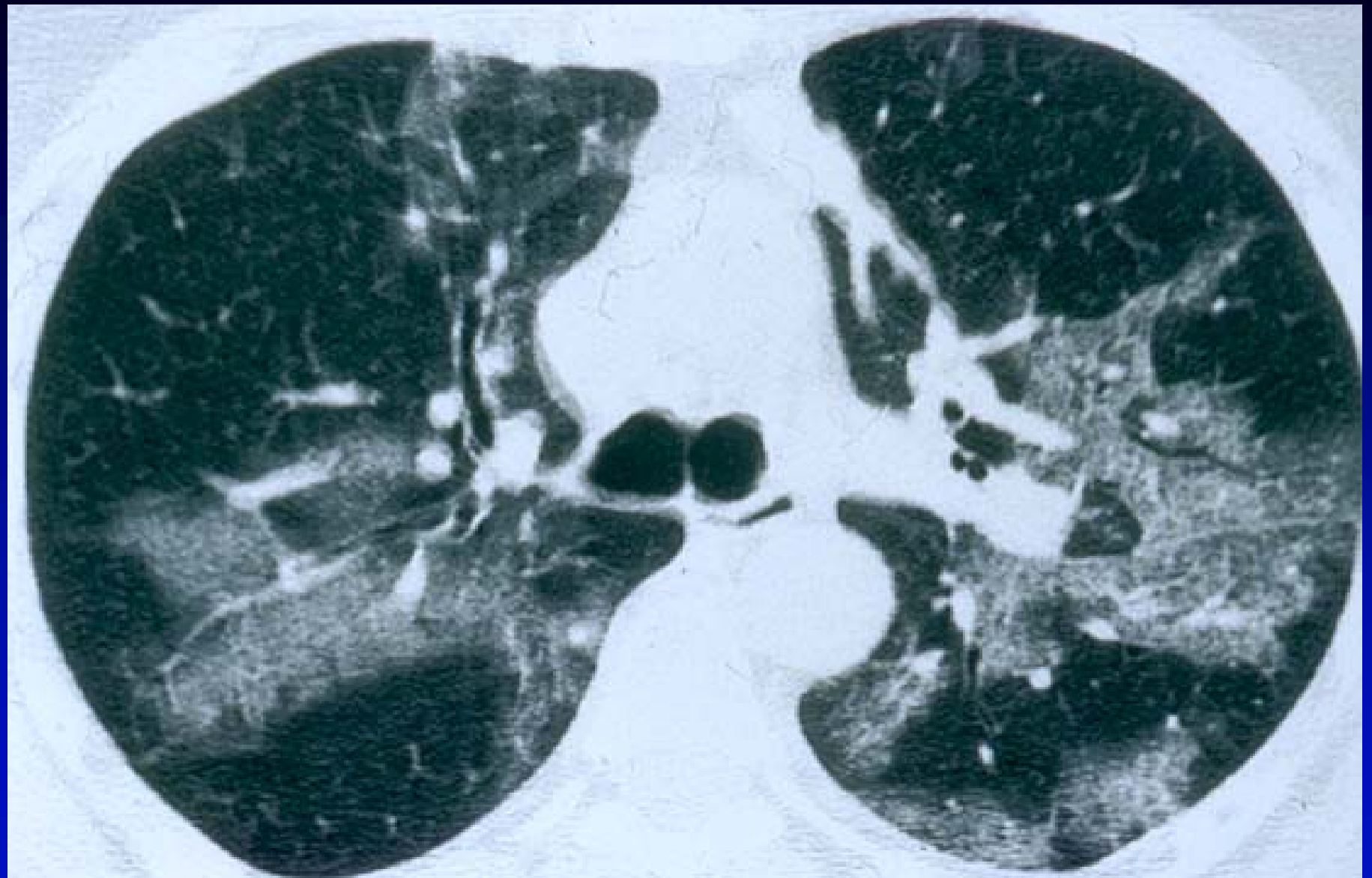
HRCT features of acute HP:

- Patchy ground glass opacities
- Peribronchiolar nodules
- Lack of honeycombing

Hypersensitivity pneumonia: Ground Glass



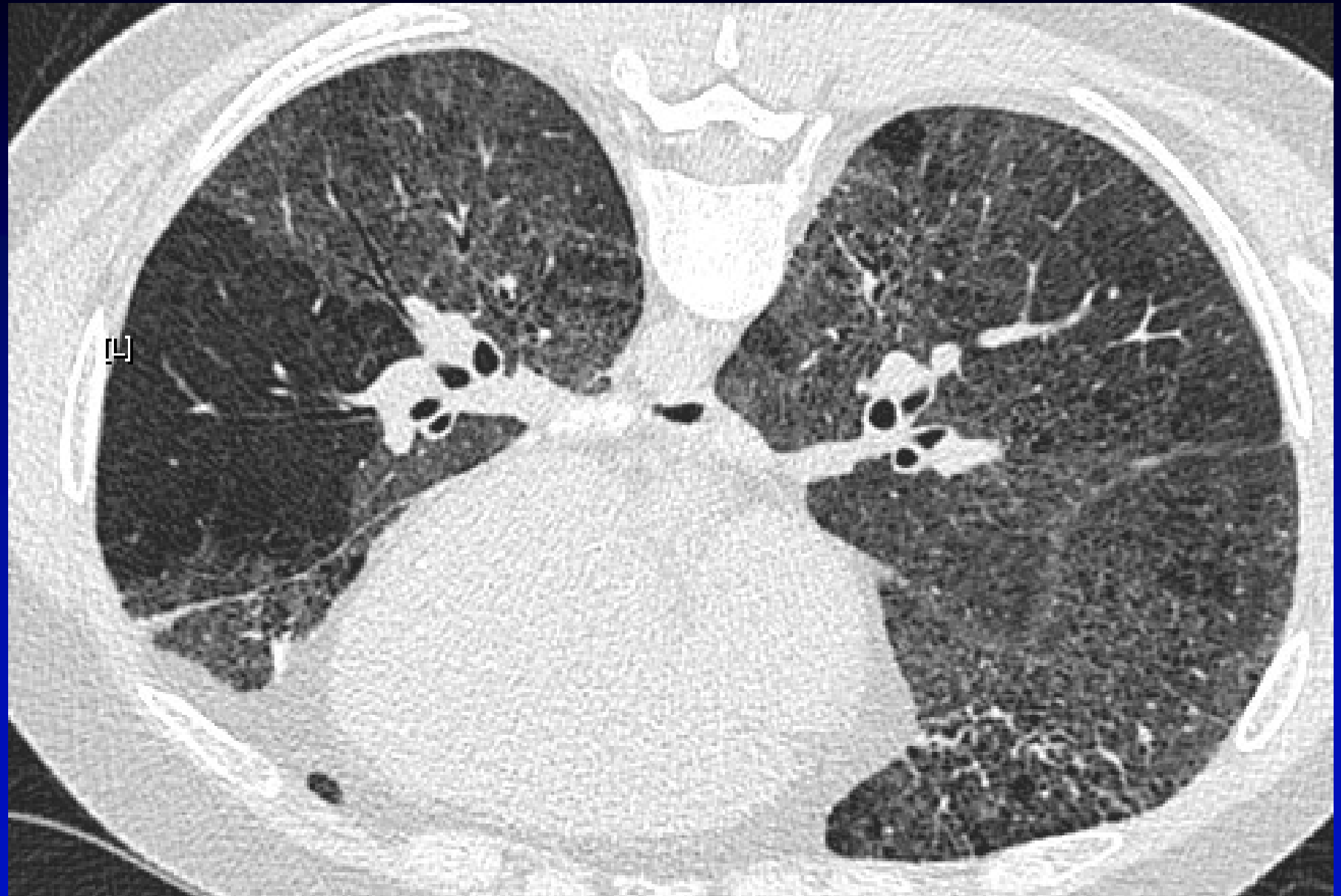


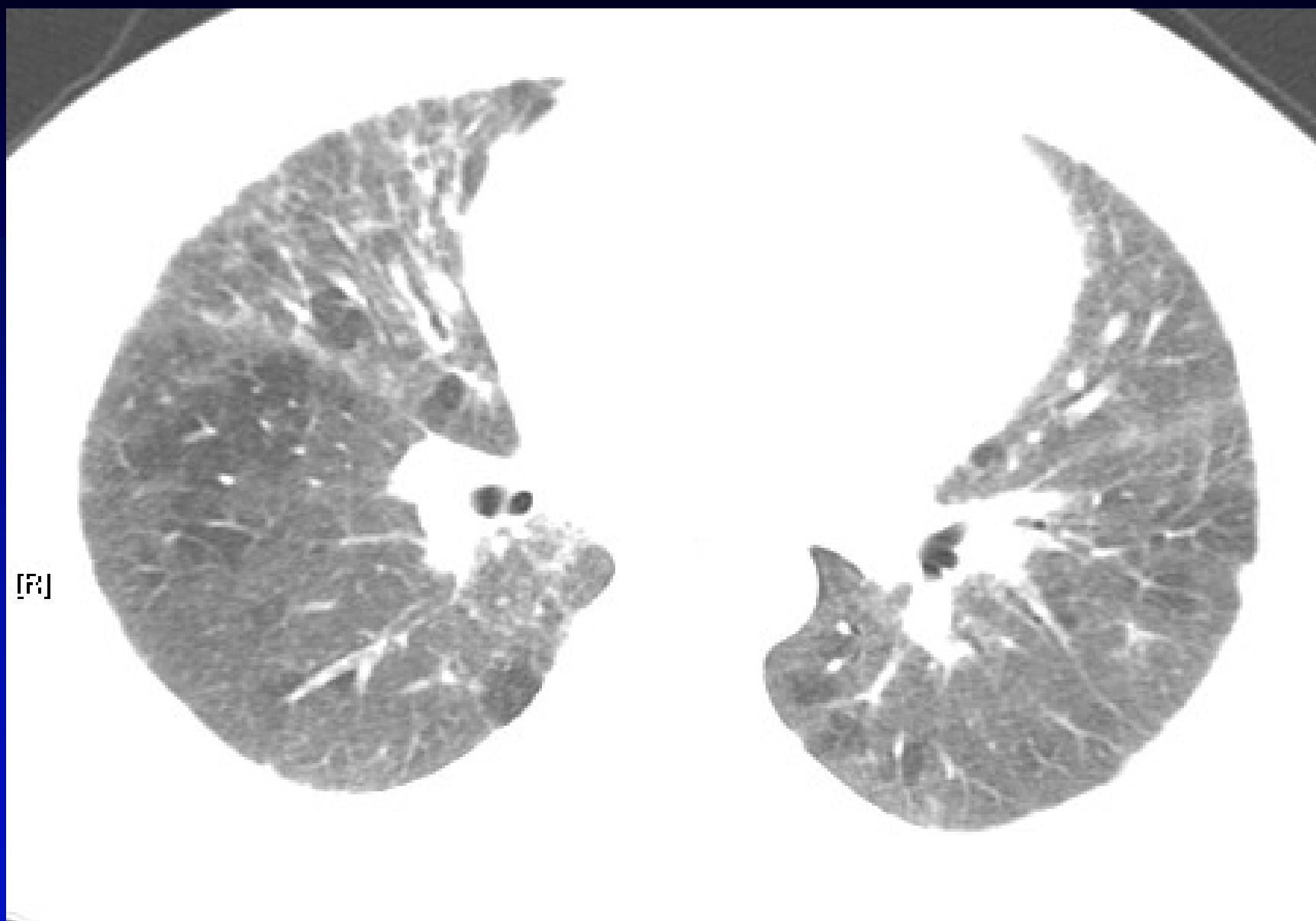


Hypersensitivity pneumonia

CT features:

- Mosaic pattern of attenuation
- Ground glass opacities

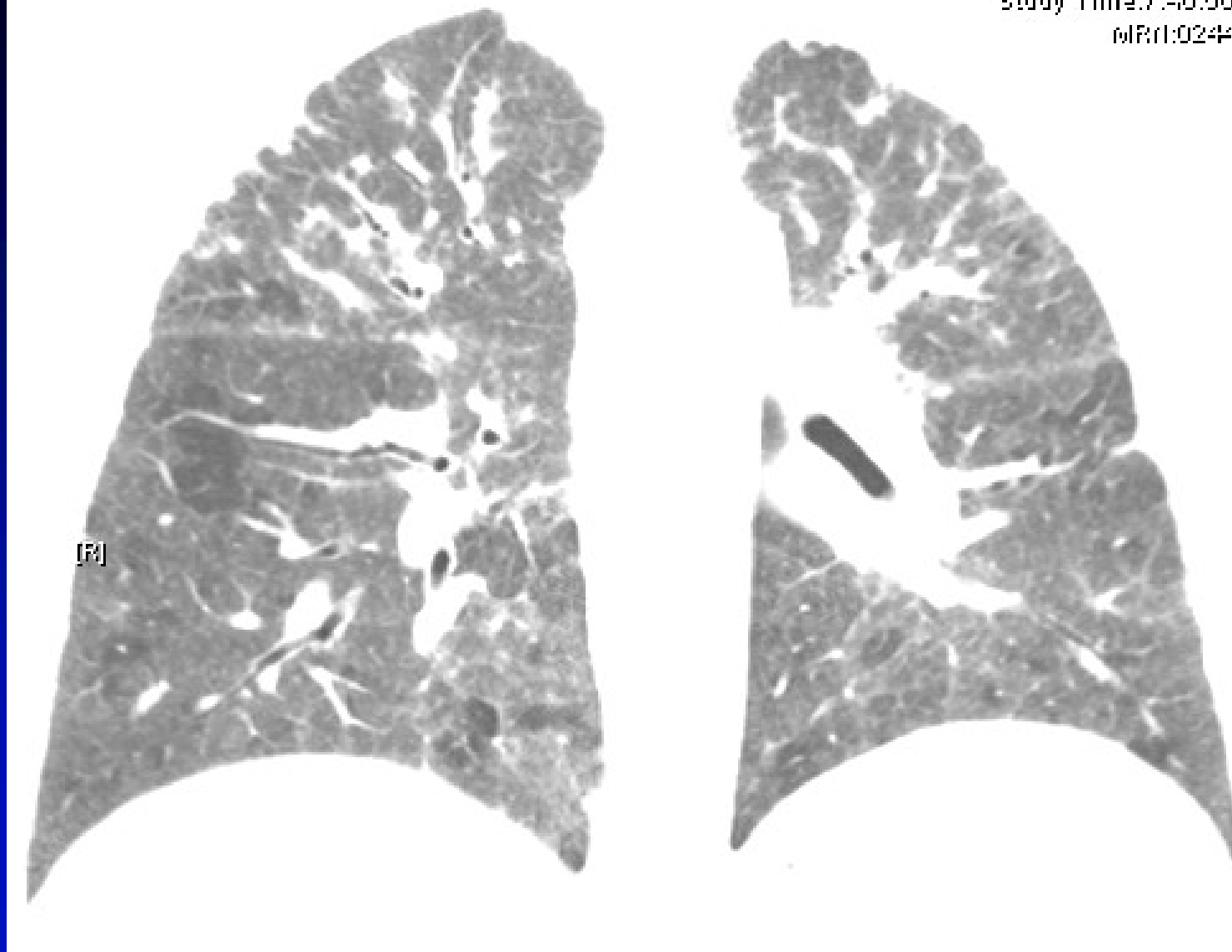




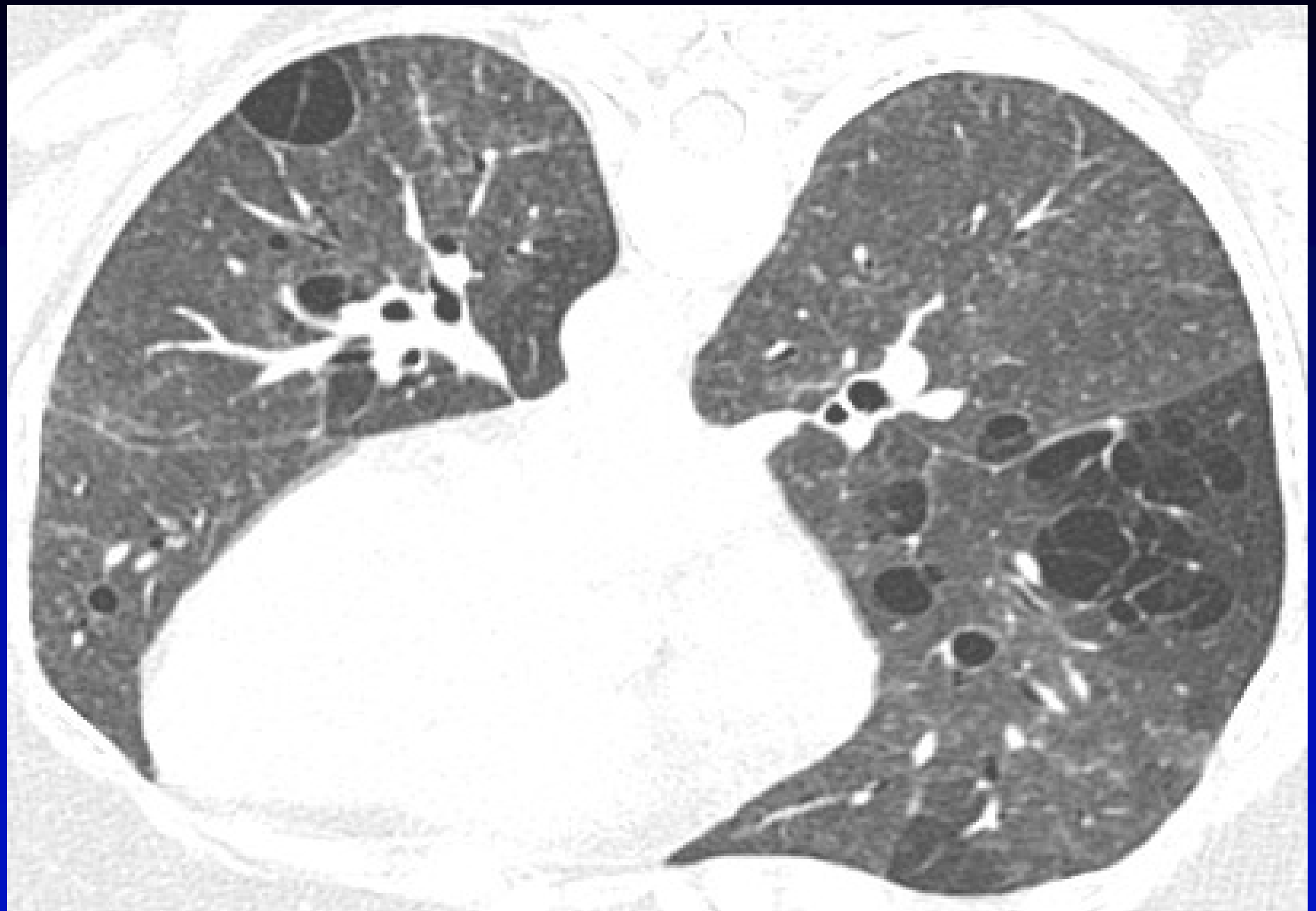
[R]

1m52

Study Date: 2/24/20
Study Time: 7:40:50
MR: 0244







Chronic Hypersensitivity Pneumonia

HRCT features of chronic HP:

- Reticular, linear
- Peribronchiolar nodules
- Honeycombing may be present

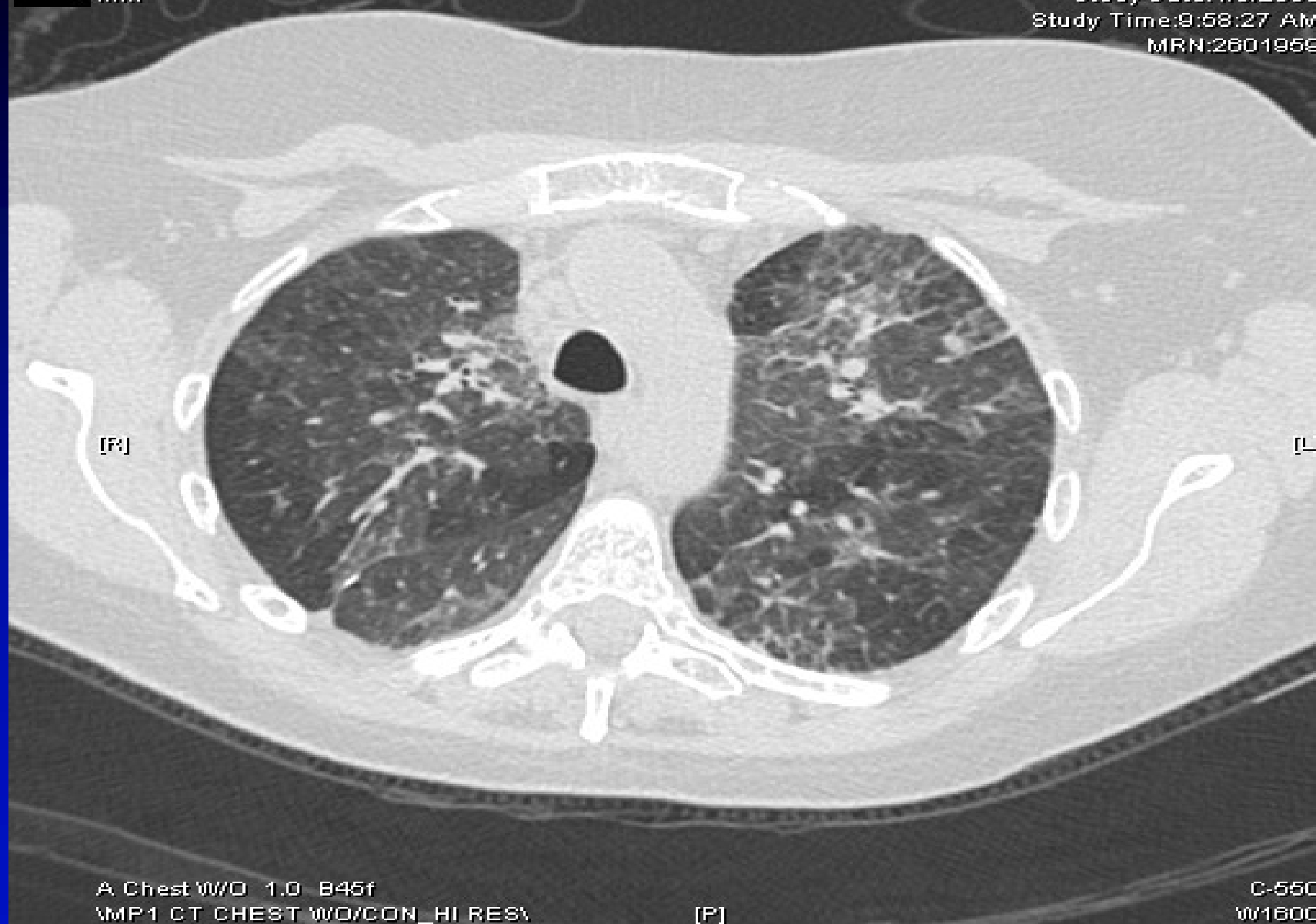


Set:3
Im:7

Progressive Wavelet Level: Full

[A]

Study Date:1/3/2008
Study Time:9:58:27 AM
MRN:2601959





Se:3
Im:8

Progressive Wavelet Level: Full
[A]

Study Date:1/3/2008
Study Time:9:58:27 AM
MRN:2601959



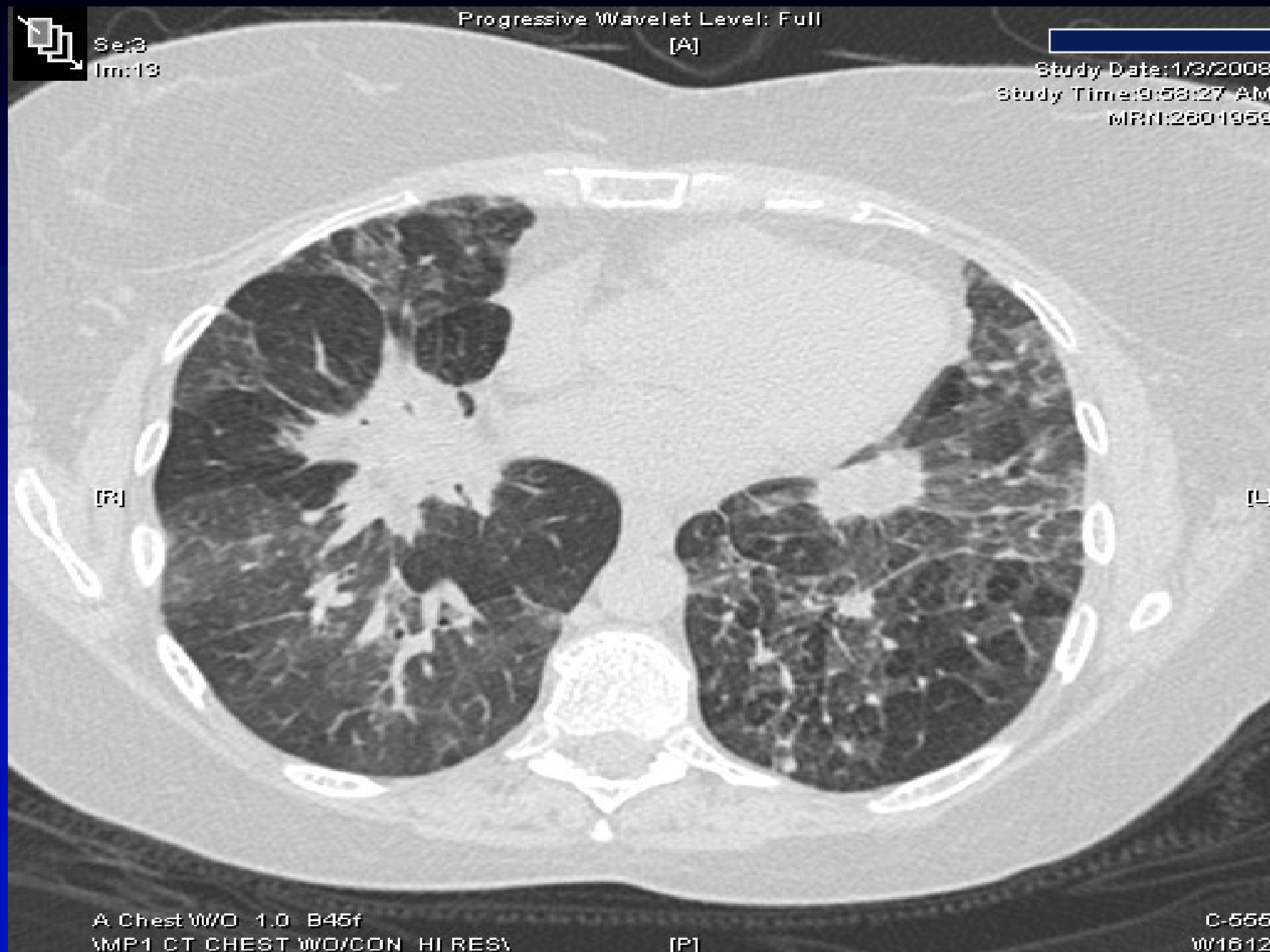


Se:3
Im:13

Progressive Wavelet Level: Full

[A]

Study Date: 1/3/2008
Study Time: 9:58:27 AM
MRN: 2601959



A Chest W/O 1.0 B45f
VMP1 CT CHEST W/O/CON_HI RES\

[P]

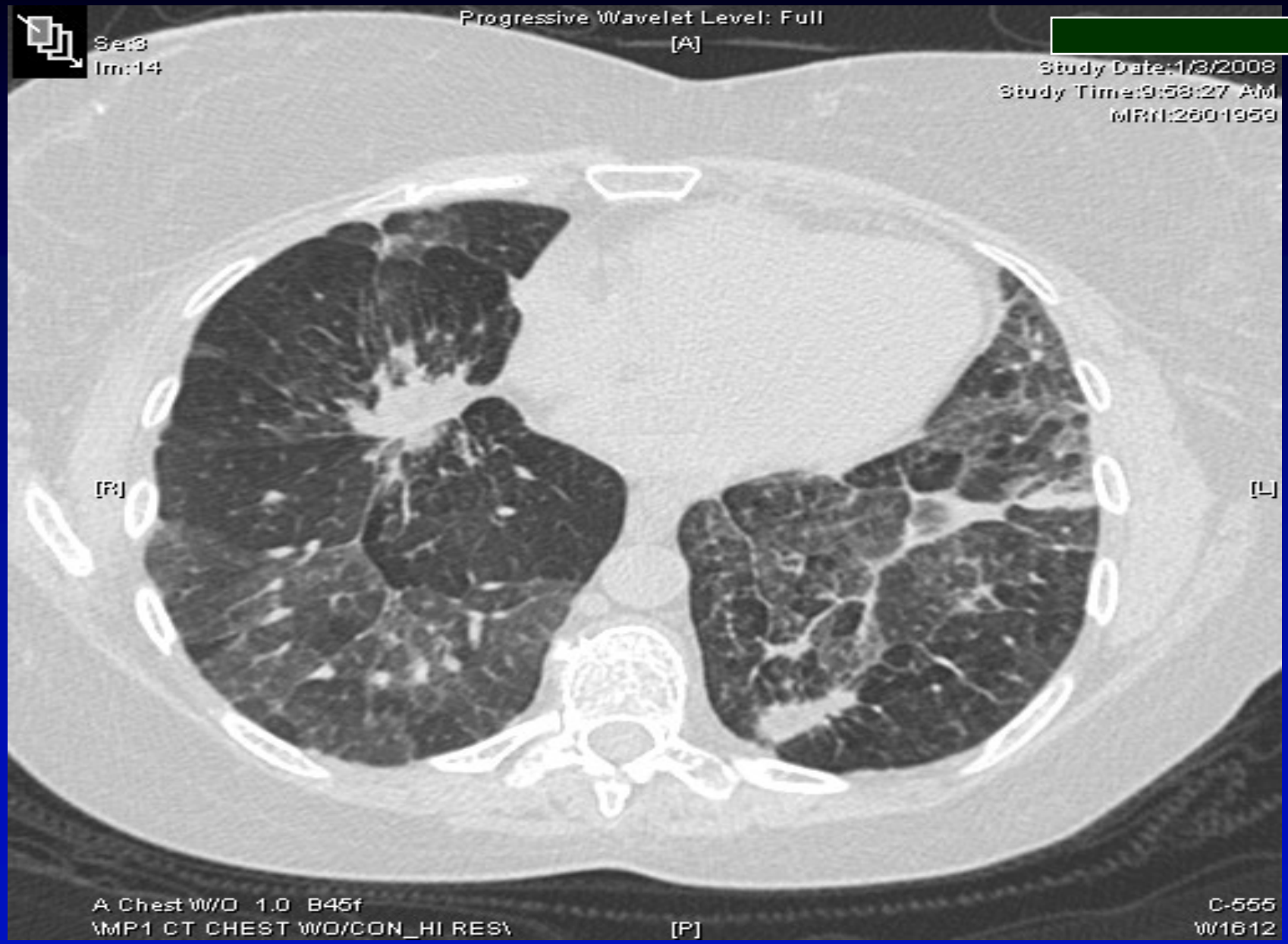
C-555
W1612



Set:
Im:14

Progressive Wavelet Level: Full
[A]

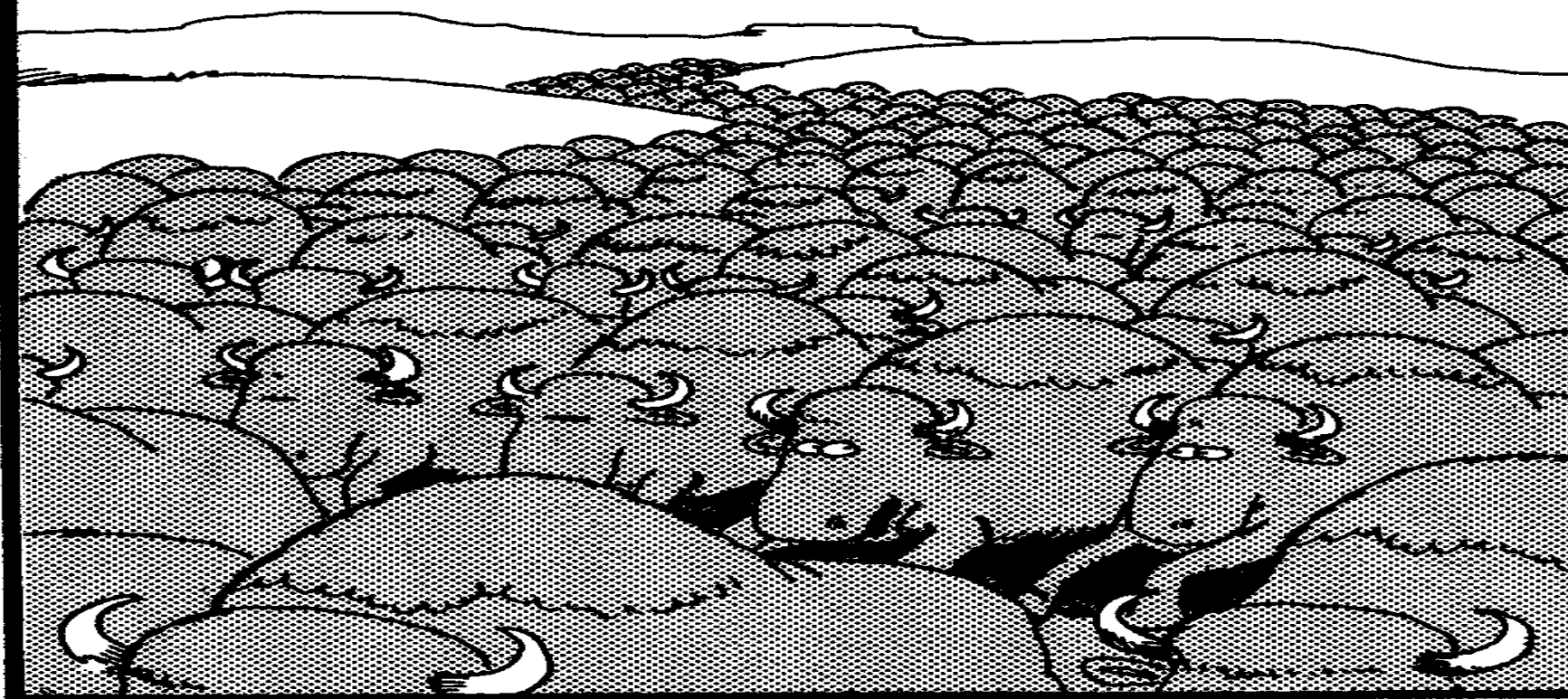
Study Date:1/3/2008
Study Time:9:58:27 AM
MRN:2601959



Treatment of HP

- **Cease exposure to offending antigen(s)**
- **Corticosteroids (severe cases)**

Larson

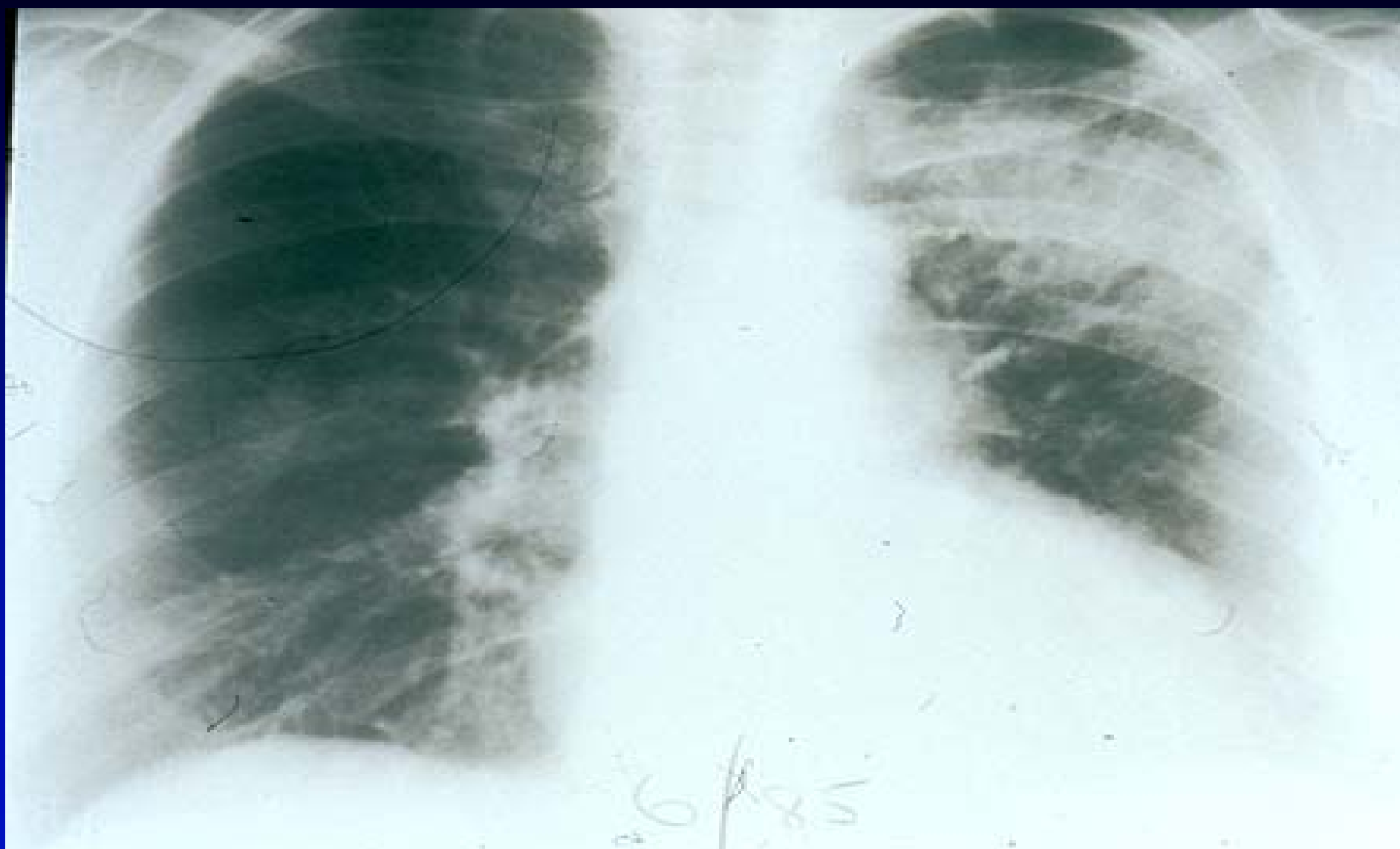


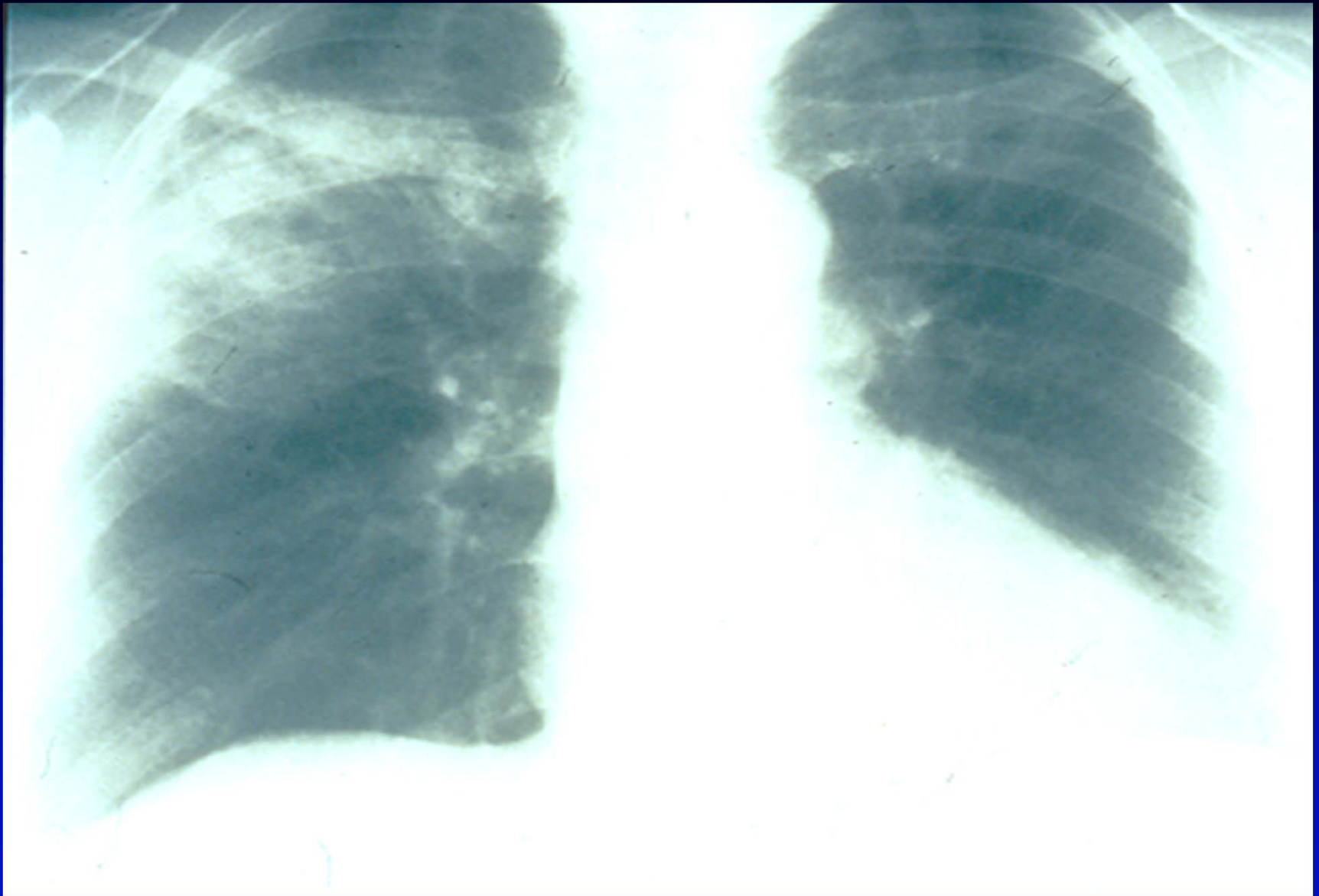
"As if we all knew where we're going."

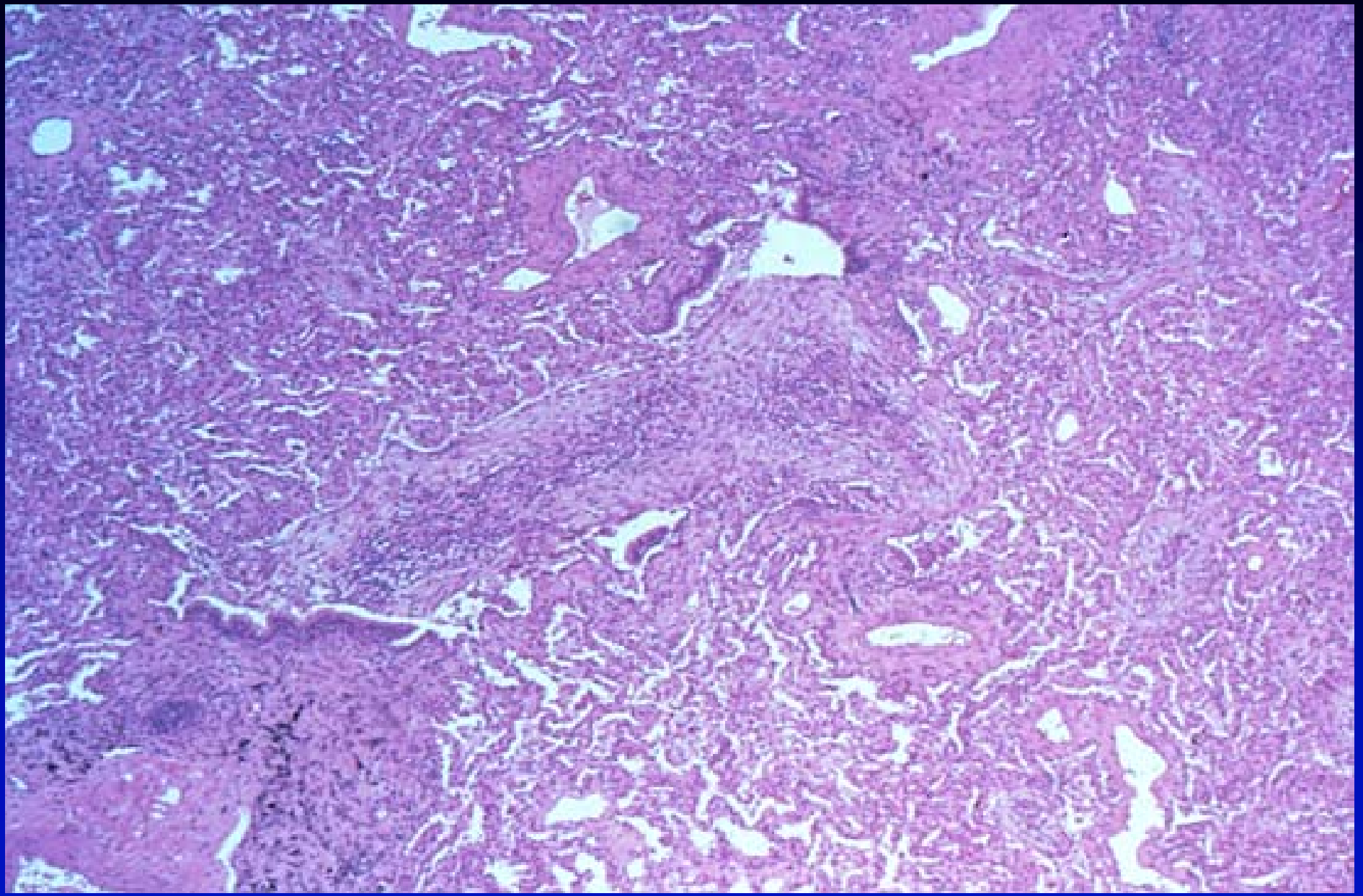
Cryptogenic Organizing Pneumonia

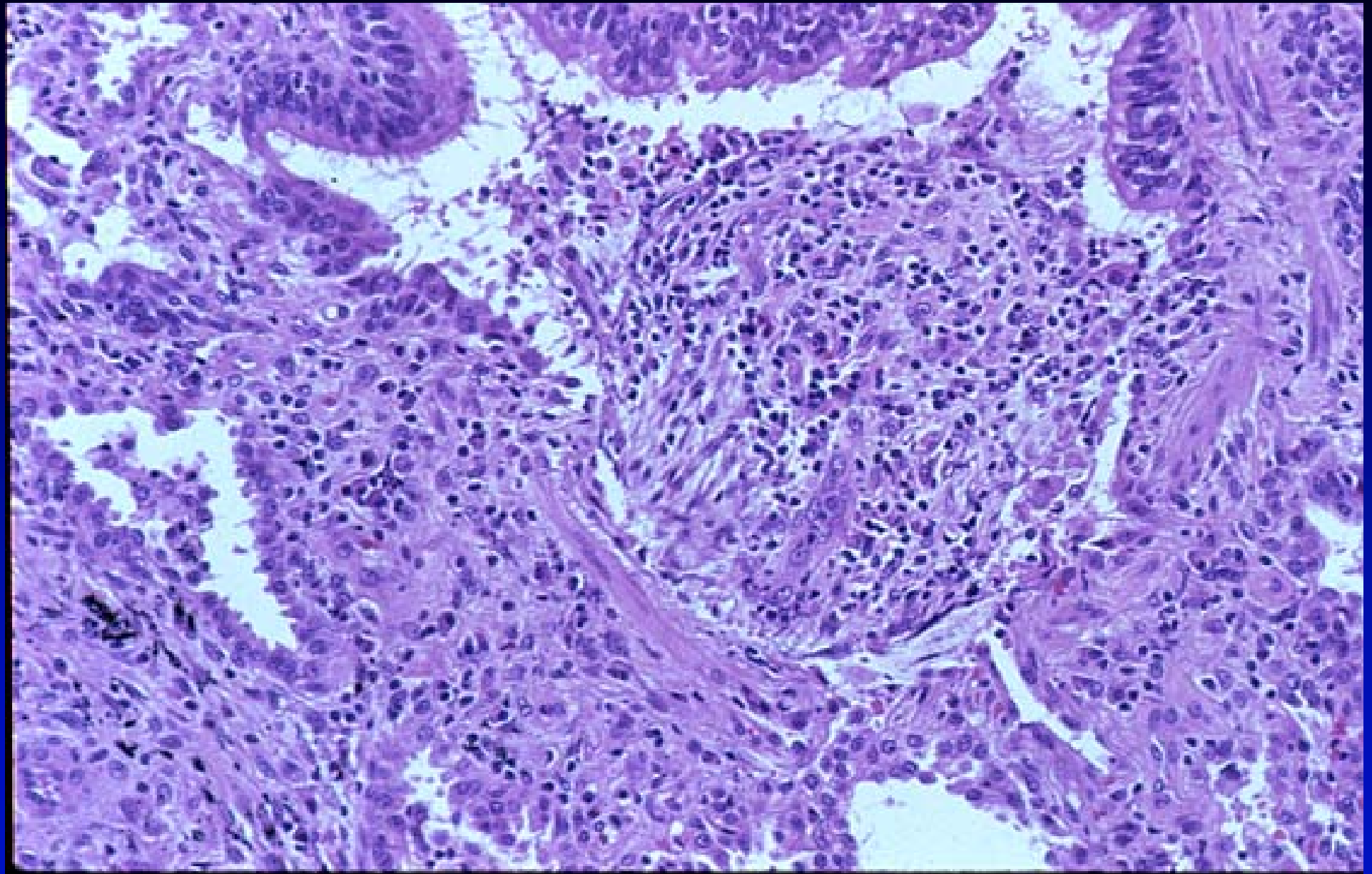
Synonymous with:

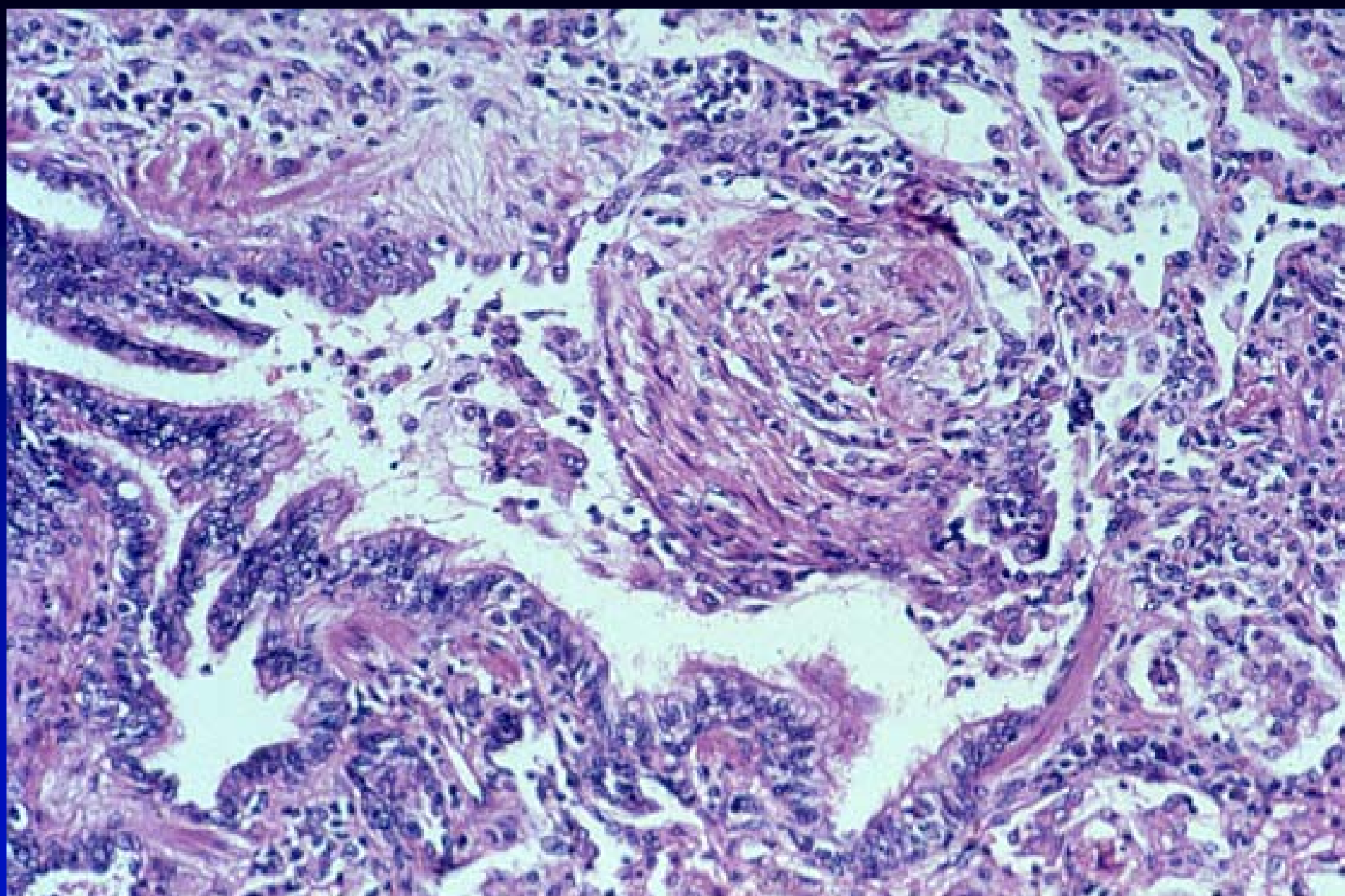
- **Bronchiolitis obliterans
organizing pneumonia (BOOP)**
- **Usually responds to steroids**

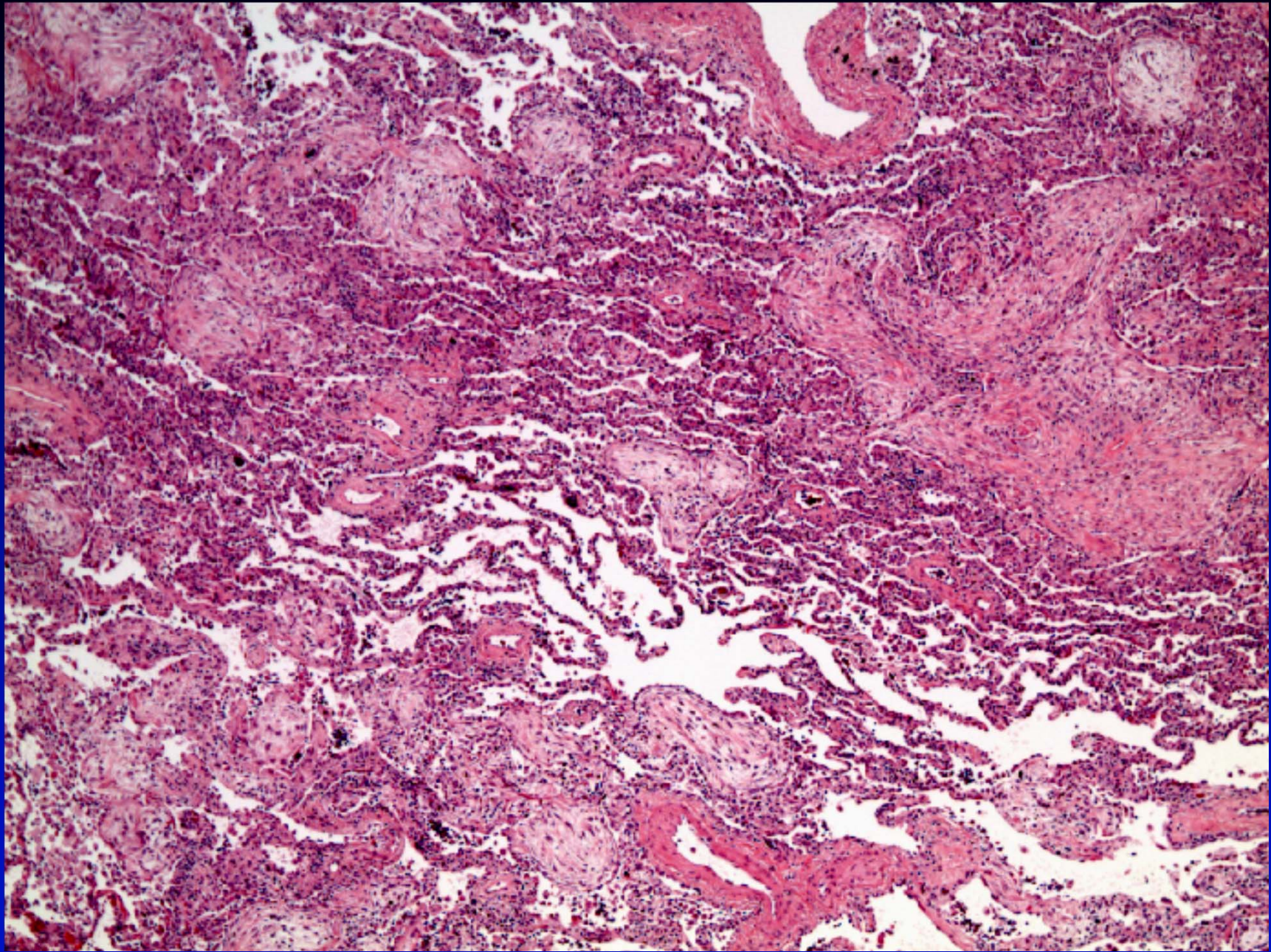


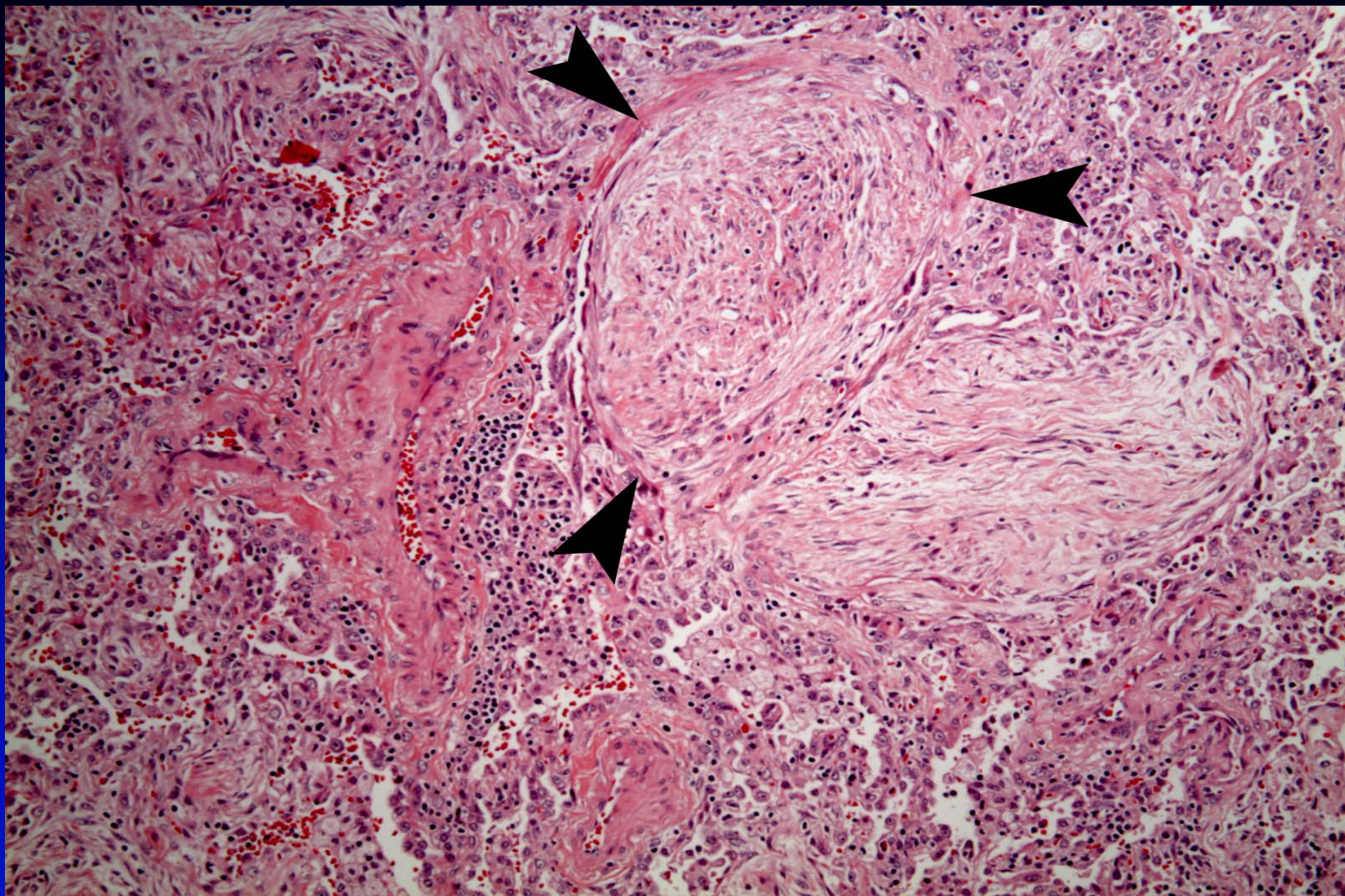










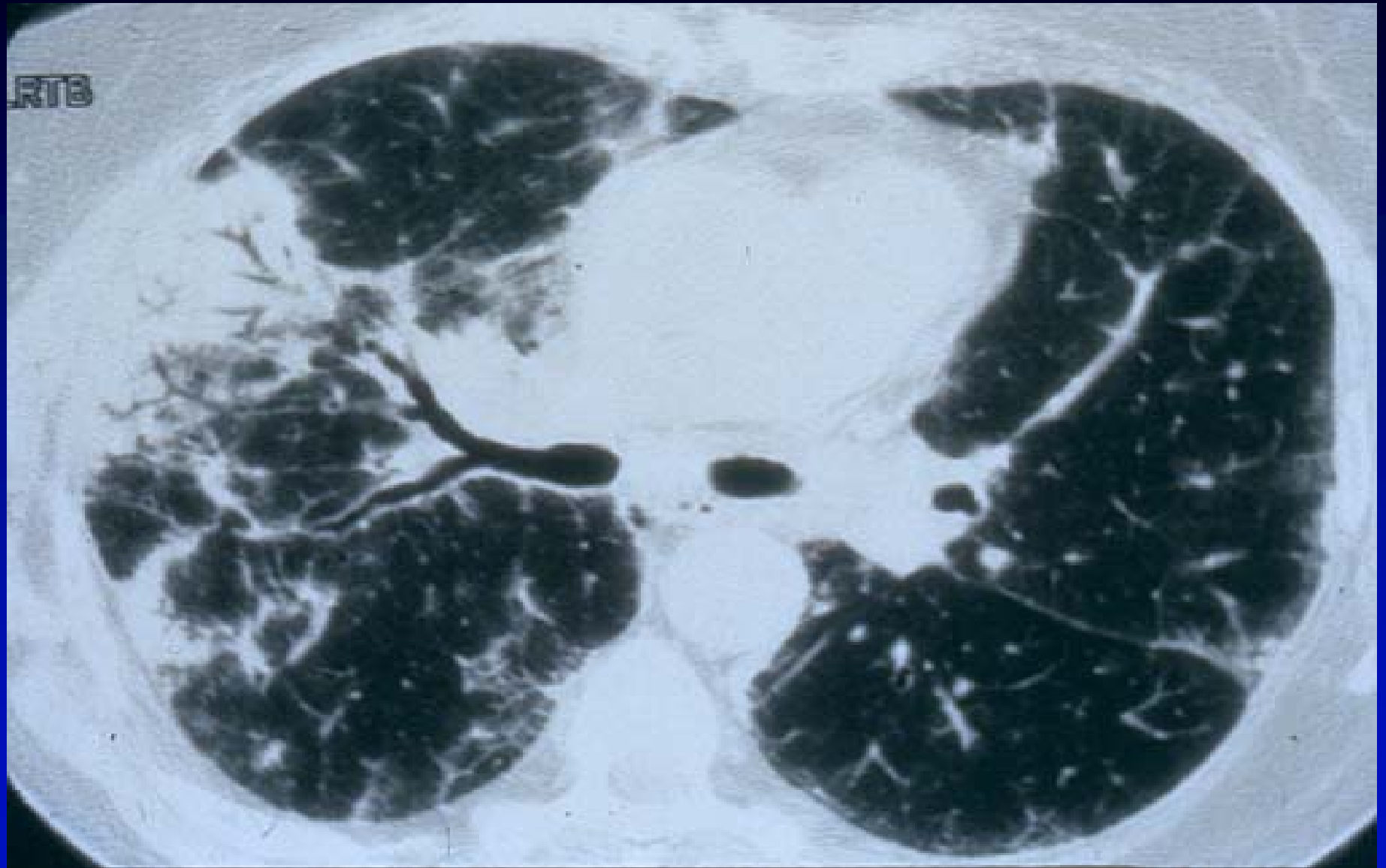


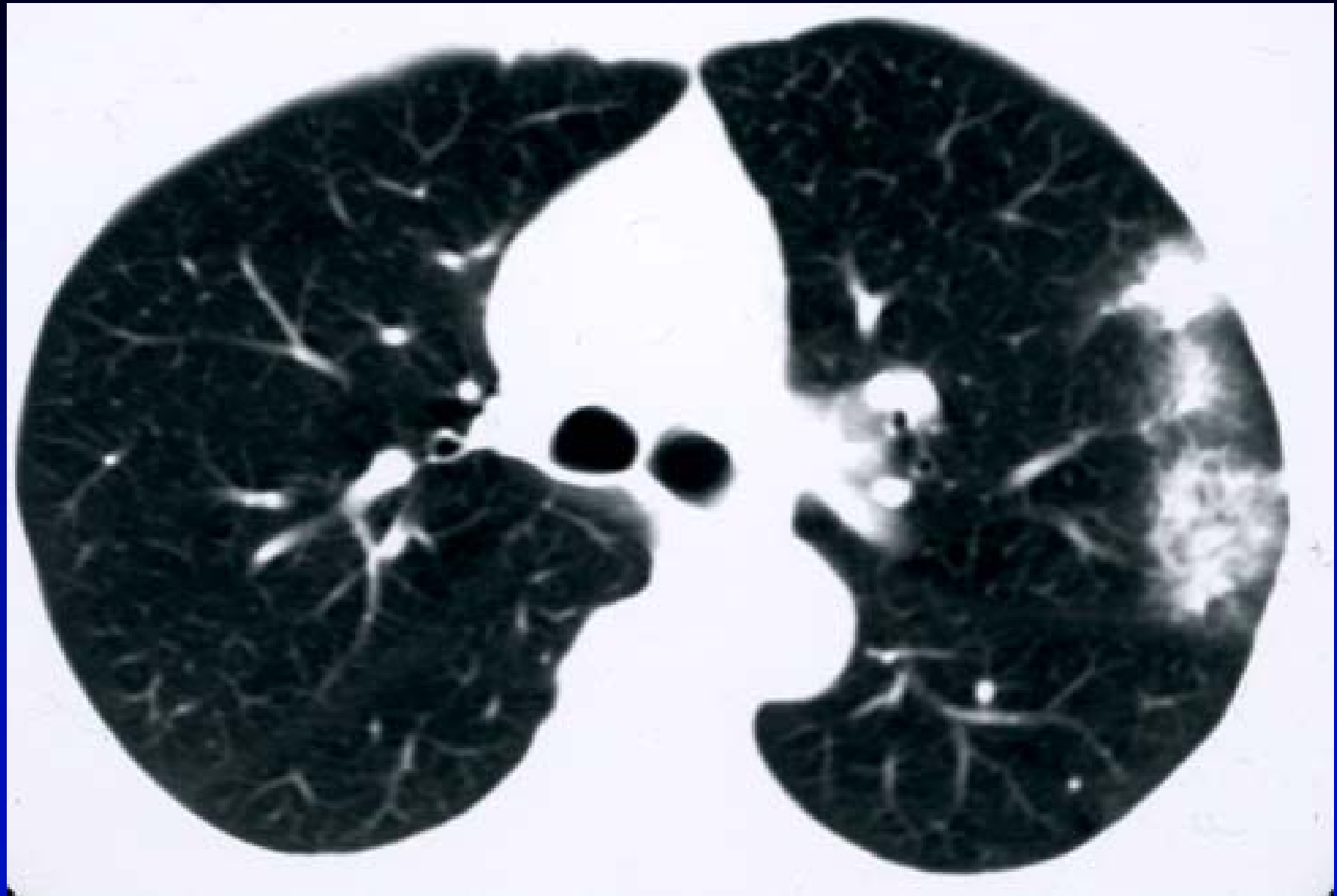
Cryptogenic Organizing Pneumonia

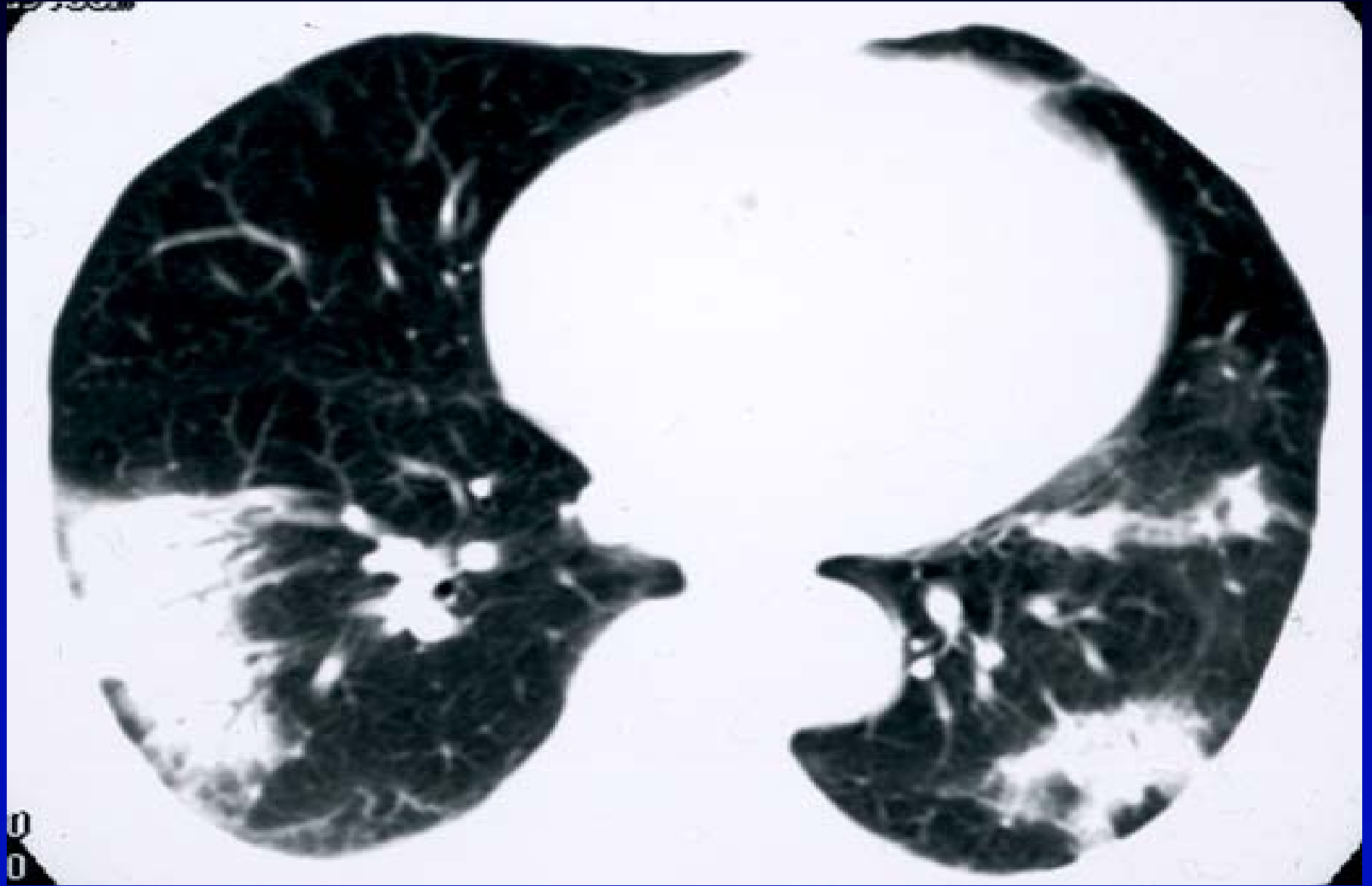
HRCT features of COP (43 pts)

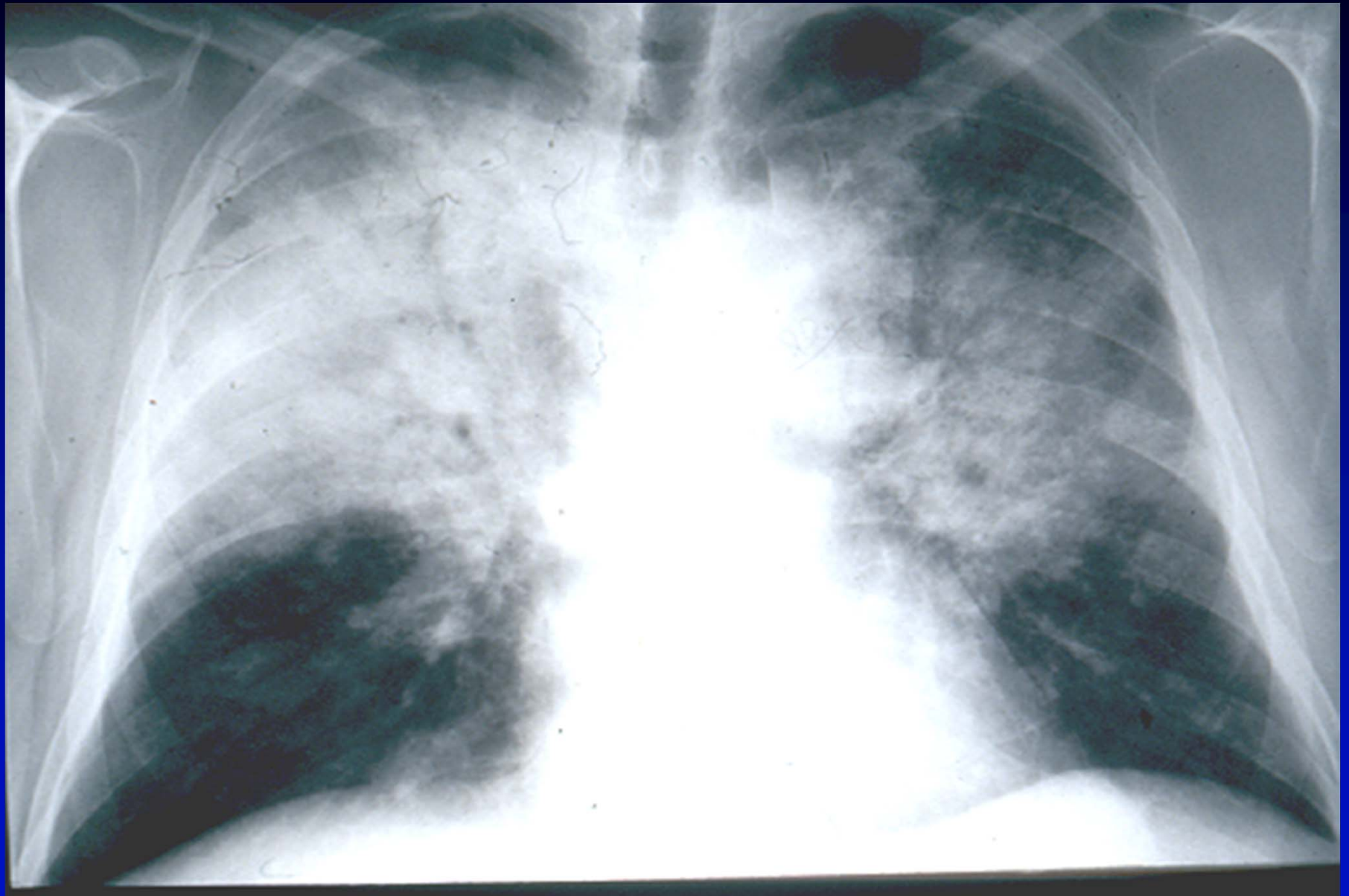
- Consolidation 34 (79%)
- Ground glass 26 (60%)
- Nodules 13 (30%)

Lee, *AJR* 1994;162:543



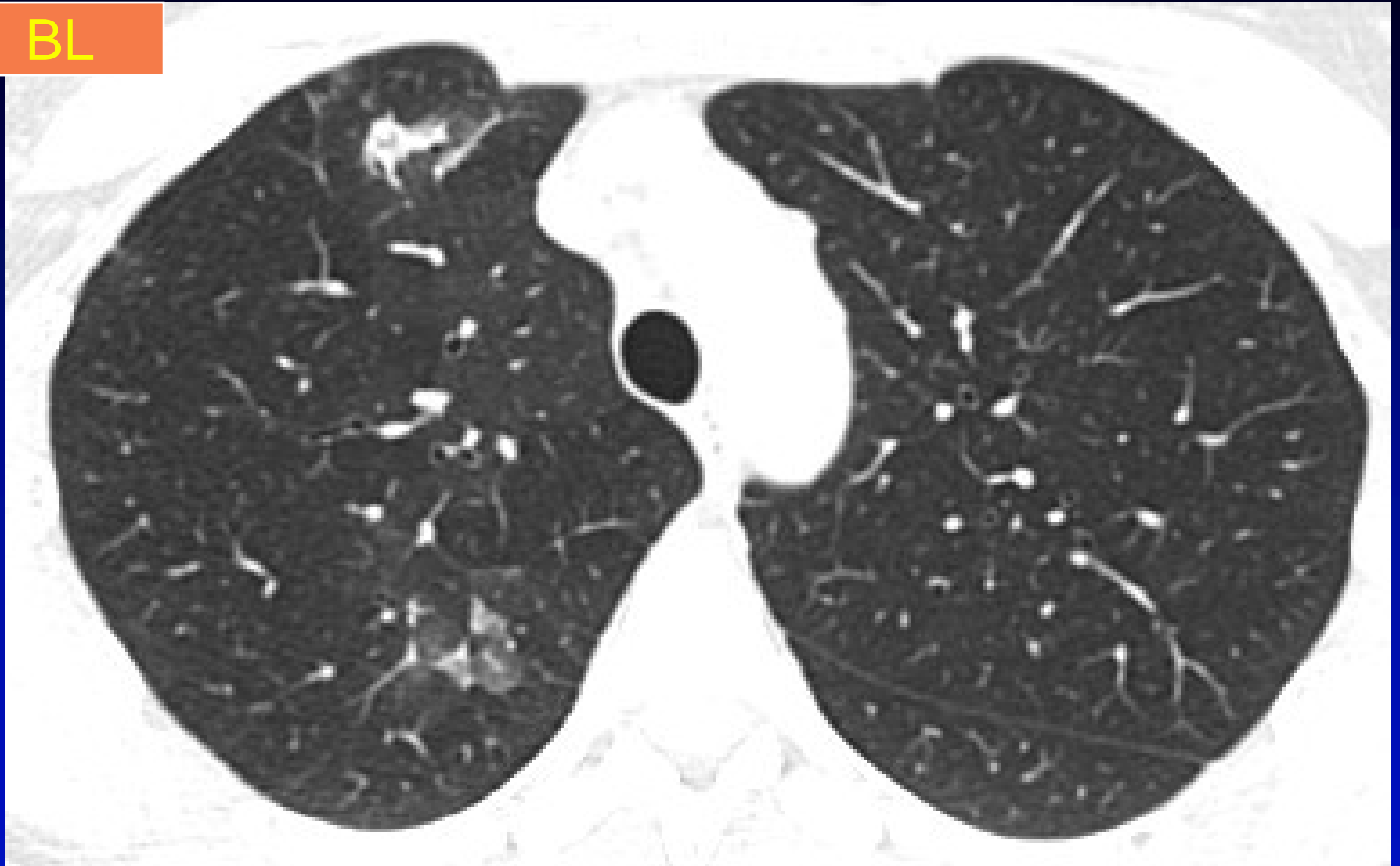


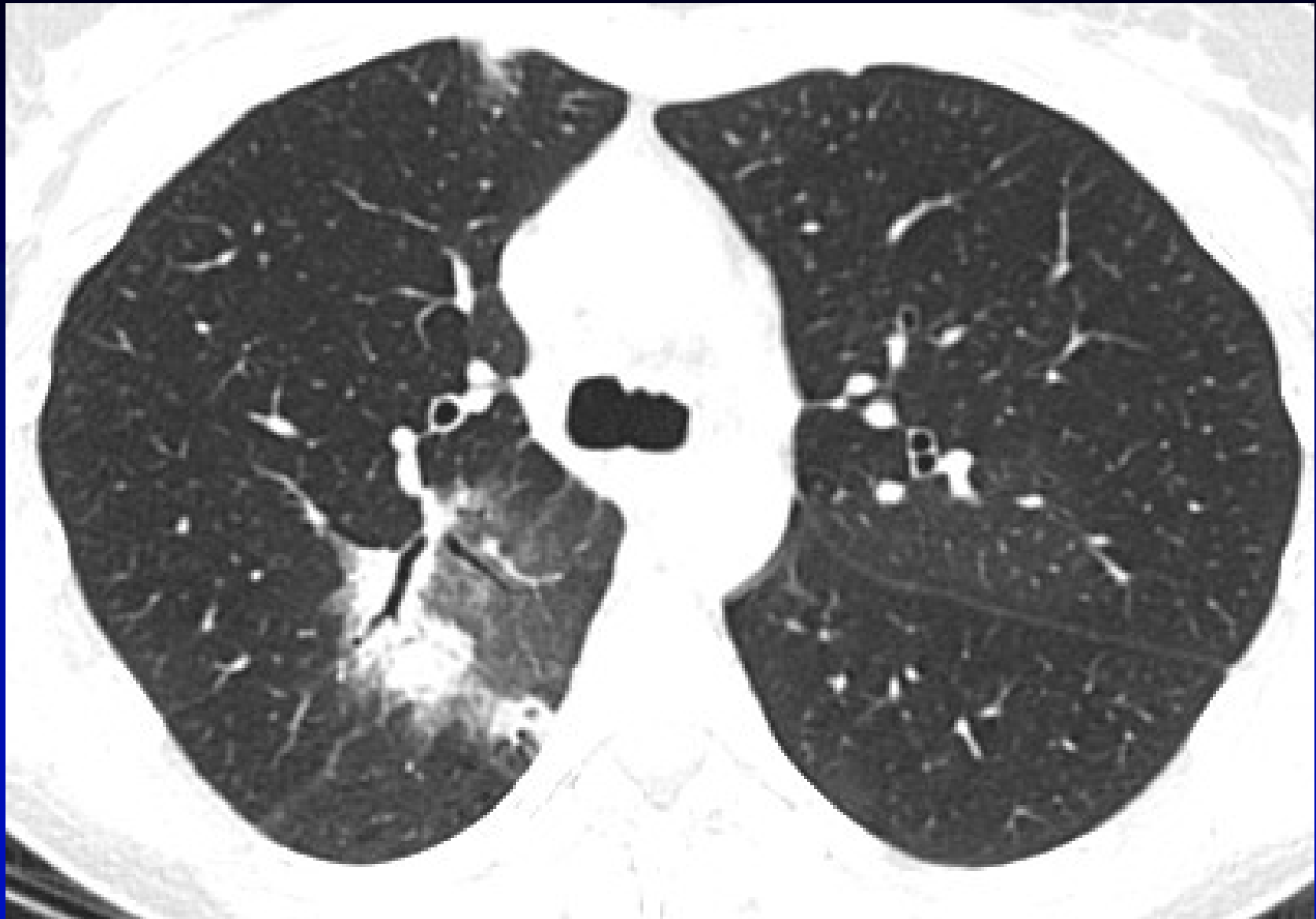


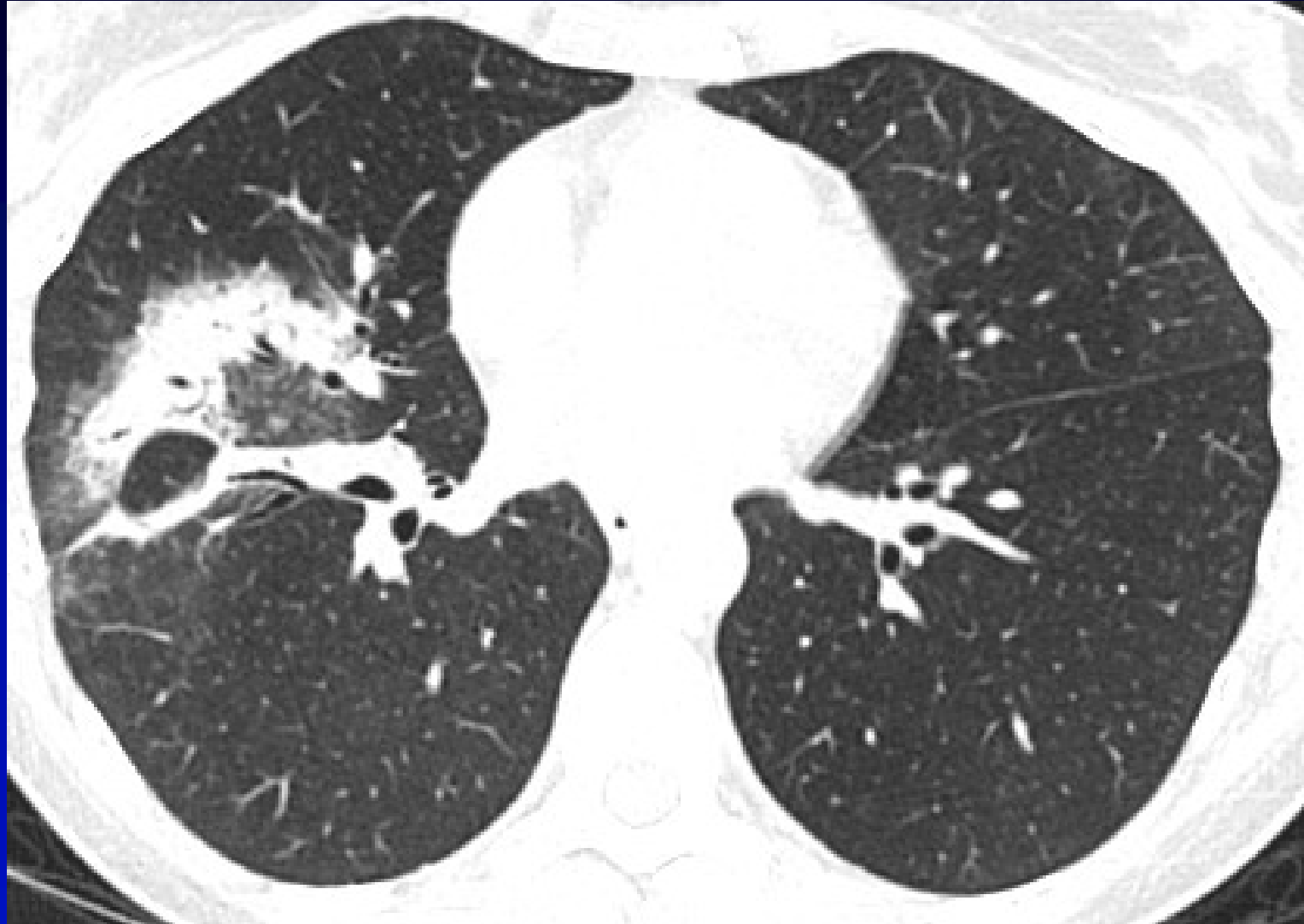




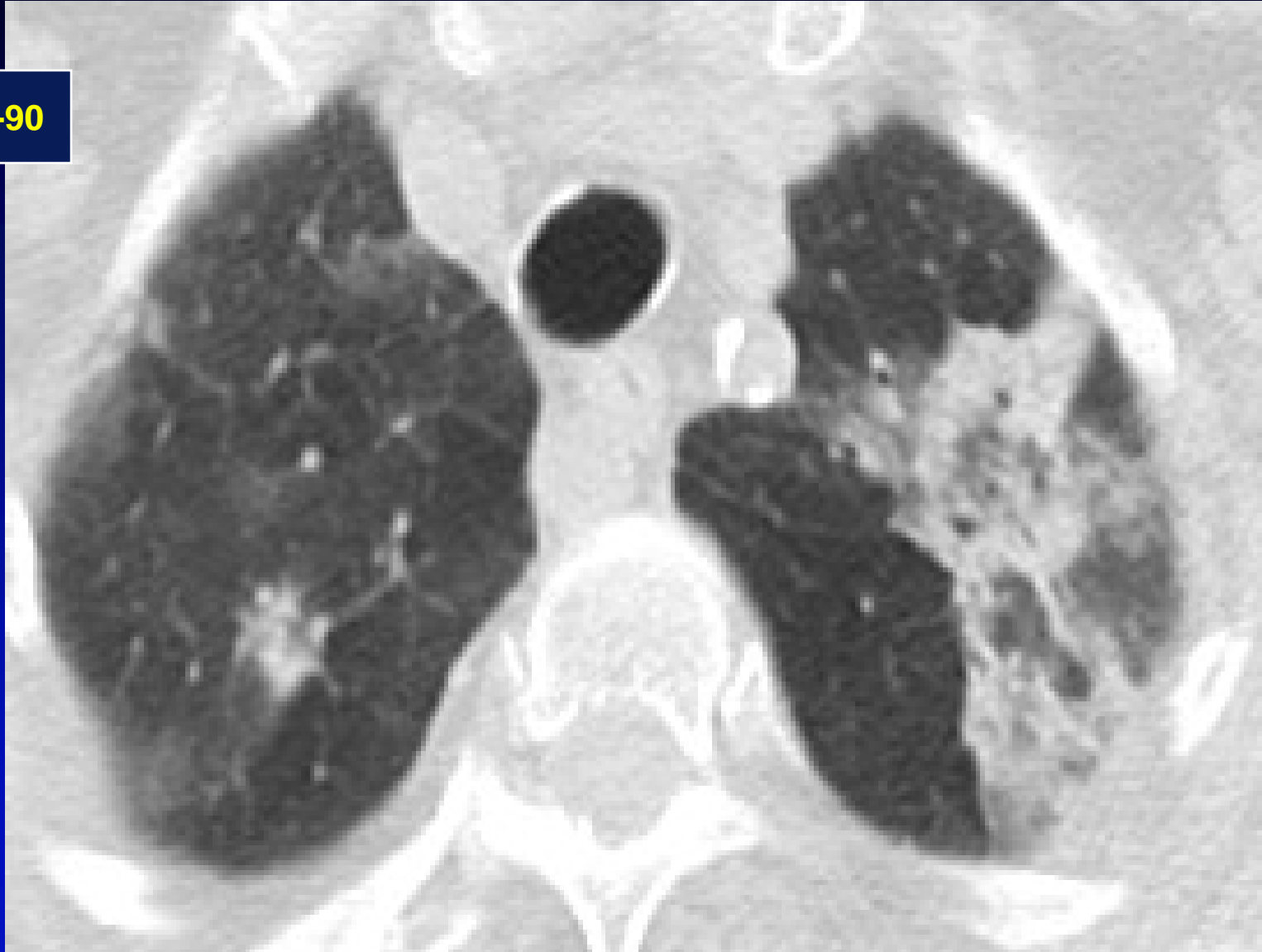
BL







KK, #222-94-90





[4]

JP



Treatment of BOOP/COP

Corticosteroids Rx of choice:

- **Responses often dramatic**
- **Complete recovery in 65%**
- **May relapse after cessation**

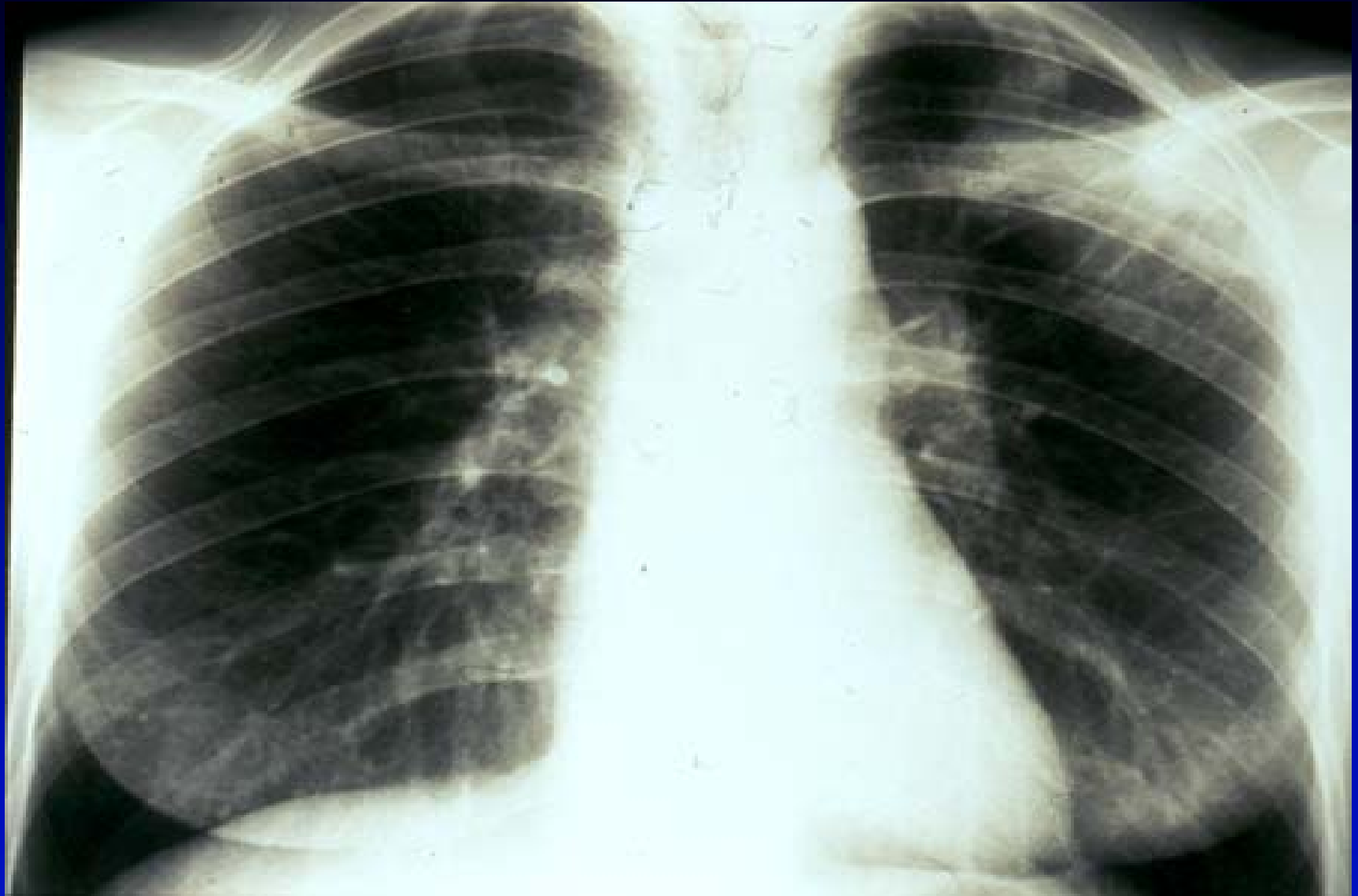
Chronic Eosinophilic Pneumonia

Chronic Eosinophilic Pneumonia

- Cough, dyspnea, wheezing
- Blood eosinophilia (> 80%)
- Migratory alveolar infiltrates
- Evolves over weeks to months

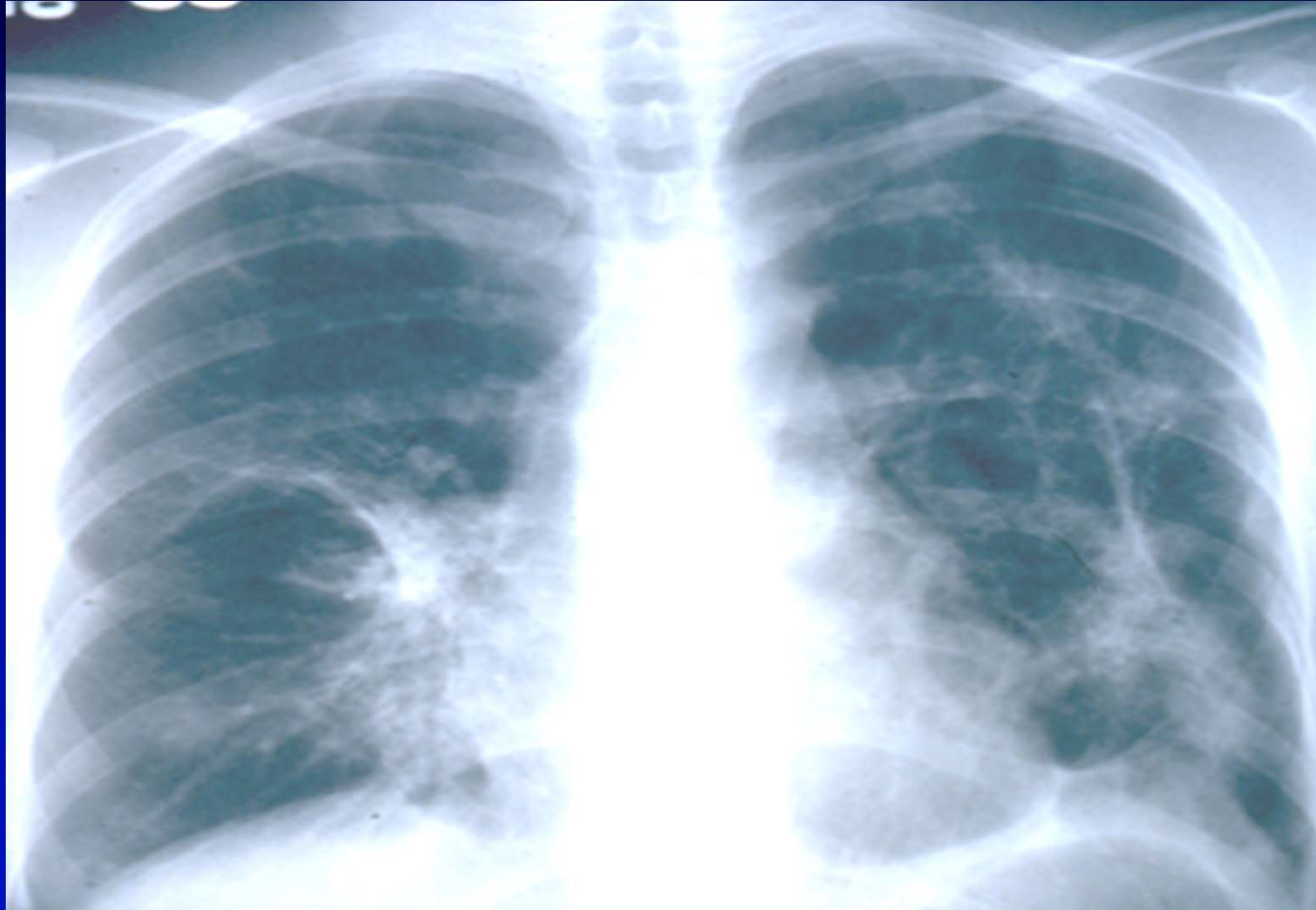
CEP: Radiographic features

- Dense alveolar infiltrates
- Predilection upper lobes, subpleural
- Central sparing (photographic negative pulmonary edema)
- Marked clearing with steroid Rx



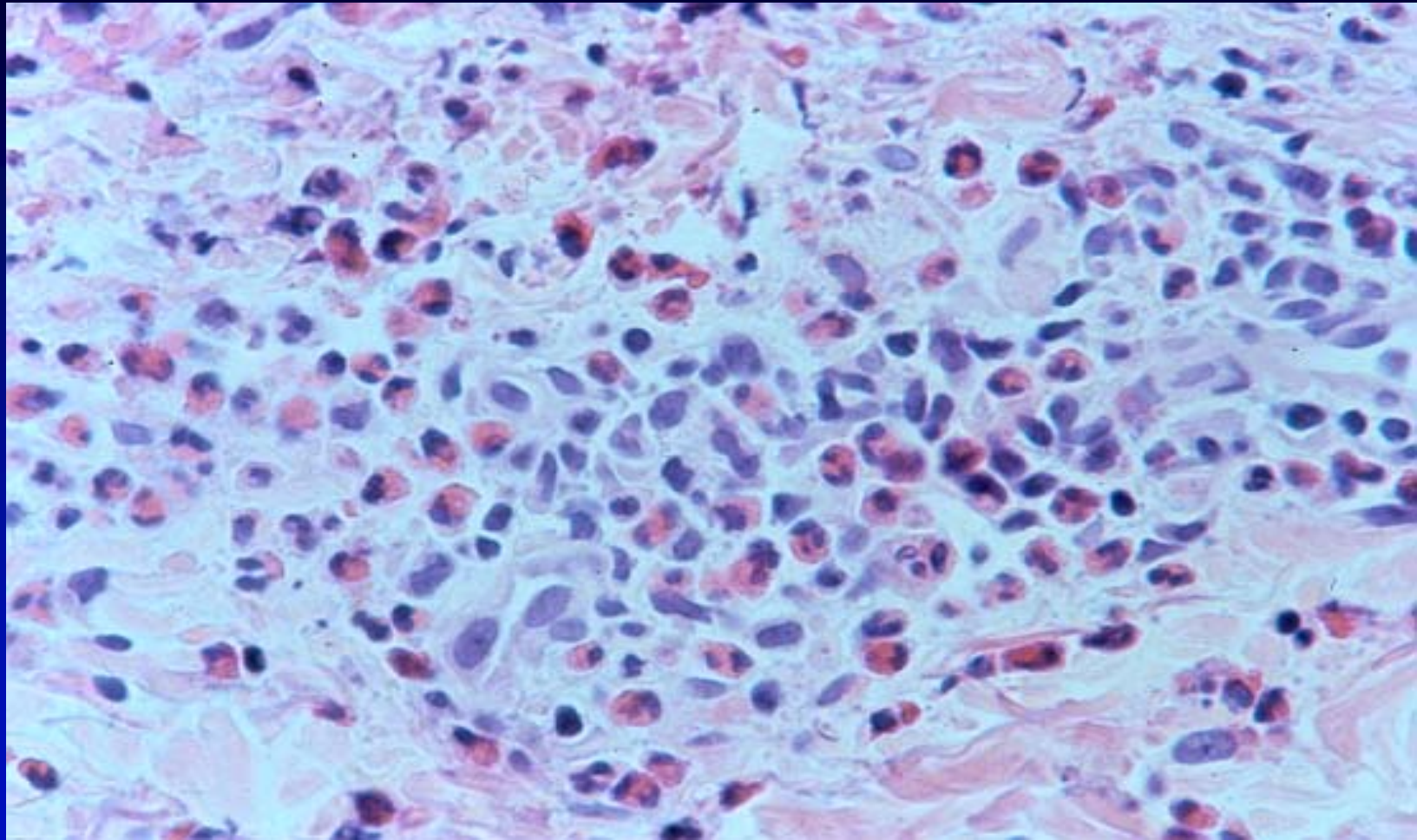


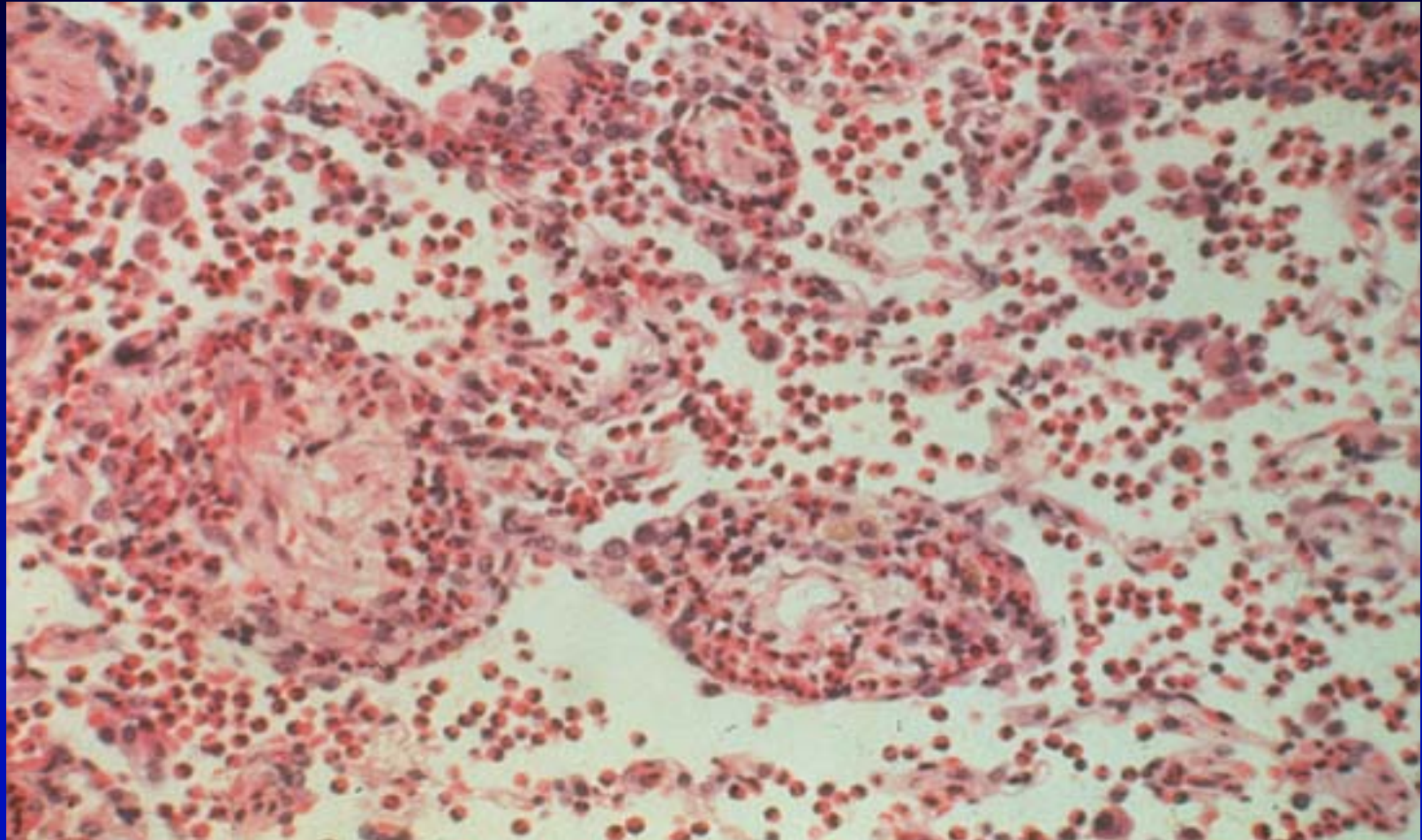
CEP (5 days after steroid Rx)

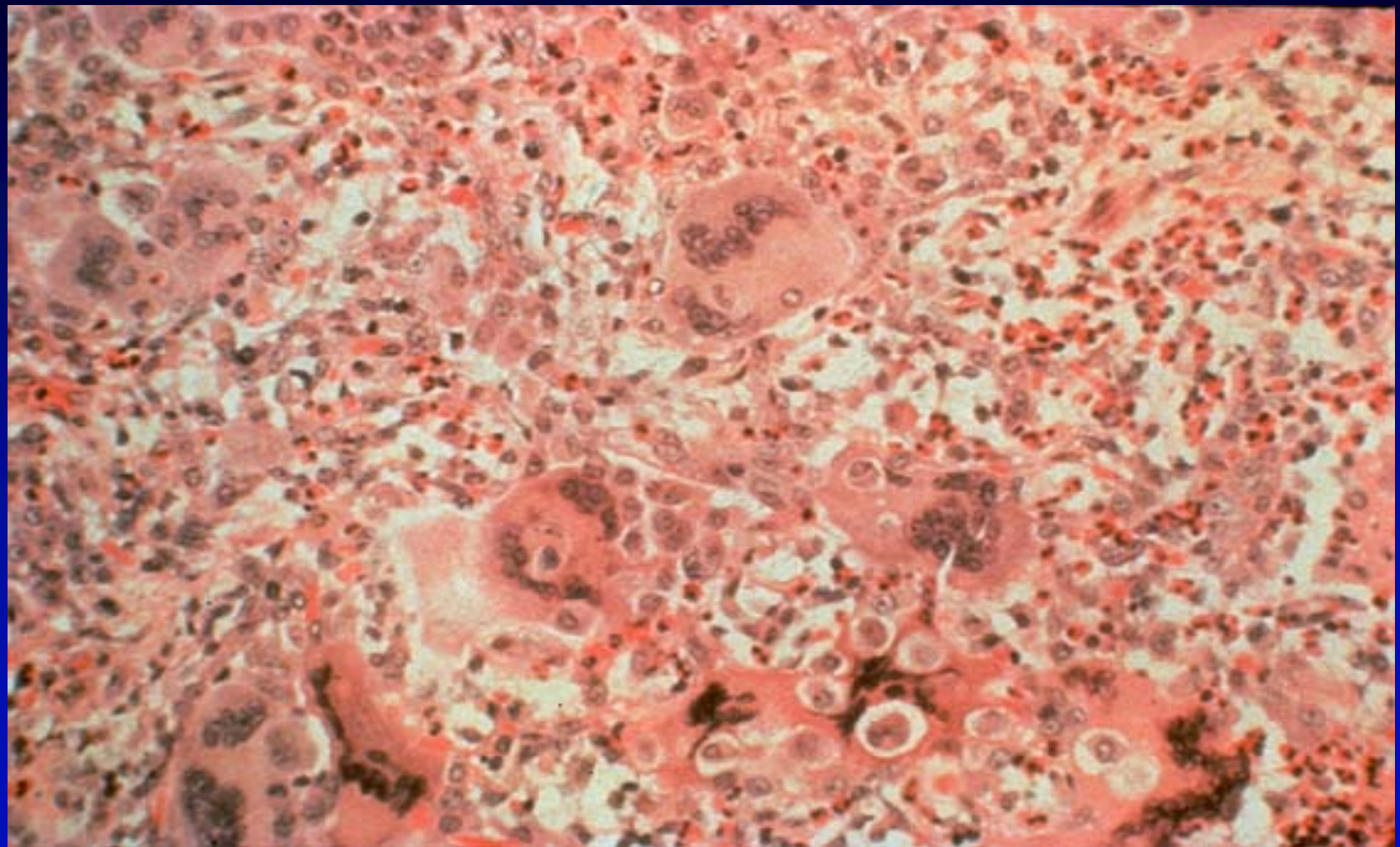


CEP (2 weeks post steroid Rx)







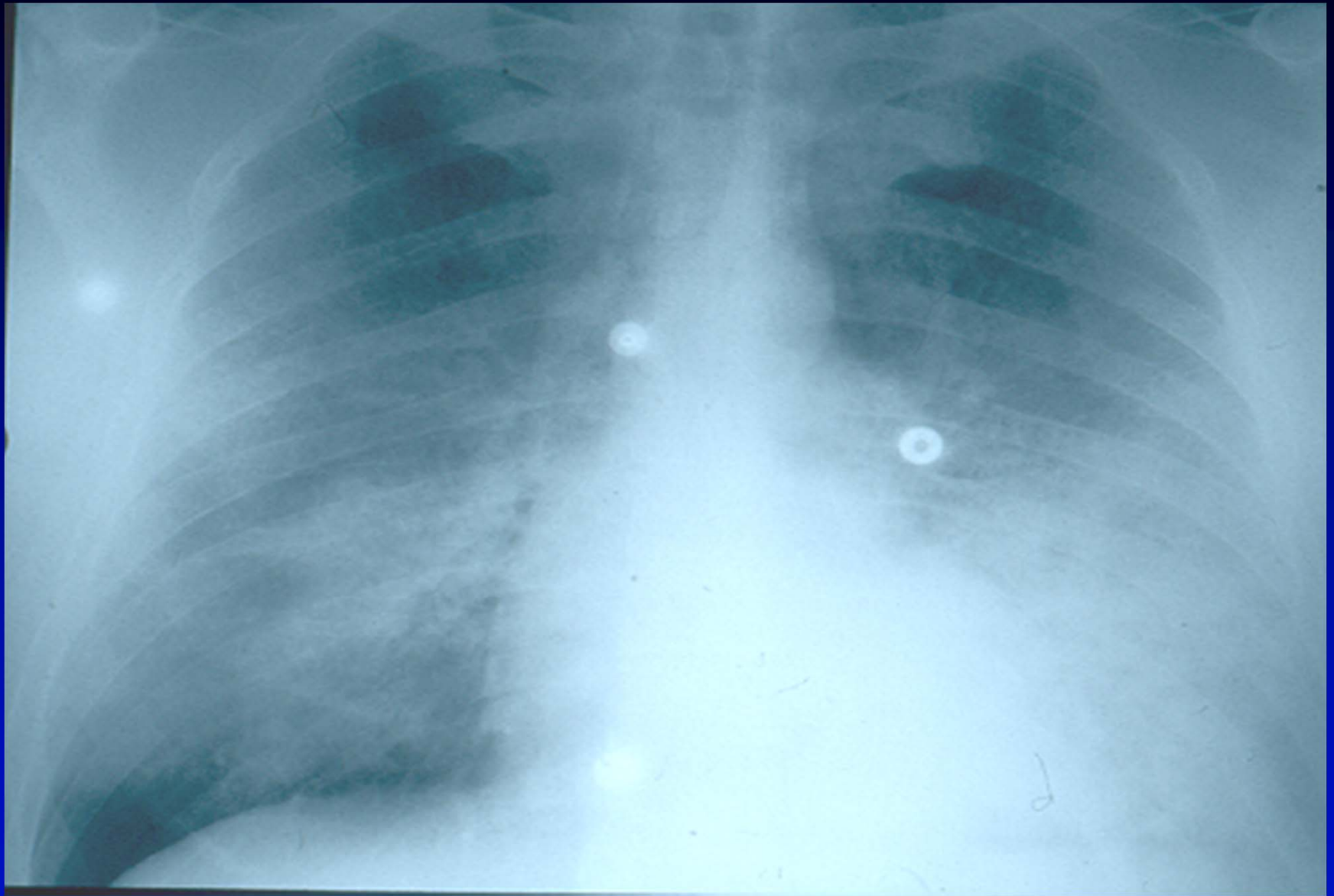


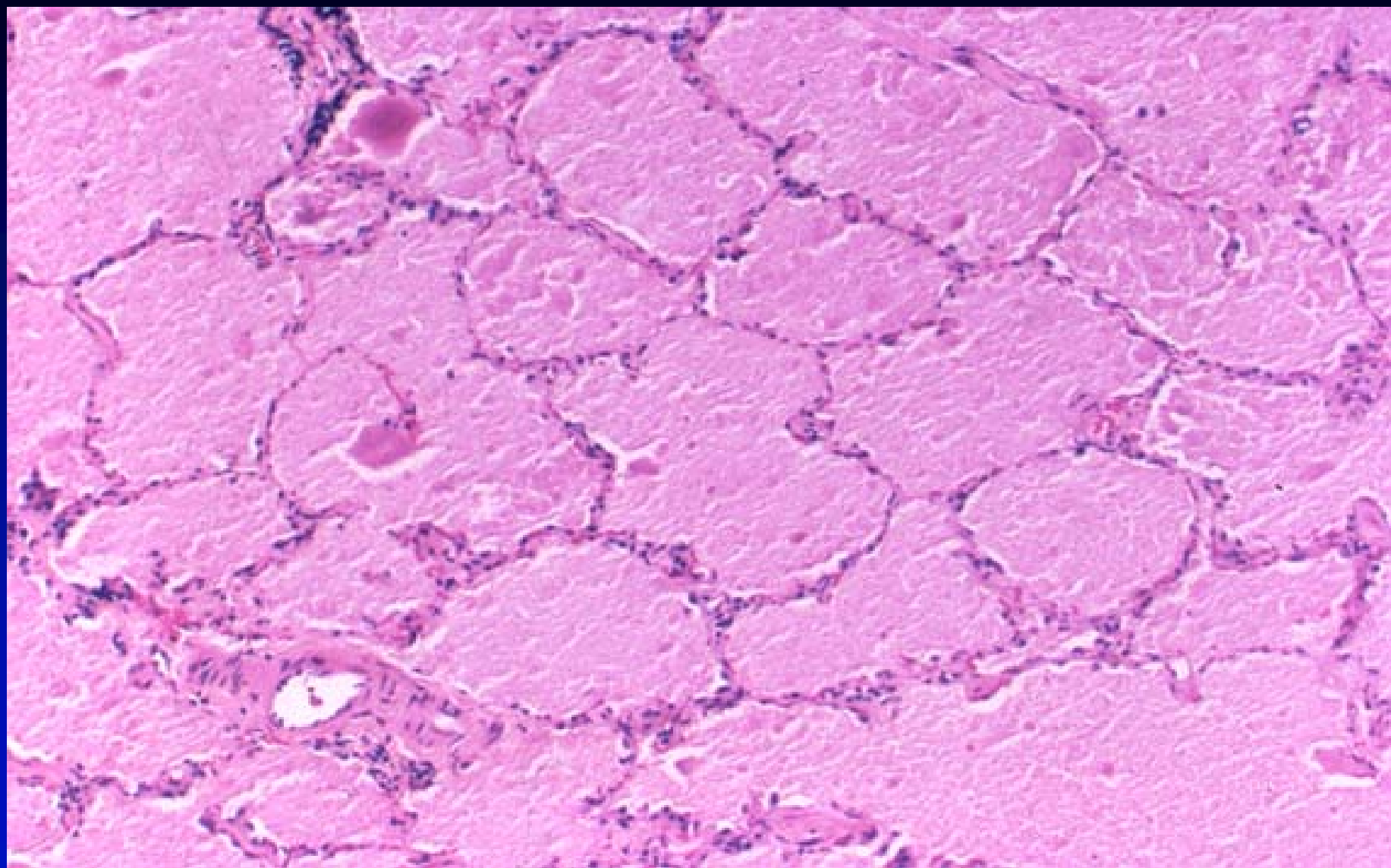
Chronic Eosinophilic Pneumonia

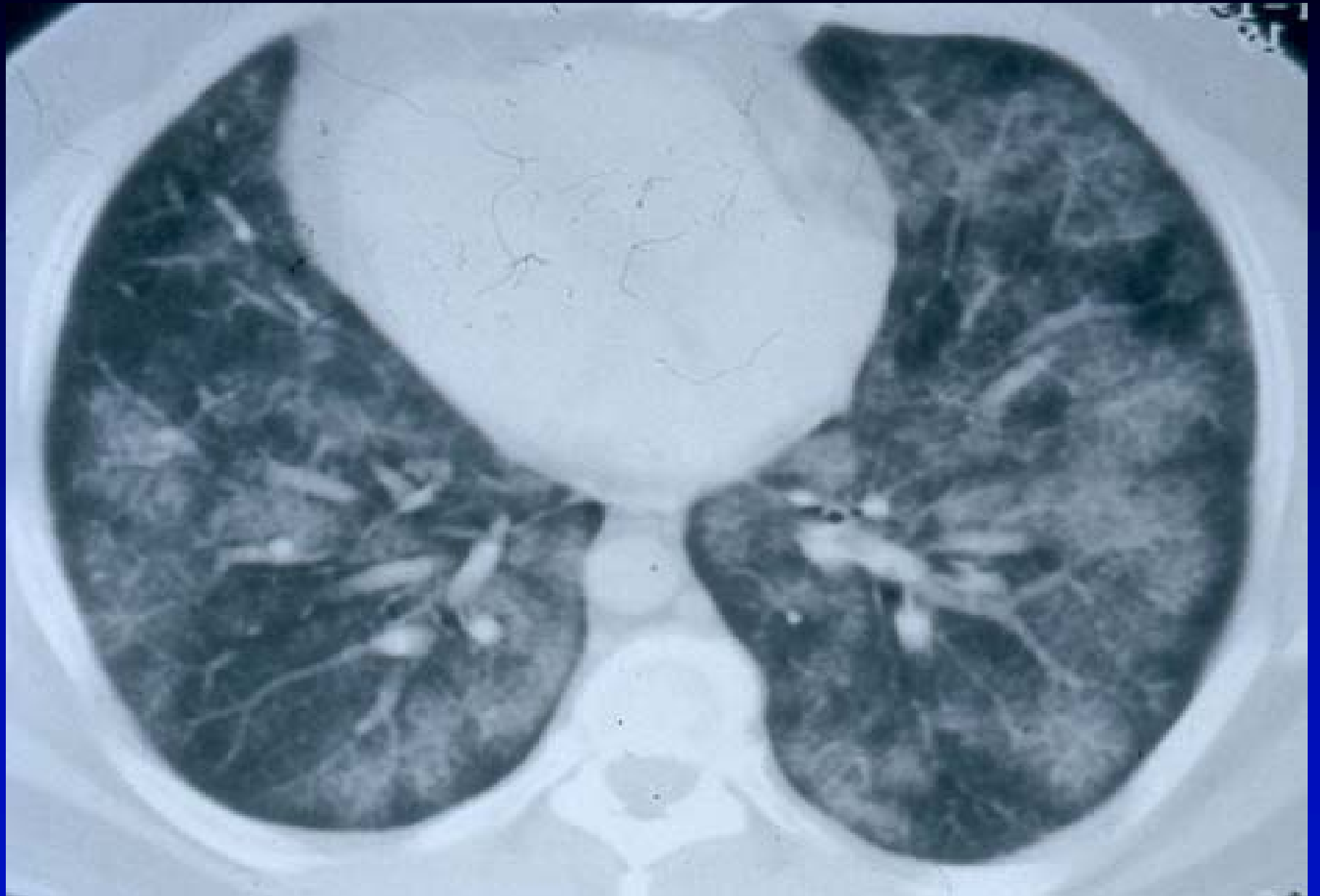
- **Response to steroids dramatic**
- **Partial clearing CXR < 48 hrs**
- **Rapid clearing supports diagnosis**

Chronic Eosinophilic Pneumonia

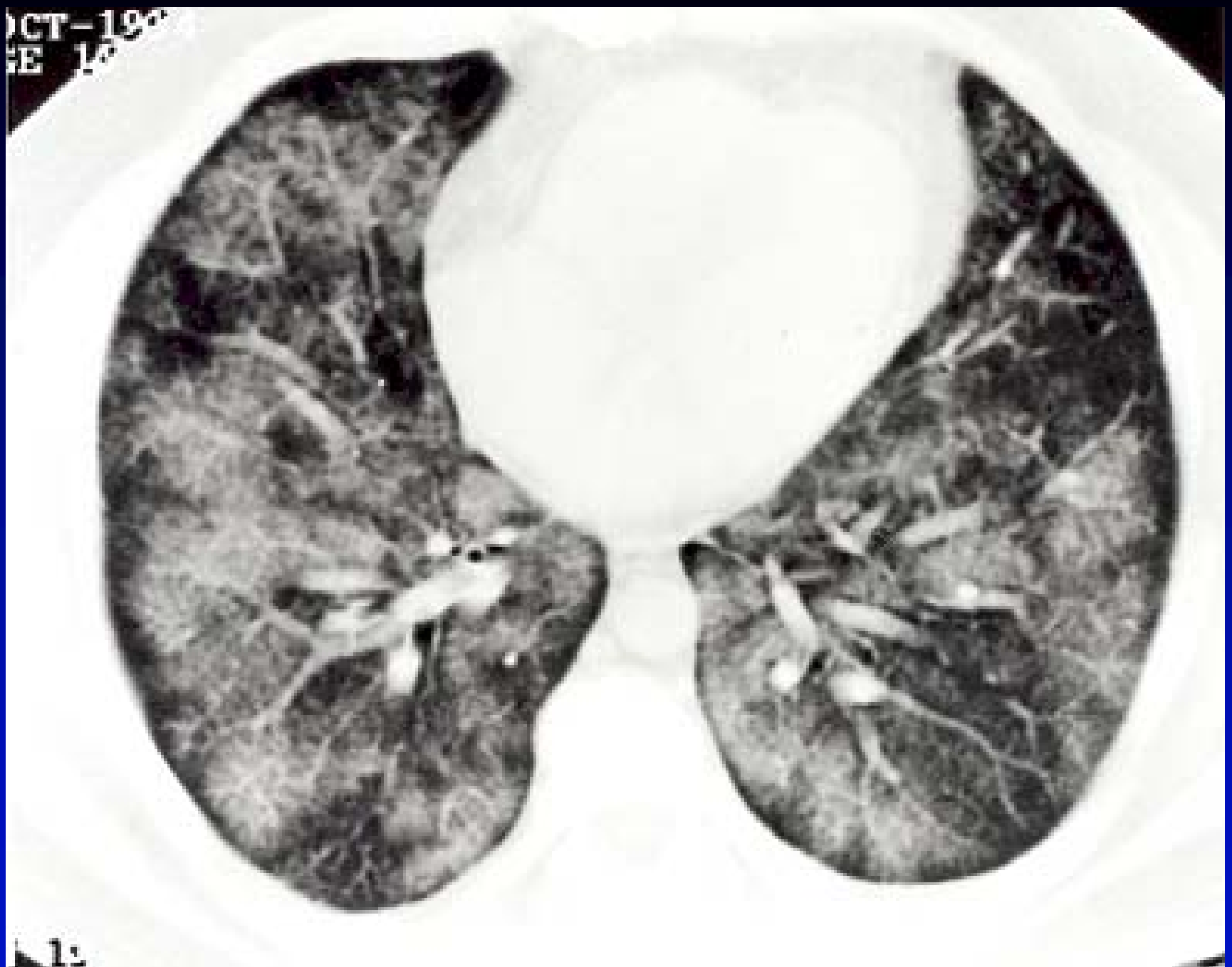
-
- **Open biopsy not required to Dx**
 - **Diagnosis by BAL (eosinophils),
dramatic response to steroid Rx**
-



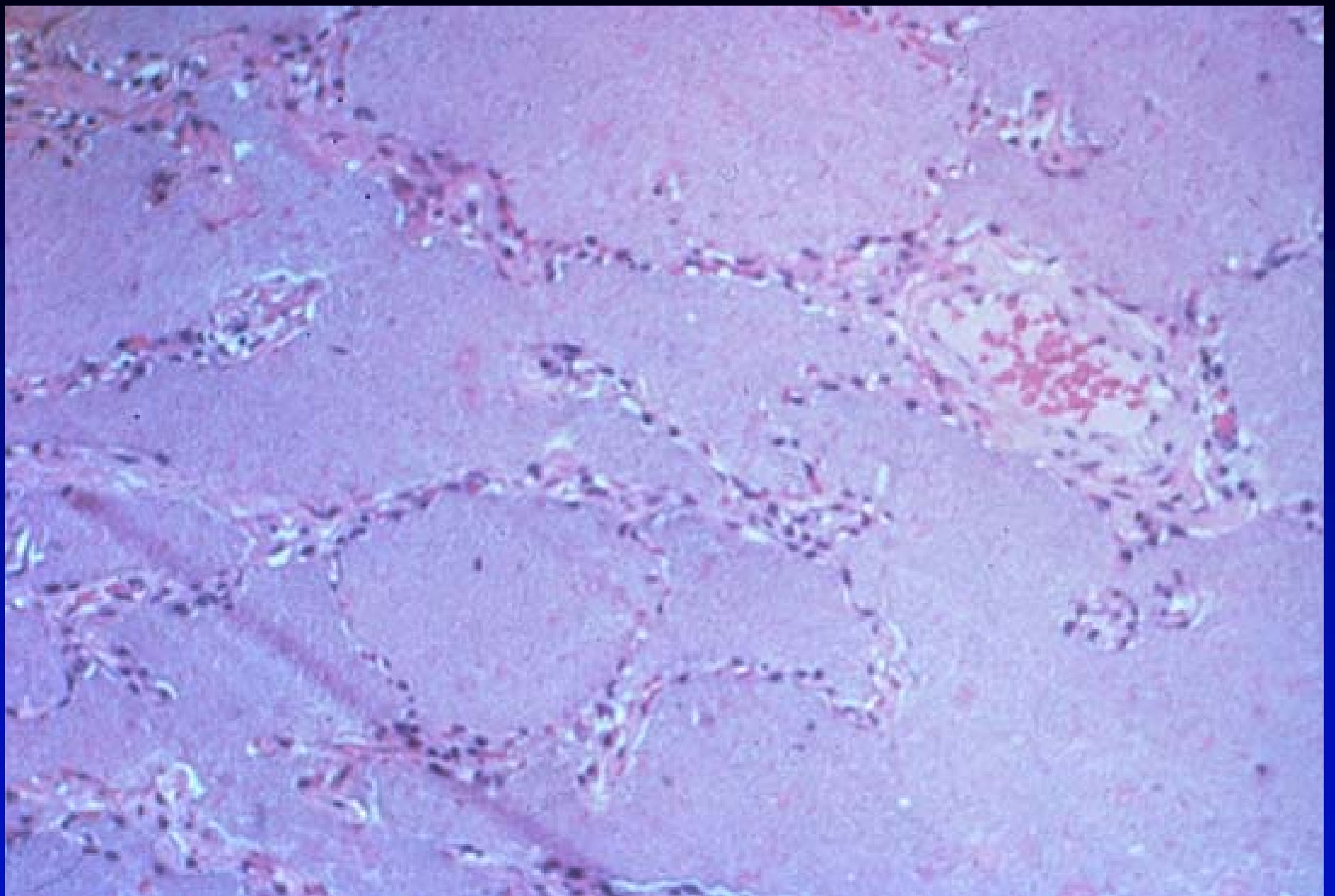




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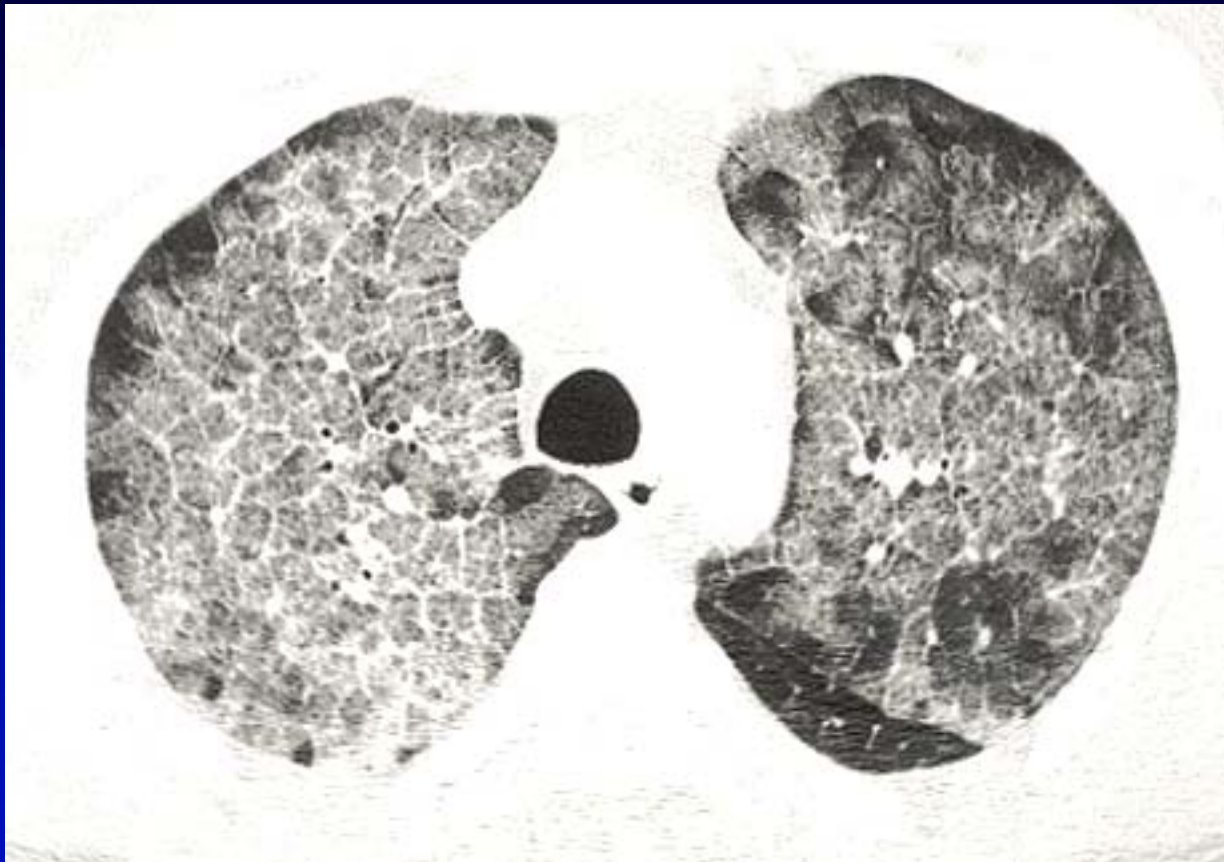


Pulmonary alveolar proteinosis

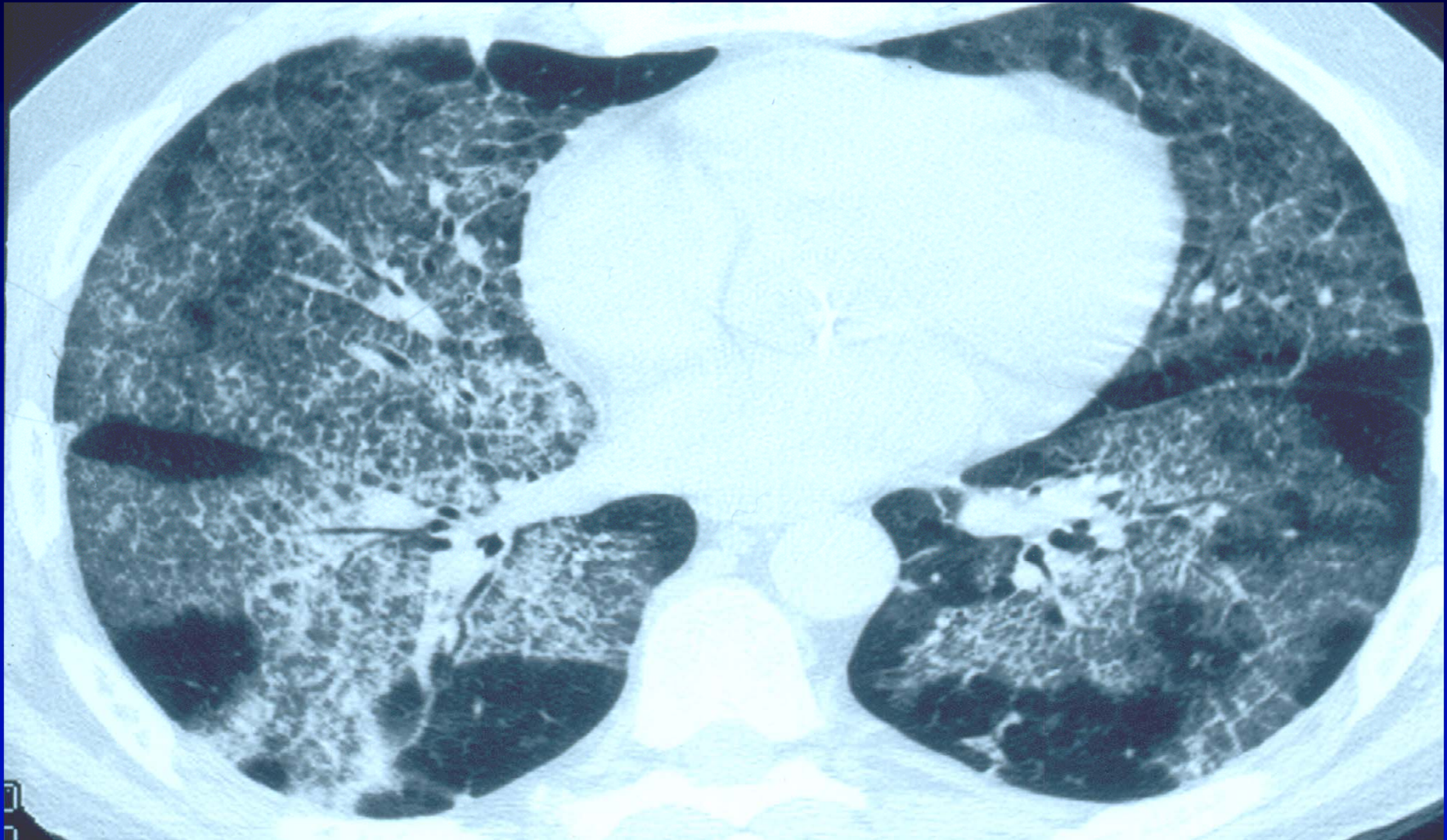
HRCT features of PAP:

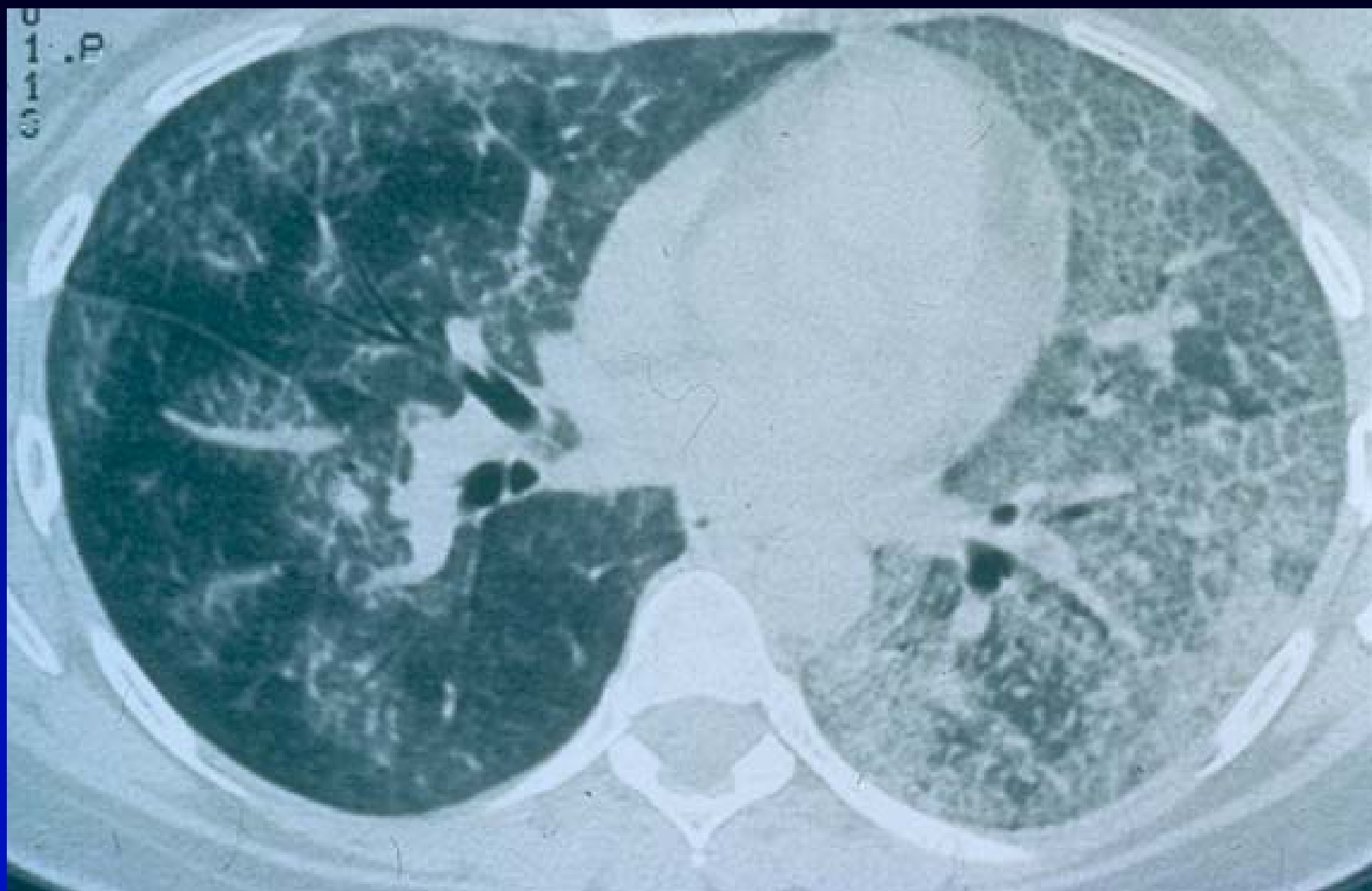
- Dense ground glass opacities
- “Crazy-paving”
- Preserved lung architecture
- Lack of honeycombing

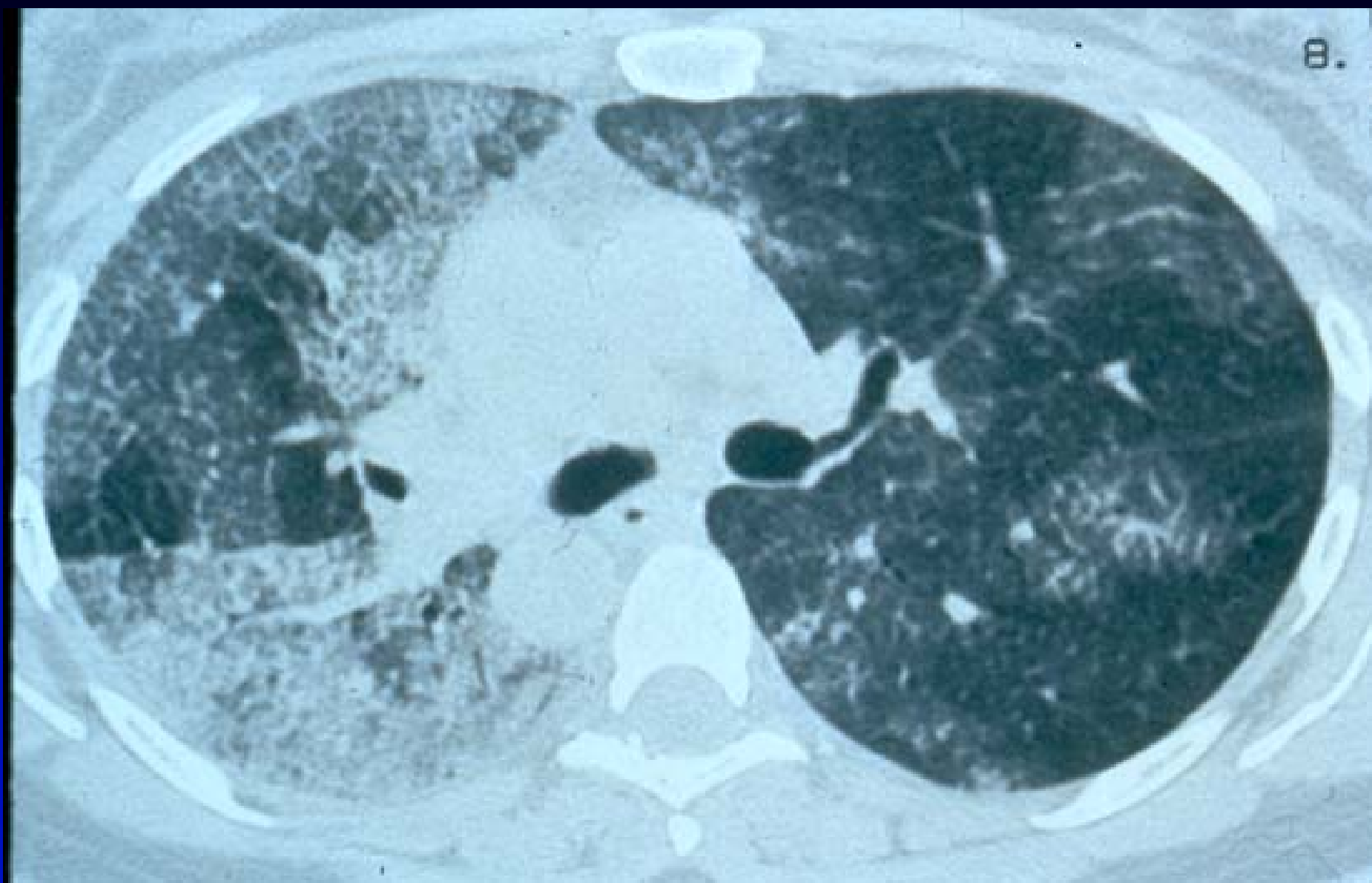
Pulmonary alveolar proteinosis (PAP)

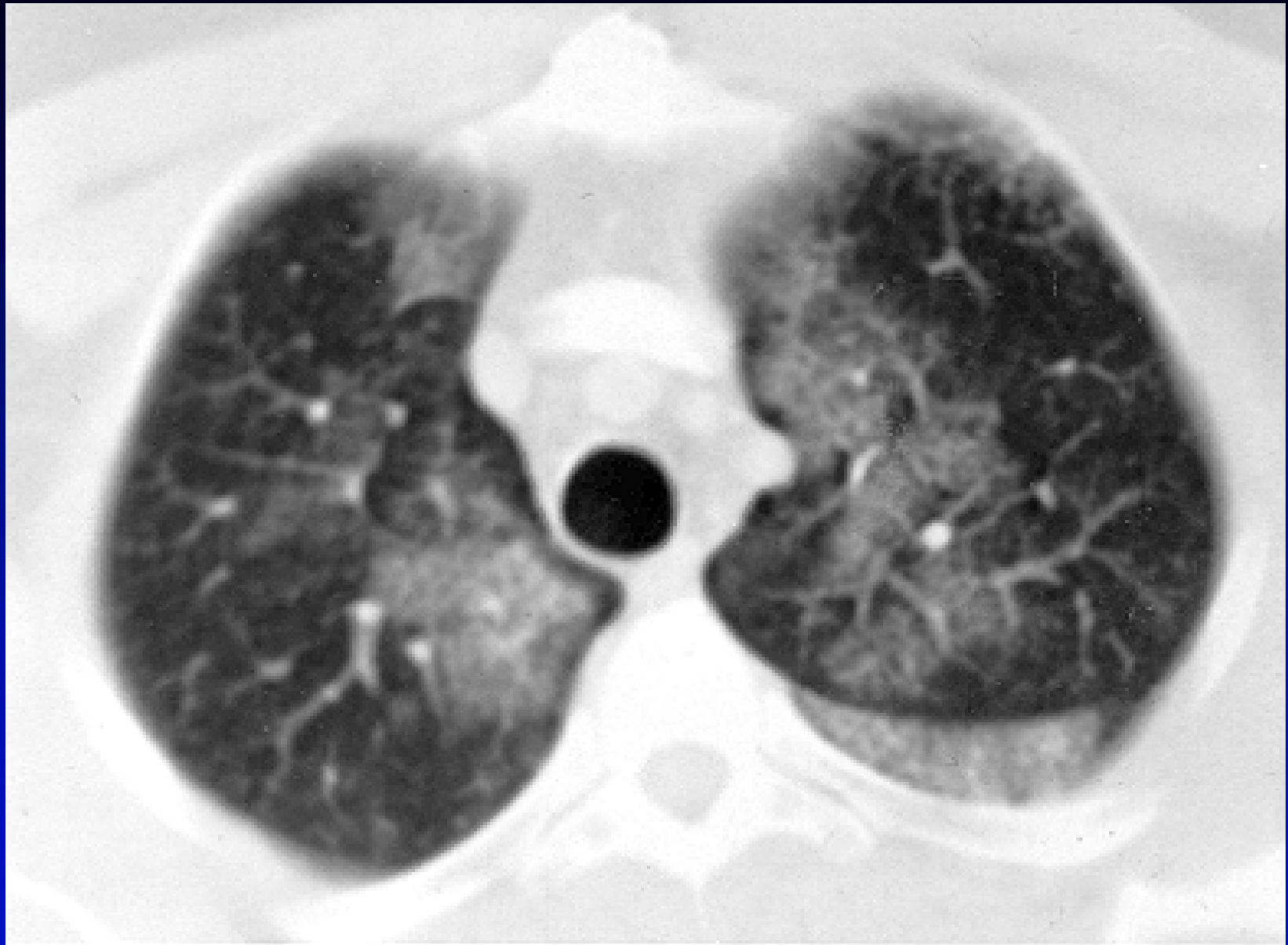


“Crazy-paving” (PAP)









Pulmonary alveolar proteinosis

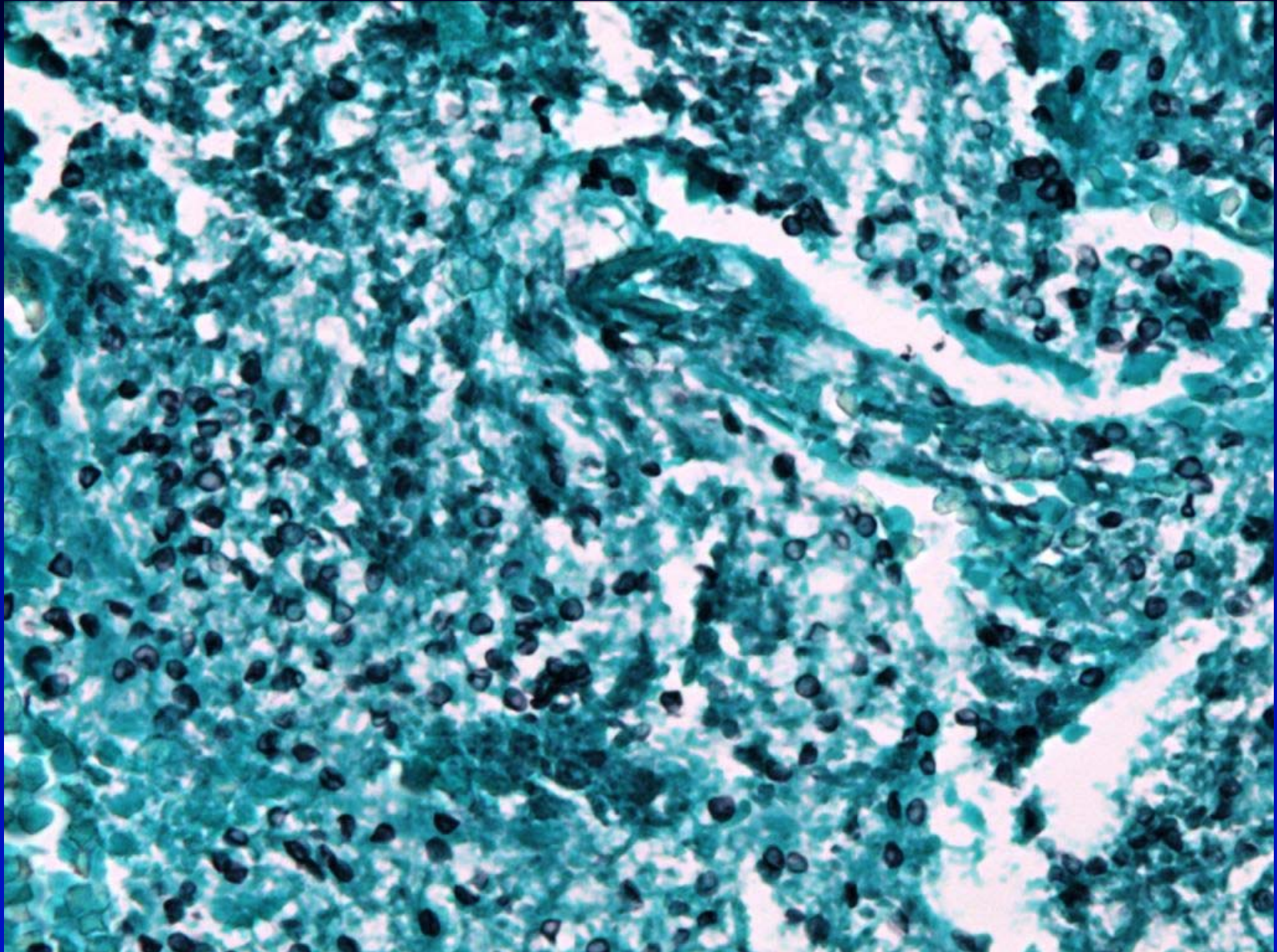
- Whole lung lavage Rx of choice
- Recurrences in 30%+

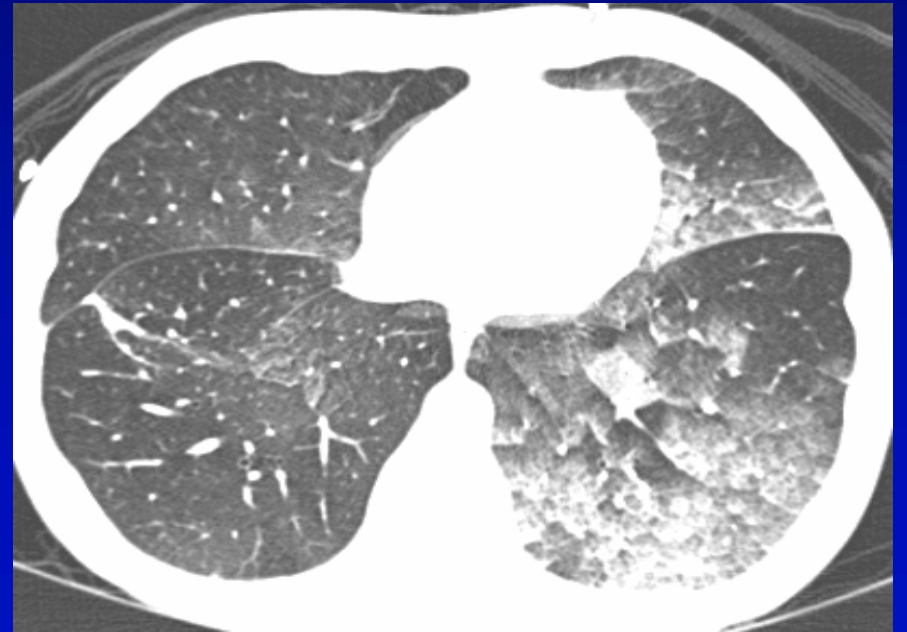
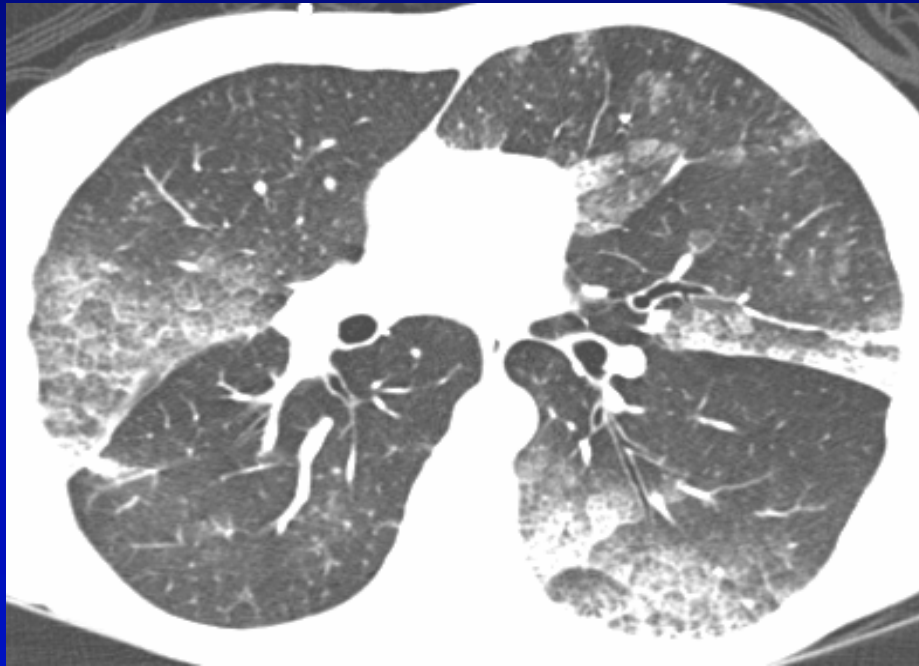
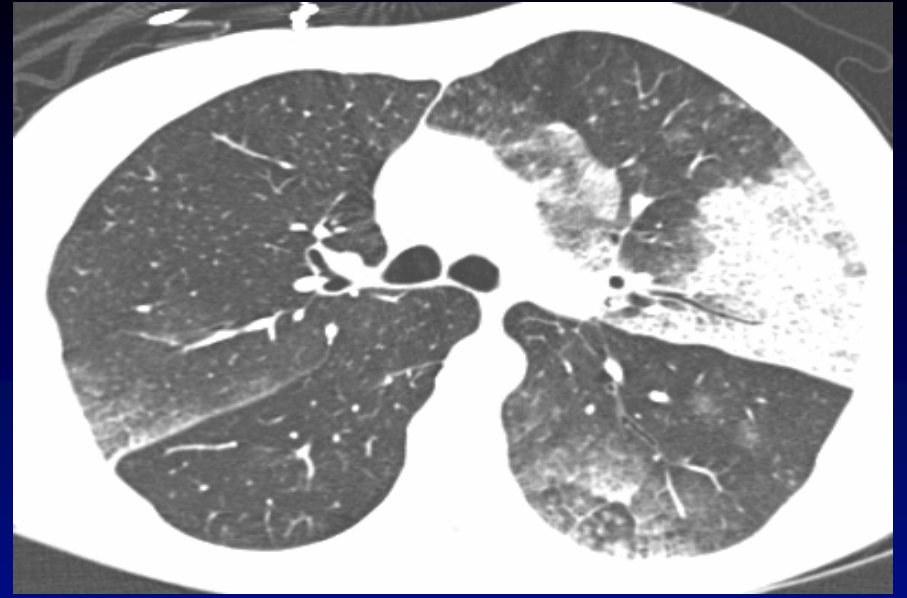
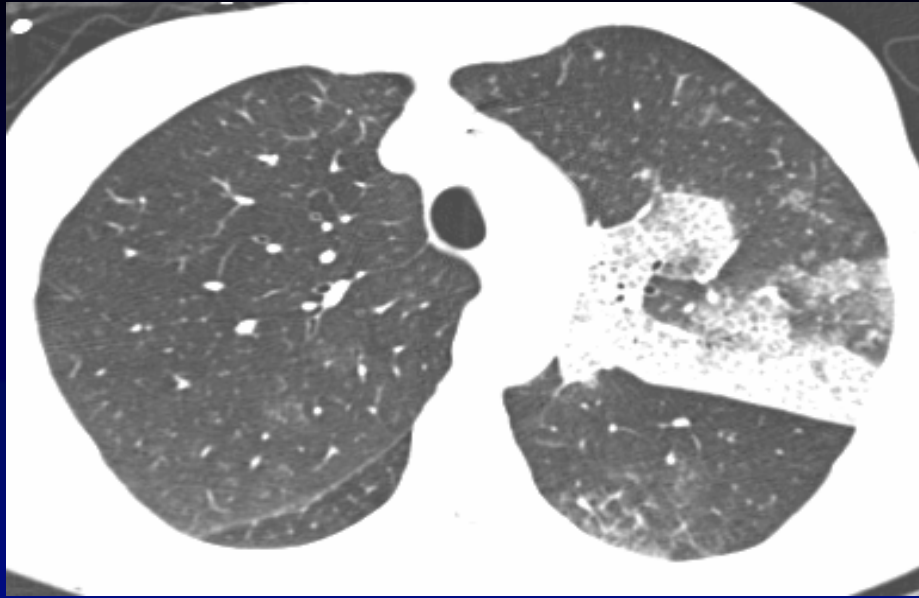


Alveolar (Ground Glass)

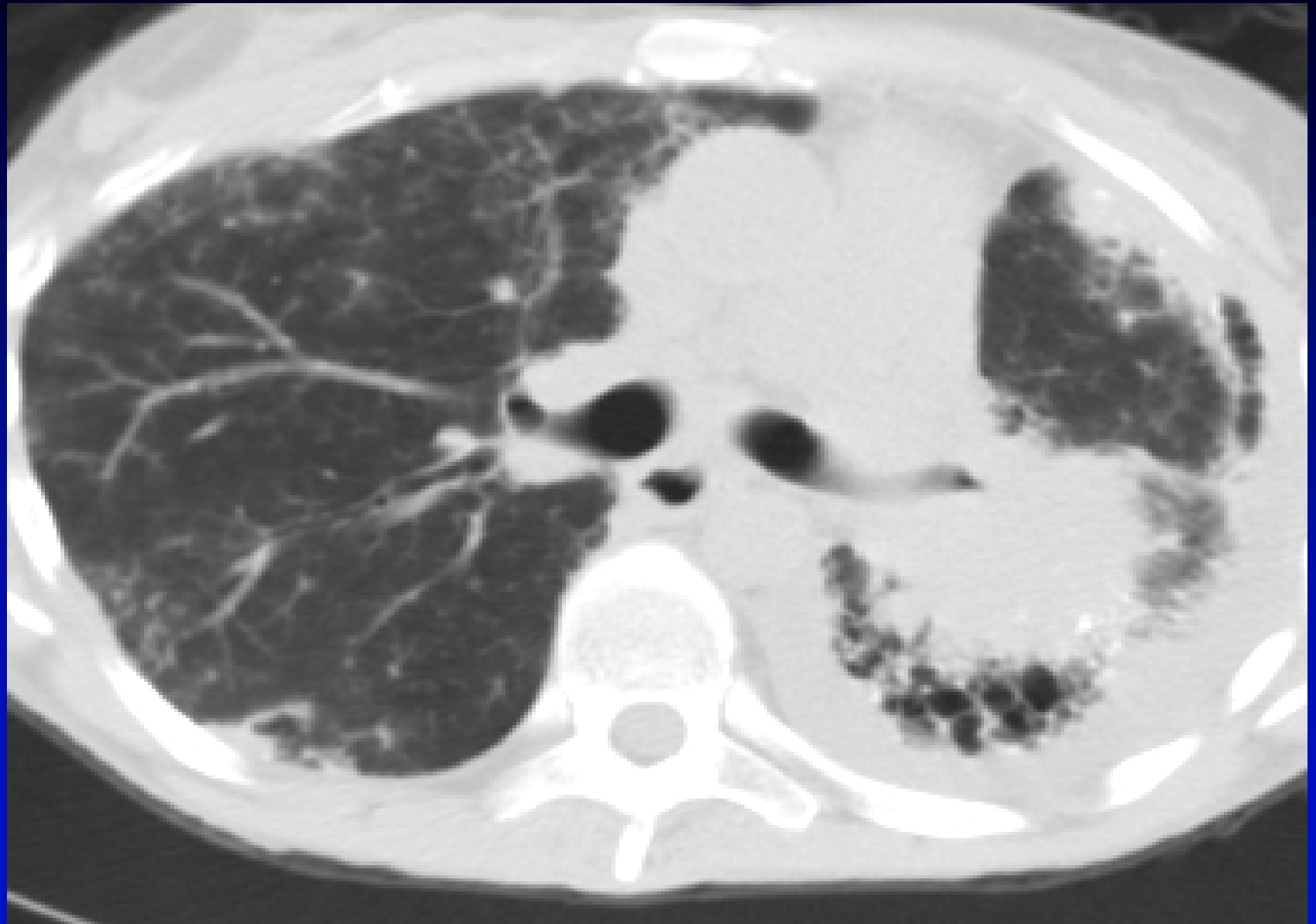
- IIPs (NSIP; AIP; DIP)
- Hypersensitivity pneumonia
- Organizing pneumonia
- Eosinophilic pneumonia
- Pulmonary alveolar proteinosis

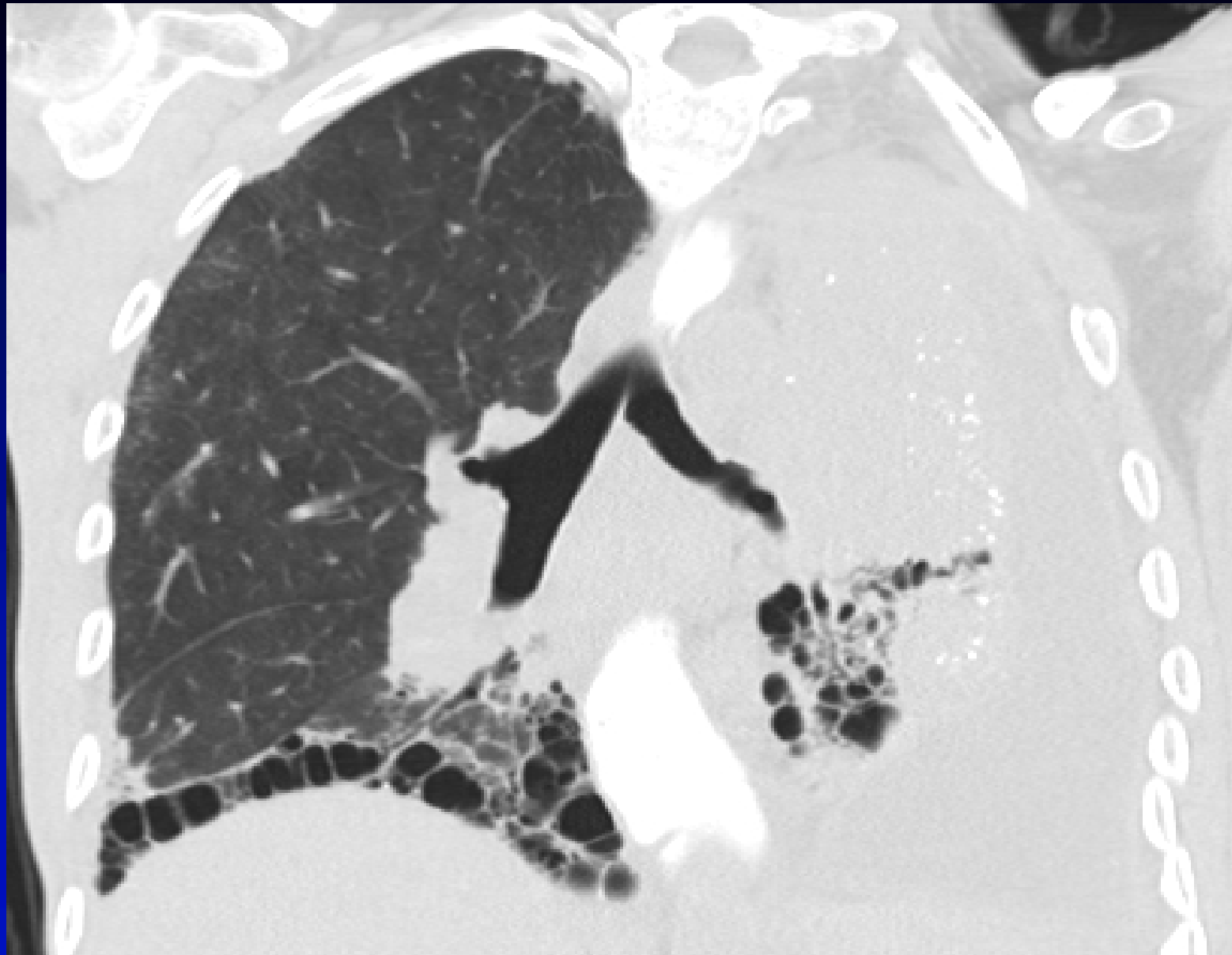
PCP

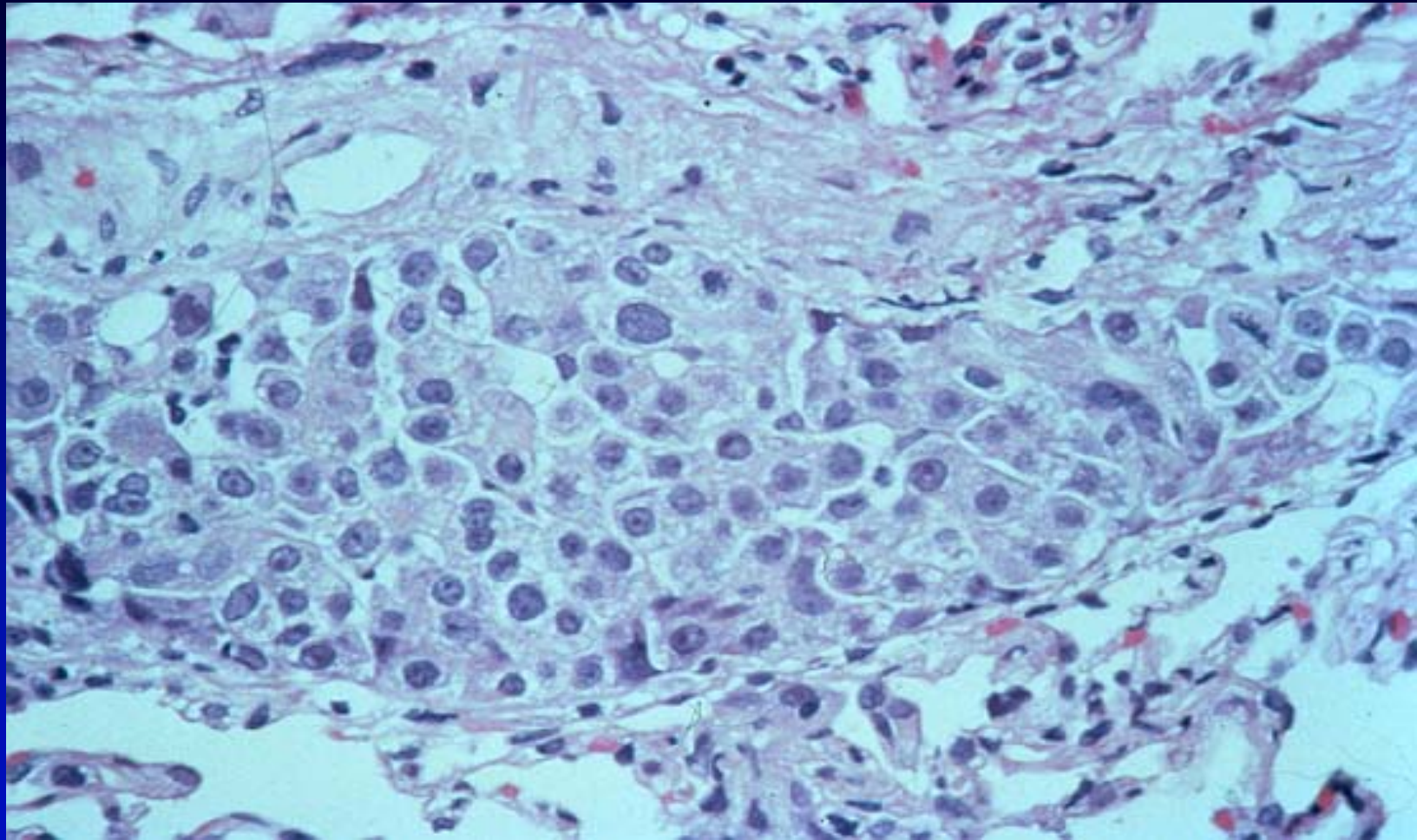




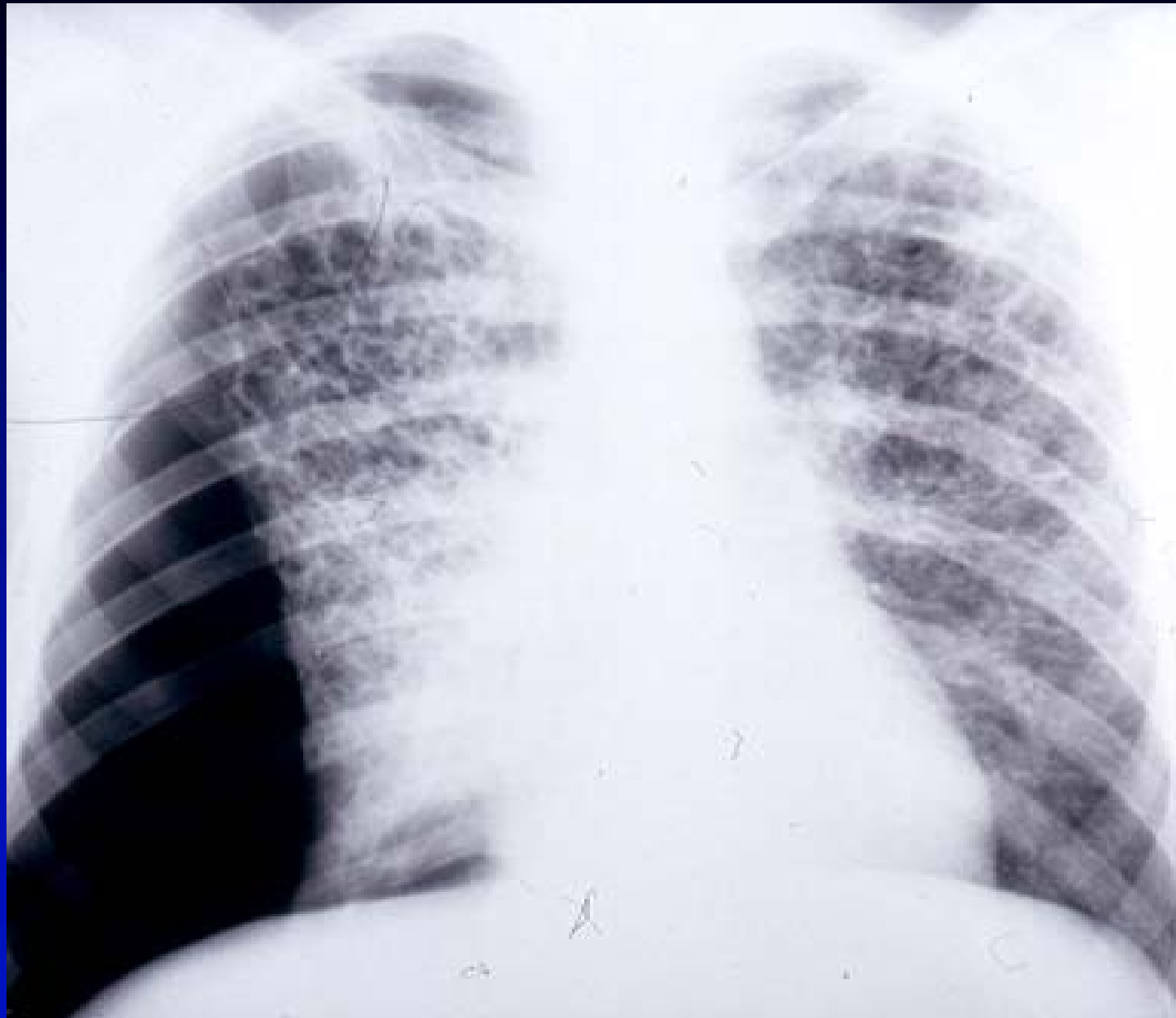
Multifocal Bronchoalveolar Cell Carcinoma

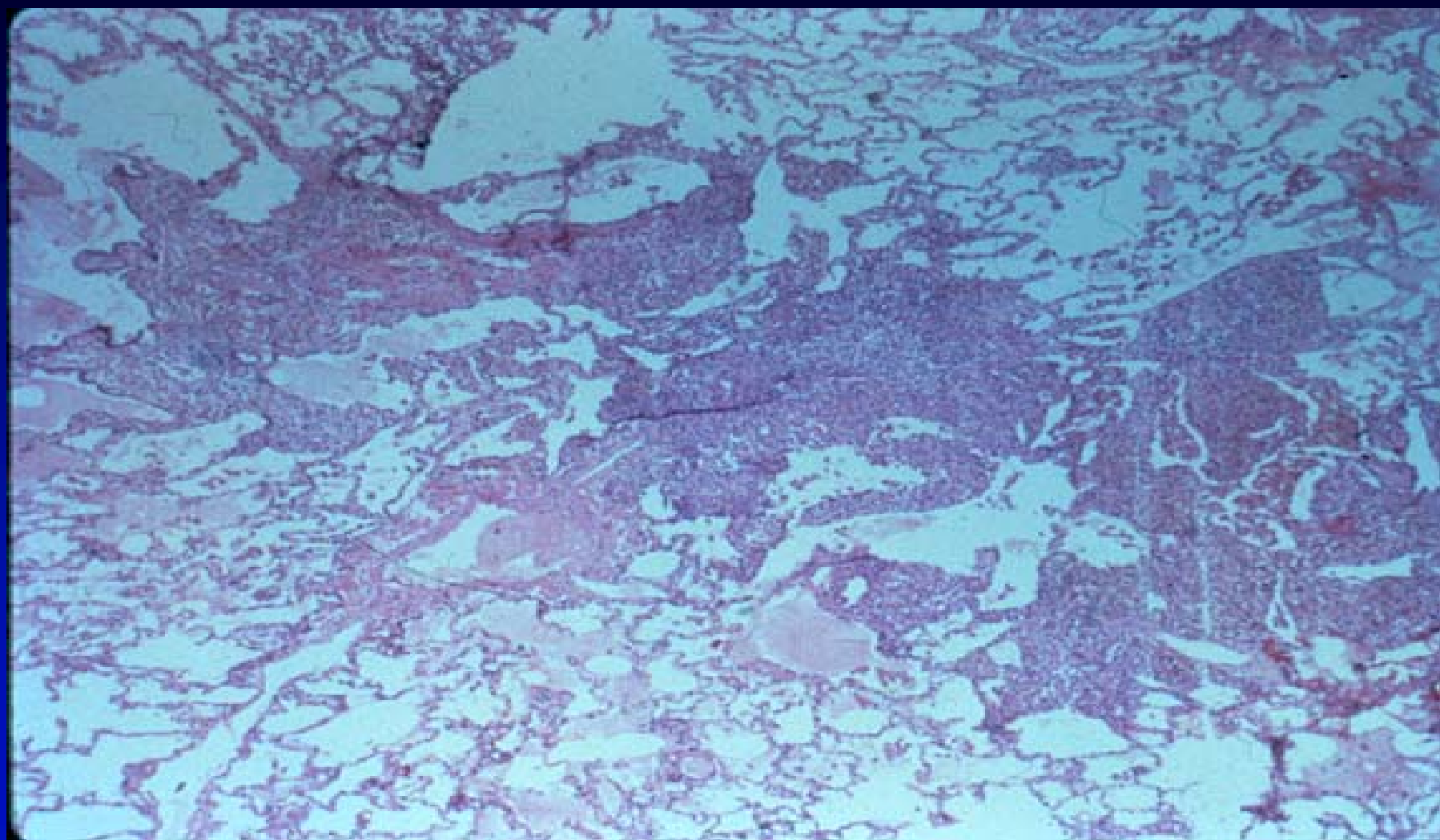


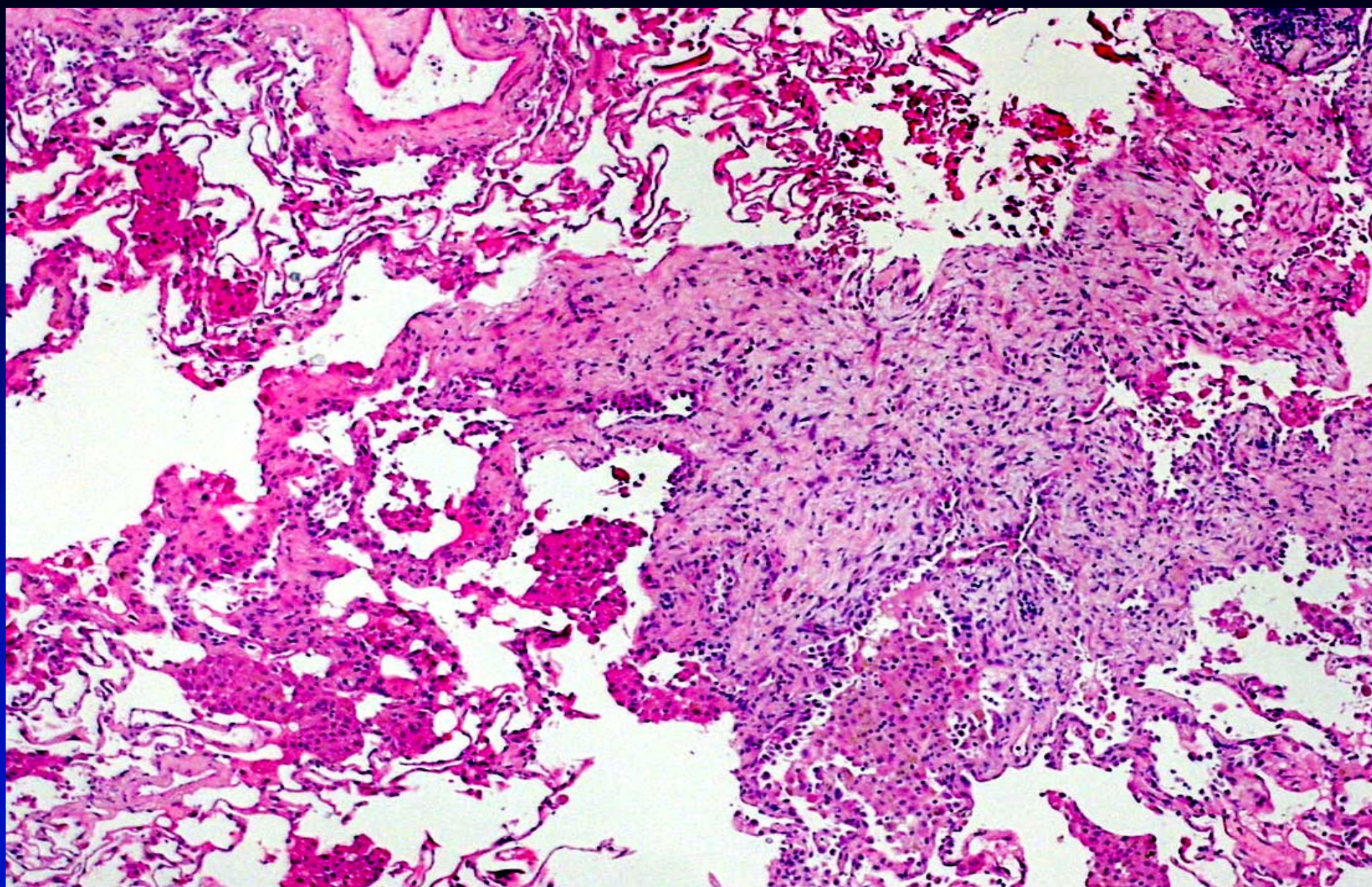


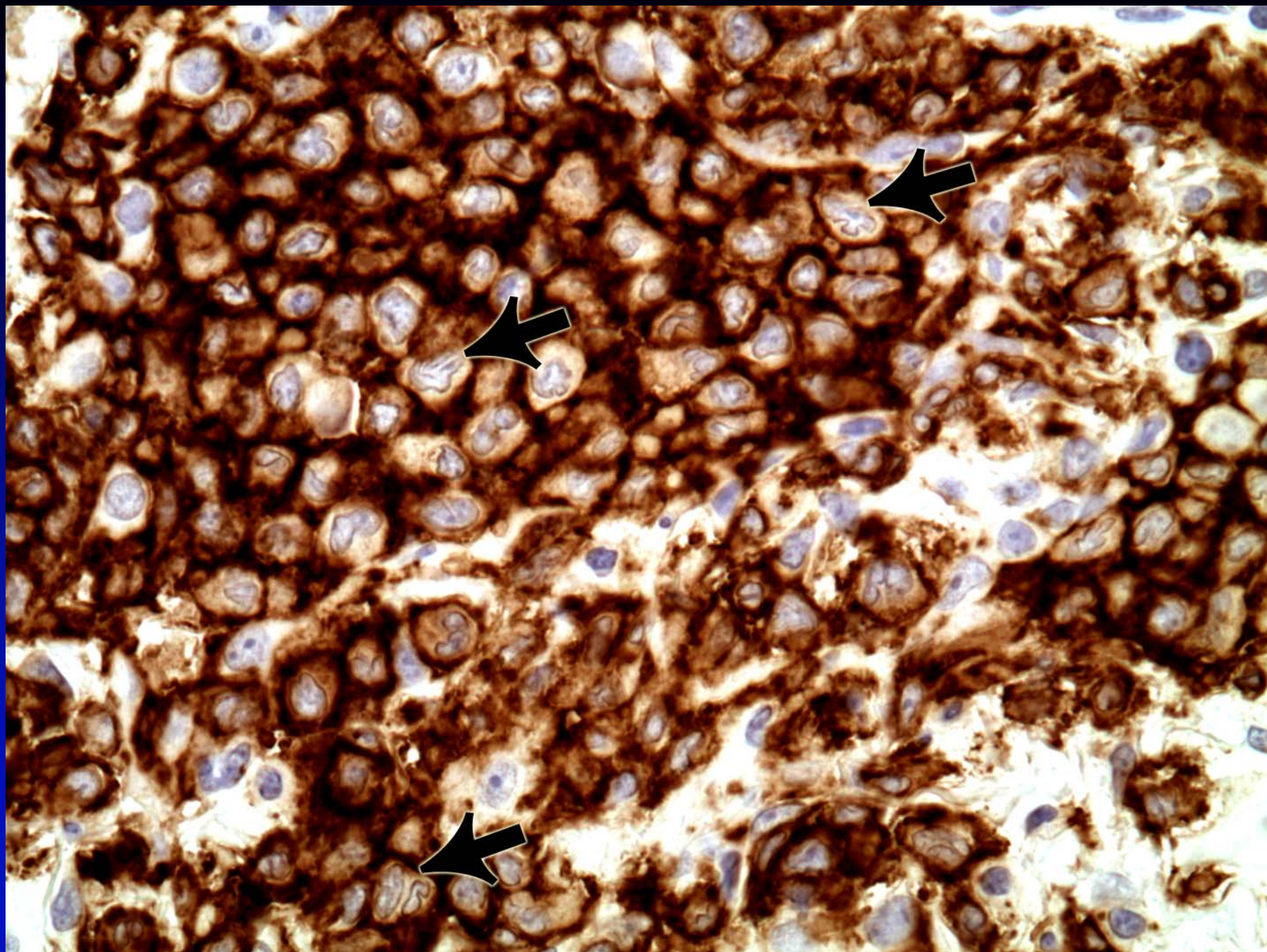


- **Cystic Lung Diseases**









Langerhans' cell granulomatosis

Formerly termed:

- Pulmonary eosinophilic granuloma
- Histiocytosis X
- Langerhans cell histiocytosis

Langerhans' cell granulomatosis

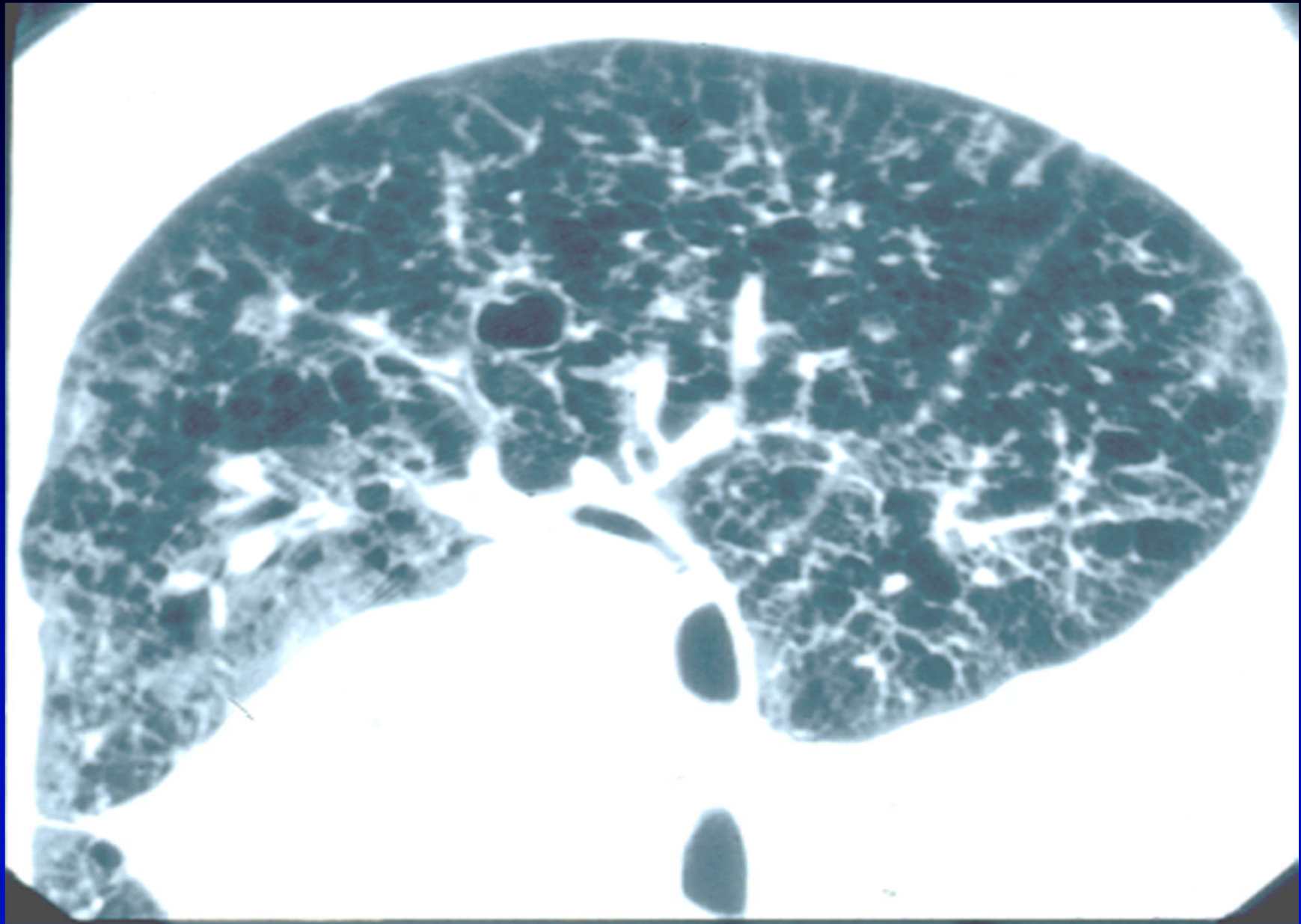
Epidemiology of LCG:

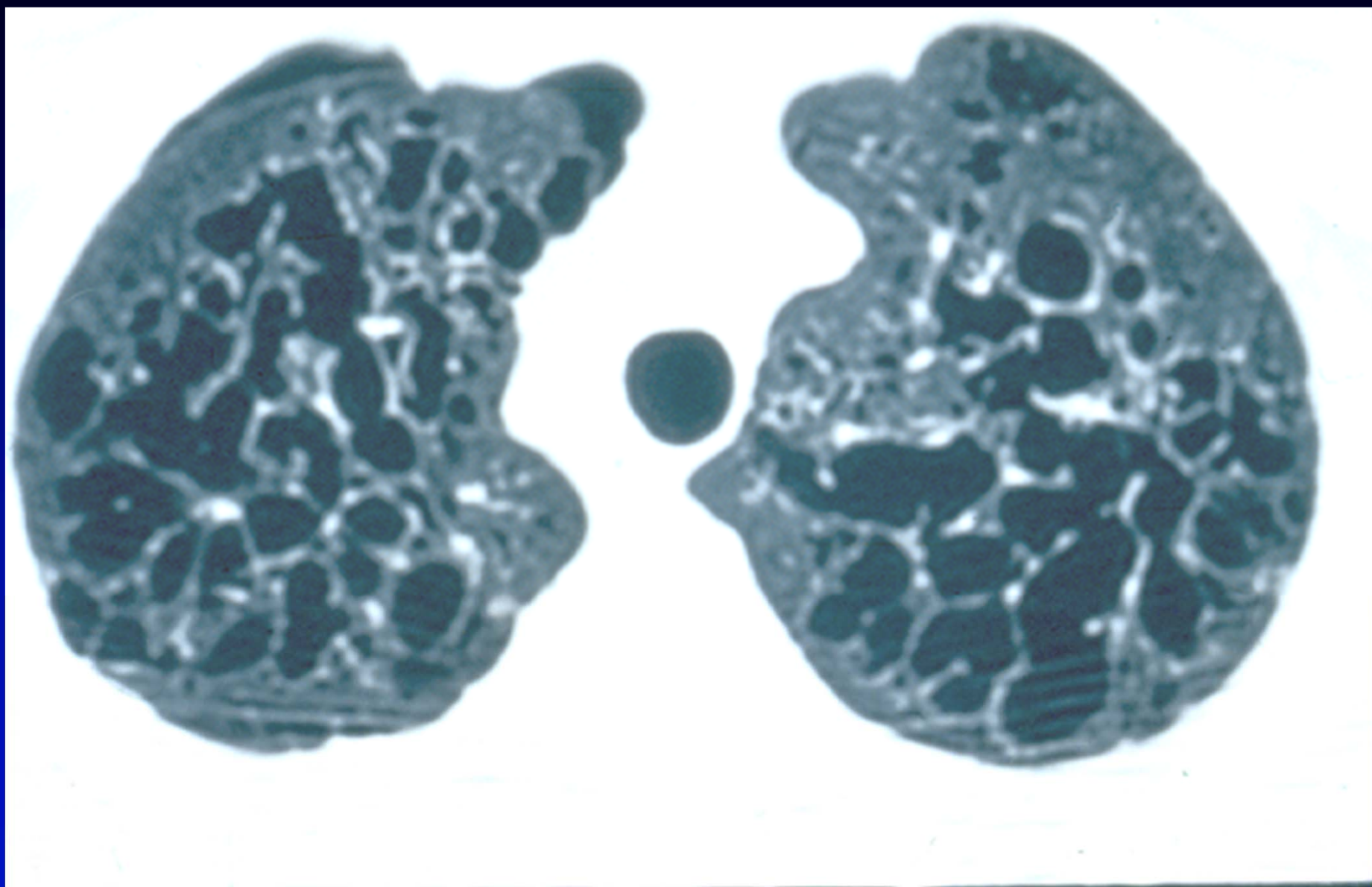
- Smokers (> 95%)
- Prevalence 2-5 cases/million

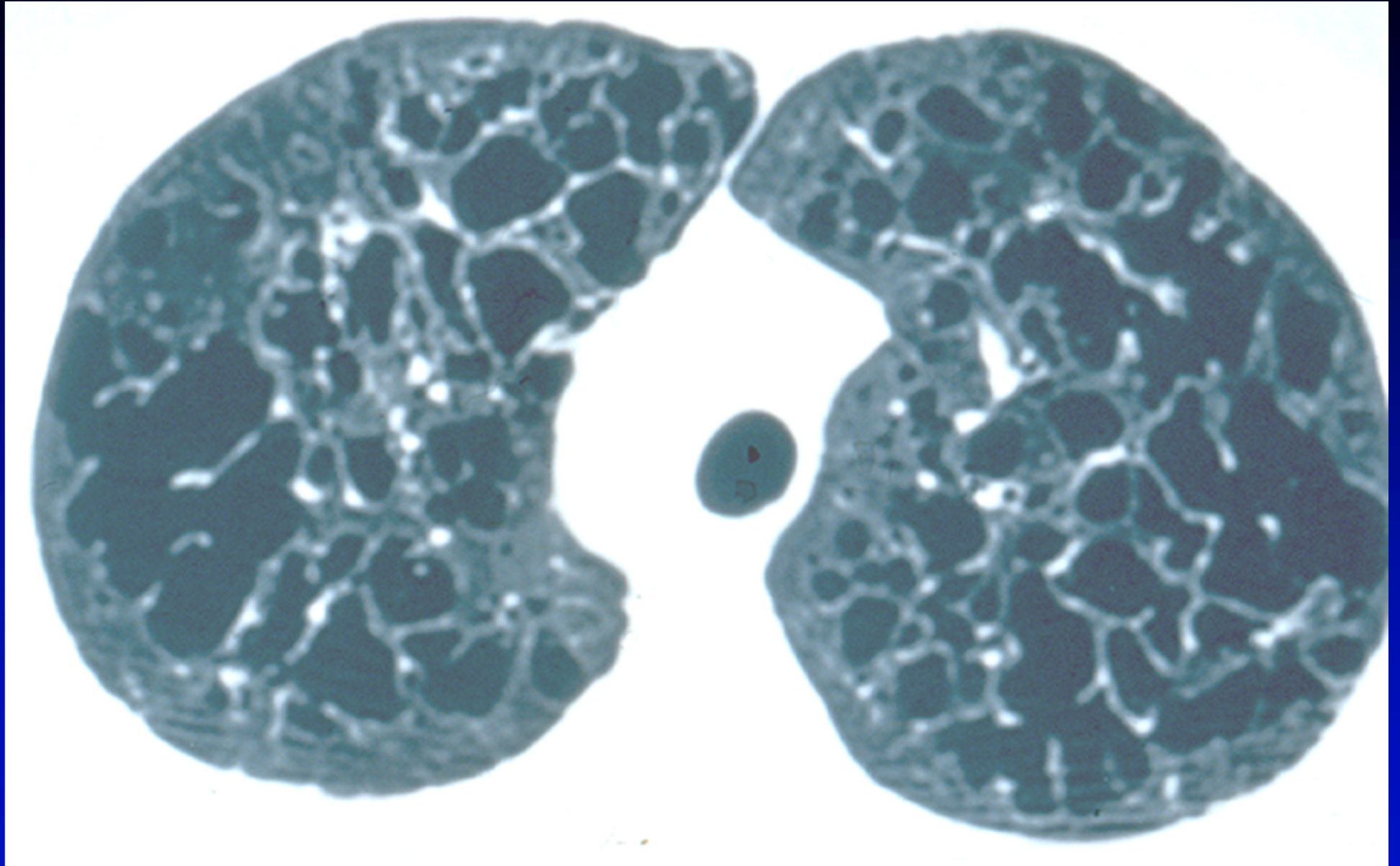
Langerhans' cell granulomatosis

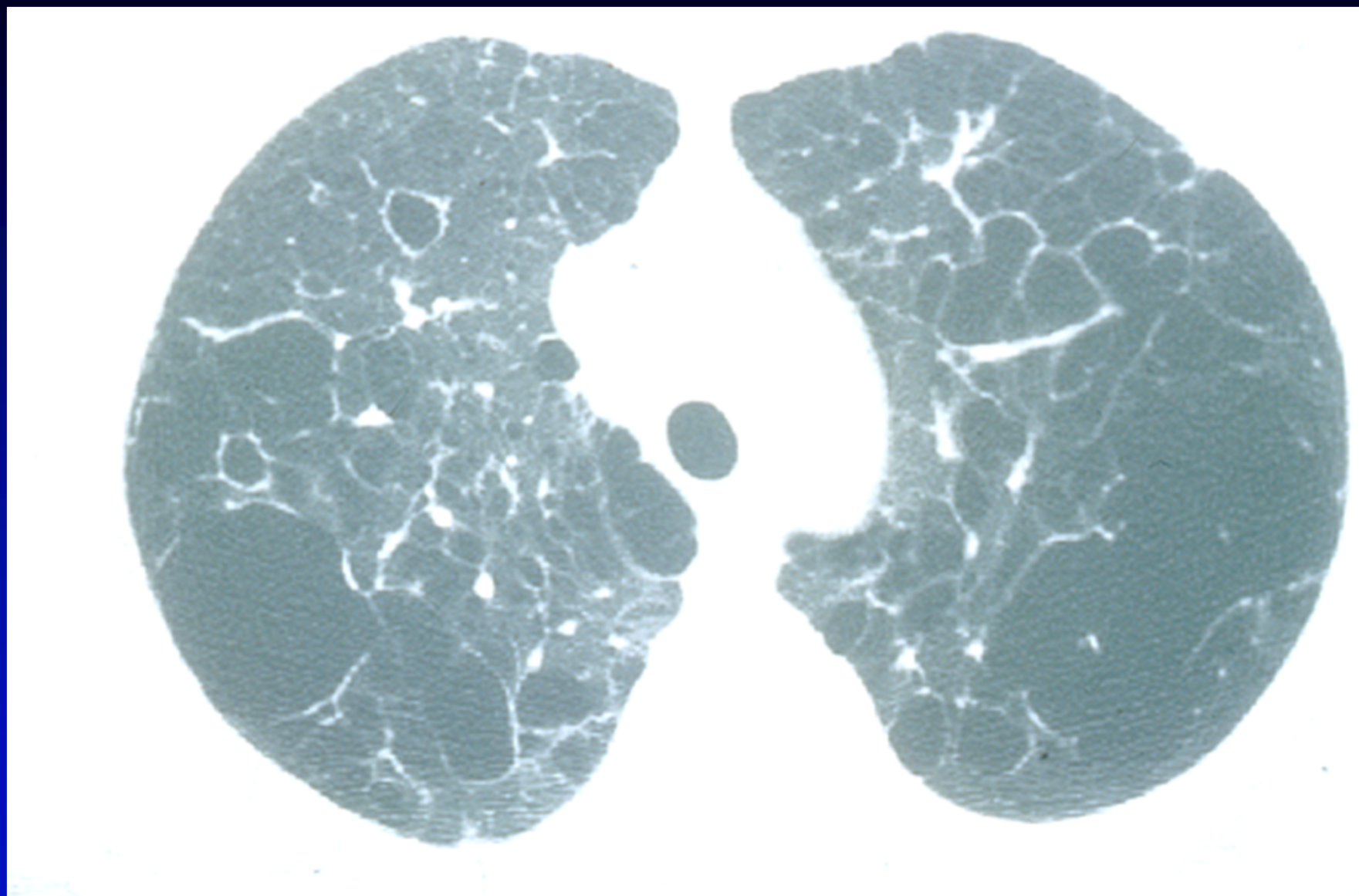
HRCT Features of LCG:

- Upper lobe predominance
- Numerous thin-walled cysts
- Peribronchiolar nodules











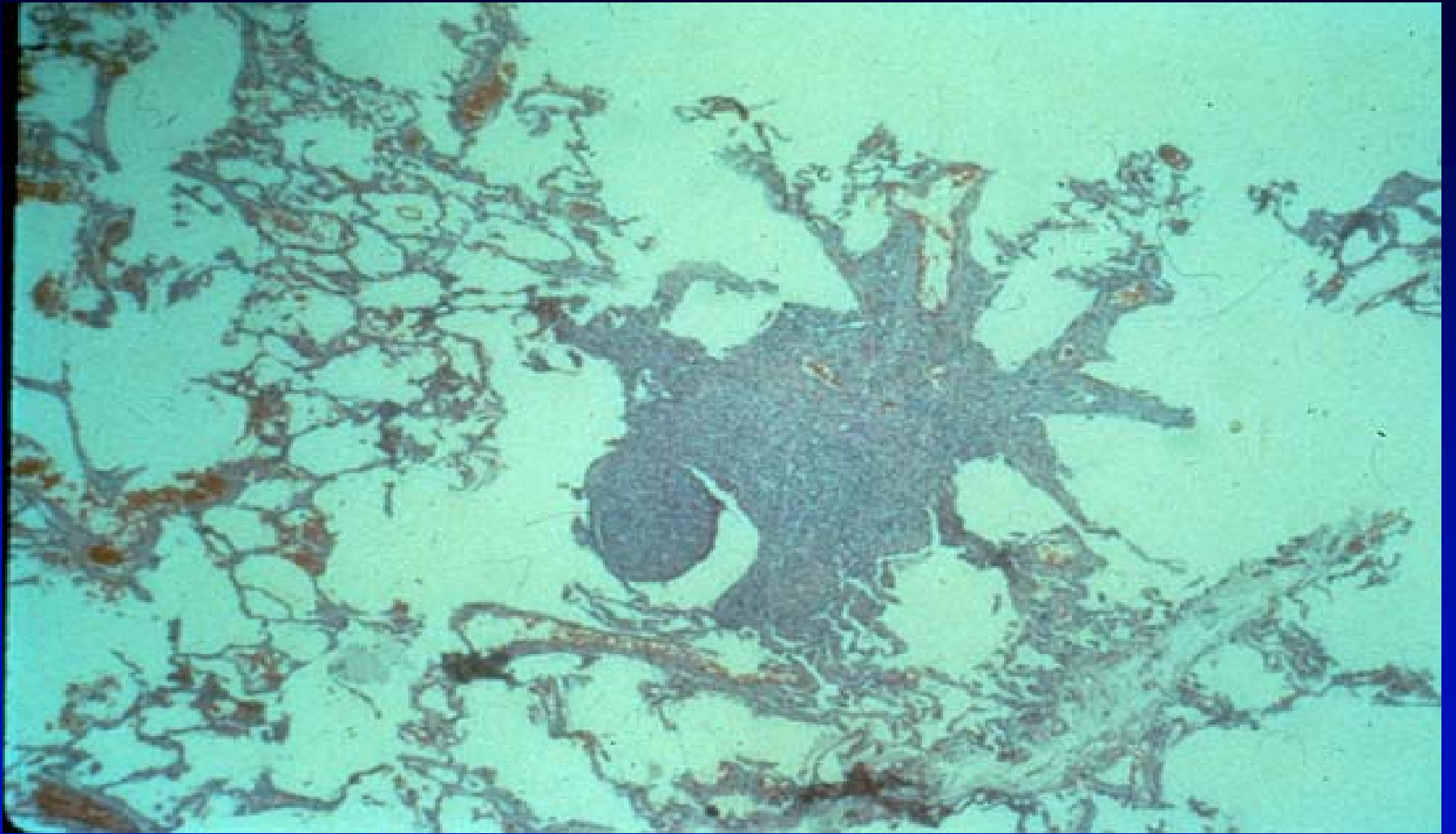


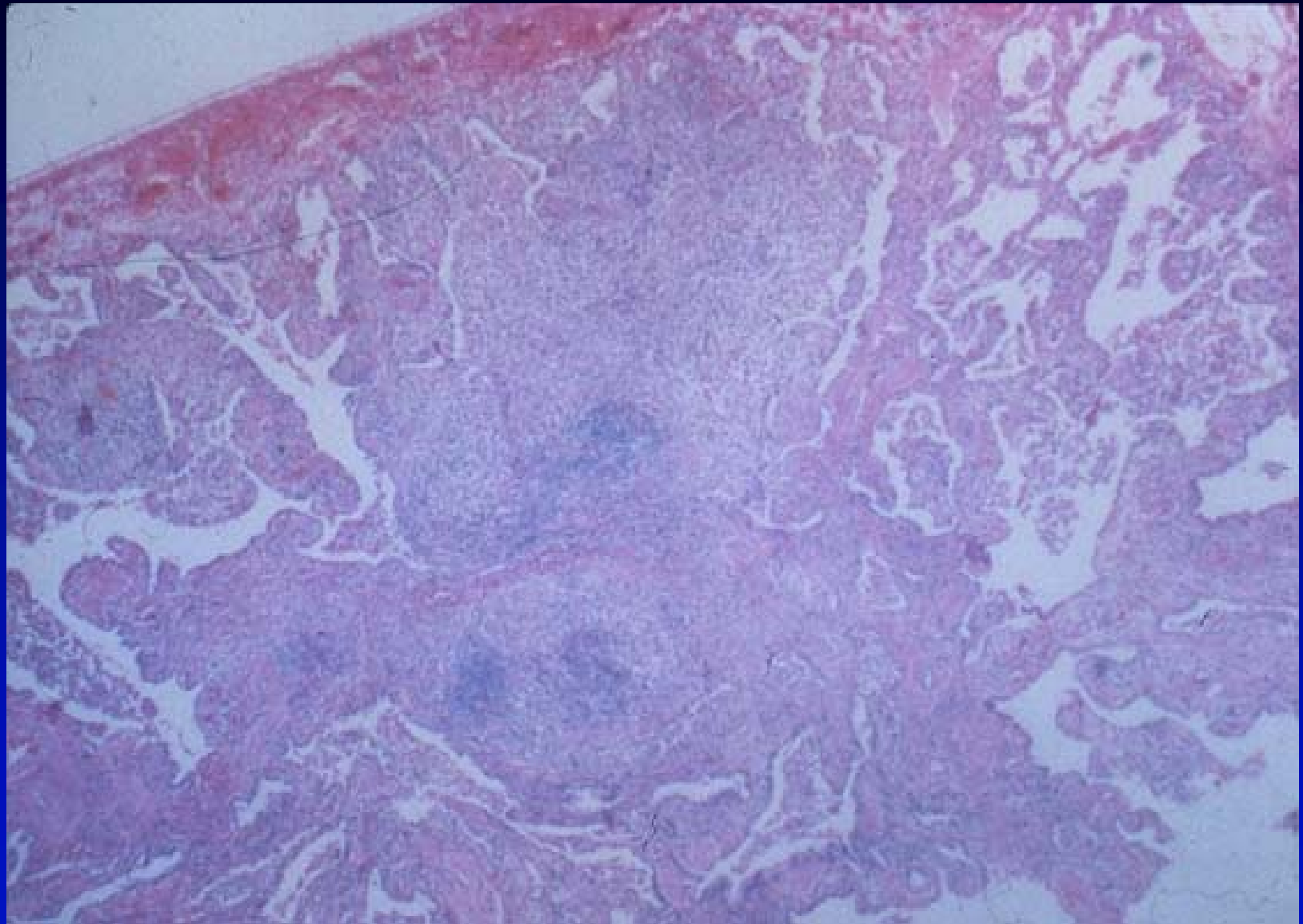


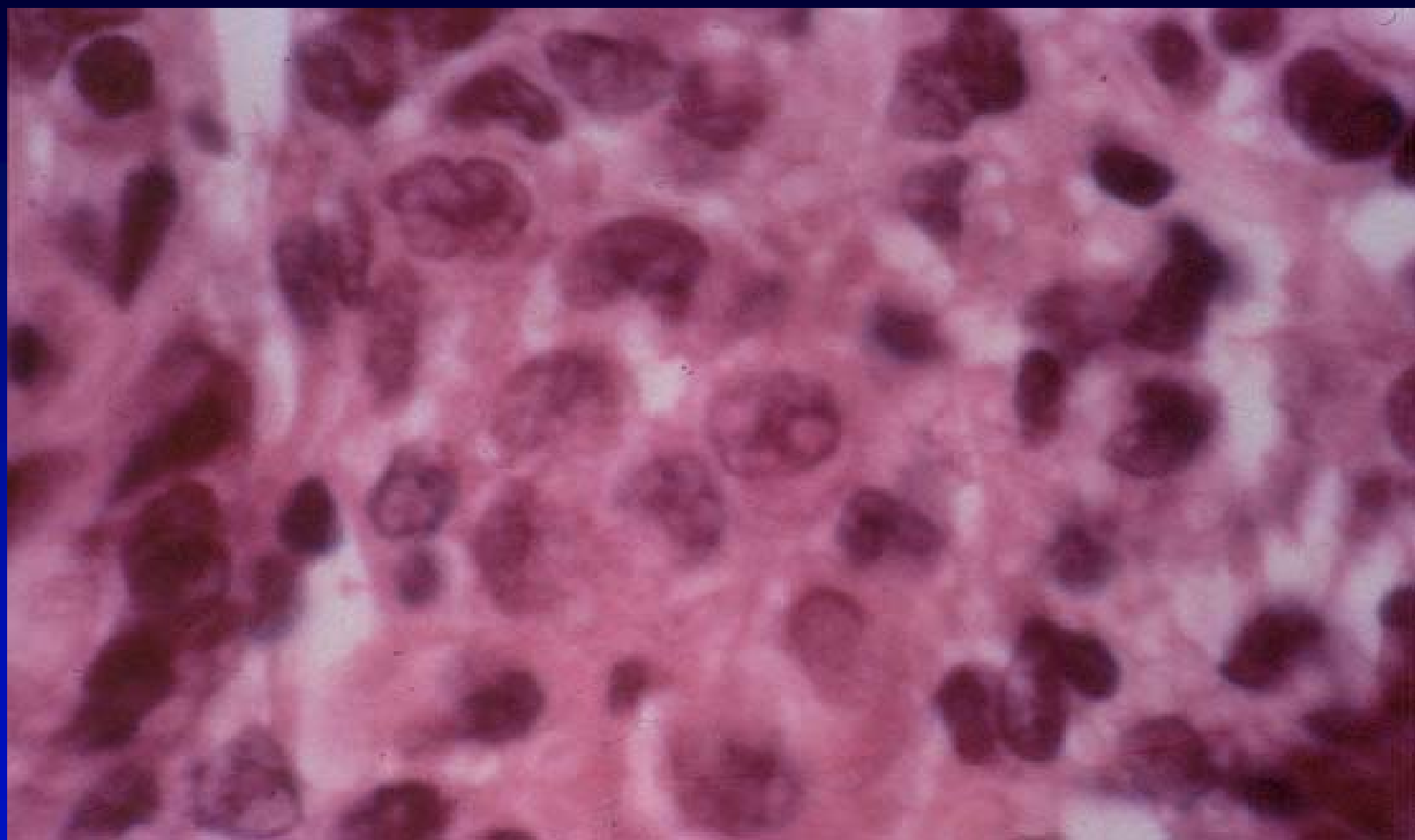
Langerhans' cell granulomatosis

Histopathology of LCG:

- Langerhans histocytes, S-100 (+)
- **Stellate pattern fibrosis** (low power)
- Inflammation; cysts, nodules







Langerhans' cell granulomatosis

Immunohistochemical stains

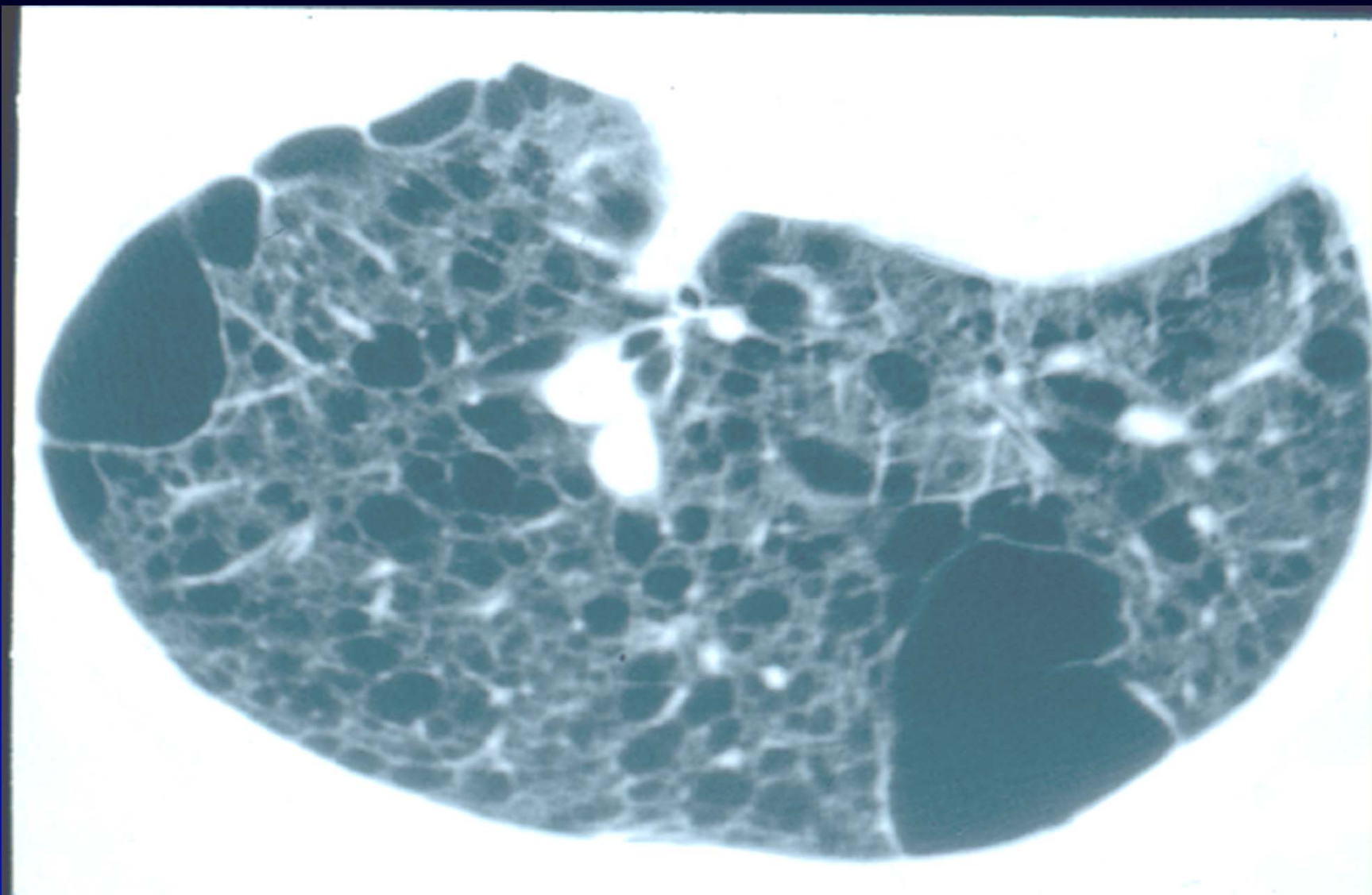
(Lung, BAL):

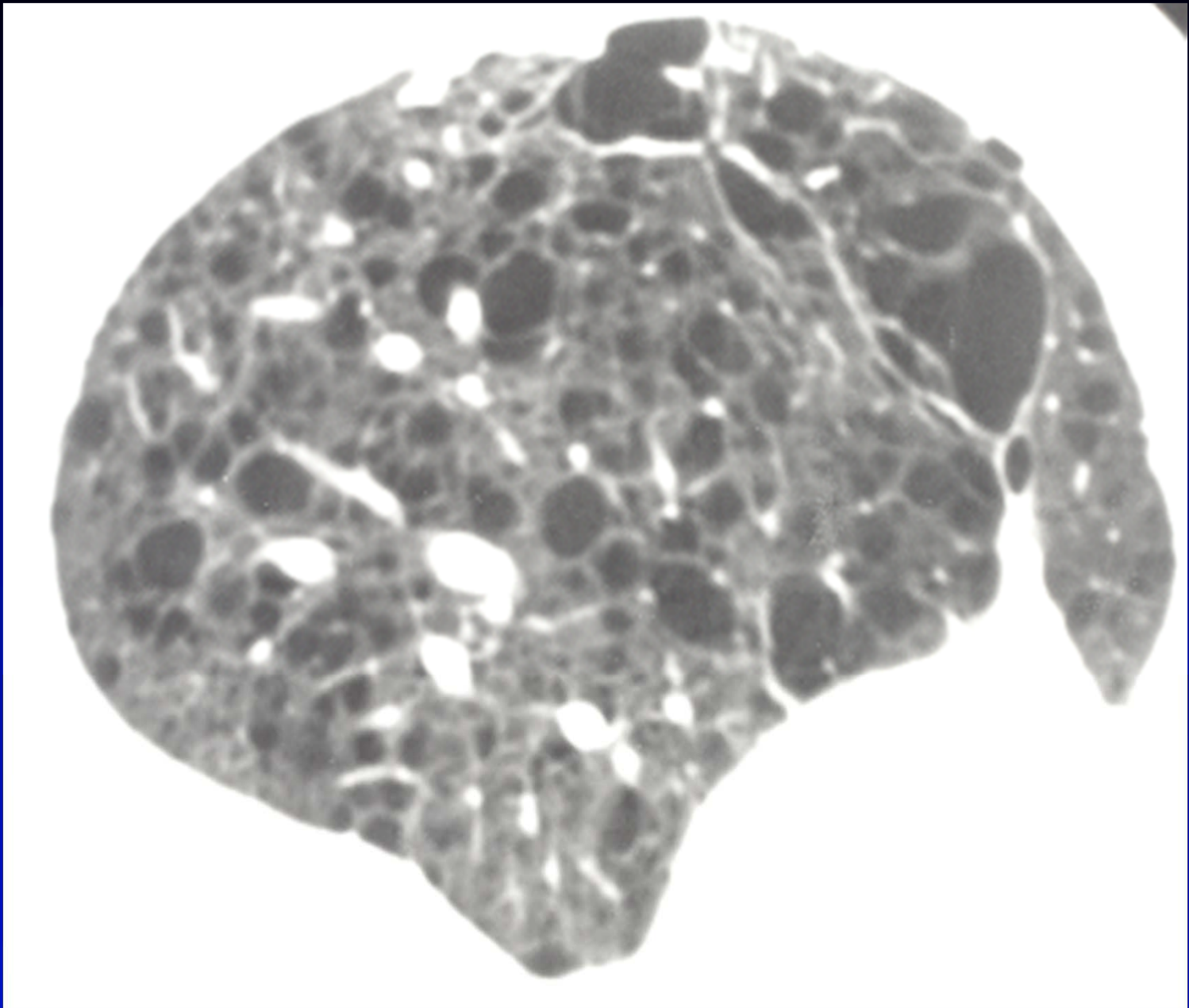
- S-100 protein
- OKT6 (CD1a antigen)

Langerhans' cell granulomatosis

Natural history variable:

- Cessation smoking paramount
- No proven therapy
- Lung transplantation (end-stage)





07-59-03 AM
BONE

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R

kV 120
mA 140

Large
1.5mm

Tilt 0.0

2 0 0 07-59-03 AM

0150

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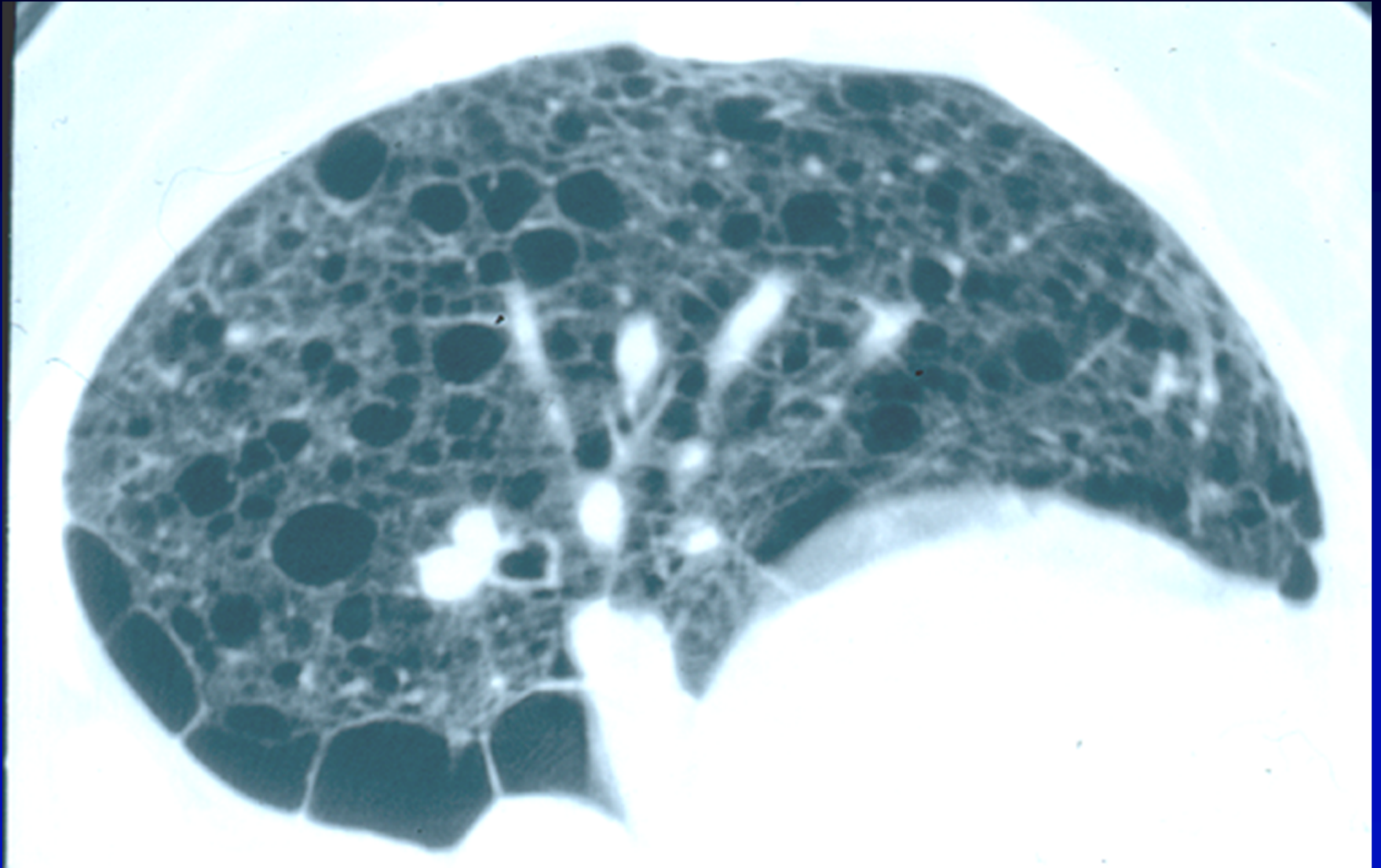
Lymphangioleiomyomatosis (LAM)

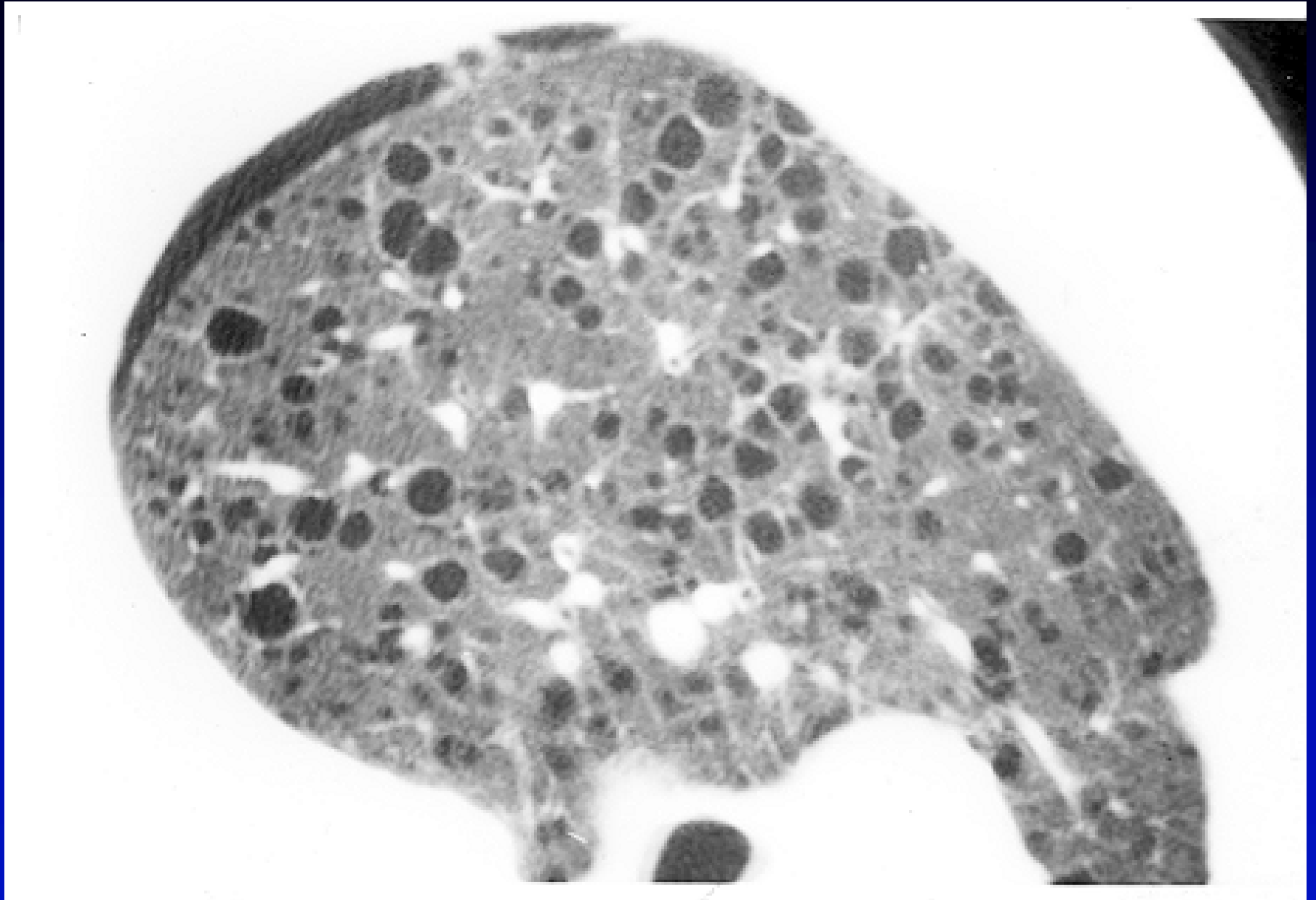
- Mean age onset 30-36 y
- Pre-menopausal women > 95%
- Prevalence 1-2 per million

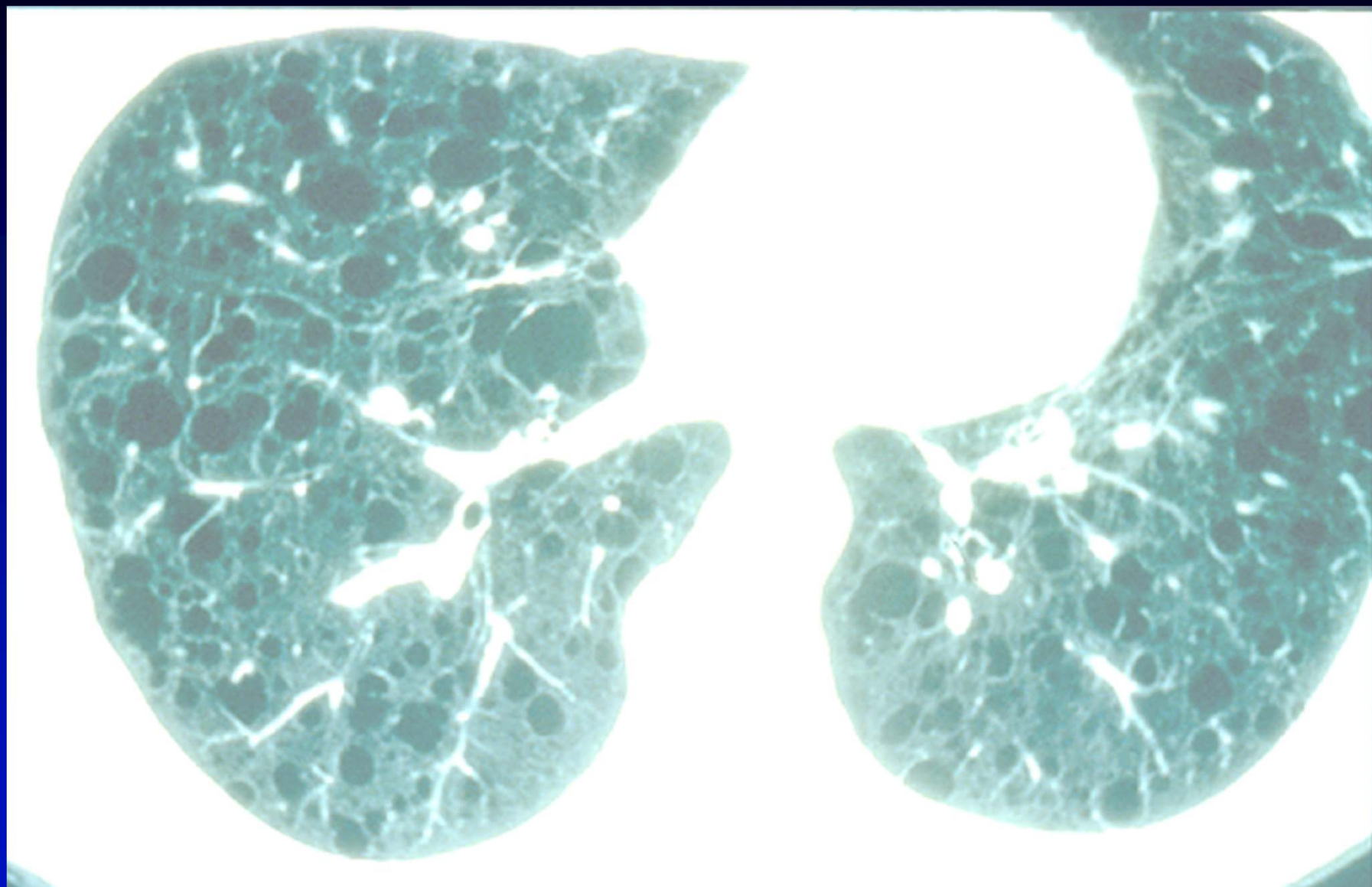
Lymphangioleiomyomatosis

HRCT features of LAM:

- Numerous thin-walled cysts
- Diffuse, no predominance
- No nodules or fibrosis









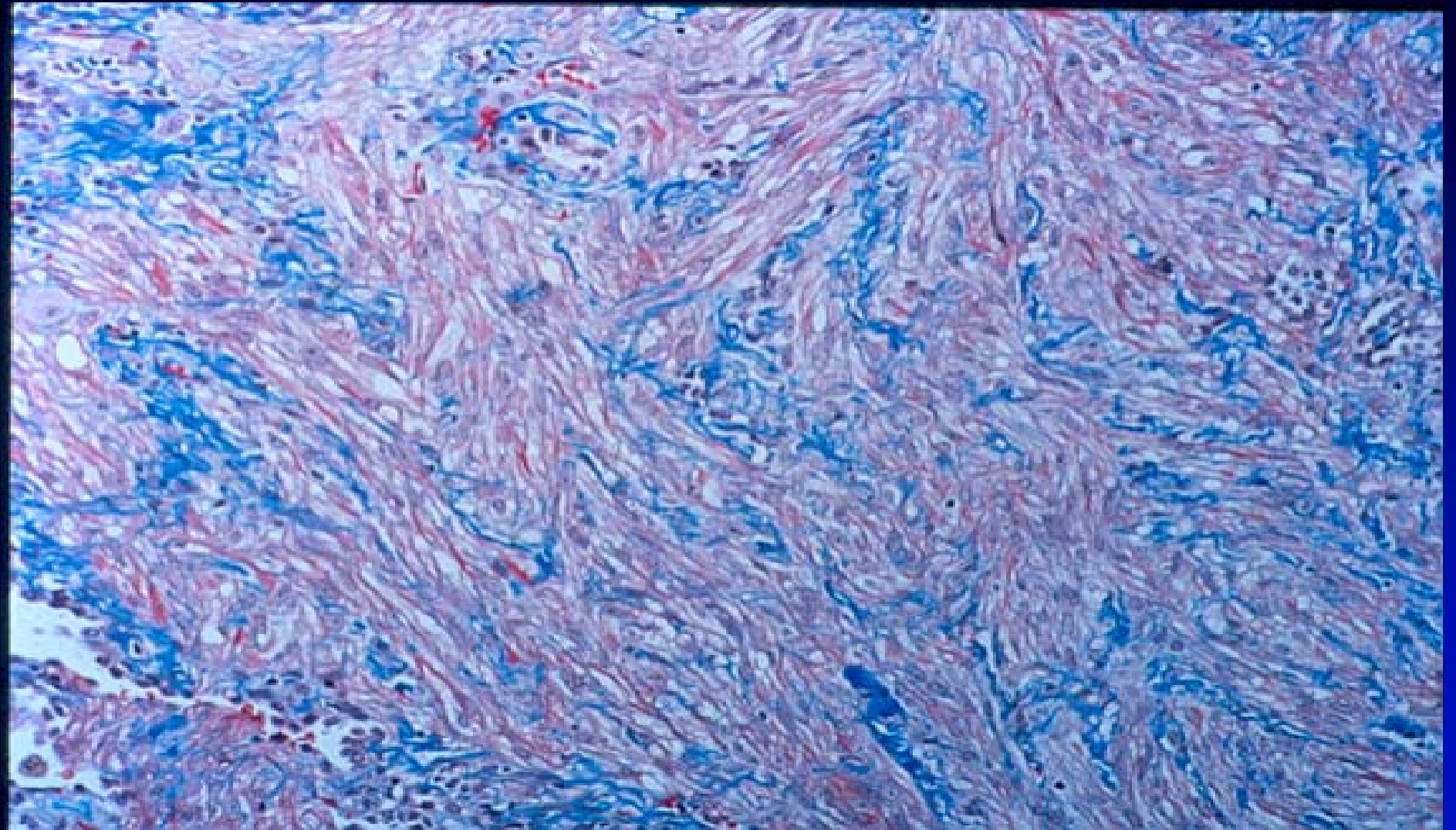




Lymphangioleiomyomatosis

Histology of LAM:

- Proliferating atypical smooth m.
- HMB-45 (+) smooth muscle cells
- Extensive cysts



Lymphangioleiomyomatosis

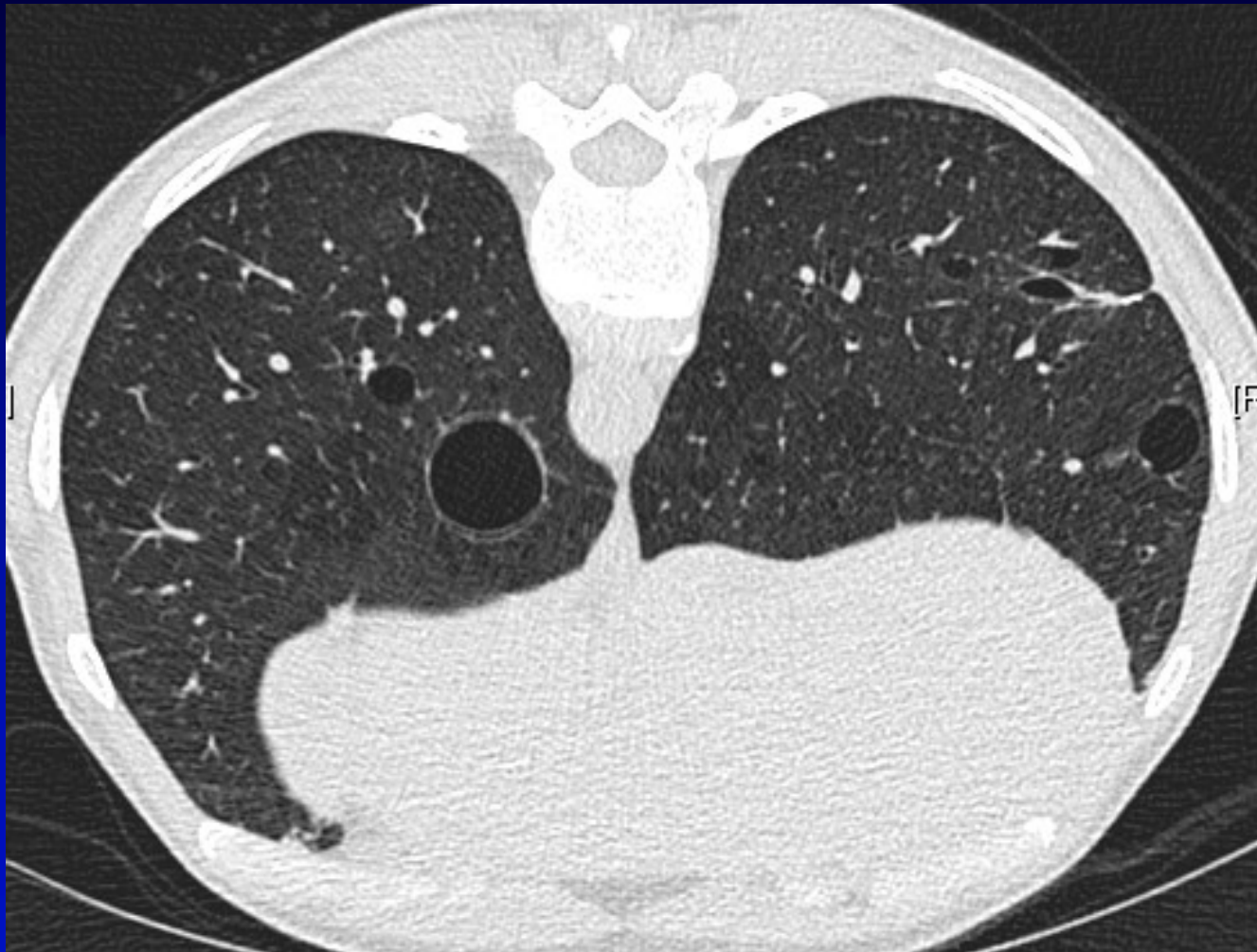
- Course indolent, but progressive
- Die of respiratory failure
- 10 y survival variable (20-79%)
- No proven effective therapy

Cystic Lung Diseases

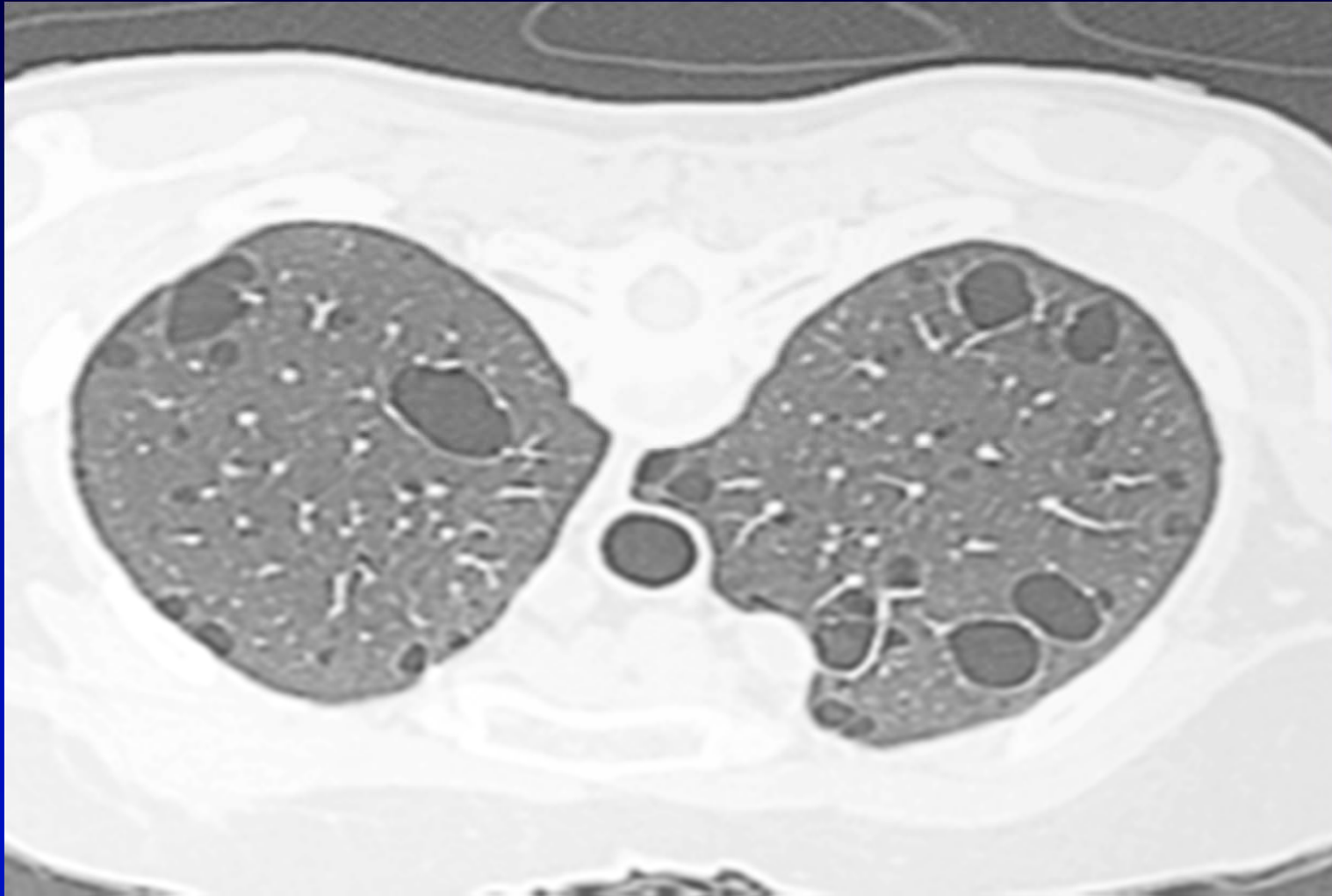
- Idiopathic pulmonary fibrosis
- Sarcoidosis
- Langerhans cell granulomatosis
- Lymphangioleiomyomatosis

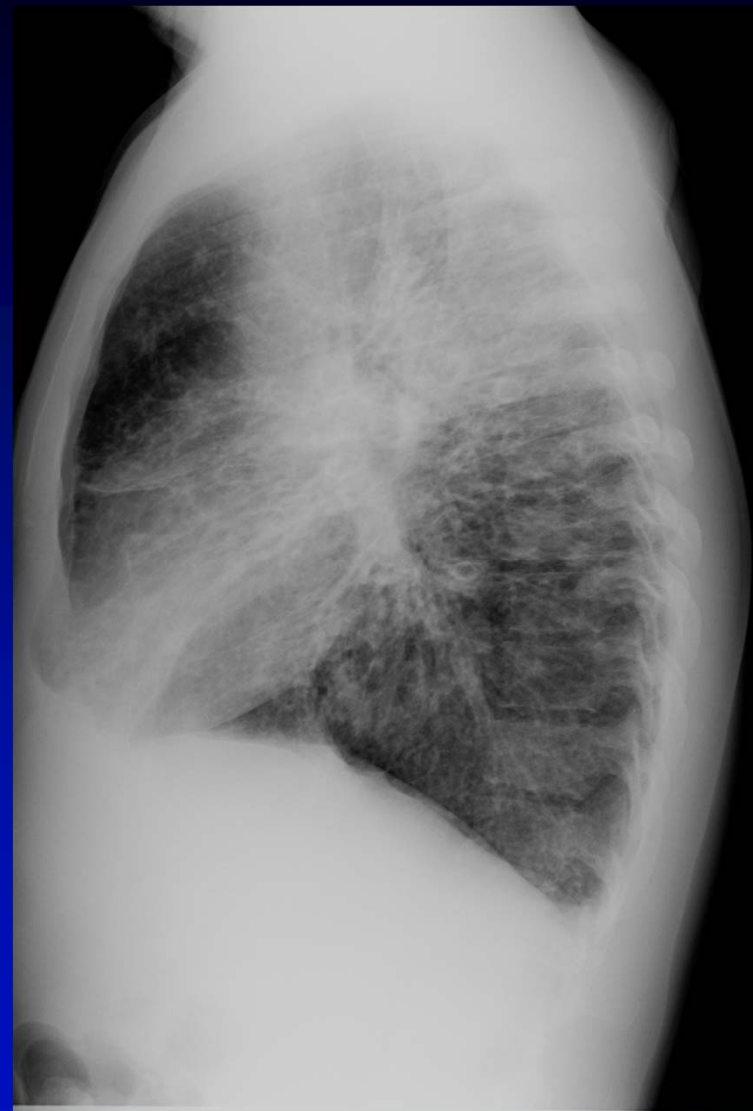
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Lymphocytic Bronchiolitis

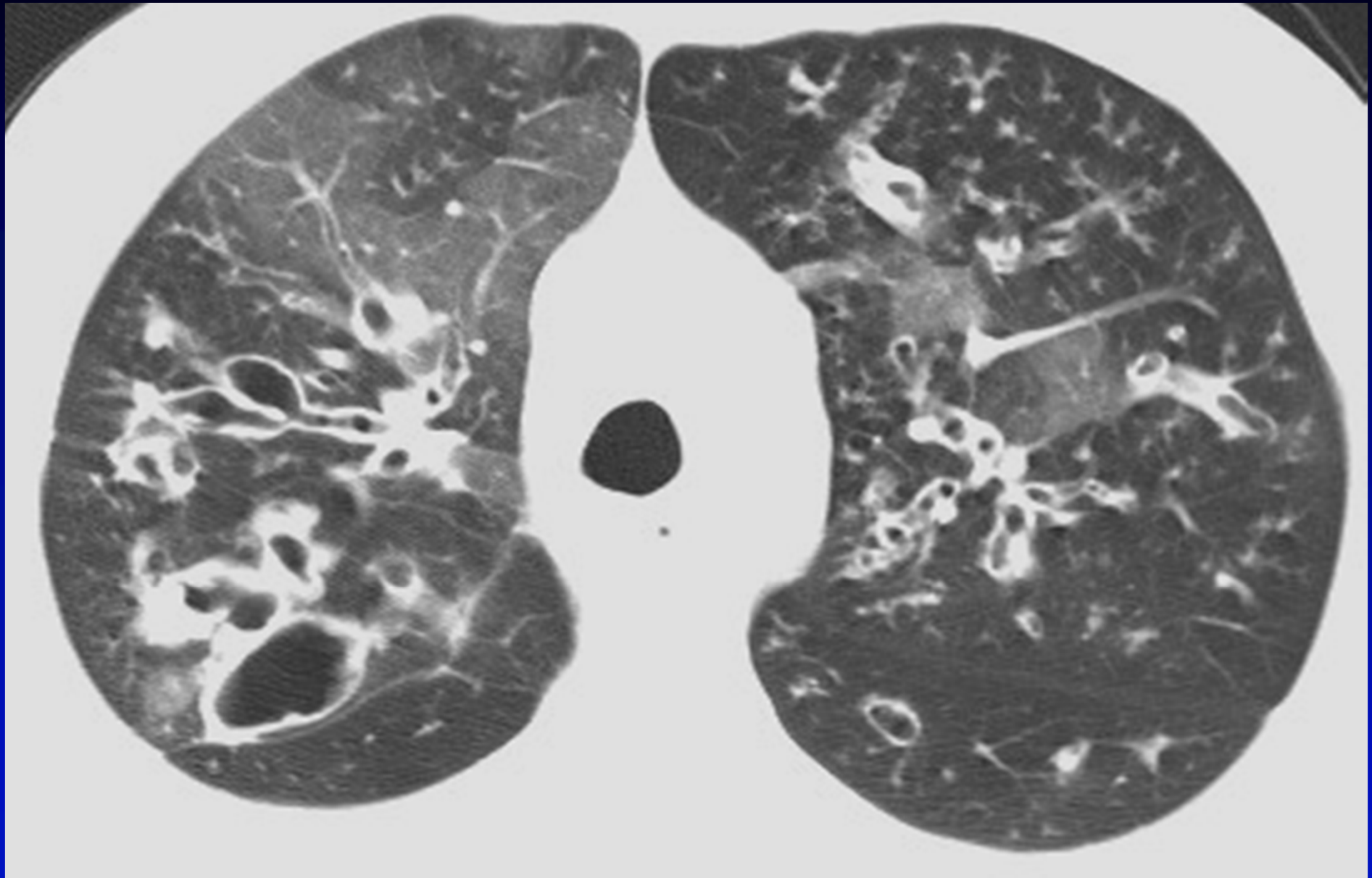


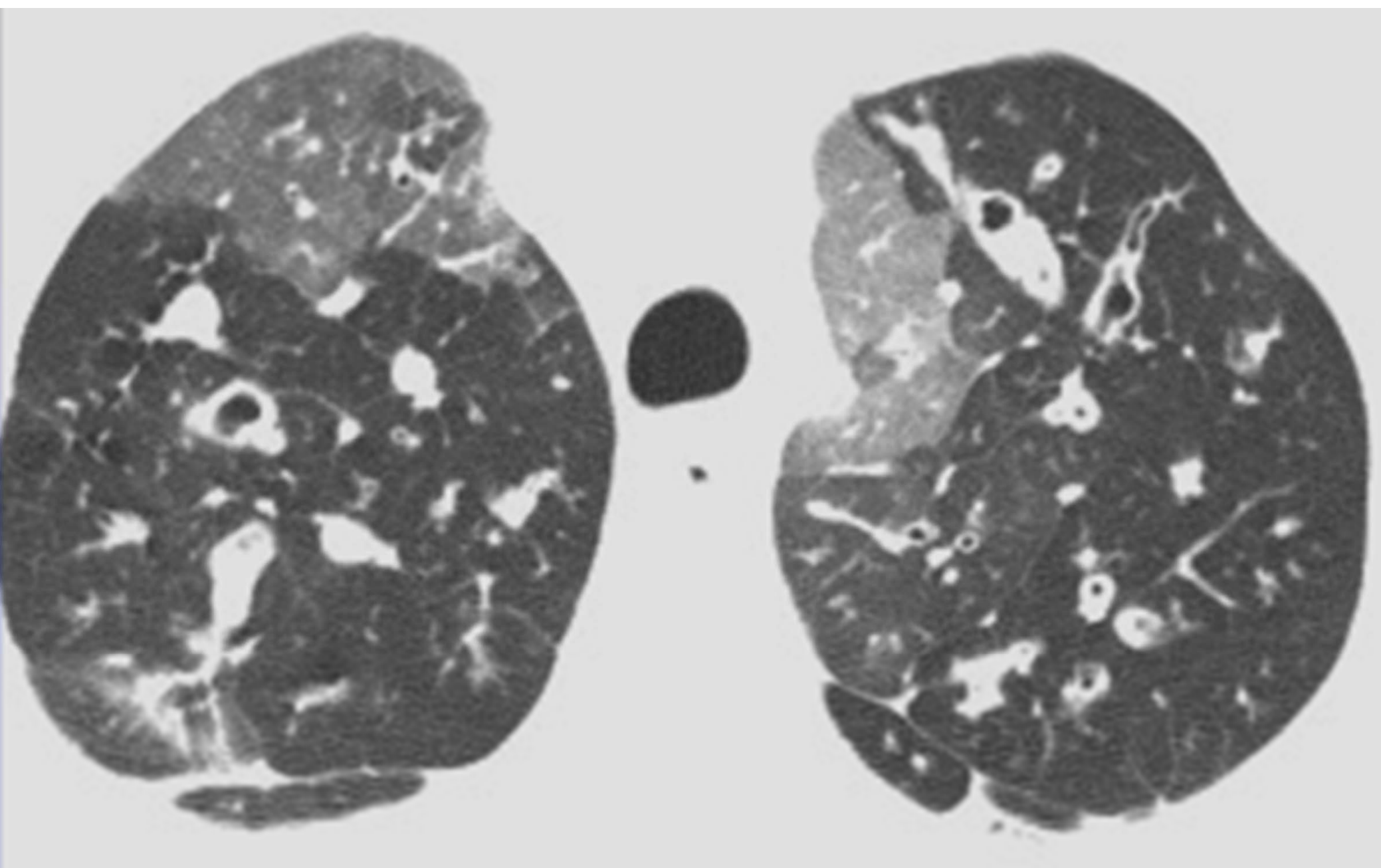
LIP (Sjogren's syndrome)

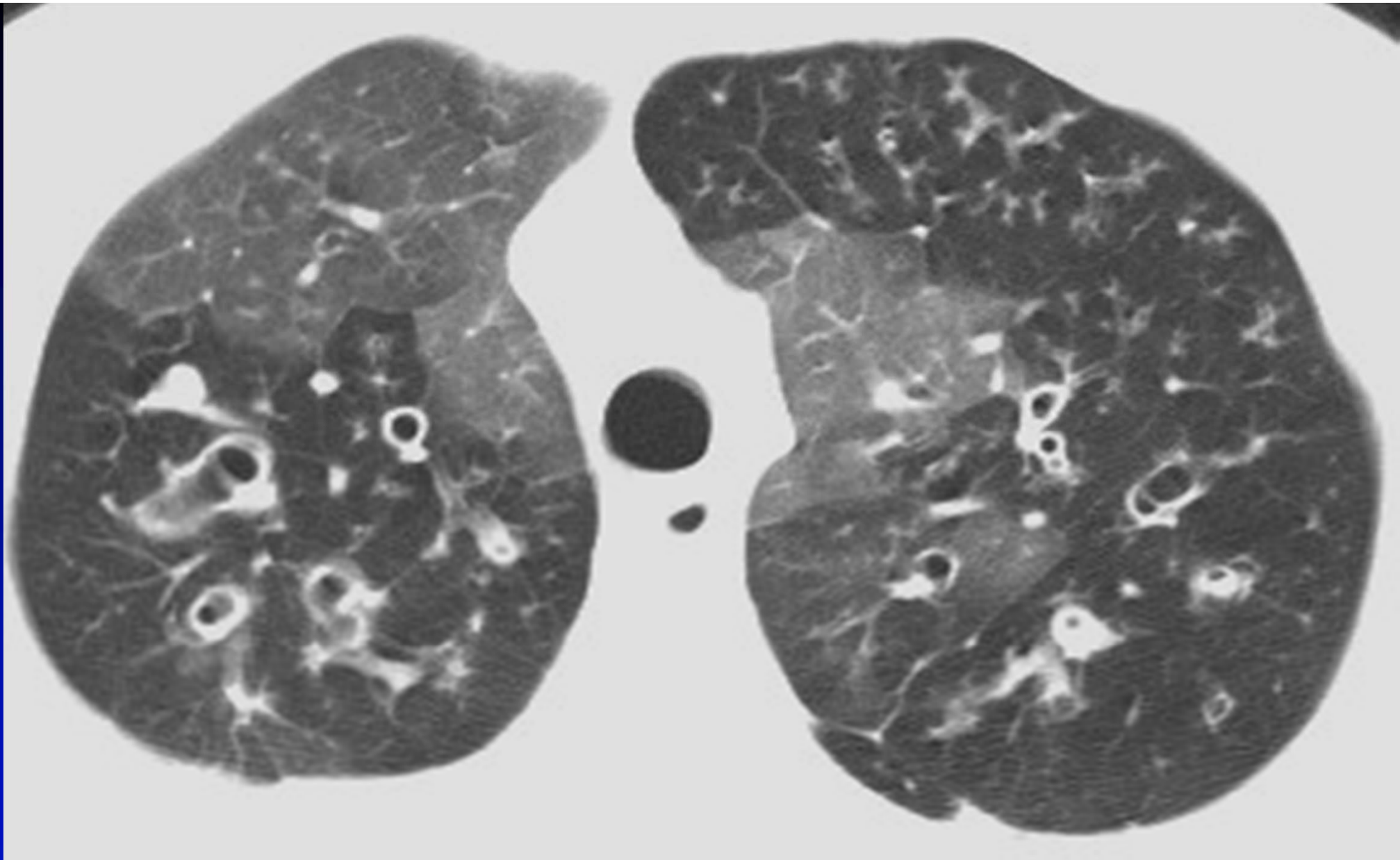




227-81-92

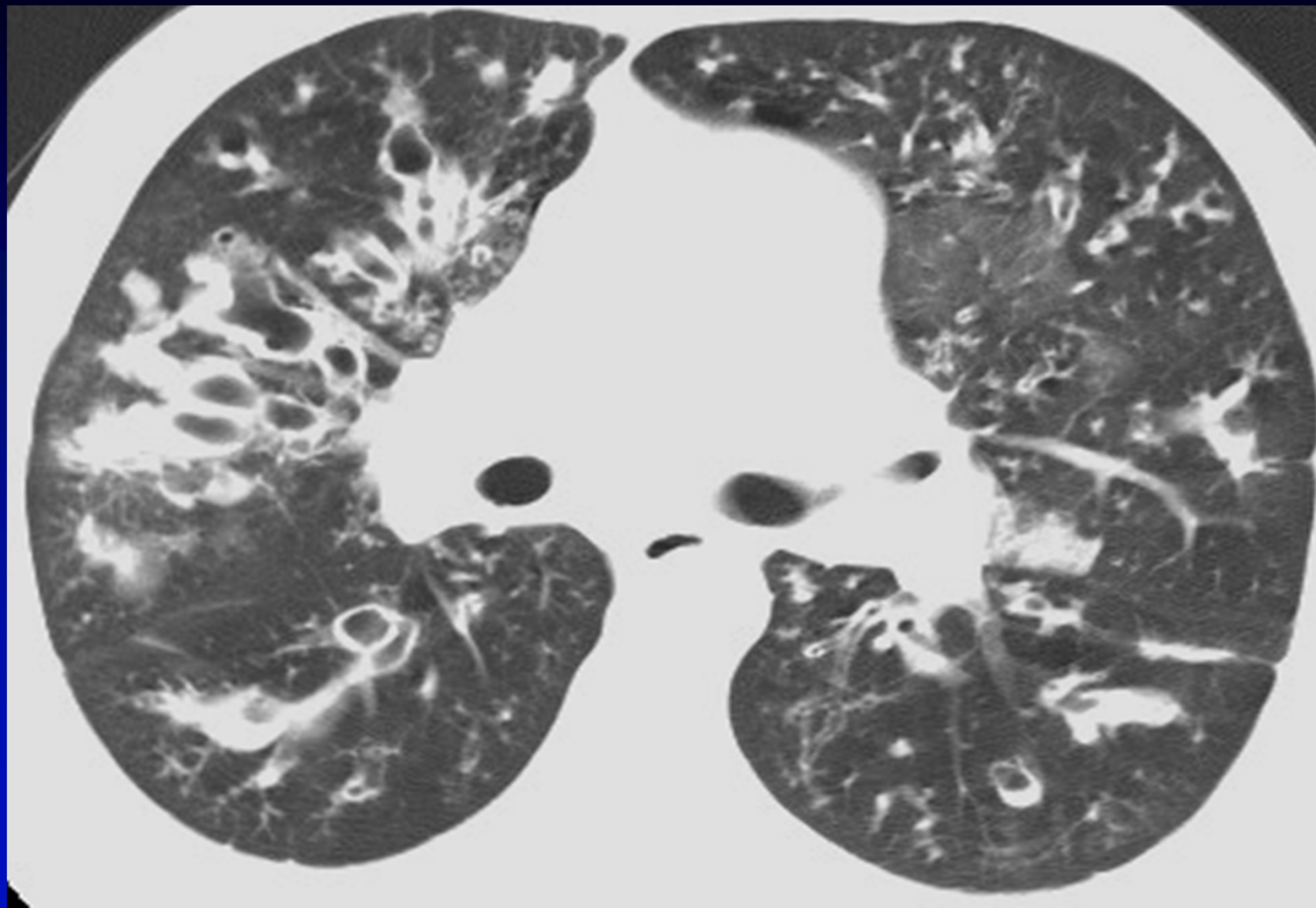


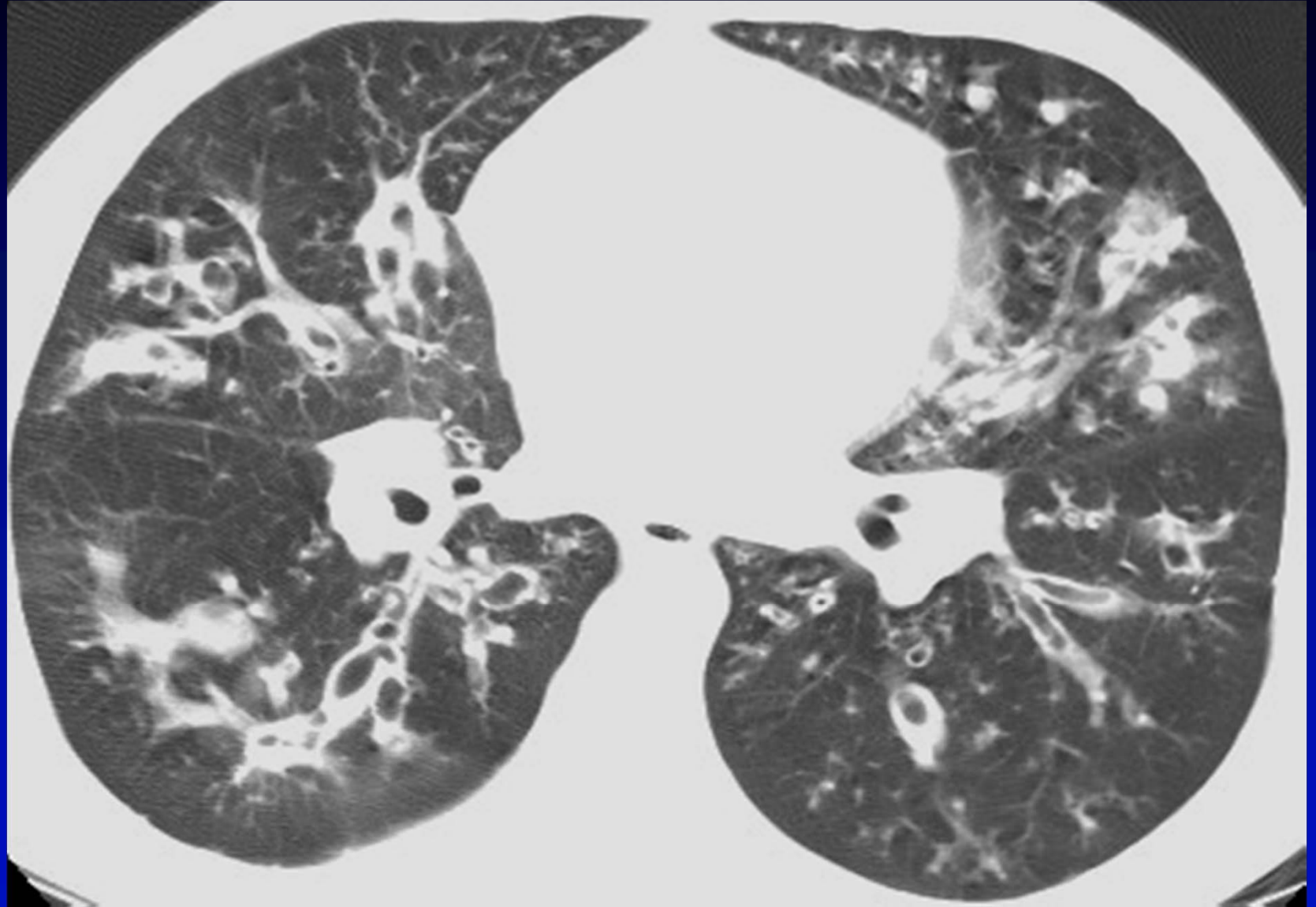


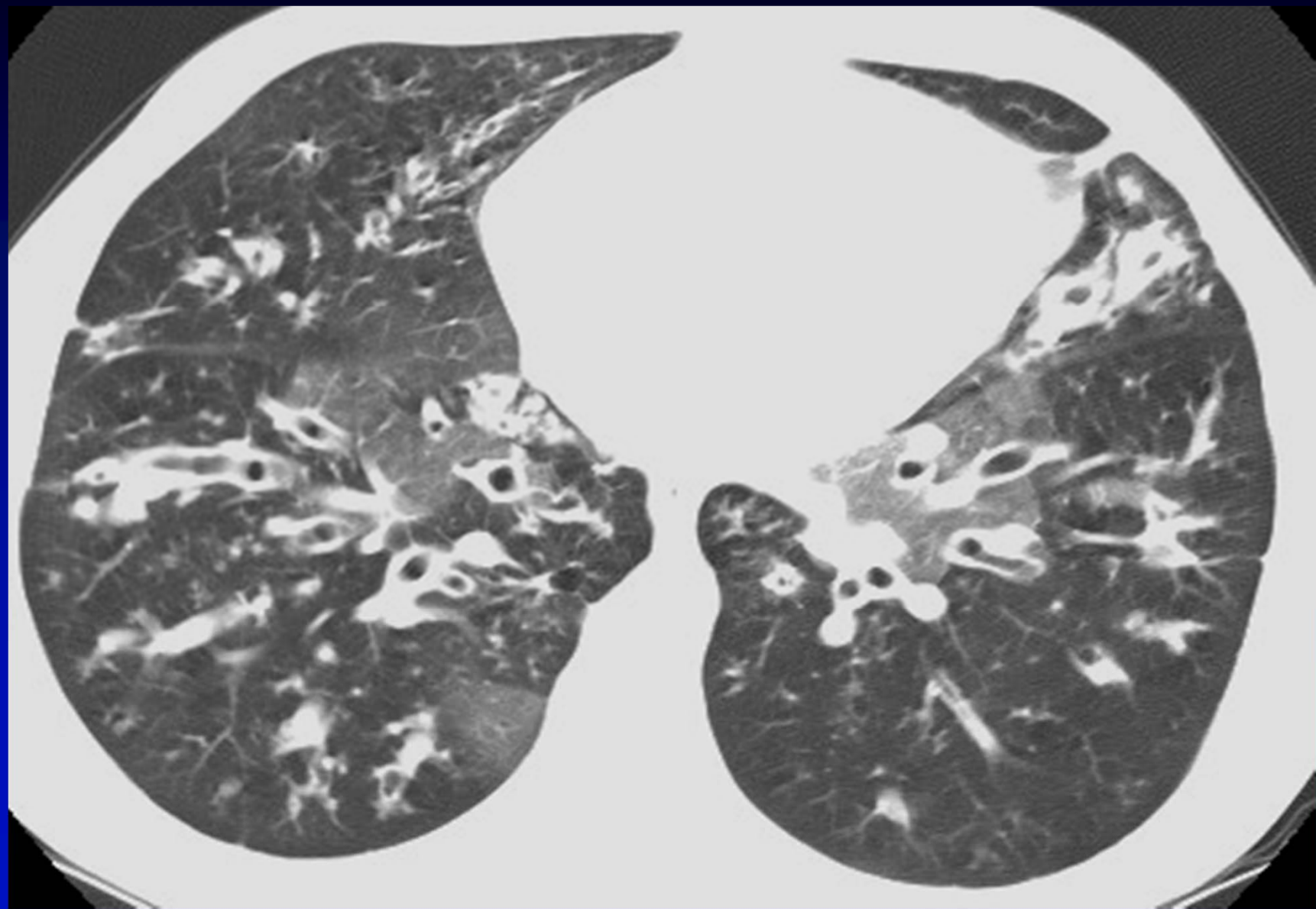


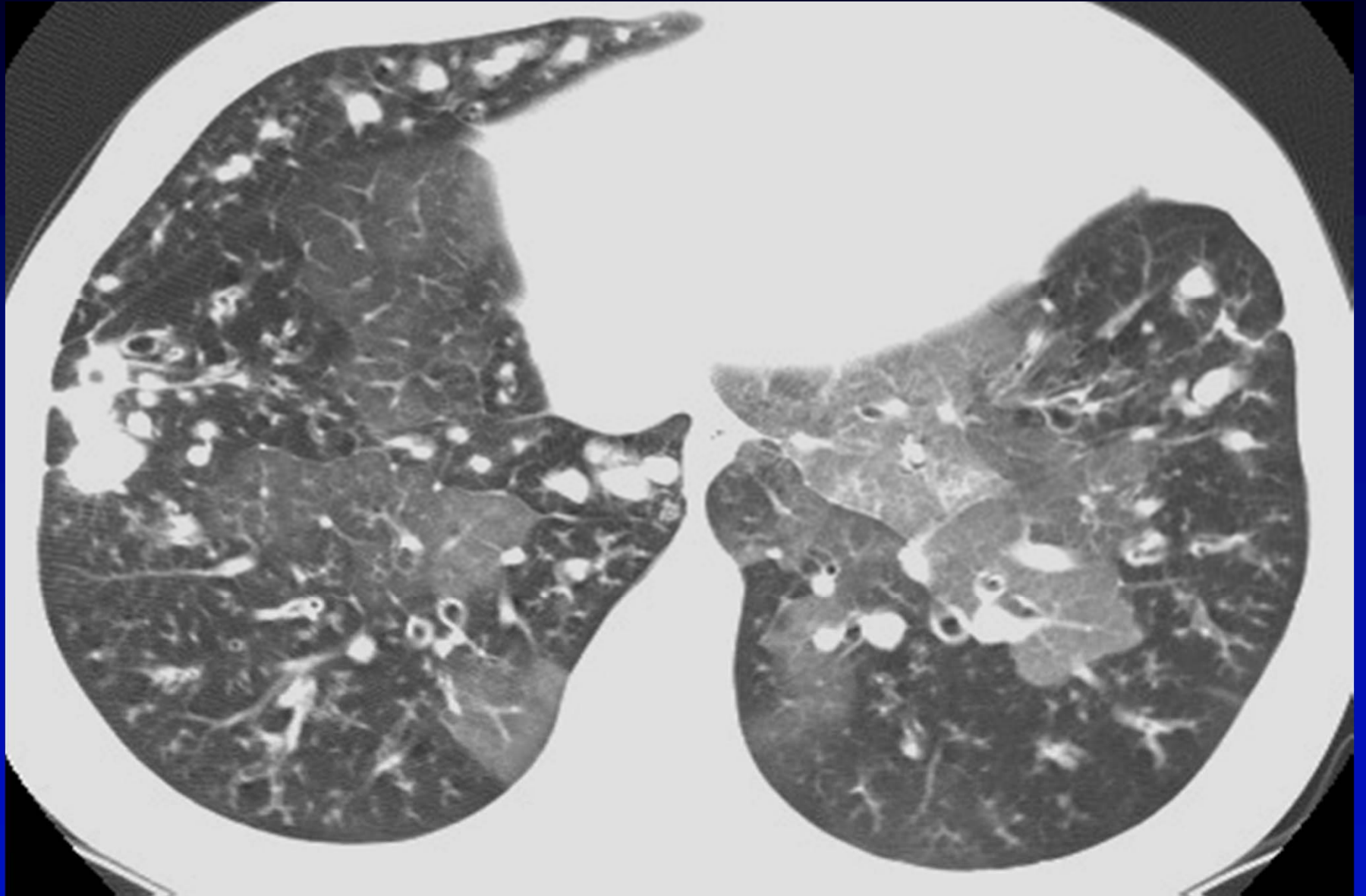
Cystic Fibrosis





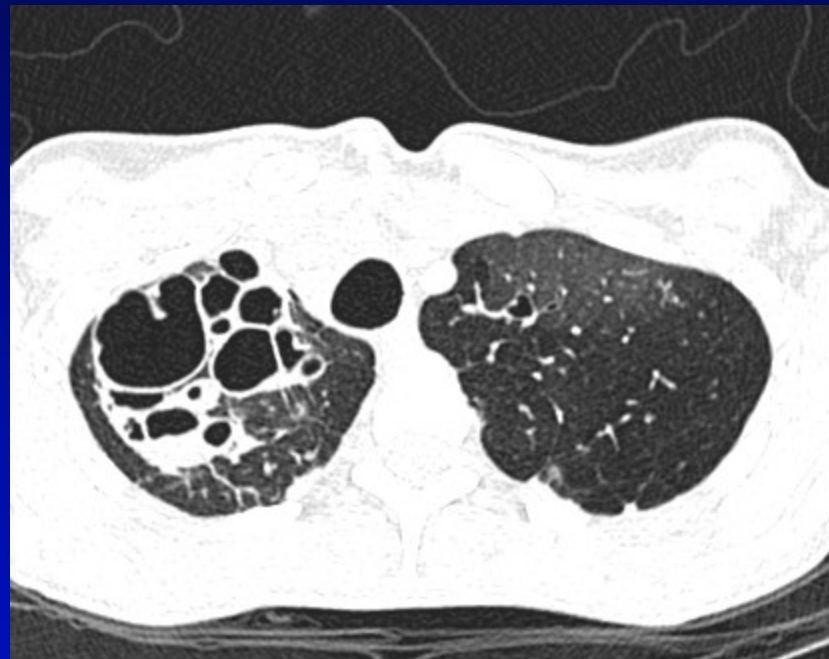


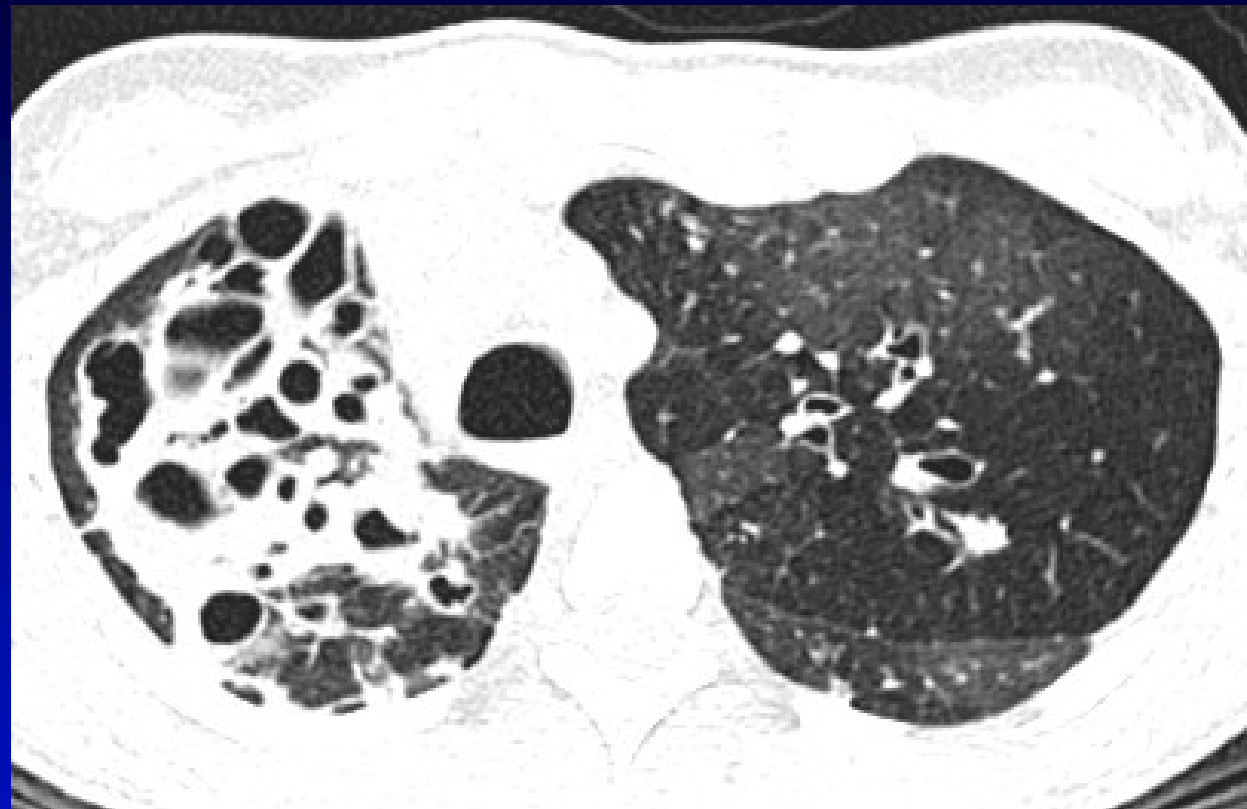


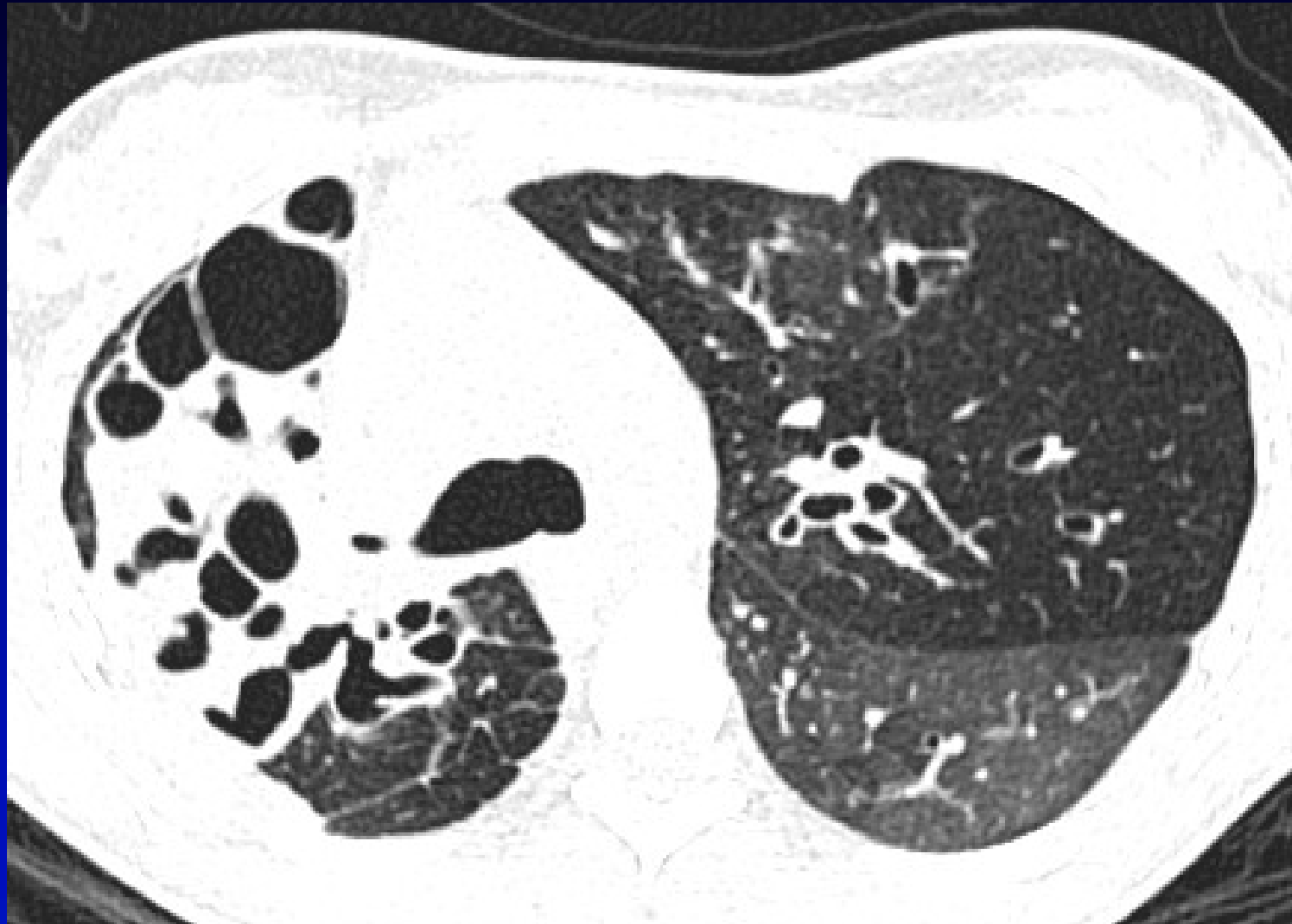


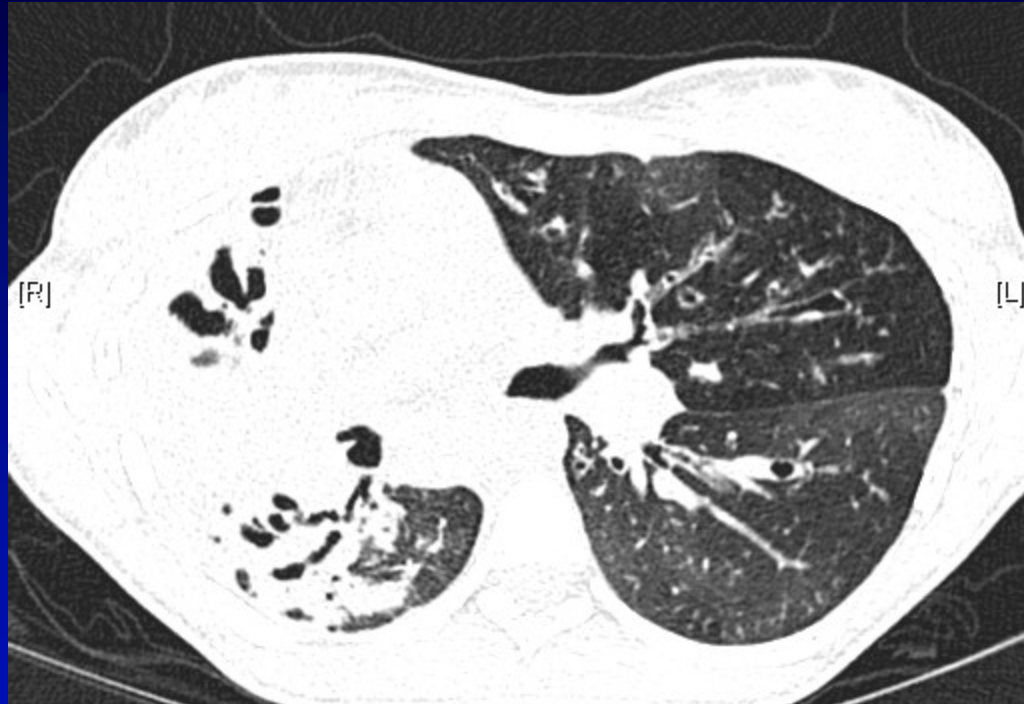
- **Cystic Fibrosis**

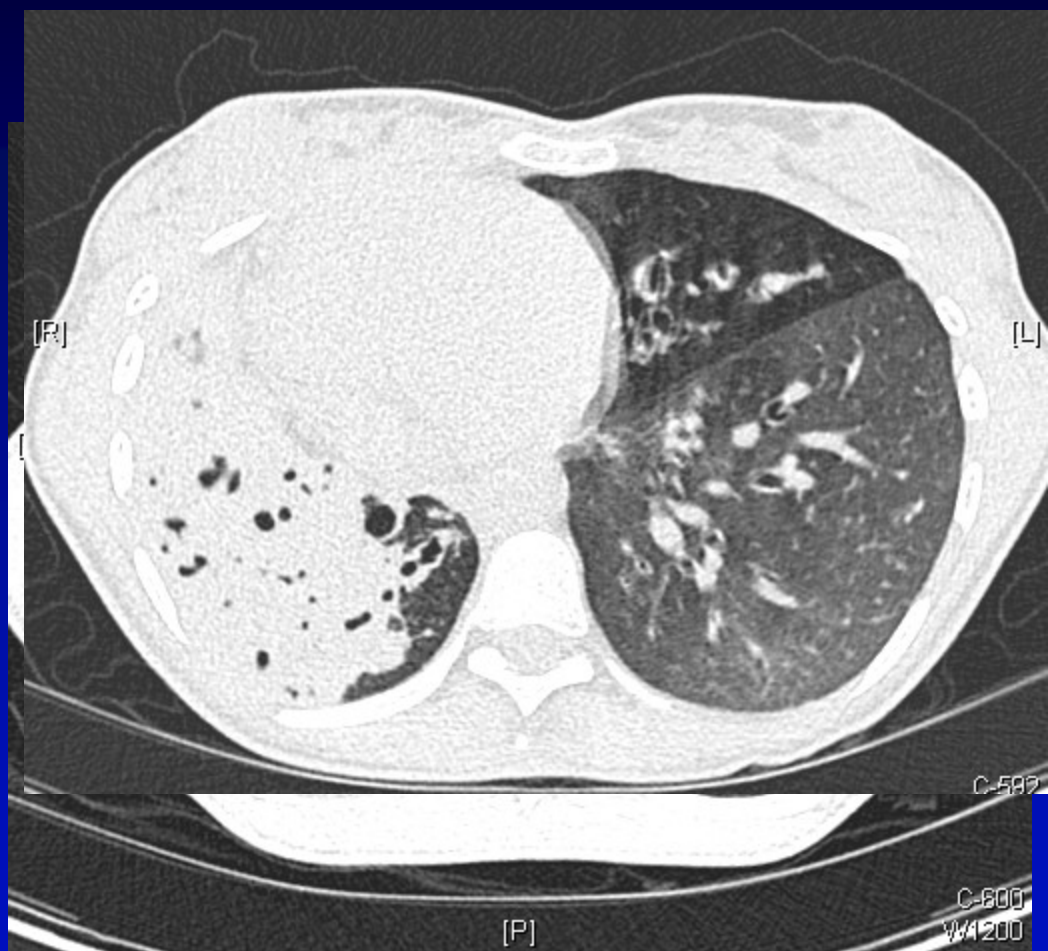
SS 400-95-90



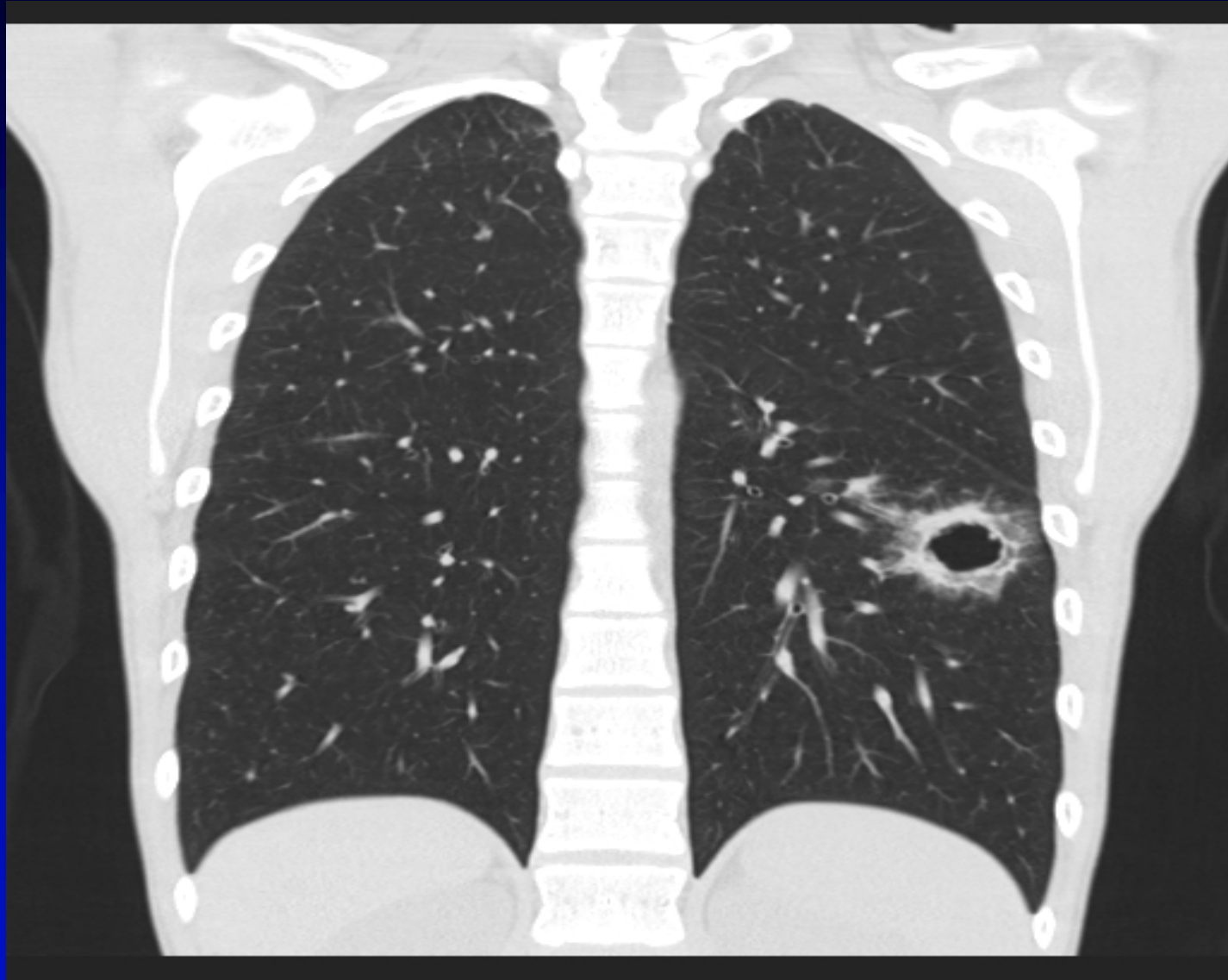






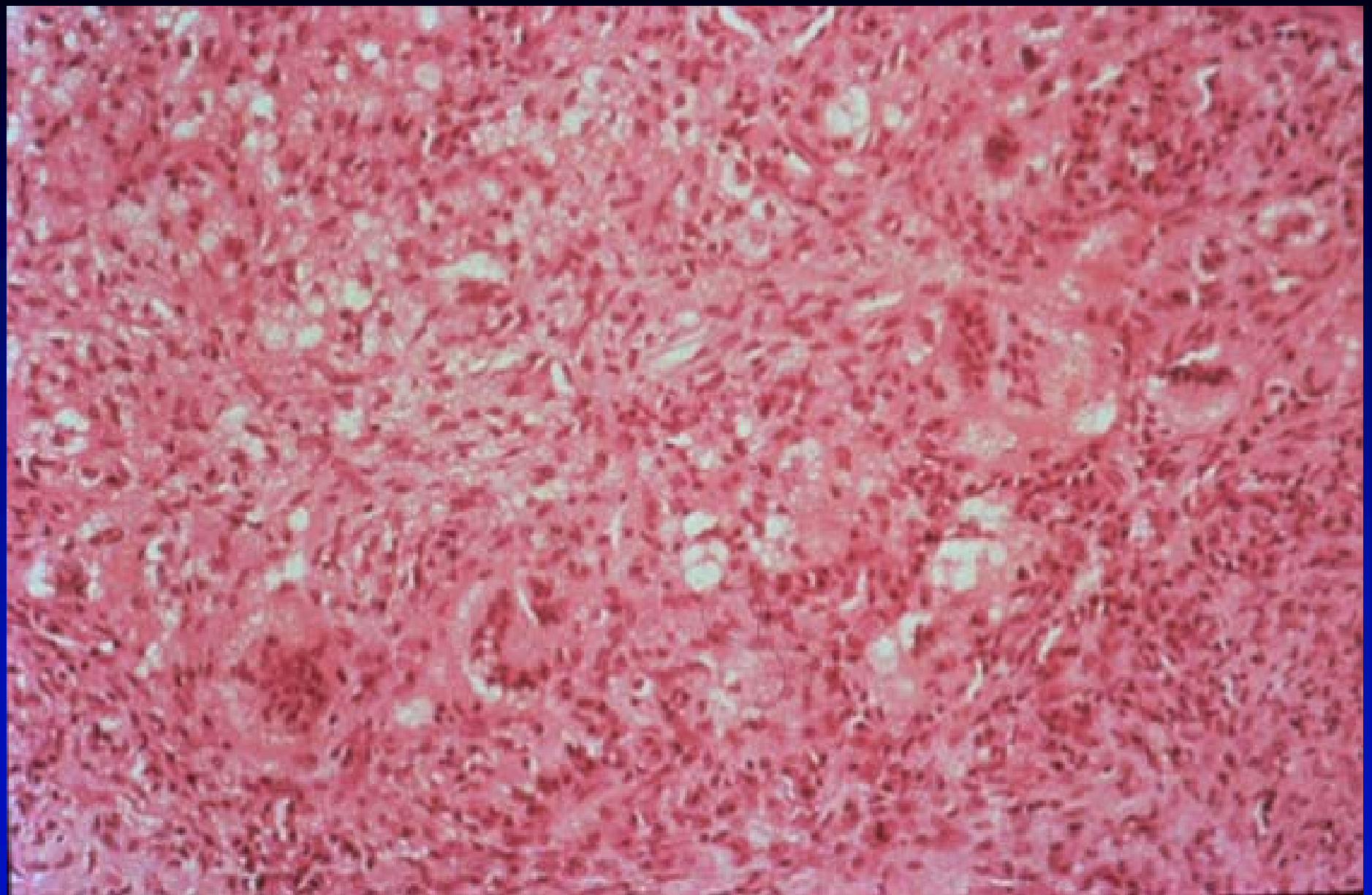


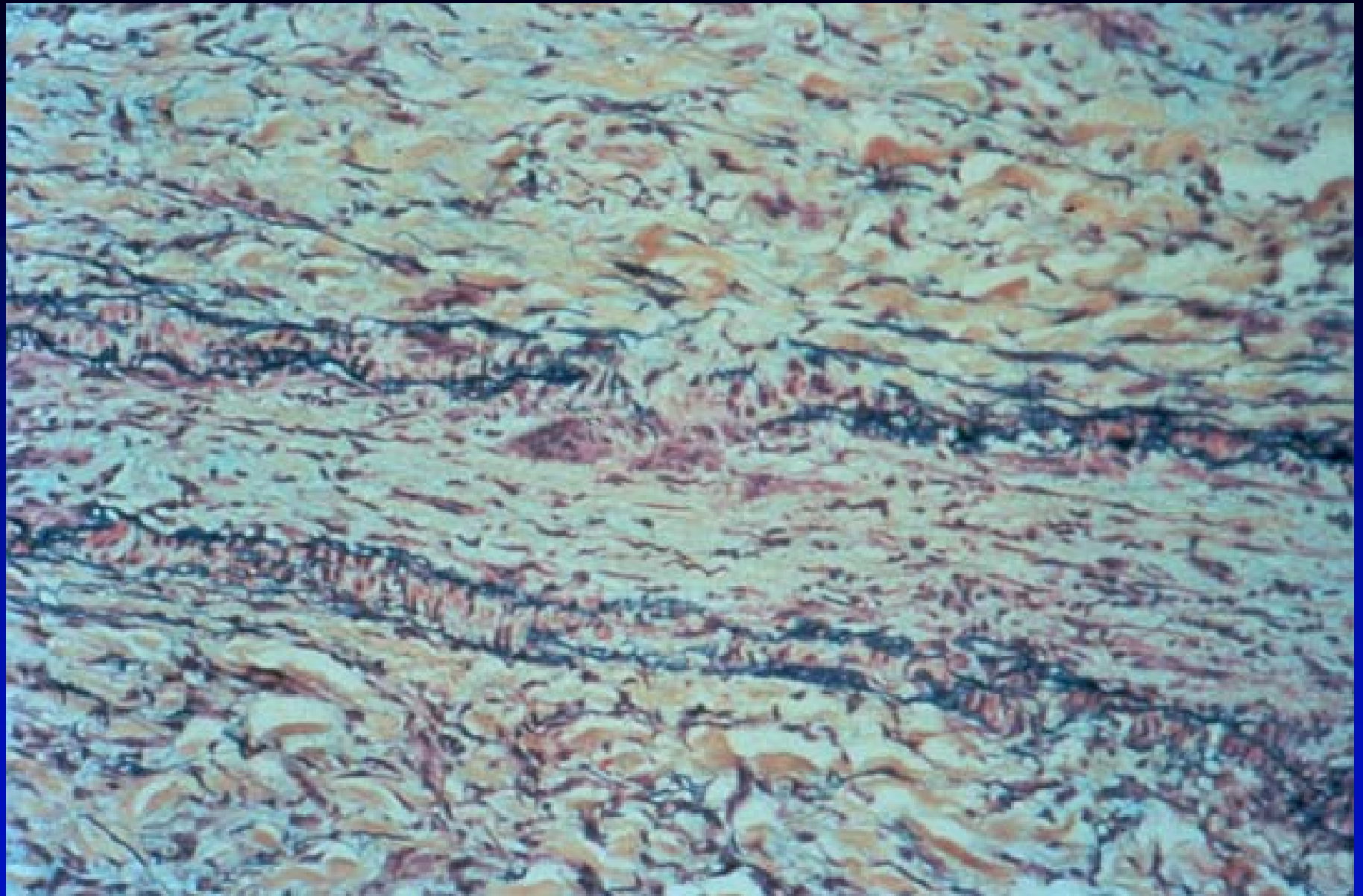
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CC

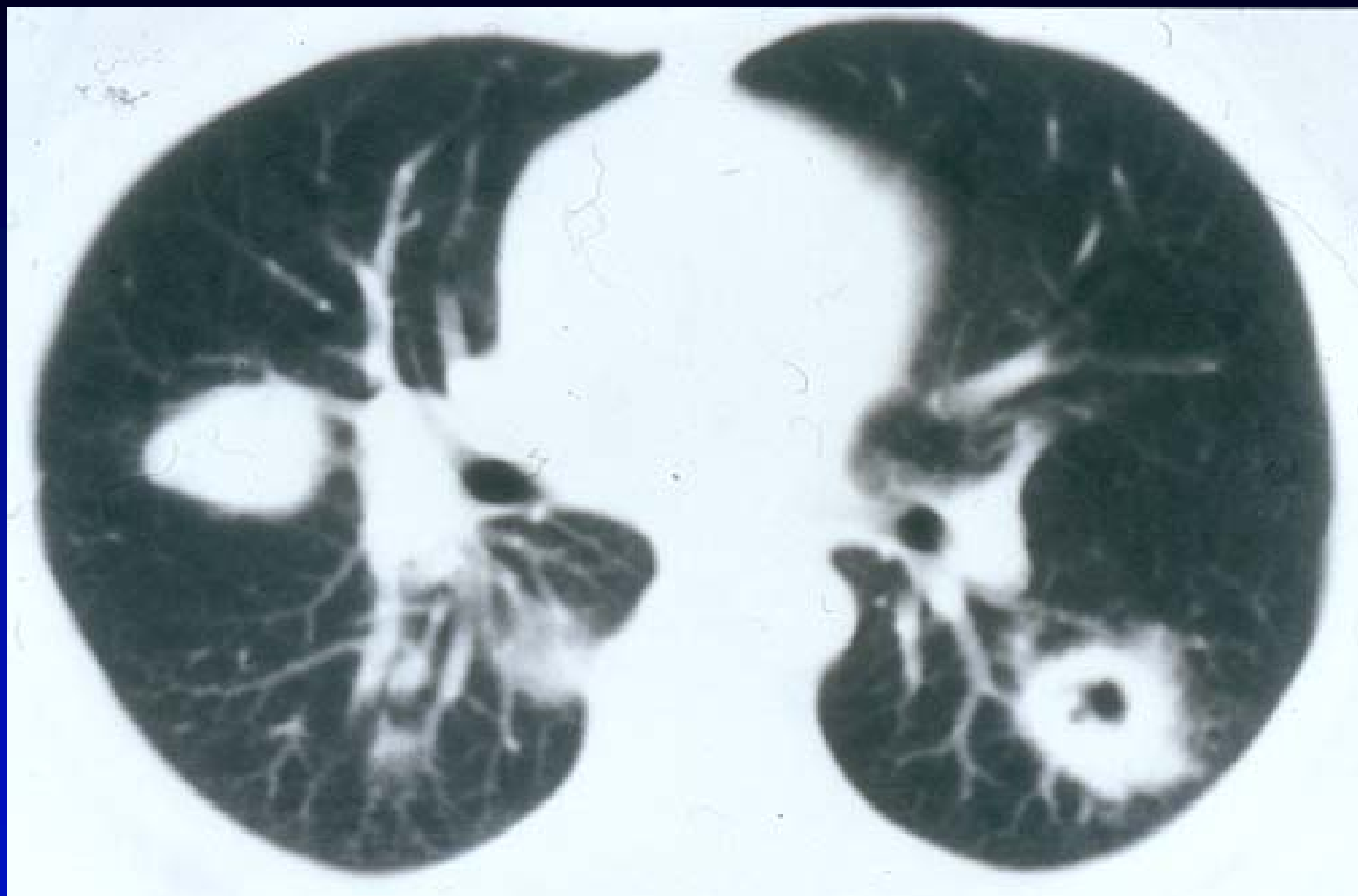




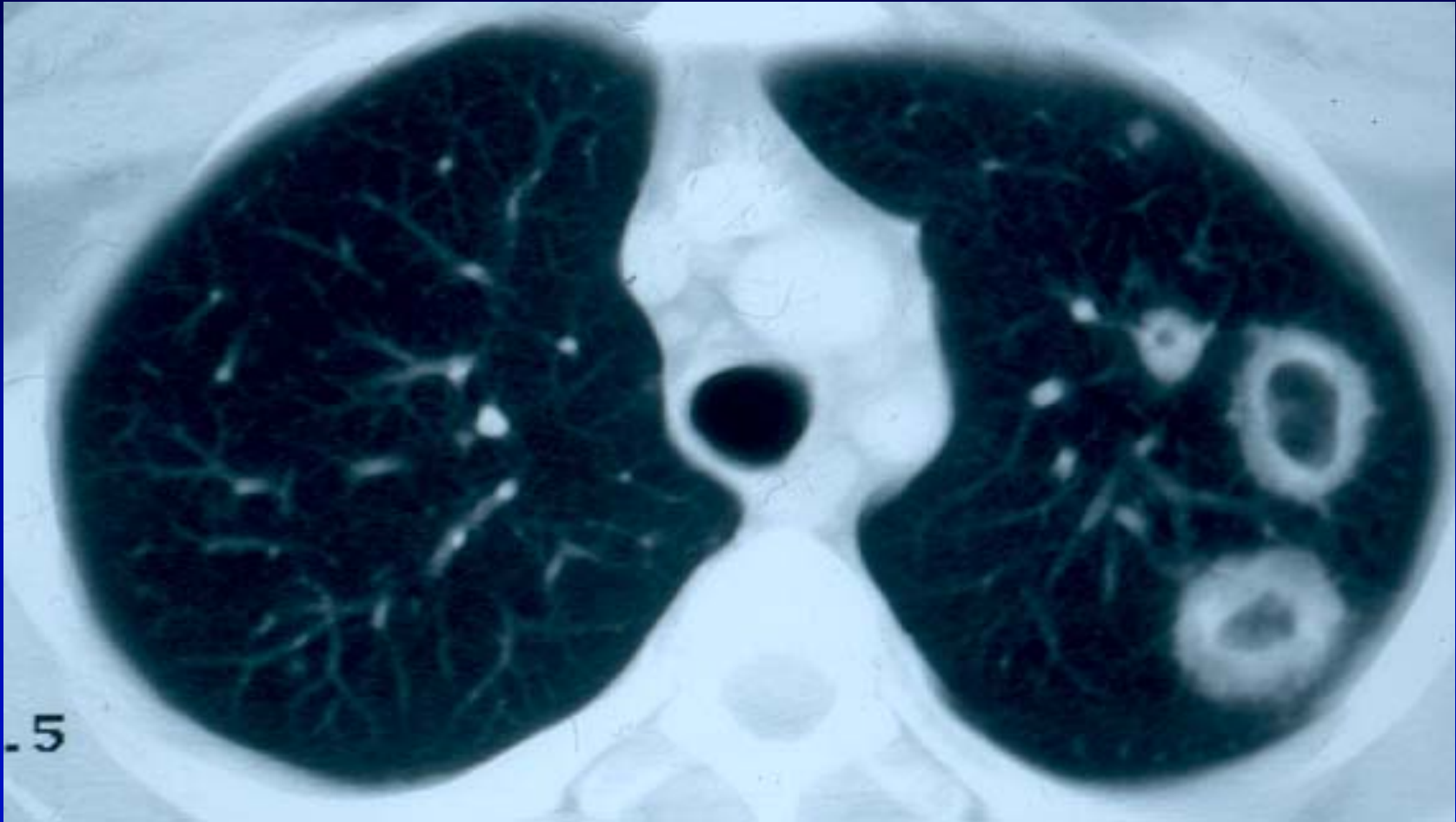


- **Wegener's Granulomatosis**





Cavitary nodules (WG)



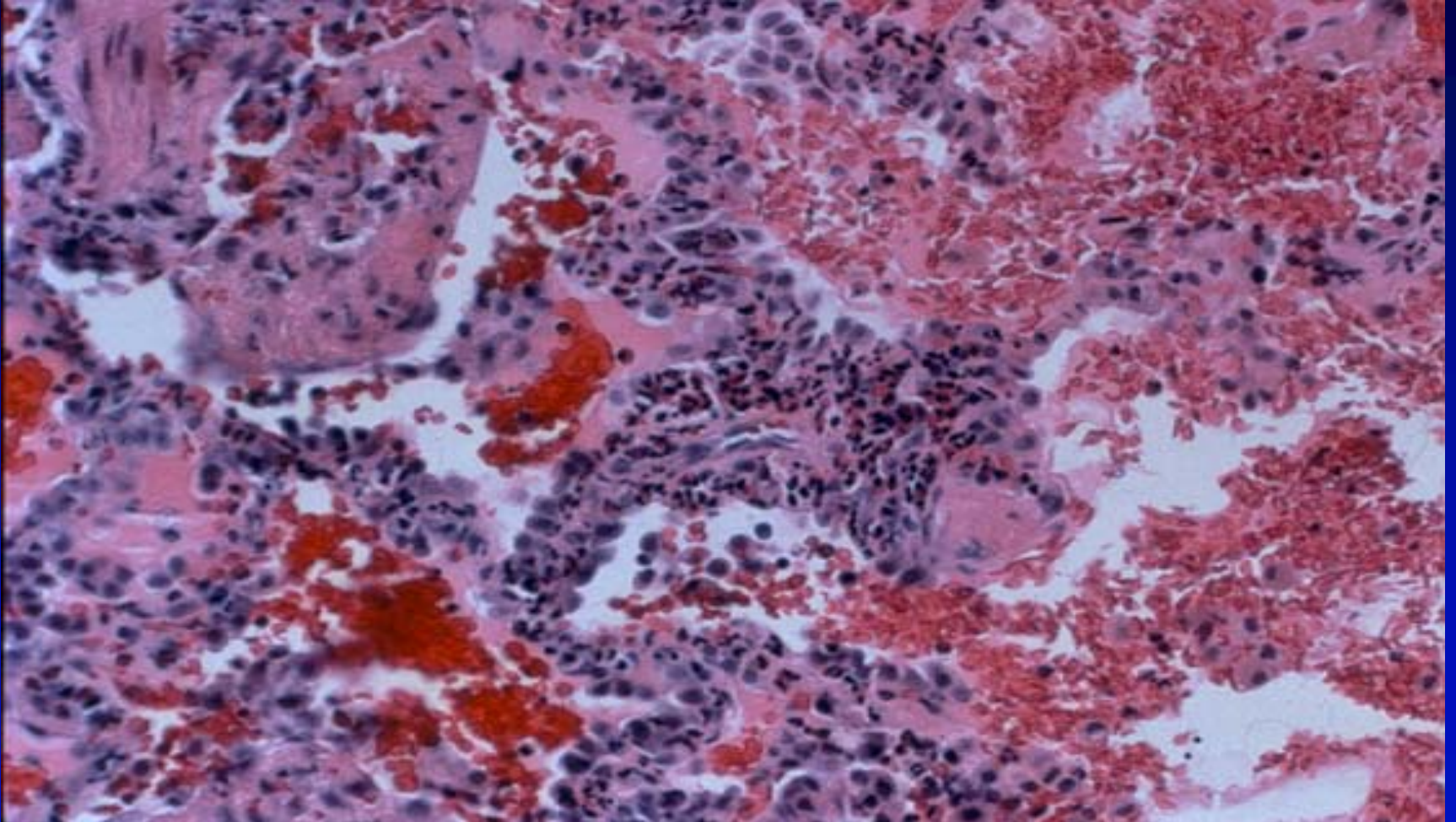


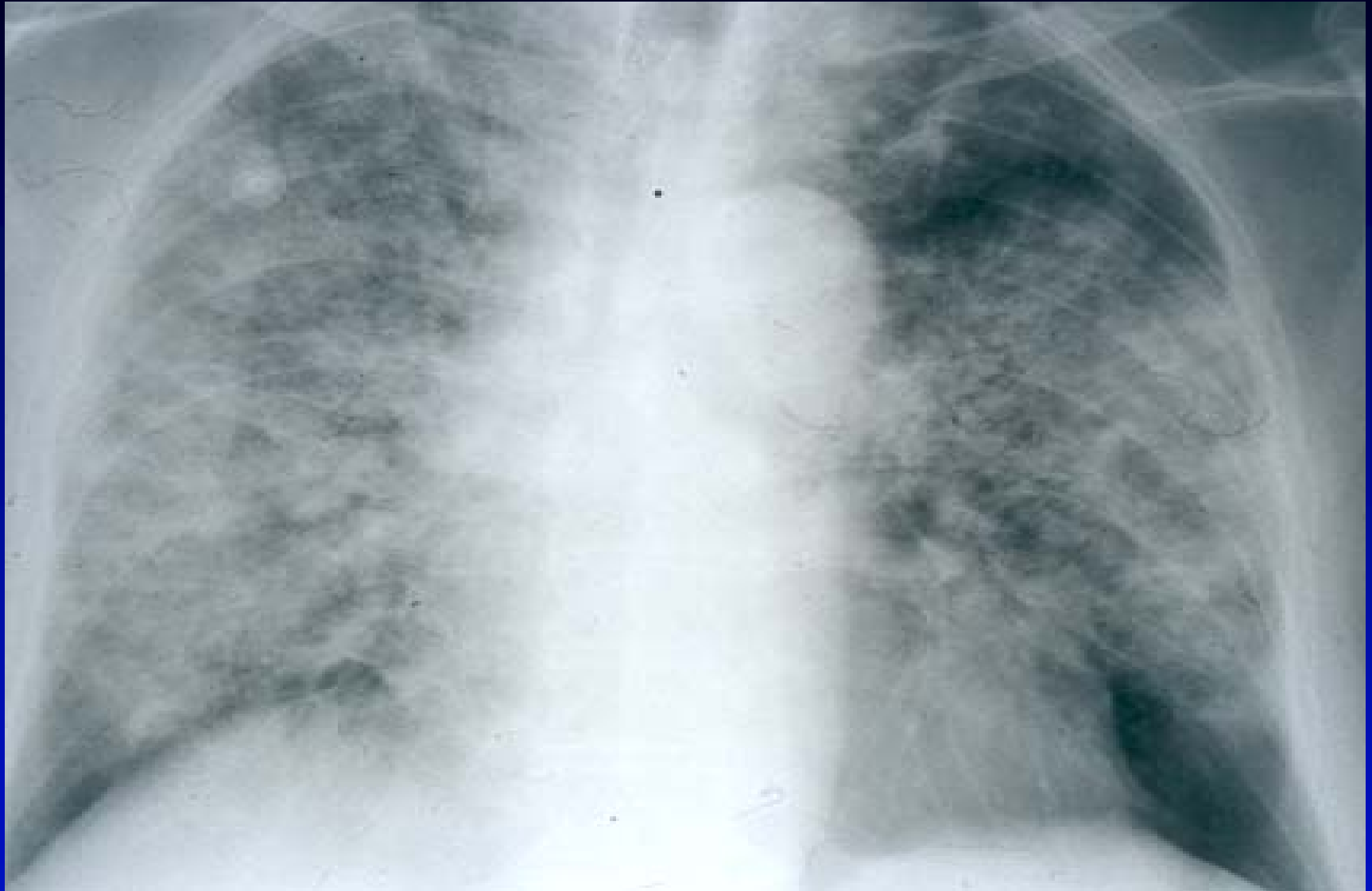


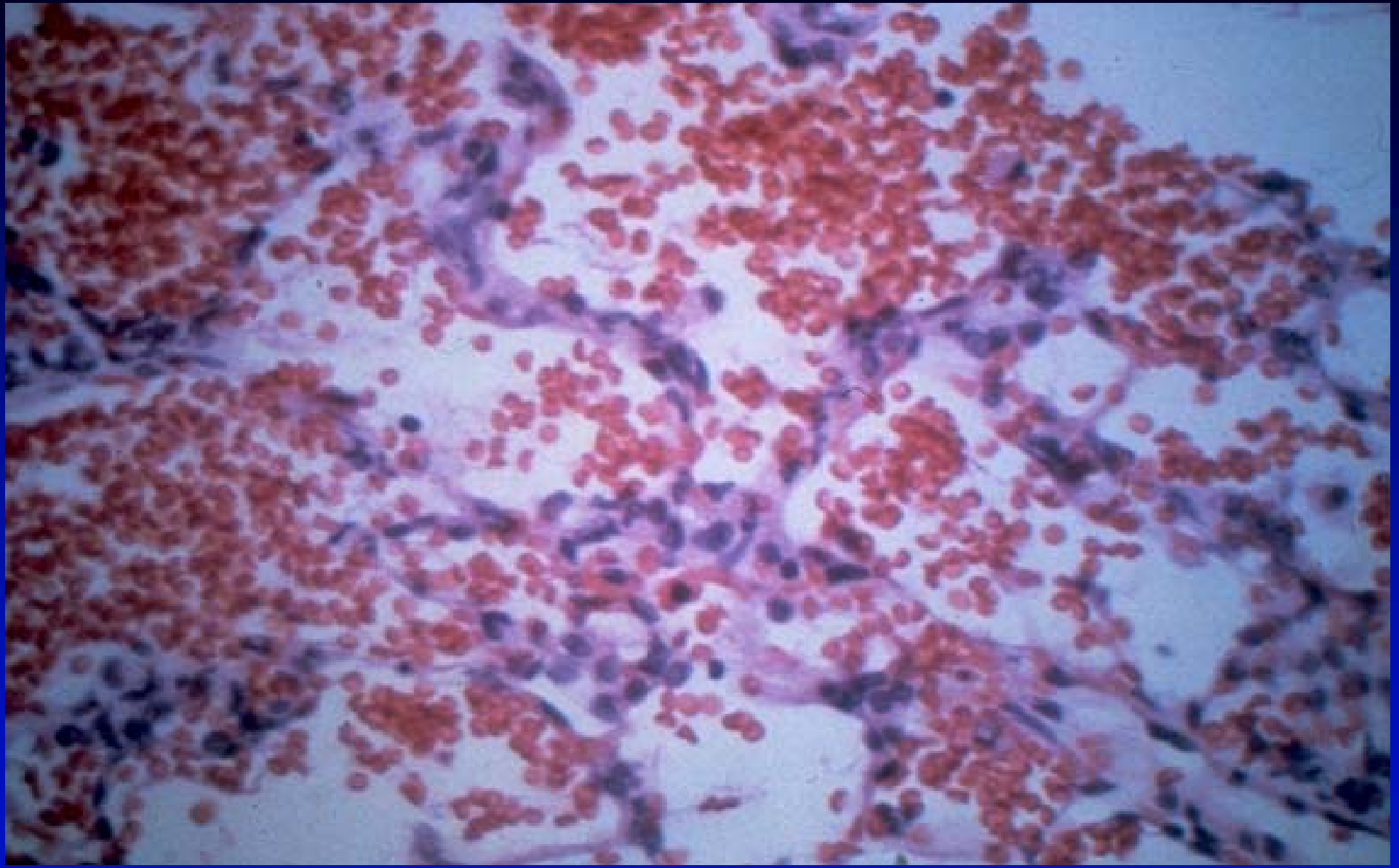


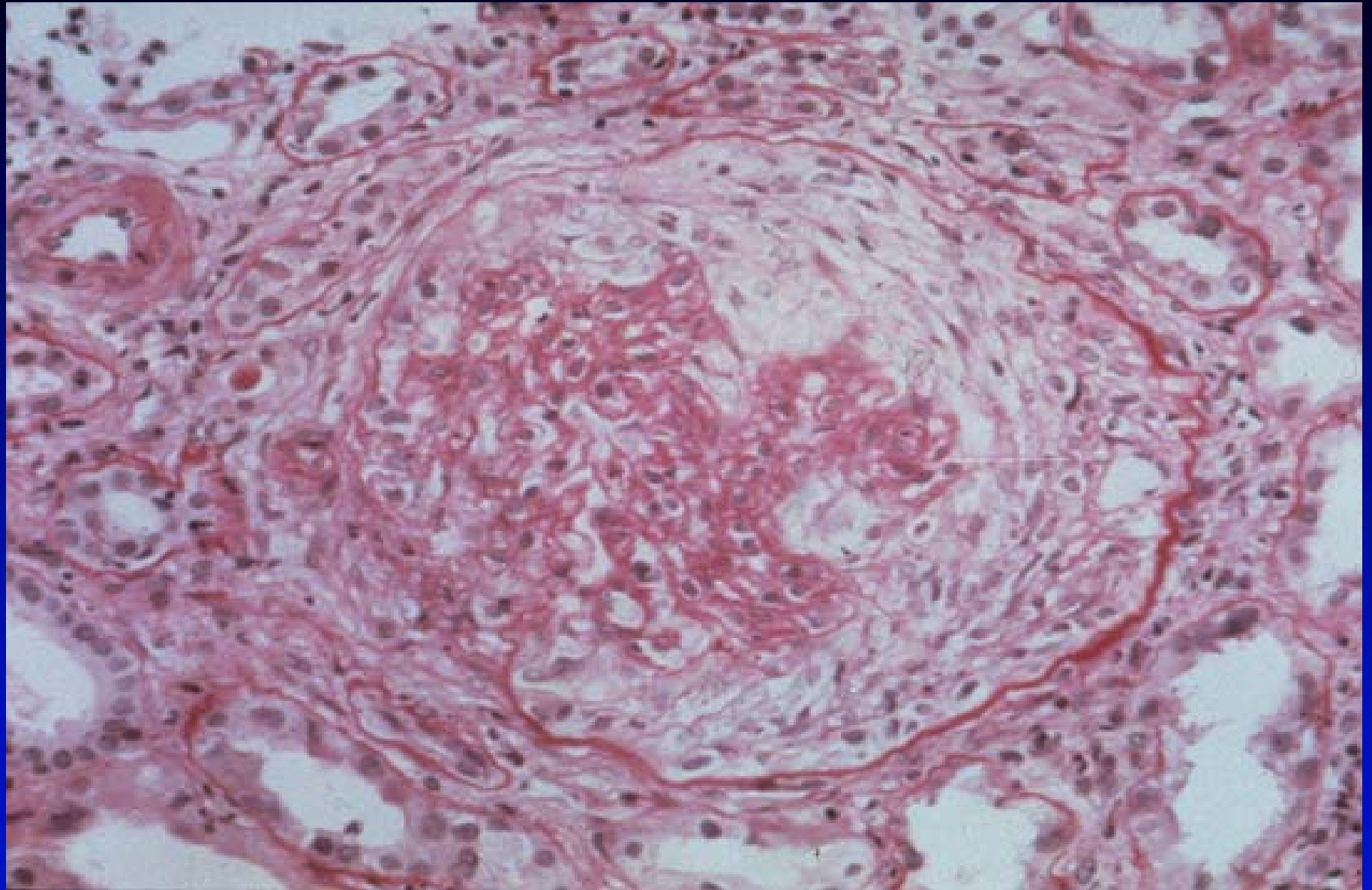


Capillaritis (neutrophilic)

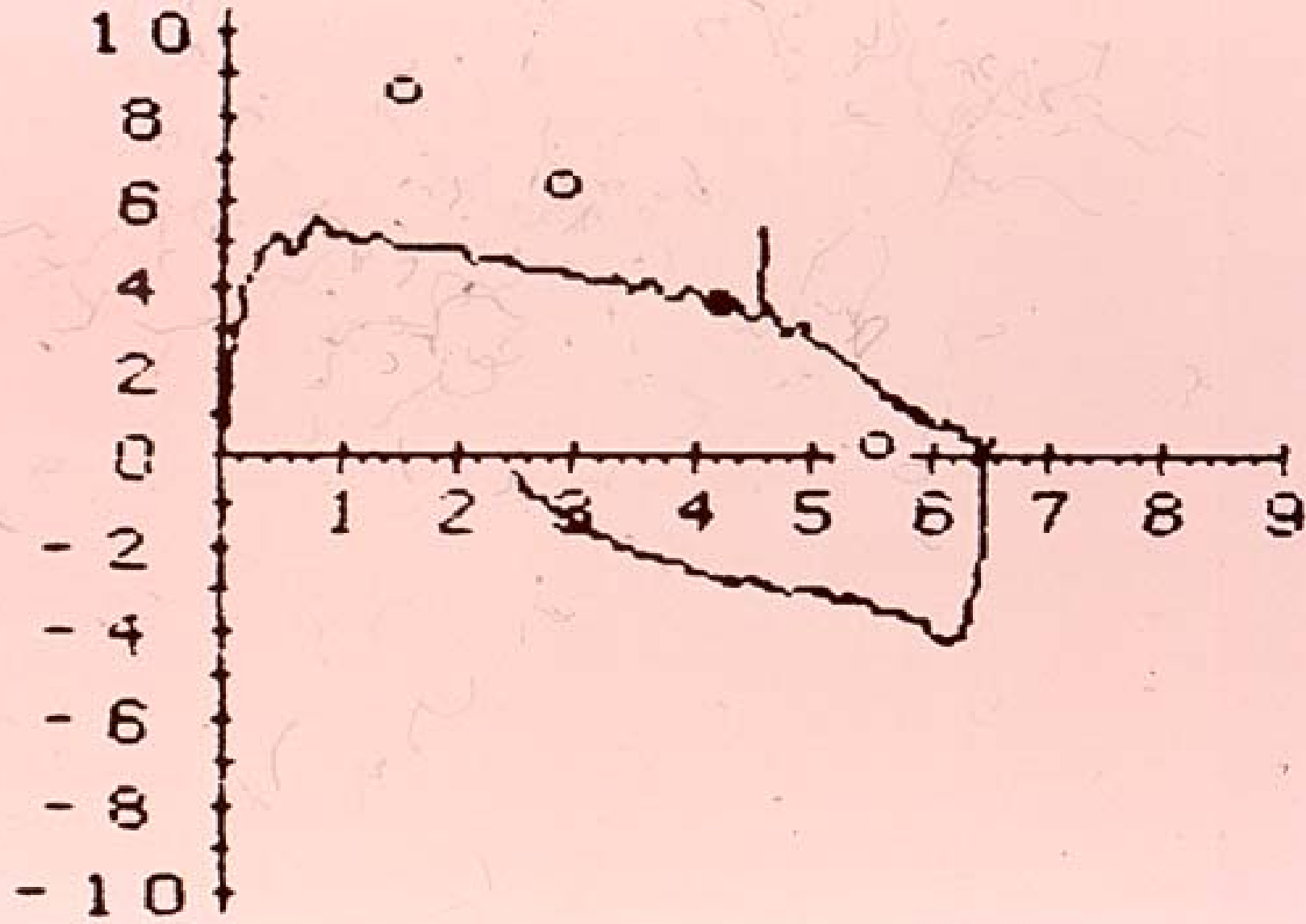




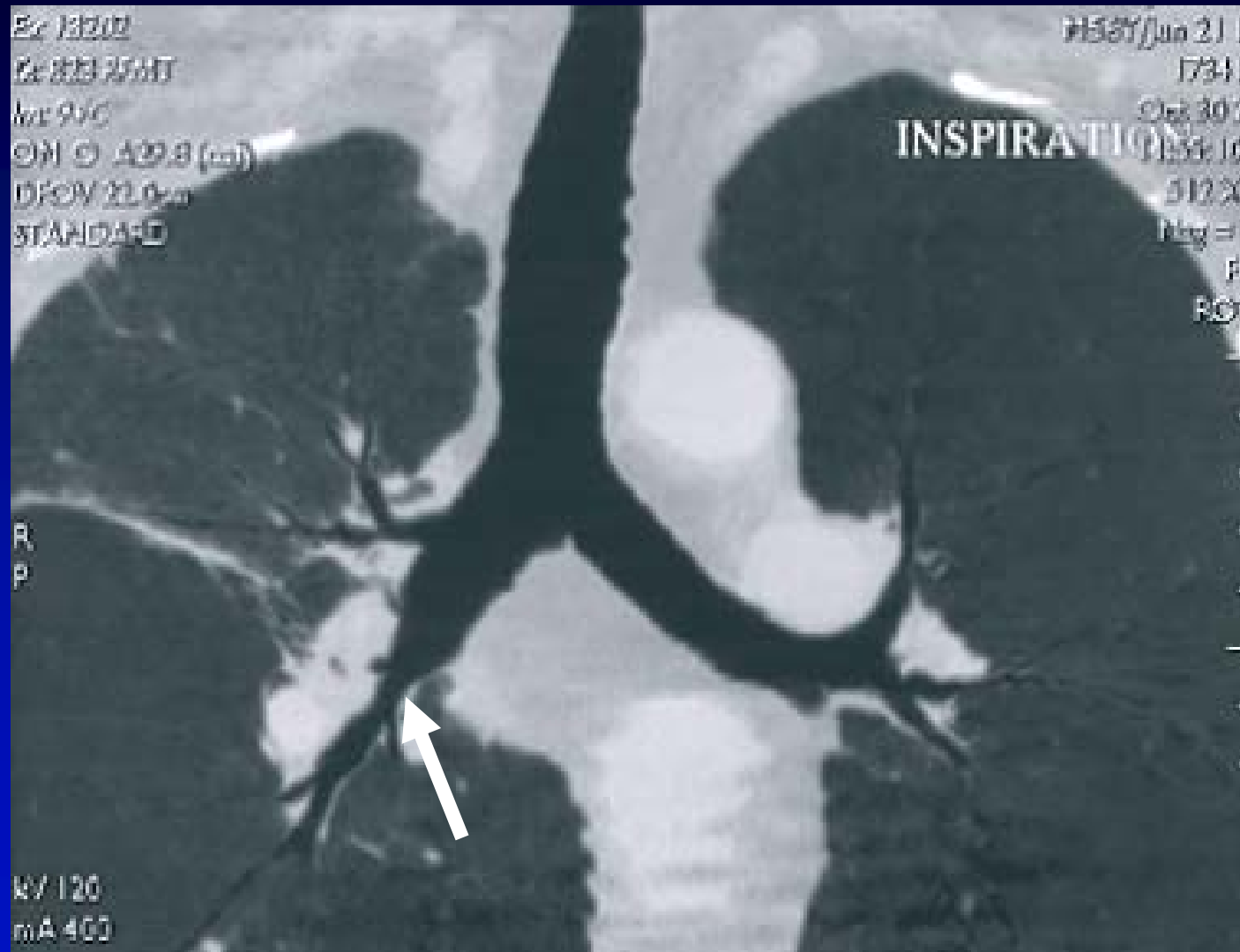




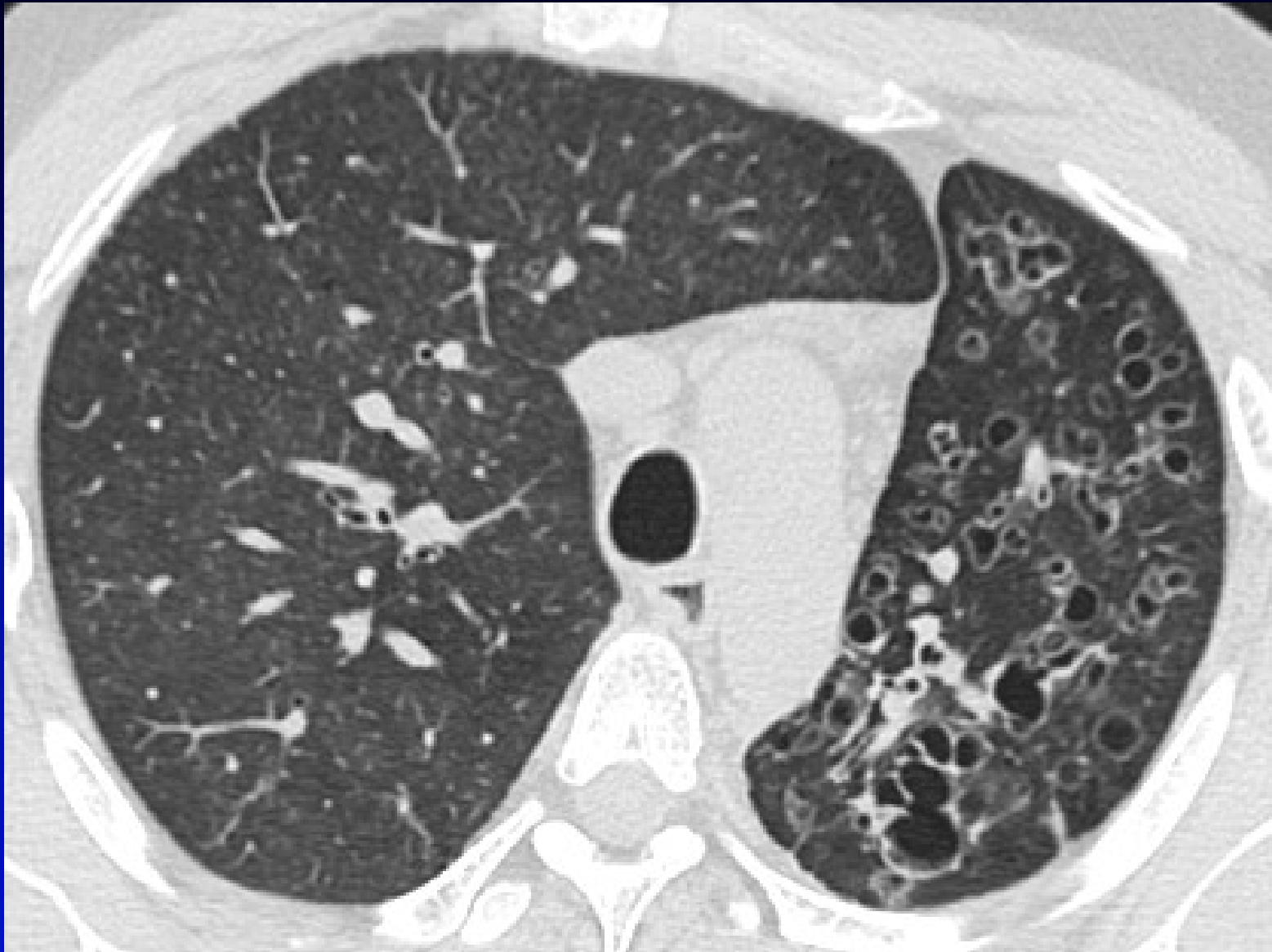
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Stenosis bronchus intermedius



SG



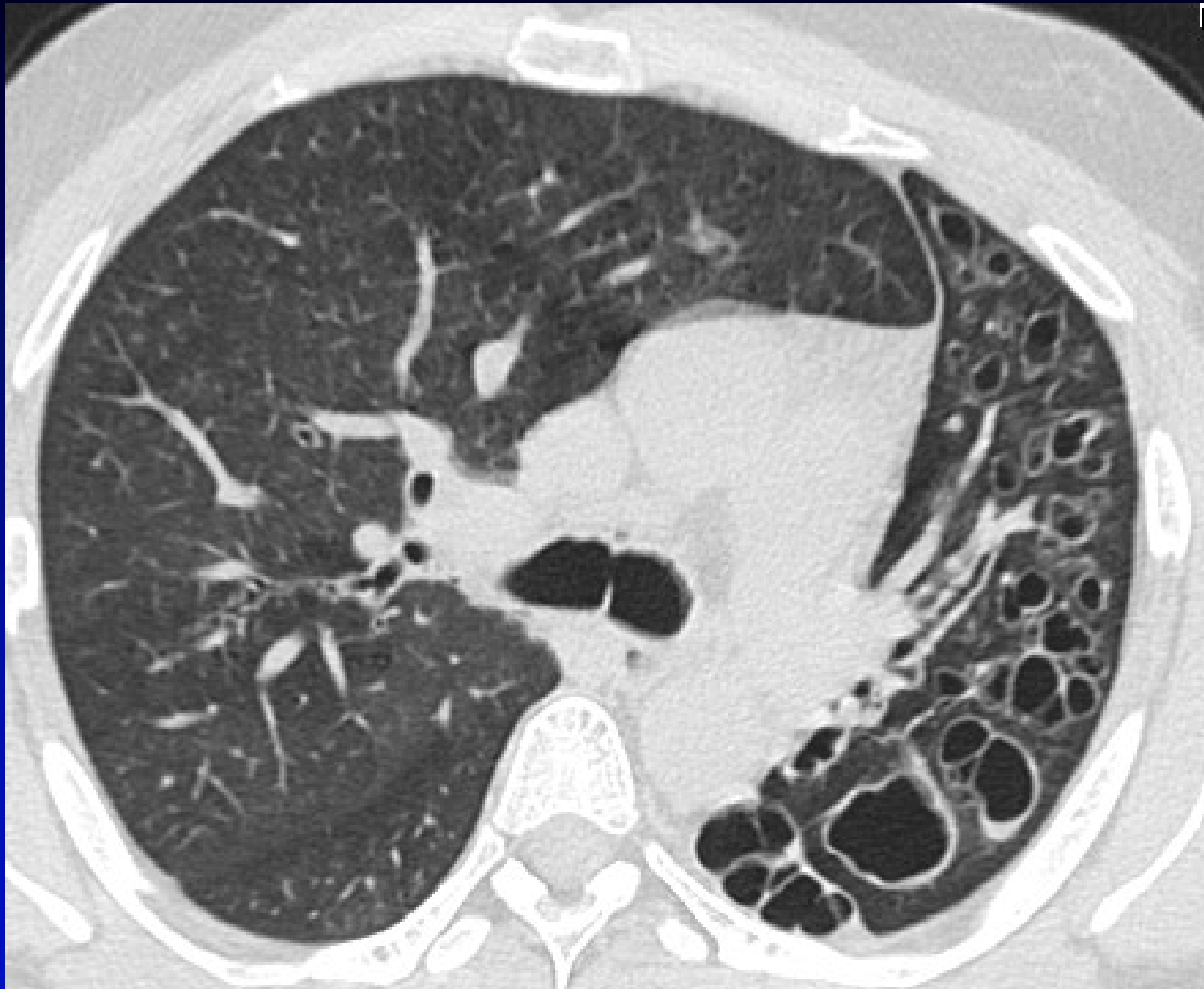
SG



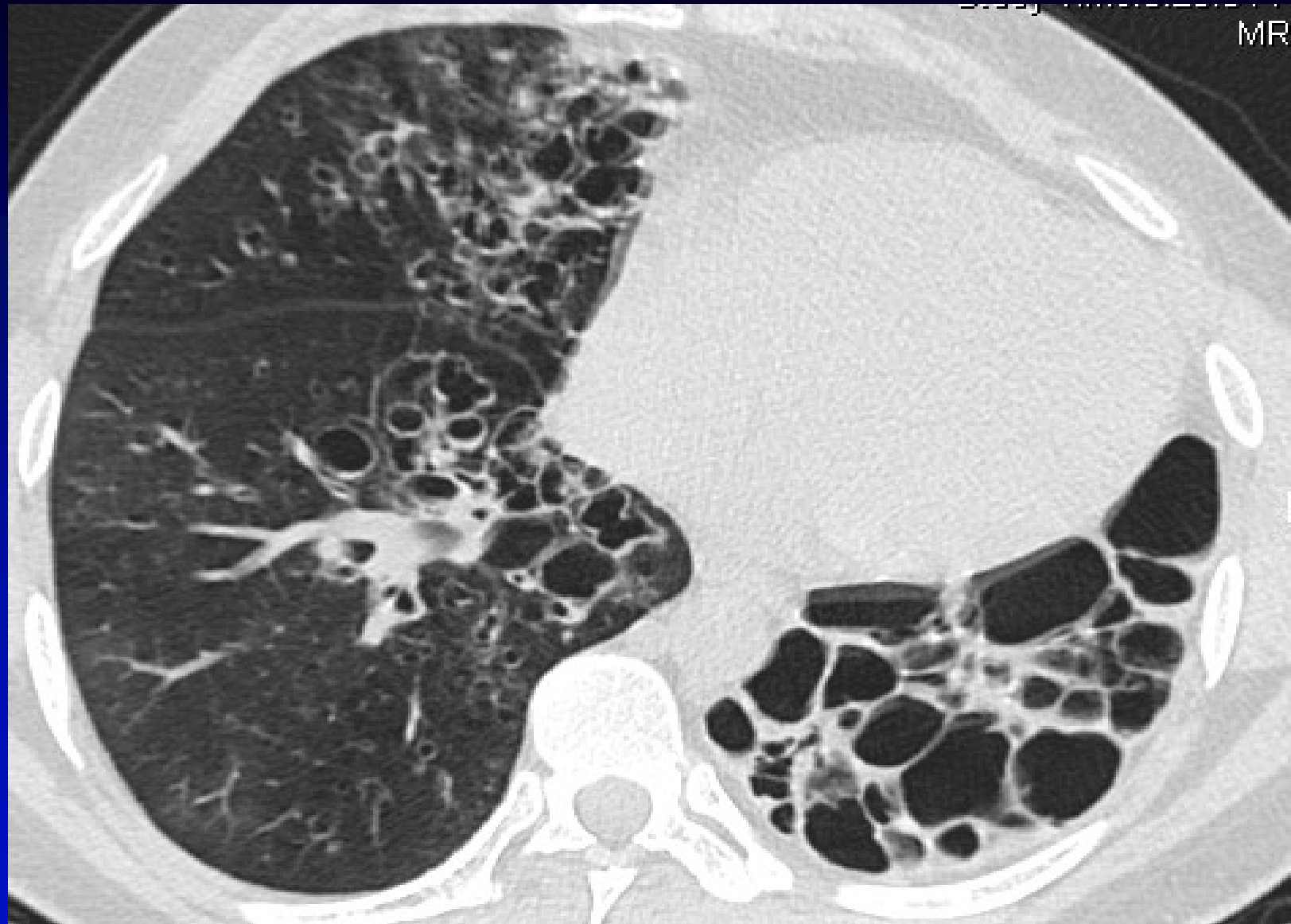
SG



SG



SG

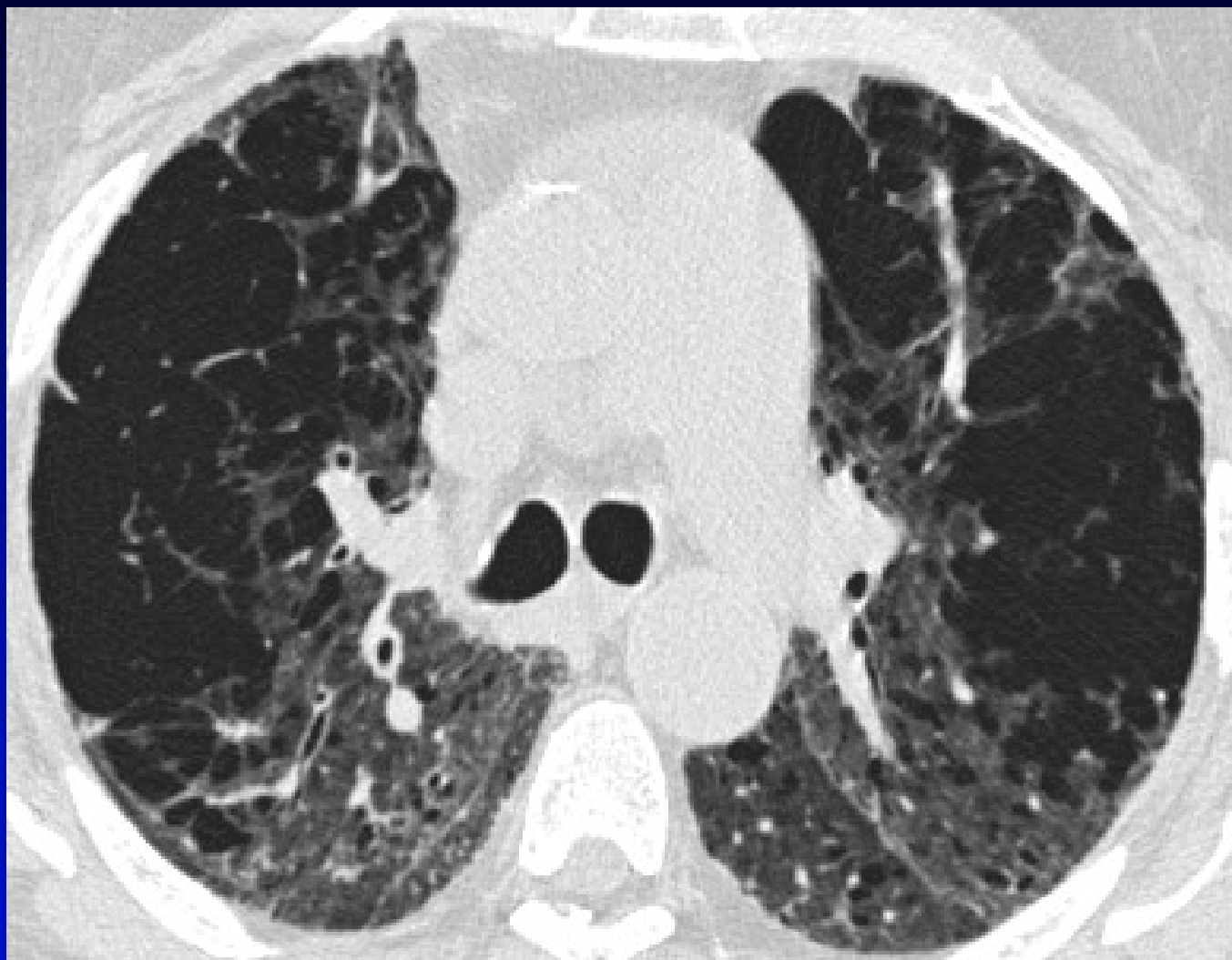


- **Centrilobular Emphysema
(upper lobe predominant)**

GW 063-71-58

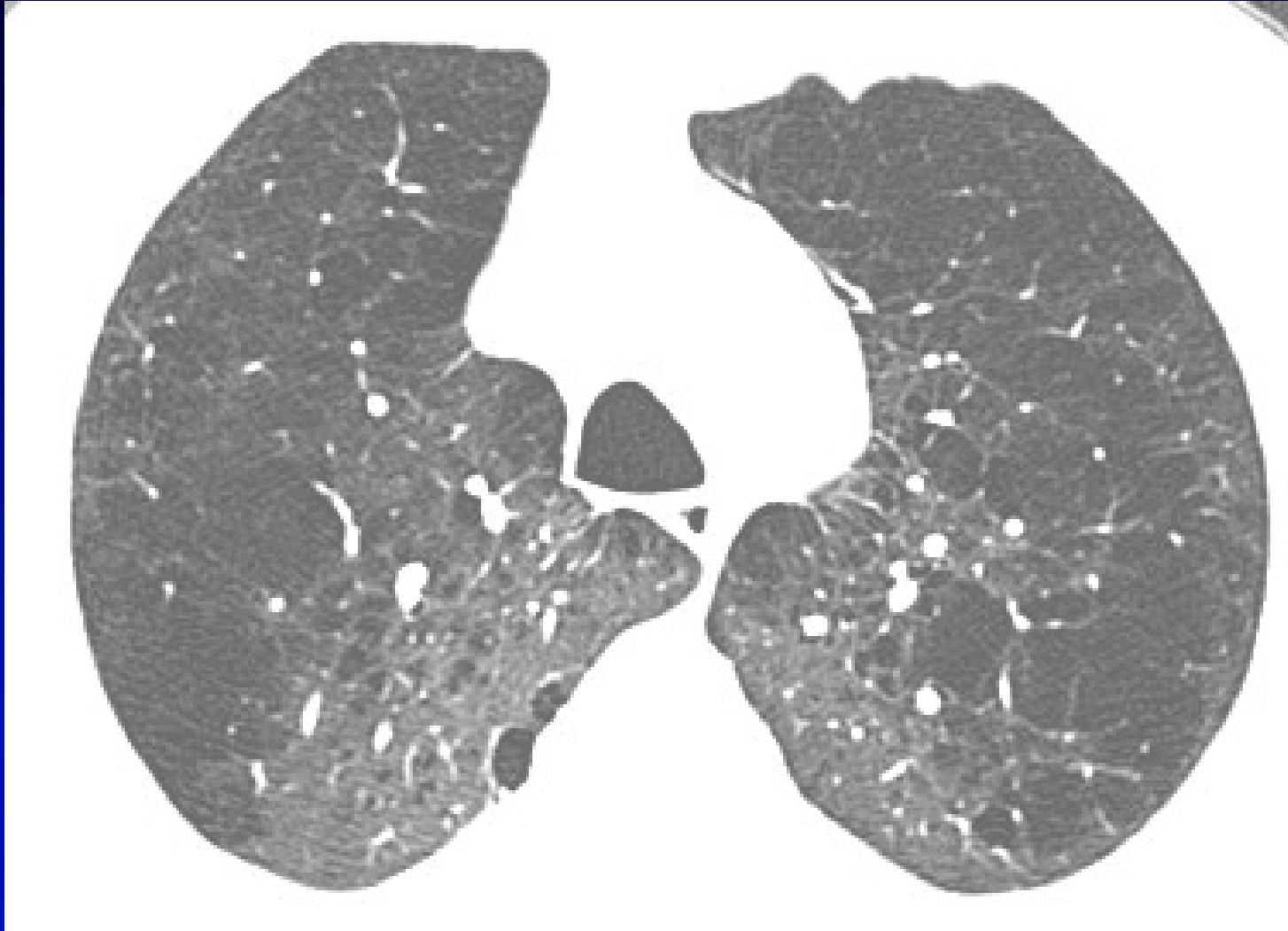


GW

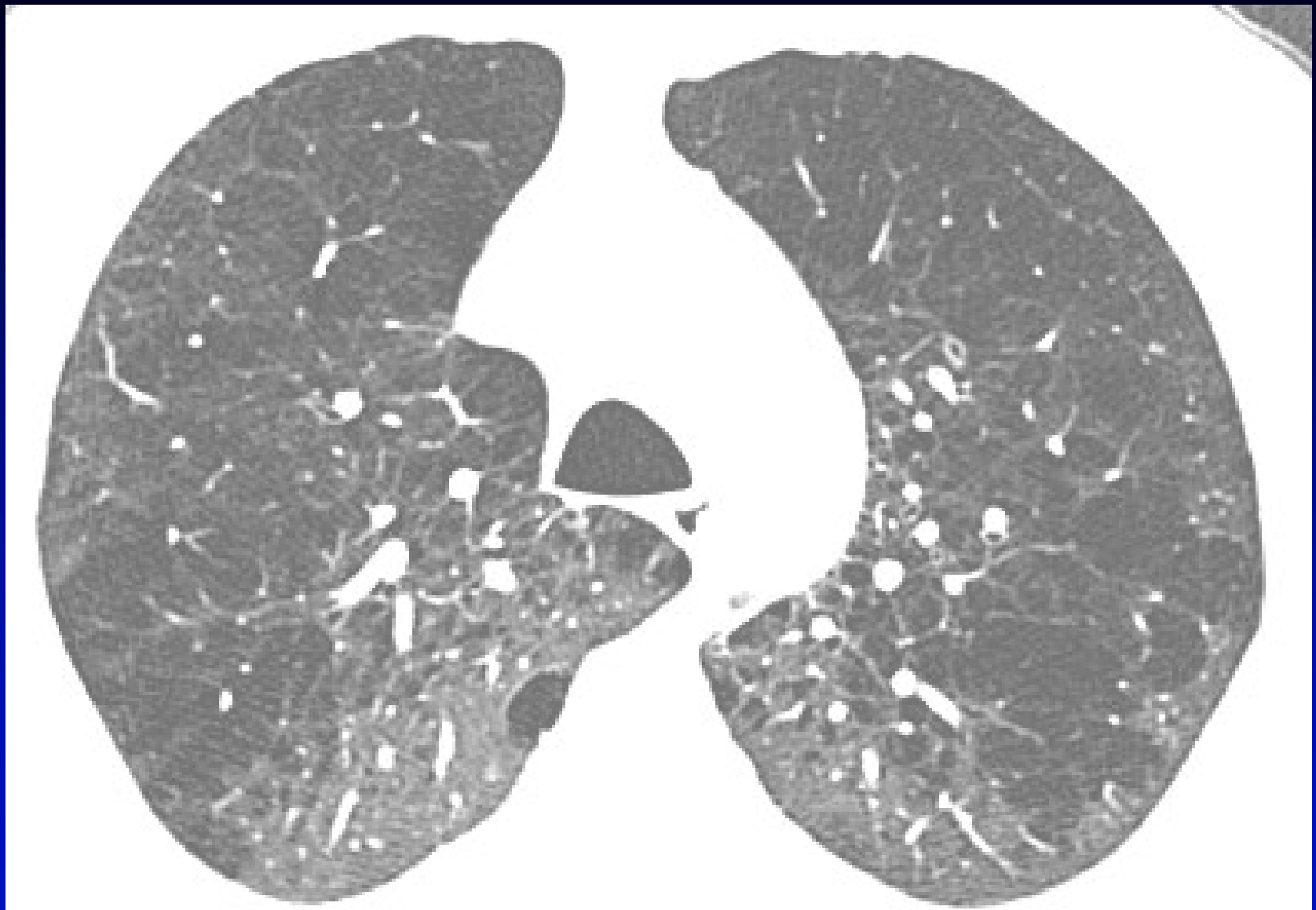


- **Panlobular Emphysema
(lower lobe predominant)**

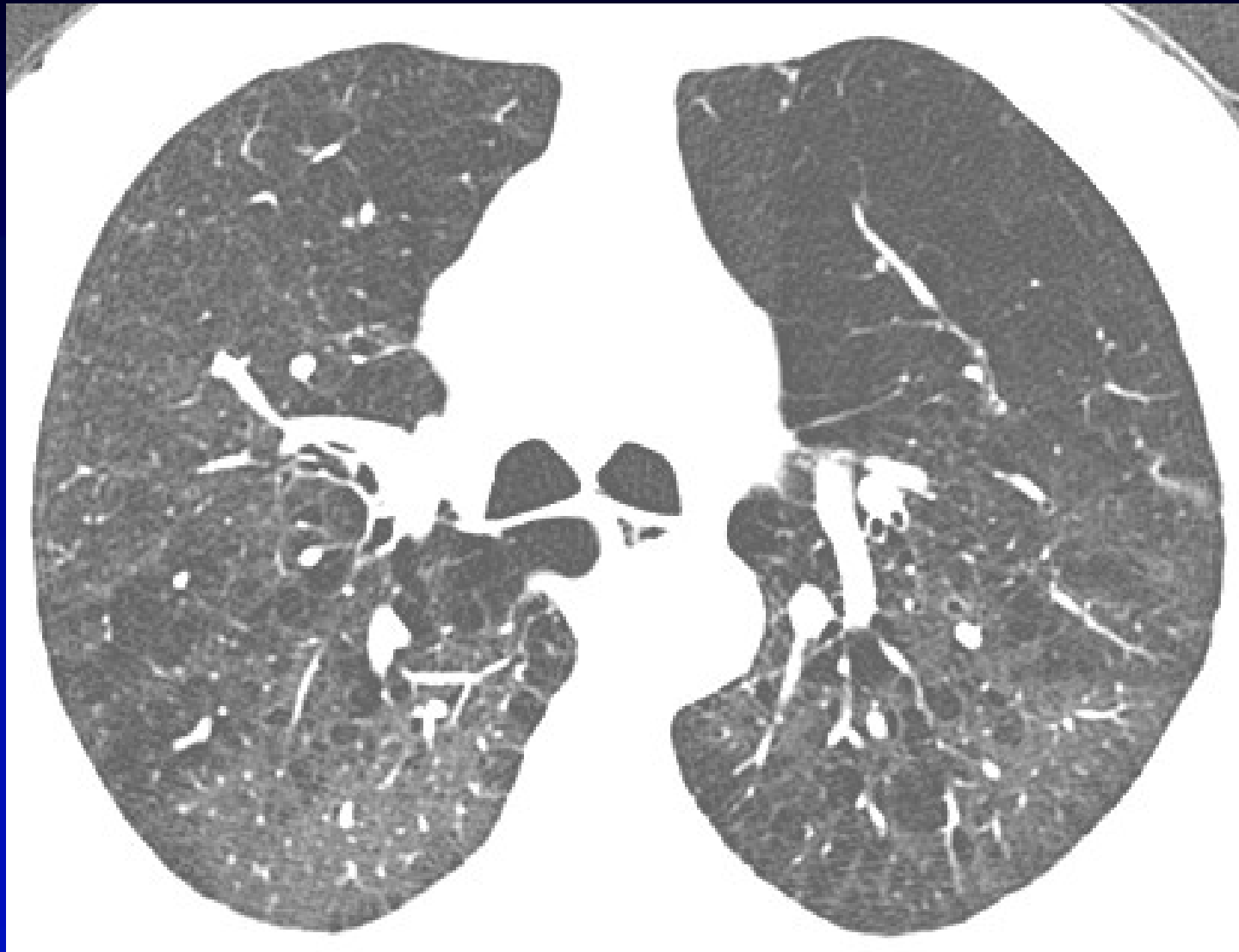
FM



FM



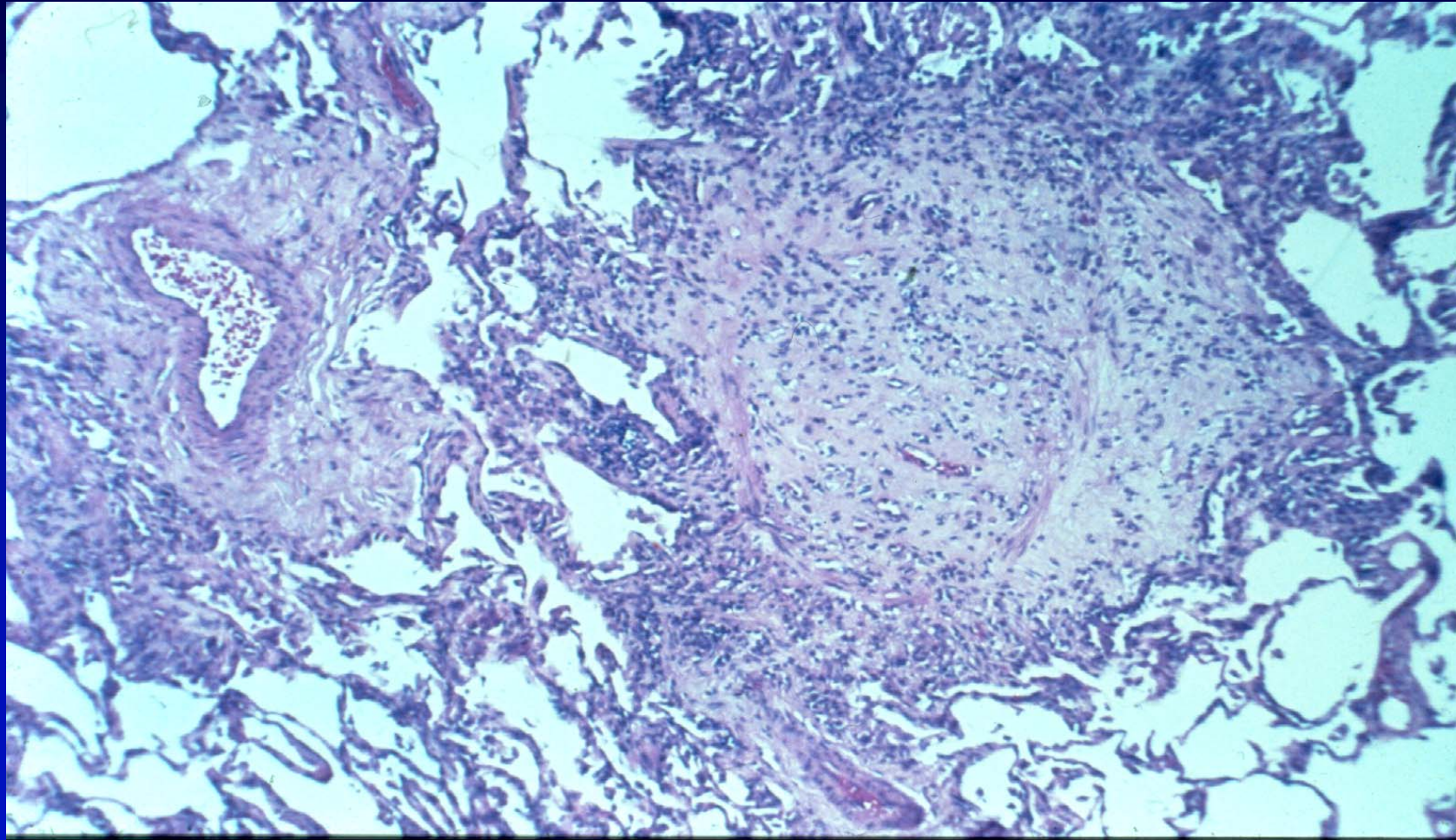
FM



FM



Obliterative Bronchiolitis (OB)



Obliterative bronchiolitis

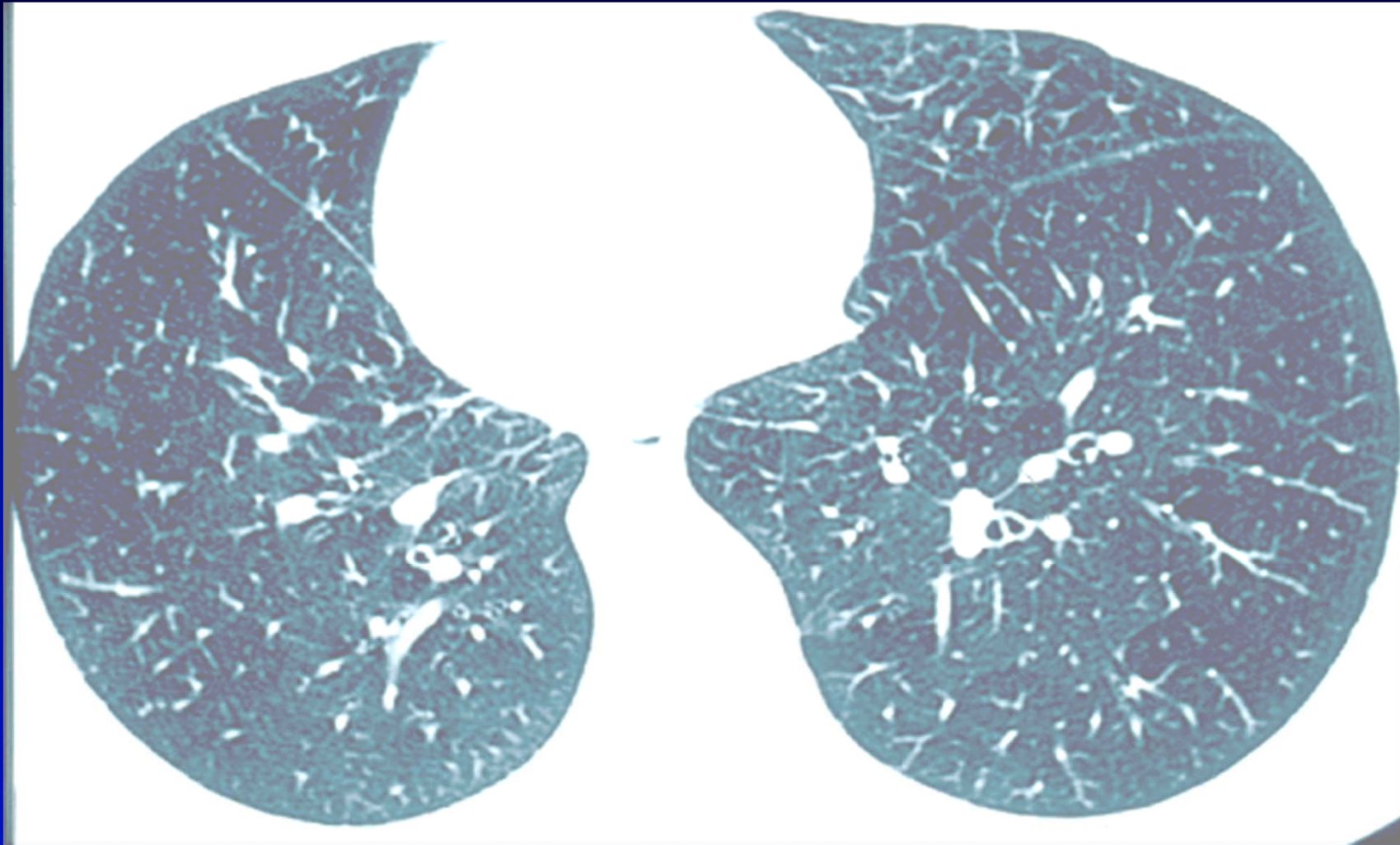
HRCT features of OB:

- **Mosaic pattern of attenuation**

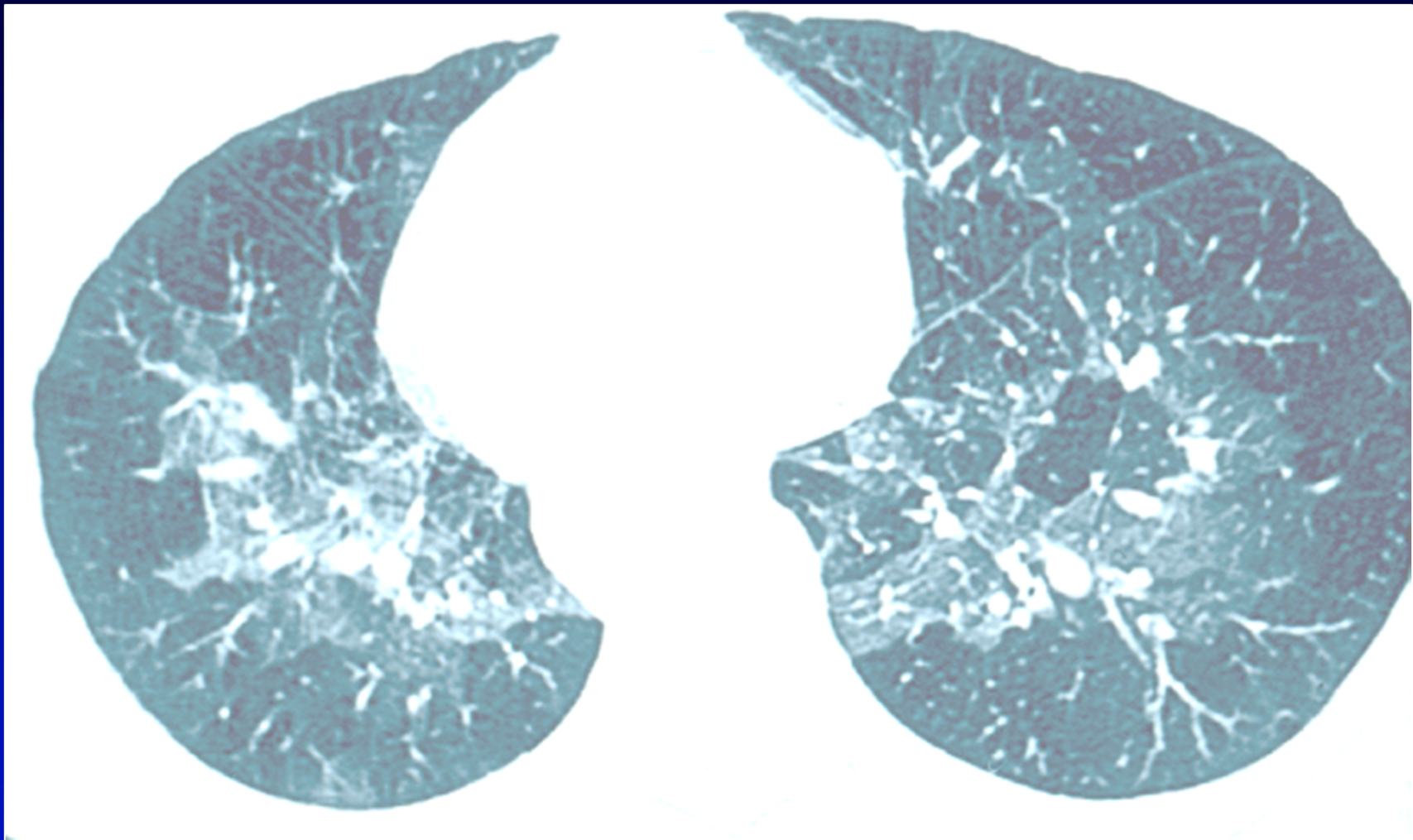
(alternating low and high attenuation areas)

- **Accentuated with expiration**
- **May be normal**

OB: Inspiratory CT scan



OB (expiratory CT-mosaic pattern)



Obliterative bronchiolitis

Expiratory CT scan:

- **Patchy, hyperlucent areas (mosaic)**
 - **Focal air-trapping** (obstructed bronchioles)
 - In normals, homogeneous increased lung attenuation with expiration

