Five Consecutive Cases when VATS Surgical Lung Biopsy Improved Outcome

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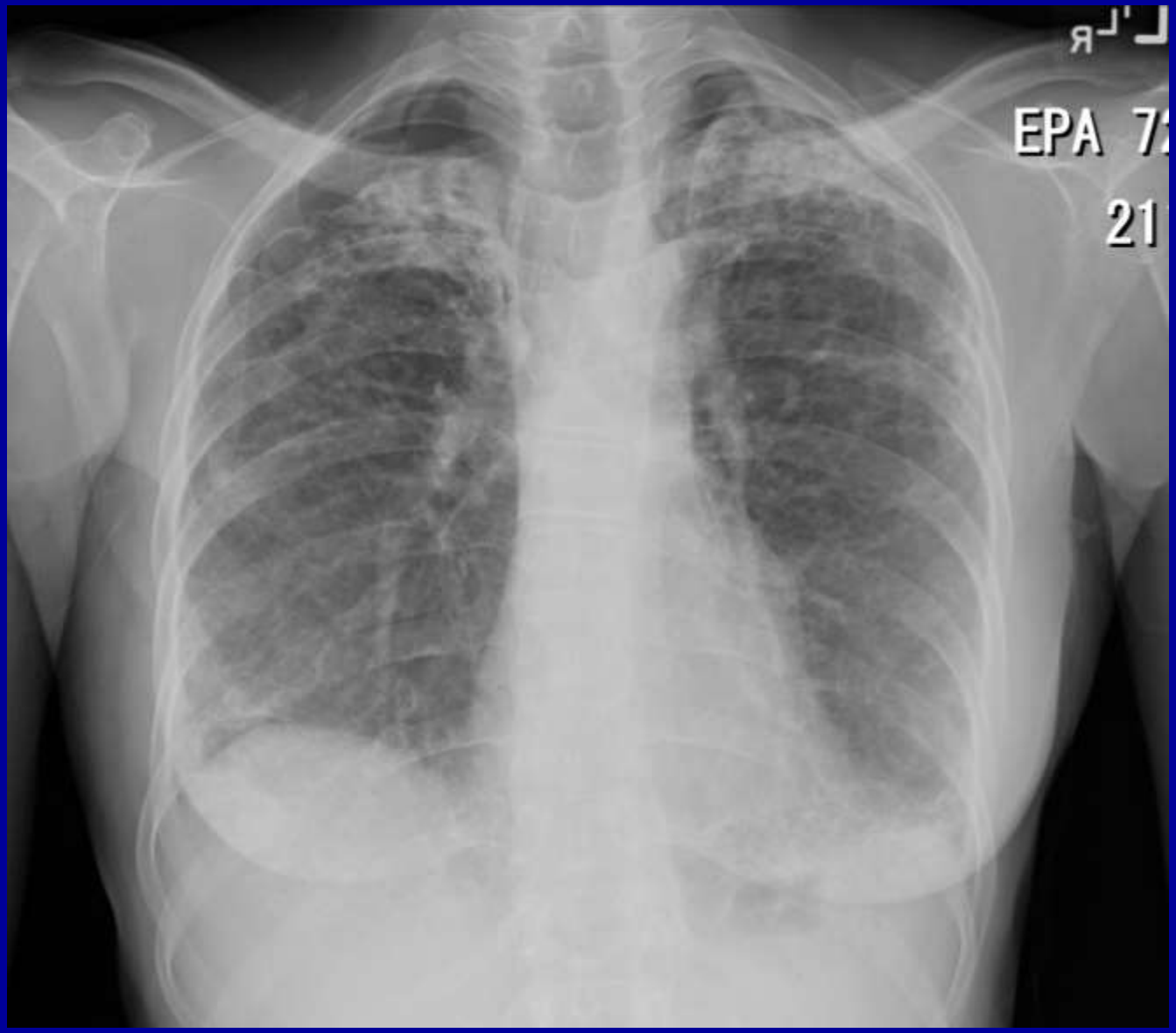
Disclosures

- Advisory Board:
  InterMune
  Boerhinger Ingelheim
  iBIOs
- Grants:
  InterMune
  Boerhinger Ingelheim
  Roche
  Gilead
- Consulting:
  InterMune
  Gilead

Case 1

Case presentation

- 54 yo ♀ with dyspnea over 1 month
- 1 week of bilateral, pleuritic chest pain
- No past medical history
- No known exposures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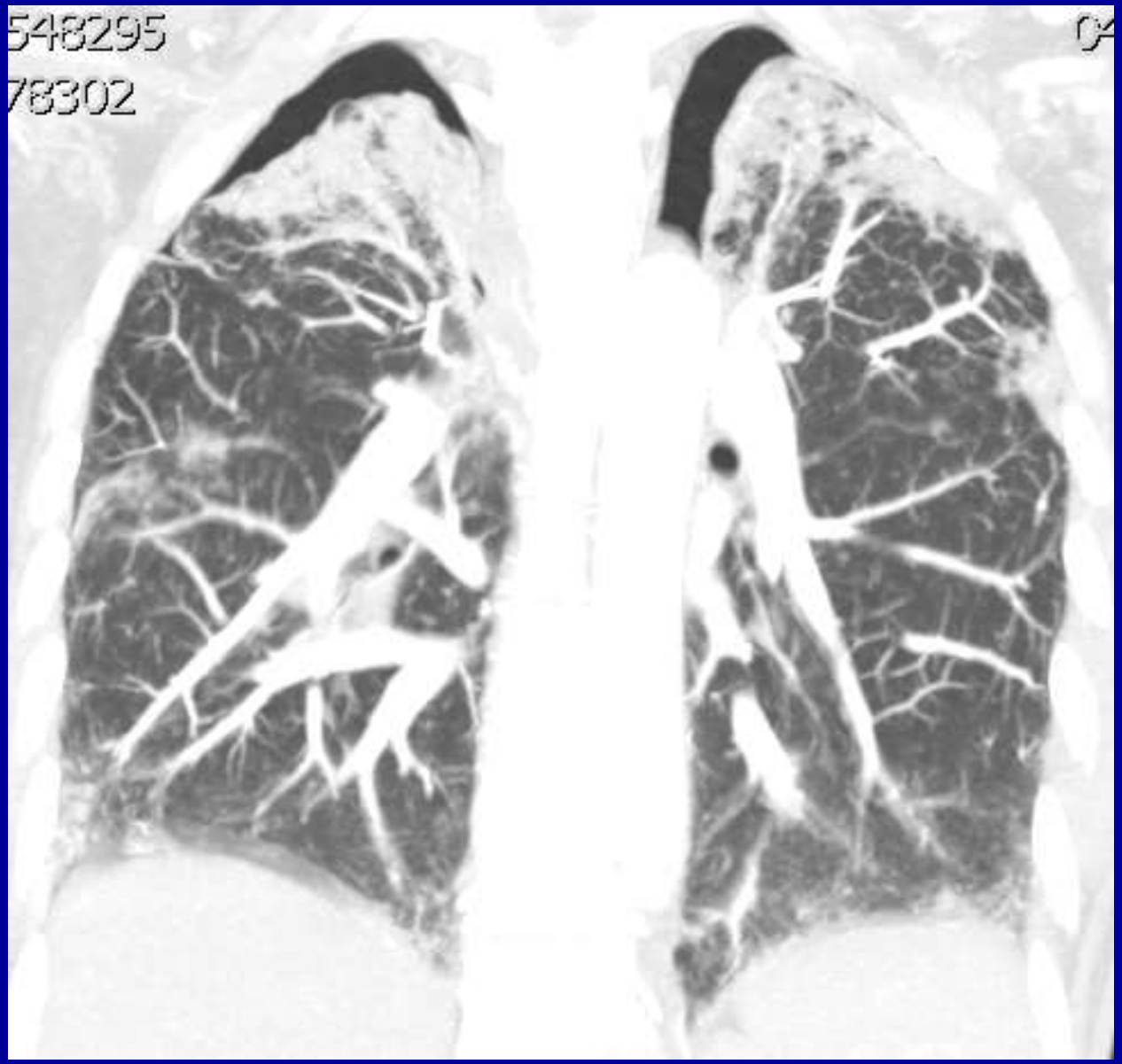
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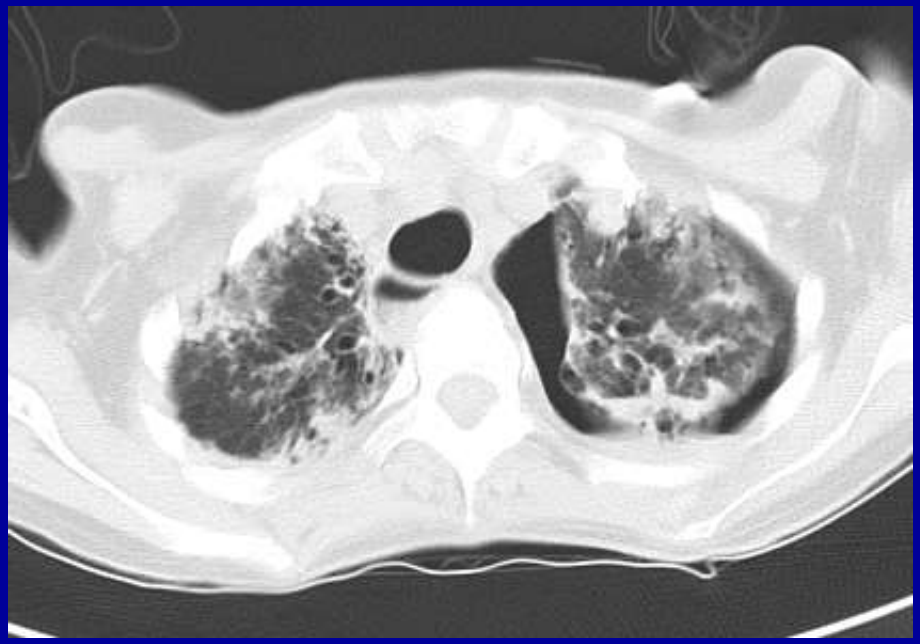
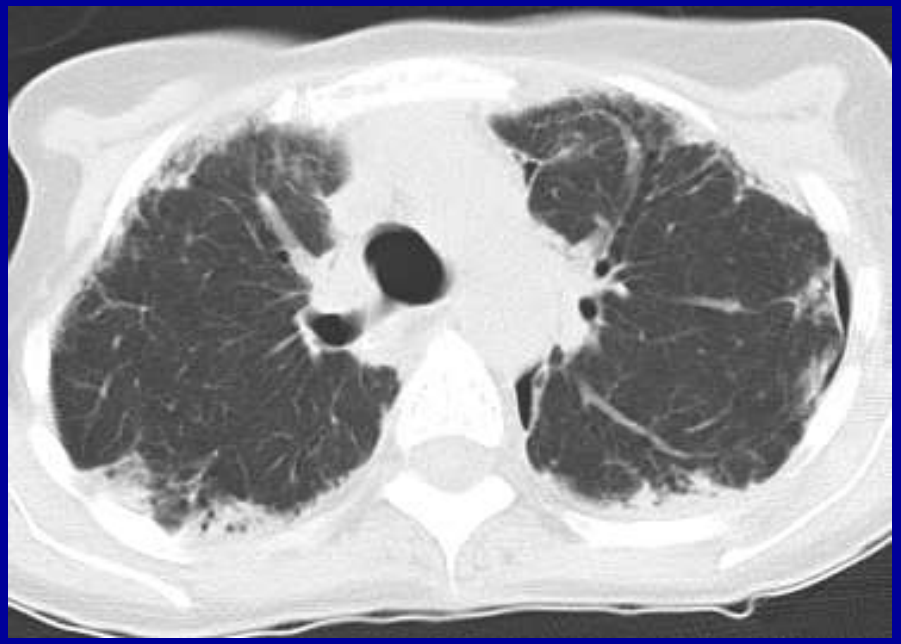
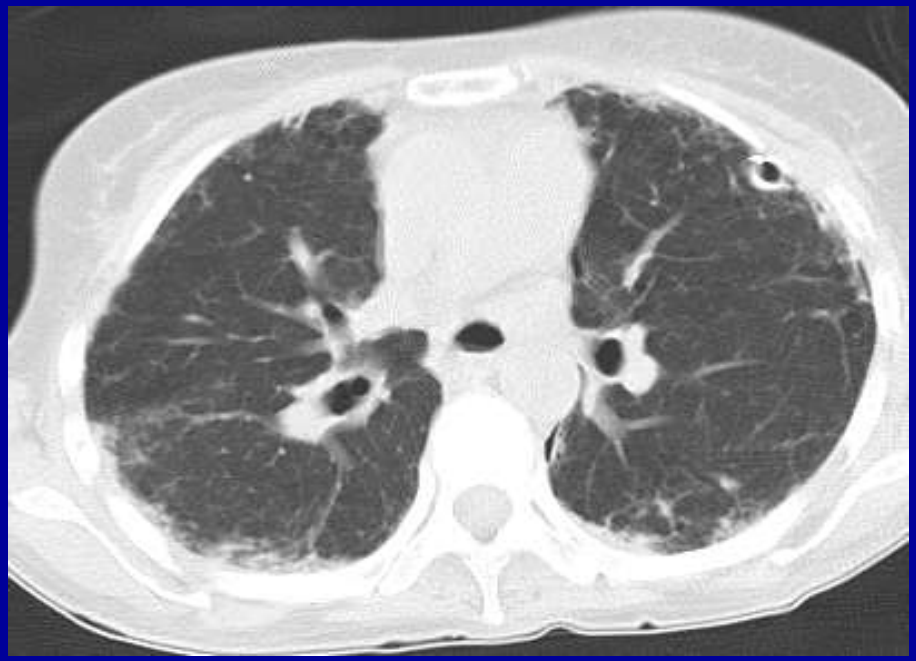
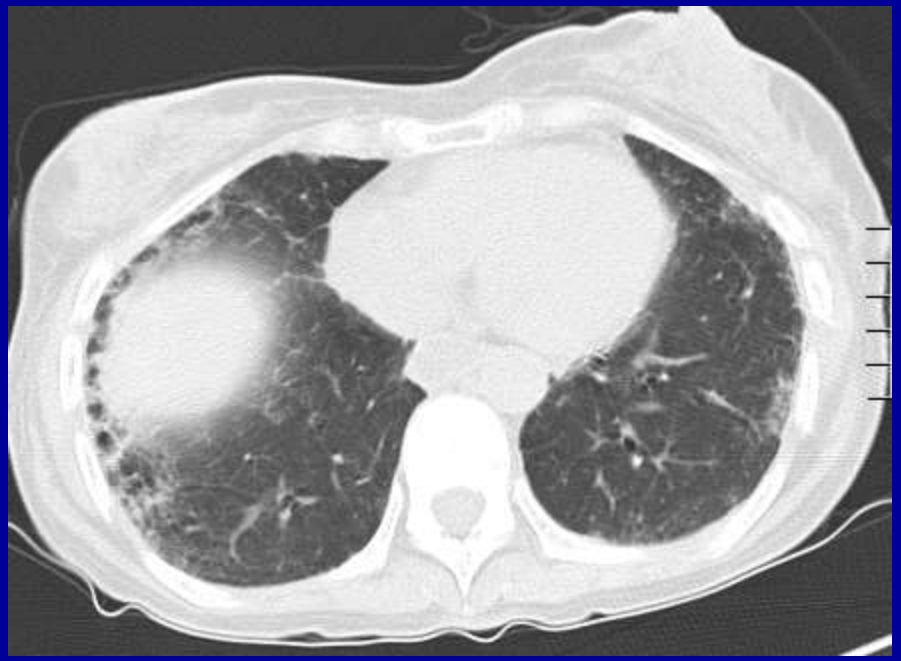
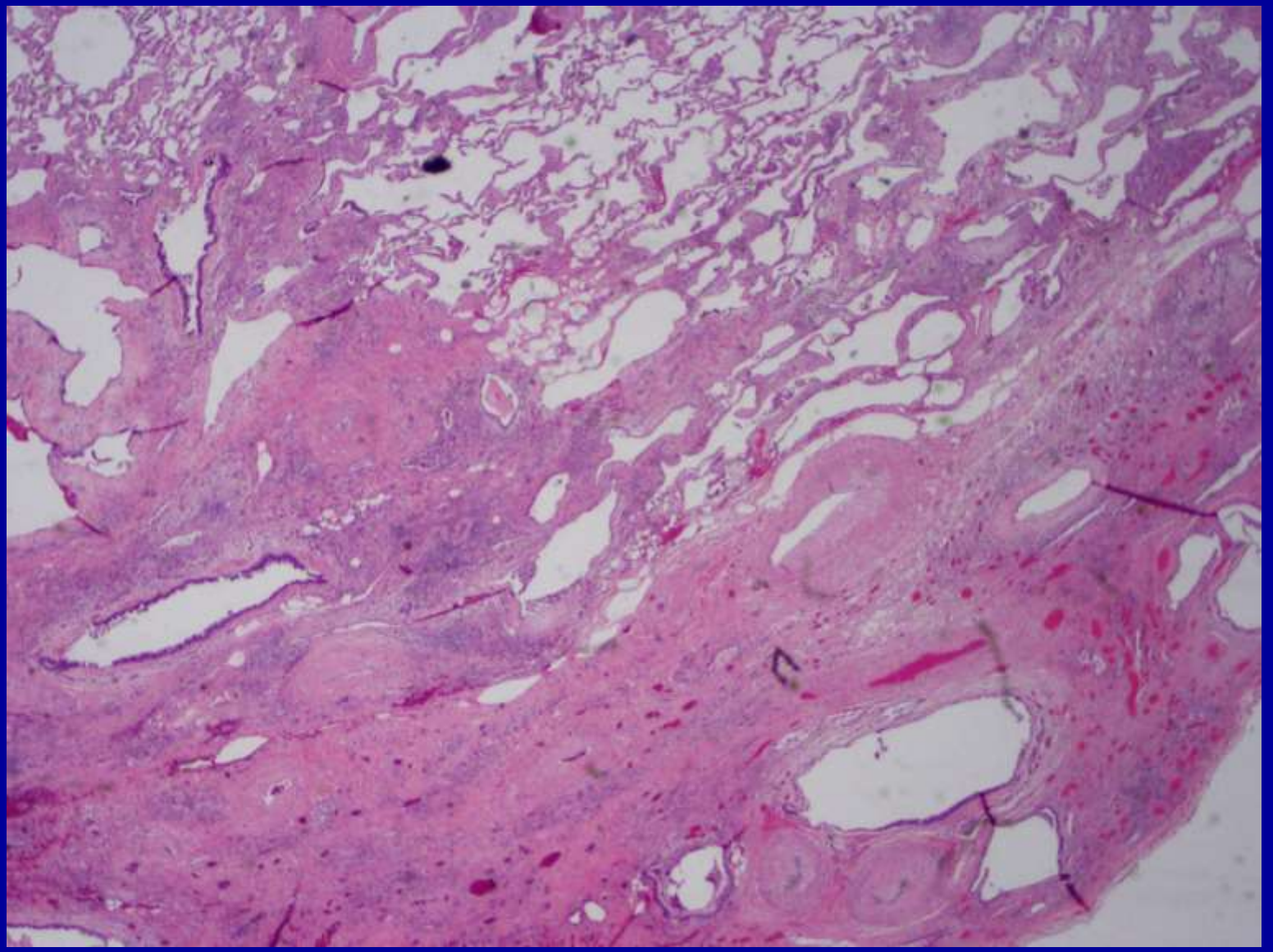





Hematoxylin-eosin stain



Riemer E.

Pleuroparenchymal Fibroelastosis (PPFE)

An Official American Thoracic Society/European Respiratory Society Statement: Update of the International Multidisciplinary Classification of the Idiopathic Interstitial Pneumonias

- "Rare IIPs"
  - Idiopathic Lymphoid Interstitial Pneumonia
  - Idiopathic Pleuroparenchymal Fibroelastosis

Travis et al. *Am J Respir Crit Care Med* 2013.

PPFE

- Amitani – “Idiopathic upper lobe fibrosis” in 1992
  - Other cases described prior in 1960’s

- Frankel in 2004, 5 patients at National Jewish → PPFE

Amitani R, et al. *Kokyu* 1992.
Frankel SK, et al. *Chest* 2004.

Pleuroparenchymal Fibroelastosis: Its Clinical Characteristics

- Idiopathic PPFE
- Radiation
- Anticancer chemotherapy
- Bone marrow- or stem cell-transplantation
- Lung transplantation
- Occupational dust exposure
  - Asbestos
  - Aluminum
- Infection
  - Aspergillus
  - Mycobacterium avium-intracellulare
- Hereditary PPFE ~ PPFE with family history
- Autoimmune diseases
  - Rheumatoid arthritis
  - Ulcerative colitis
  - Psoriasis
  - Ankylosing spondylitis
- Hypersensitivity pneumonitis

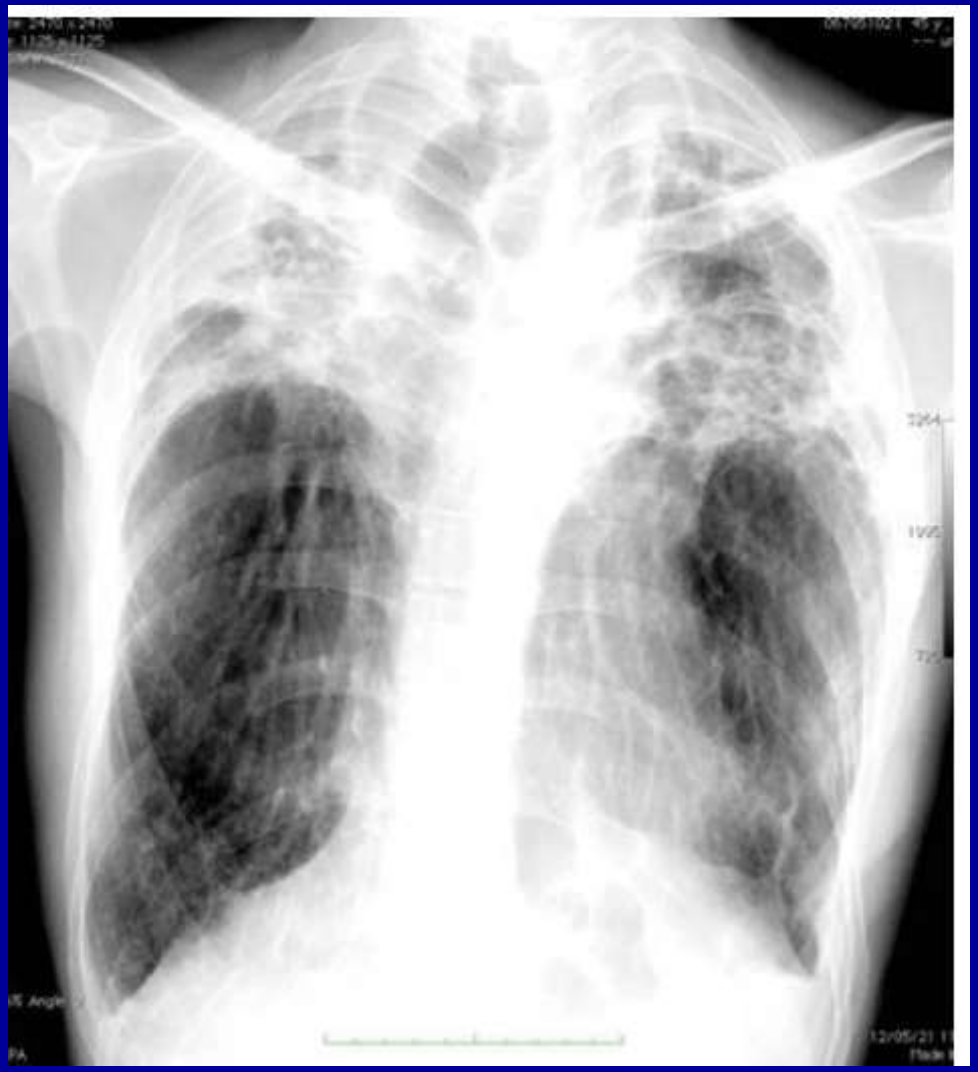
Watanabe K. *Curr Respir Med Rev* 2013.

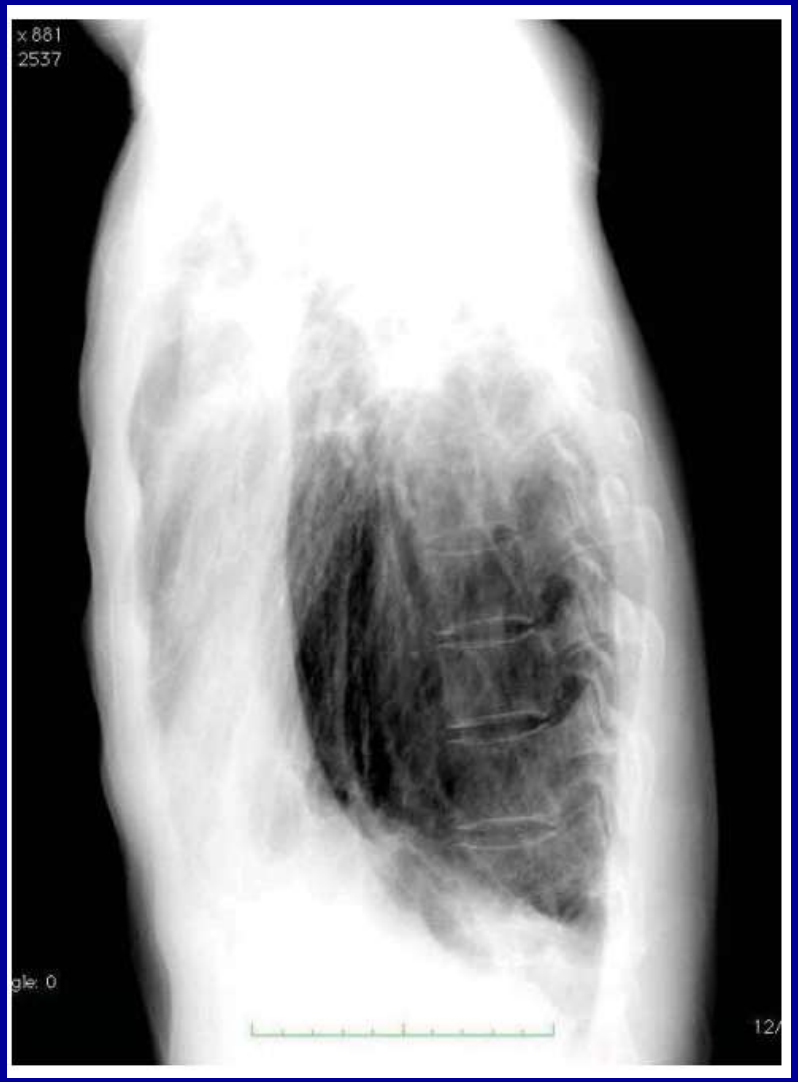
Pleuroparenchymal Fibroelastosis: Its Clinical Characteristics

No gender preponderance
Age at onset
Wide-ranging, younger than in idiopathic pulmonary fibrosis (IPF)
Smoking history
Unrelated to the incidence of PPFE
Clinical symptoms
Exertional dyspnea and dry cough with insidious onset
Chest pain due to pneumothorax
Loss of body weight
Physical findings
Slender stature and flattened thoracic cage
Crackles sometimes audible
Serum biomarkers
KL-6 within the normal or around the upper normal limit
Elevated Surfactant protein D (SP-D)
Autoantibodies such as rheumatoid factor and antinuclear antibody sometimes elevated
Prognosis
Wide-ranging in each case studies from slowly progressive with 10 - 20 years of presentation to rapidly progressive course
Poorer prognosis of secondary PPFE such as transplantation-associated PPFE

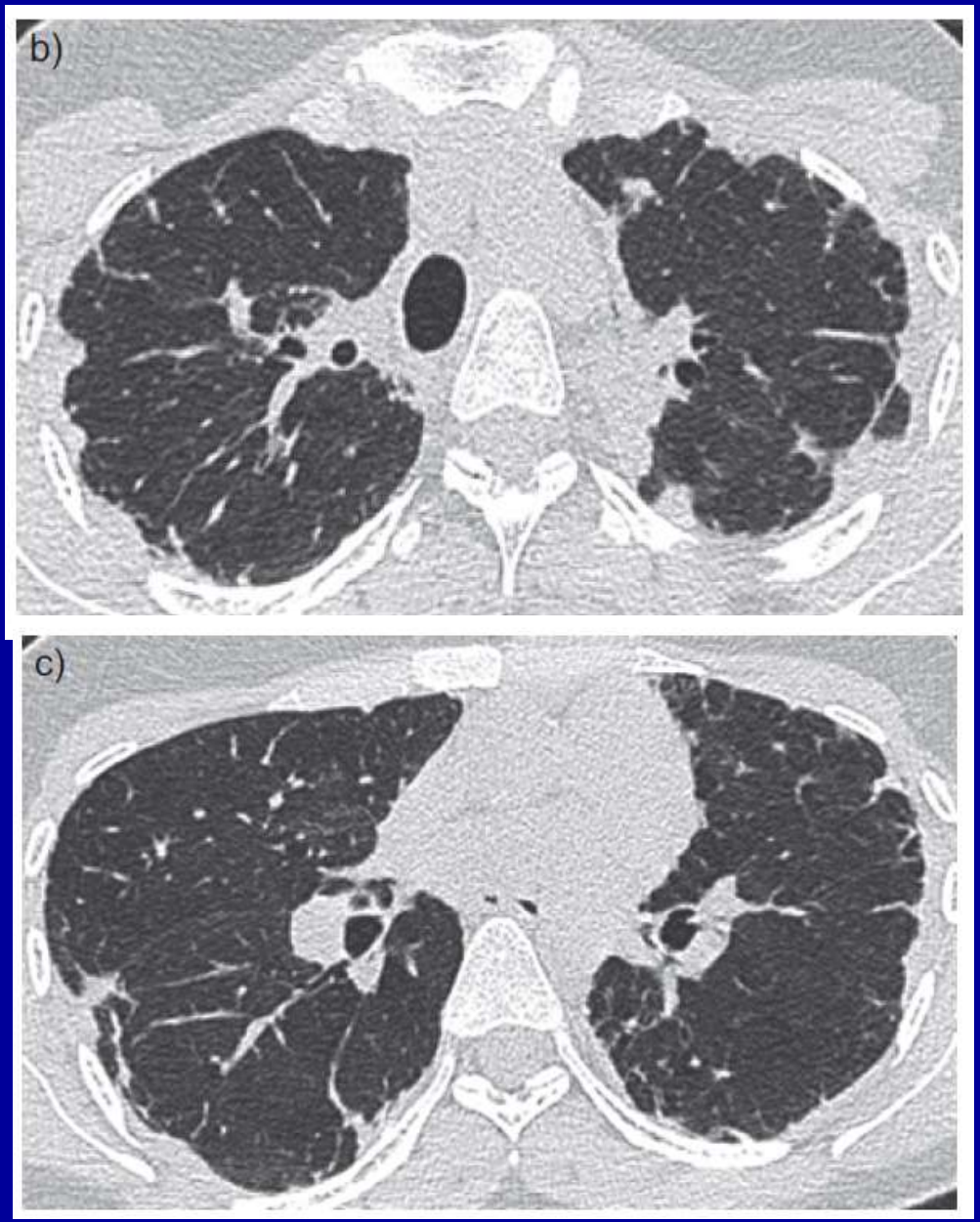
Watanabe K. *Curr Respir Med Rev* 2013.

PPFE

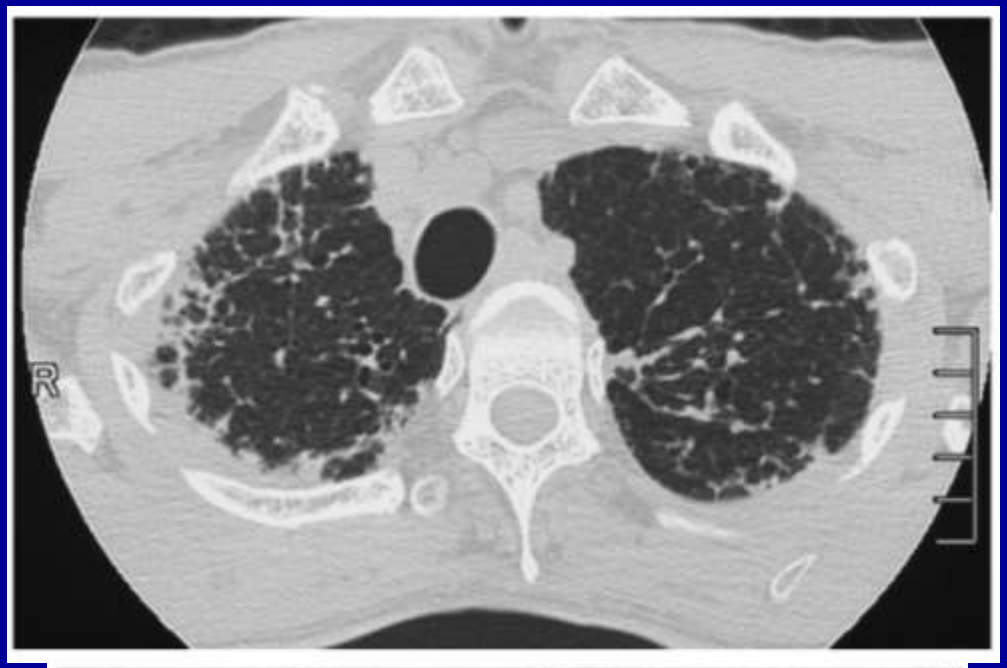


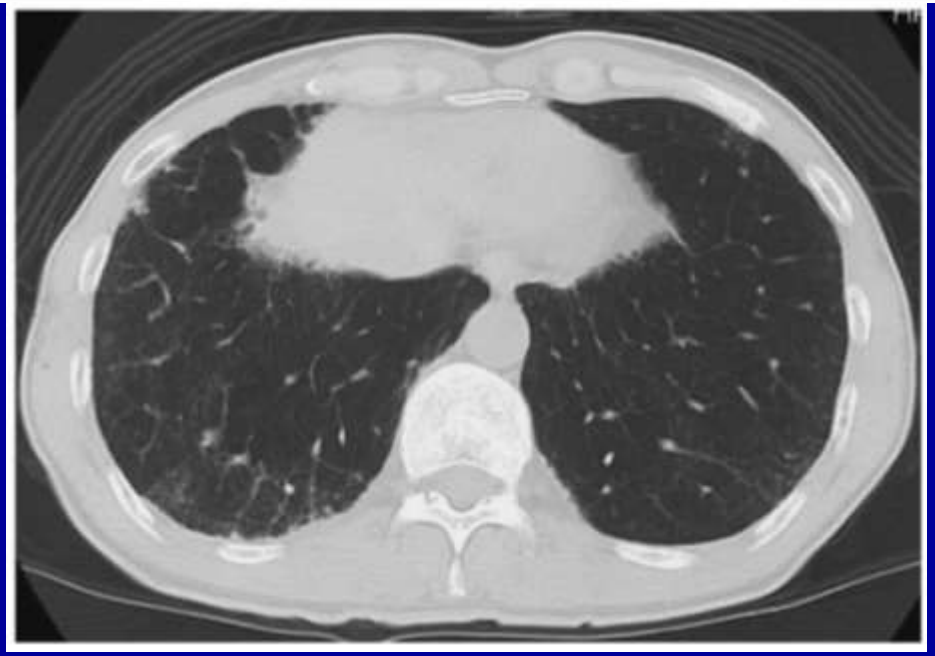


Watanabe K. *Curr Respir Med Rev* 2013.



Reddy TL, et al. *Eur Respir J* 2012.





Watanabe K, et al. *Respir Investig* 2012.

Pleuroparenchymal Fibroelastosis: Its Clinical Characteristics

Chest Radiograph

Abnormally narrowed anterior-posterior thoracic dimension (flattened thoracic cage)

Elevated hilar opacities

Reticular and nodular opacities in the bilateral upper lung fields

Fibrocystic opacities in the upper lung fields and occasional reticular opacities in the lower lung fields in the advanced stage

Chest CT

Initial stage:

Subpleural nodular and reticular opacities in the apex, but minimal changes in the middle and lower lobes

Advanced stage:

Fibrotic opacities with traction bronchiectasis extending to adjacent lobes with multiple bulla and large cysts at the upper lung fields

Occasional subpleural reticular opacities in the bilateral lower lobes resembling usual interstitial pneumonia (UIP)

Watanabe K. *Curr Respir Med Rev* 2013.

Pleuroparenchymal Fibroelastosis: Its Clinical Characteristics

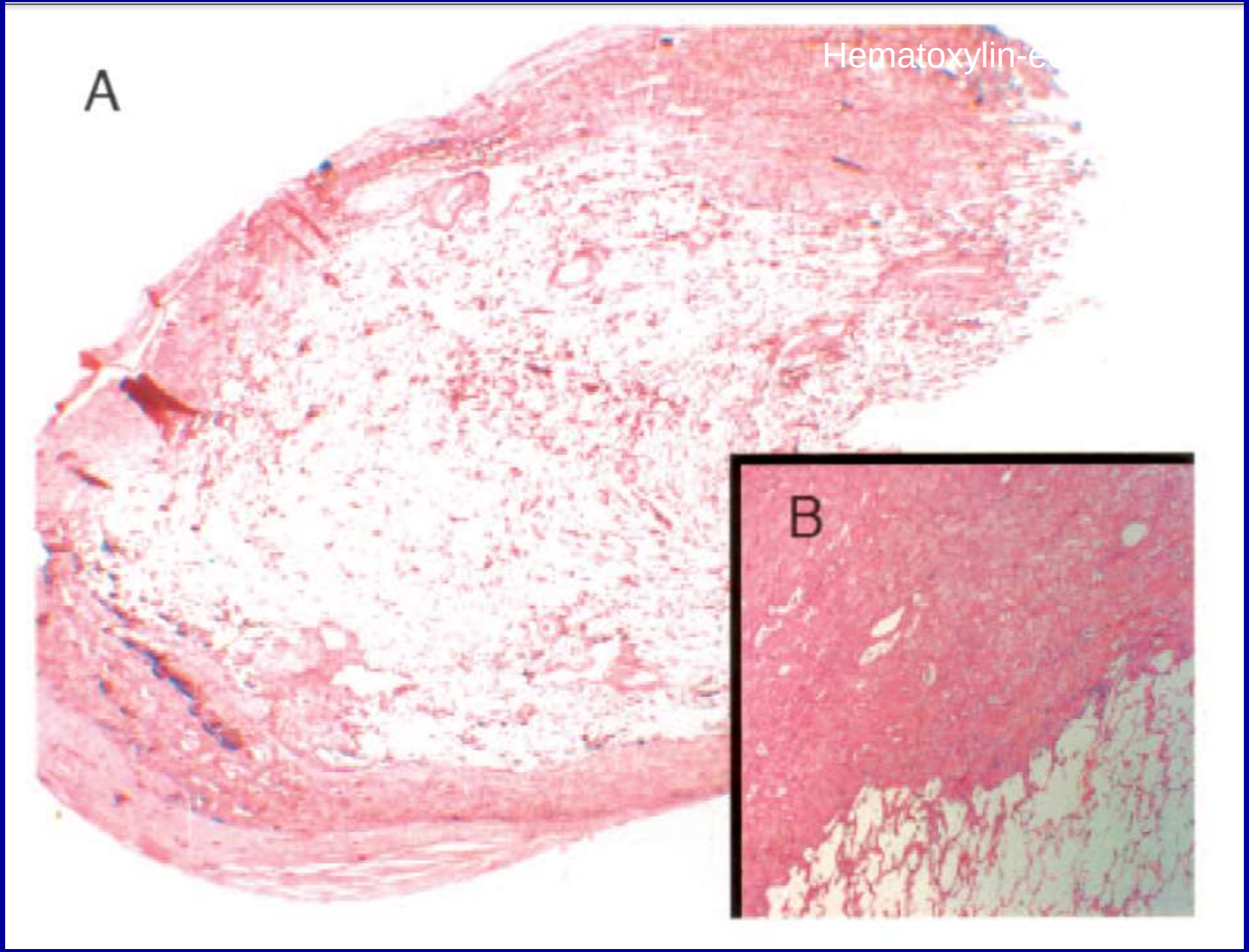
Ventilatory Impairment

- Decreased FVC
- Increased FEV_1/FVC (%)
- Decreased TLC
- Increased RV/TLC (%)

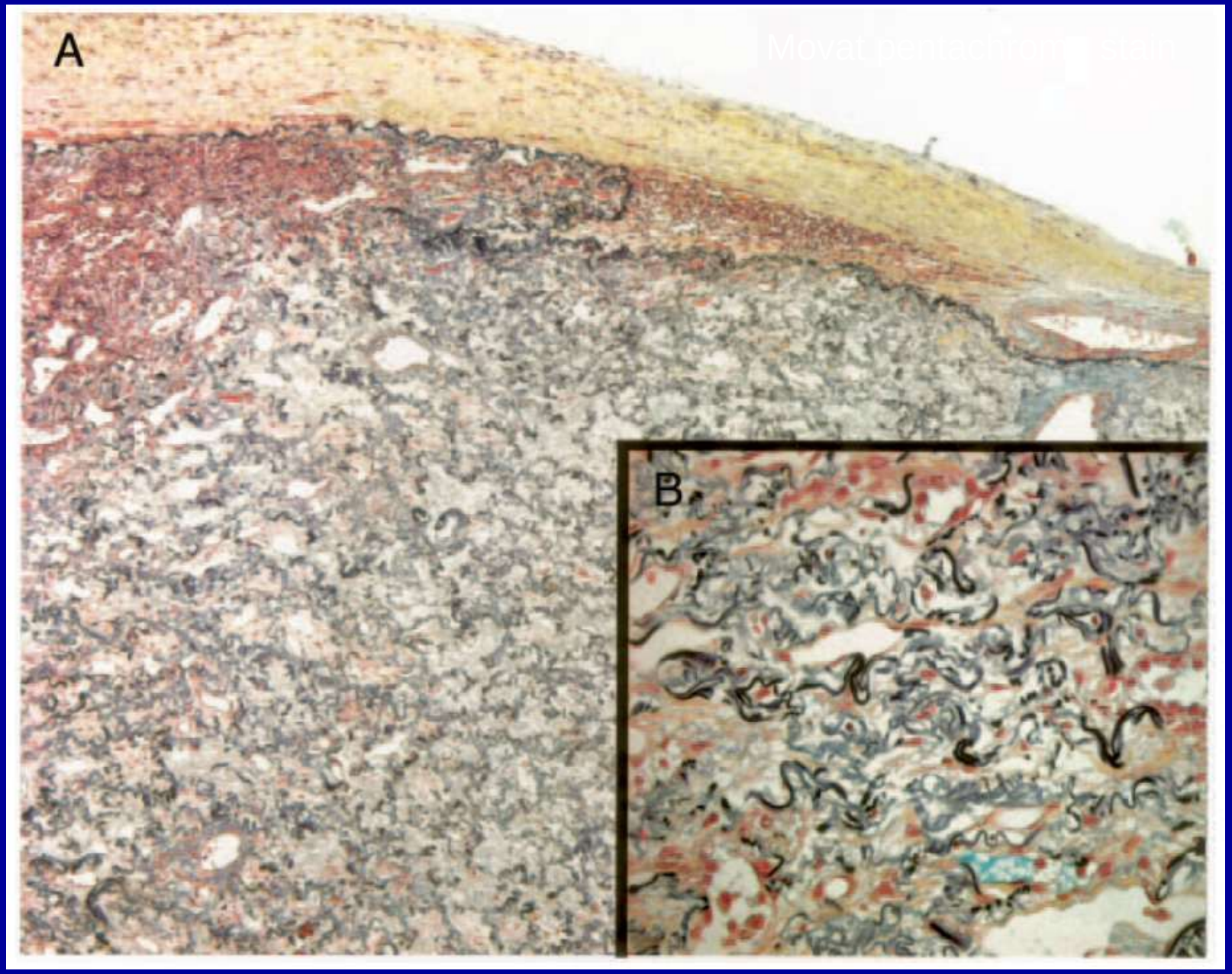
Gas Exchange Impairment

- Decreased DLco
- Normal or minimally decreased DLco/VA

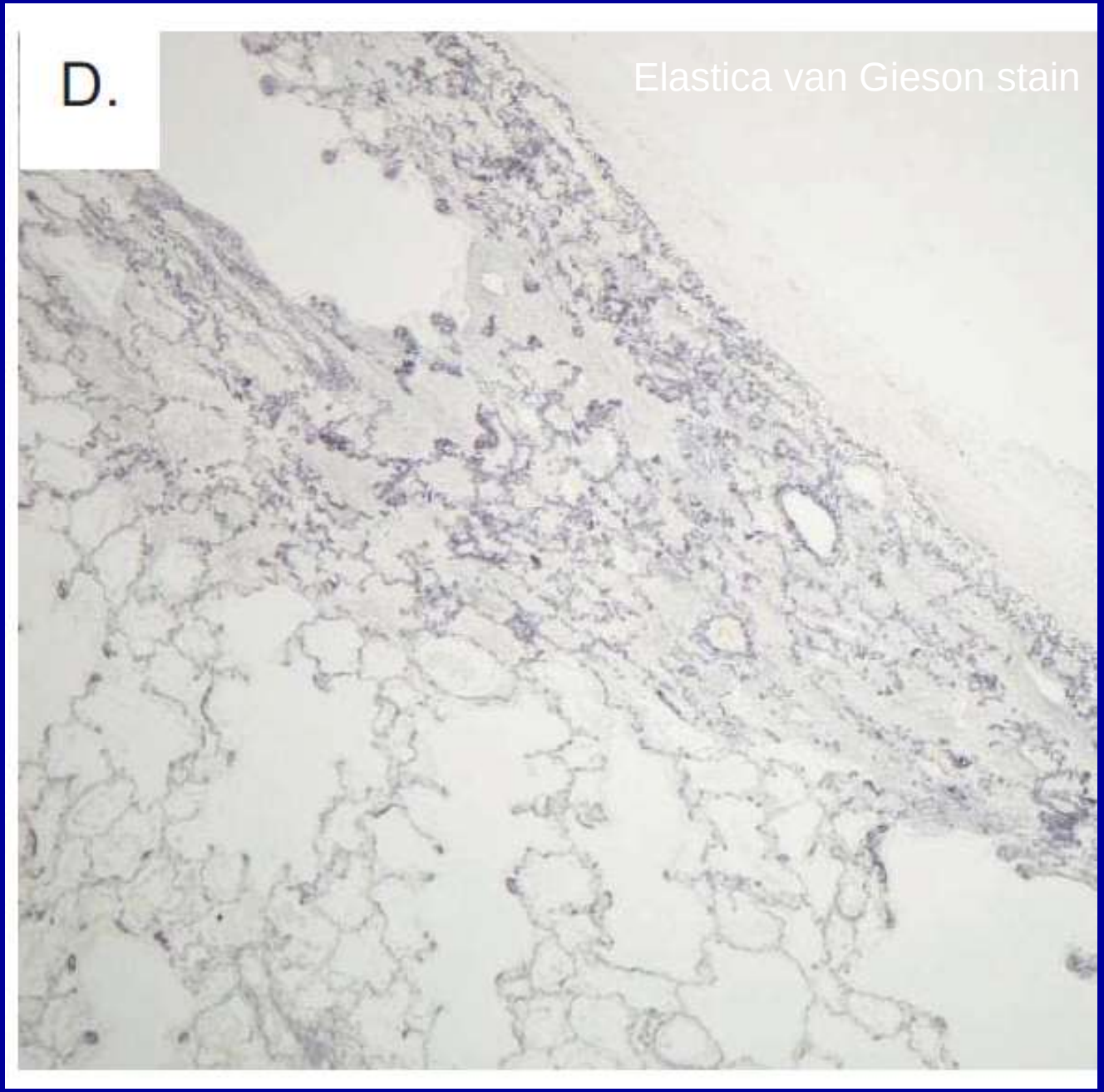
Watanabe K. *Curr Respir Med Rev* 2013.



Frankel SK. *Chest* 2004.



Frankel SK. *Chest* 2004.



Von der Thusen. *Curr Respir Med Rev* 2013

PPFE

- Histologic criteria
  - Upper zone fibrosis of visceral pleura
  - Prominent, homogenous, subpleural fibroelastosis
  - Sparing of parenchyma distant from pleura
  - Mild patchy lymphoplasmocytic infiltrates
  - Small numbers of fibroblastic foci

Frankel SK. *Chest* 2004.

PPFE vs IPF

- UIP pattern found in lower lobes of PPFE
- 2X more elastic fibers in PPFE than IPF
- Upper lobe
- Alveolar architecture preservation
- Lack of fibroblastic foci adjacent to fibrosis

Enomoto N, et al. *BMC Pulm Med*. 2014
Reddy TL, et al. *Eur Respir J*. 2012

PPFE

Treatment:

- Refractory to steroids & immunosuppression
- O2
- ?Pirfenidone
- ?Target inhibition of elastosis

Watanabe K. *Curr Respir Med Rev* 2013.

PPFE - pearls

- One of the rarest forms of IIP
- Mid-lung & upper lung zone pleural and parenchymal abnormalities
- Distinctive histopathologic findings
  - Intense pleural fibrosis
  - Subpleural parenchymal fibroelastosis
  - Upper lobe predominance
  - Sparing of lung distant to pleura
- May see UIP at bases in advanced disease
- No good therapy

Case 2

Case Presentation

- HPI:
  - 70 y/o male
  - Bx proven NASH cirrhosis and heptocellular carcinoma
  - Abnormal Chest CT
  - Needs a liver transplant

Case Presentation

- PMHx: Type 2 DM, HCC, NASH cirrhosis, hypothyroidism, HTN
- Surgical Hx: Rotator cuff, inguinal hernia
- Family History: N/C
- Social History:
  - Life-long non-smoker
  - No alcohol or illicit drugs
  - No TB, pets, birds or other exposures

Case Presentation

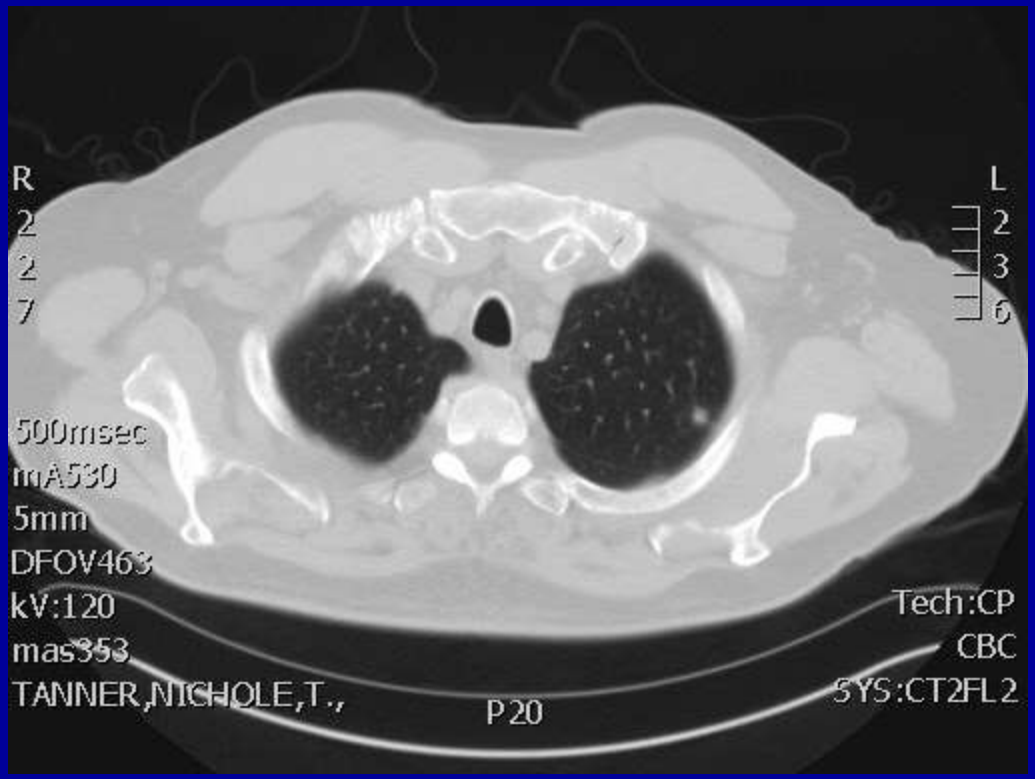
- Home medications:
  - Atenolol 25 mg daily
  - Levothyroxine 50 mcg daily
  - Vitamin E 400 IU daily
  - Lisinopril 20 mg daily

Case Presentation

- P/E:
  - V/S: 98.0 F, 56, 16, 98% on RA
  - Findings noted only for ascites, palmar erythema, and gynecomastia

- Chest CT 8 mm and 6 mm, non-calicfied nodules in LUL (noted 3 months prior and stable)

Chest CT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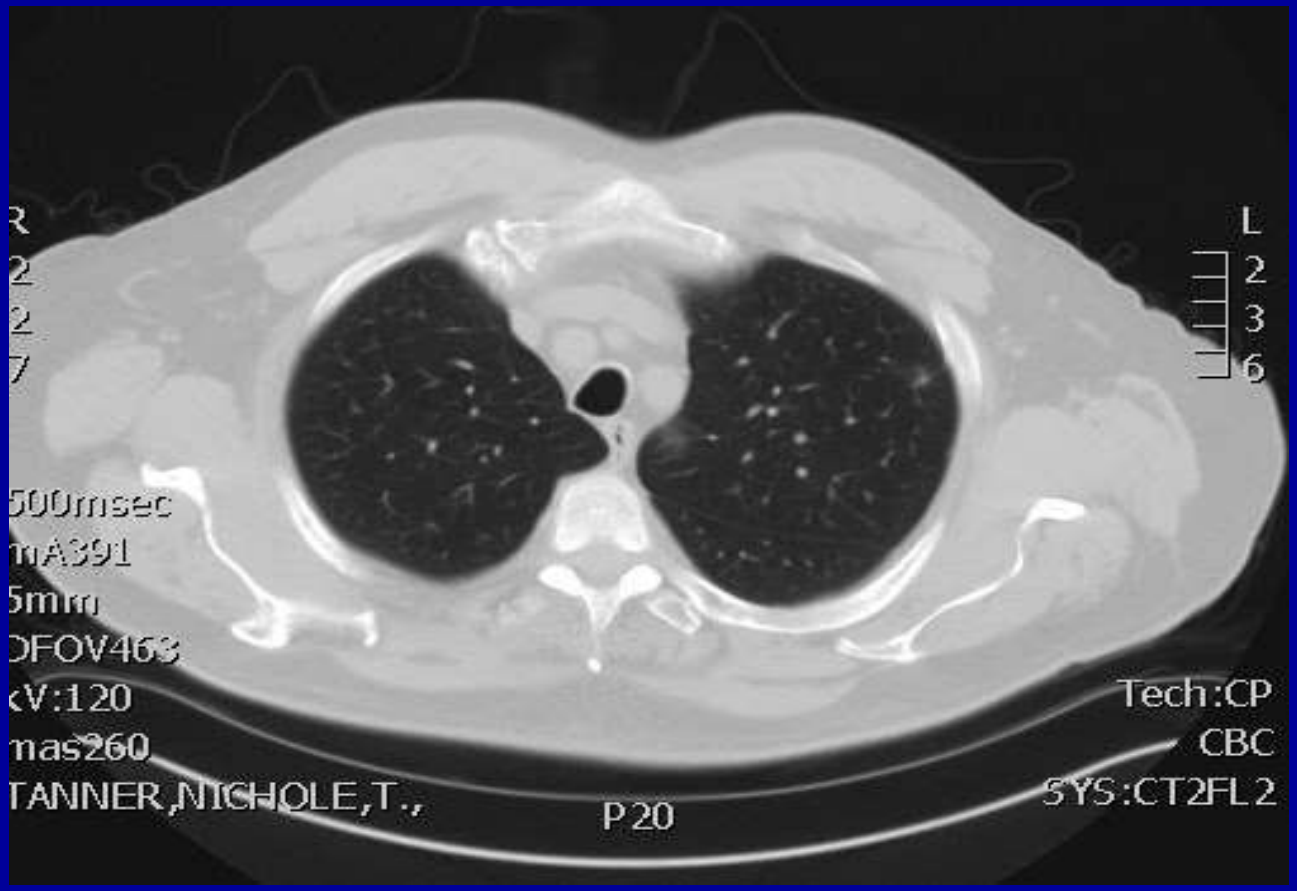
Chest CT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kV:120
mas260
TANNER,NICHOLE,T.,
P20
Tech:CP
CBC
SYS:CT2FL2

Case Presentation…

- PFTs:
  - FVC: 123% of predicted
  - FEV1: 104% of predicted
  - DLCO: 100% of predicted

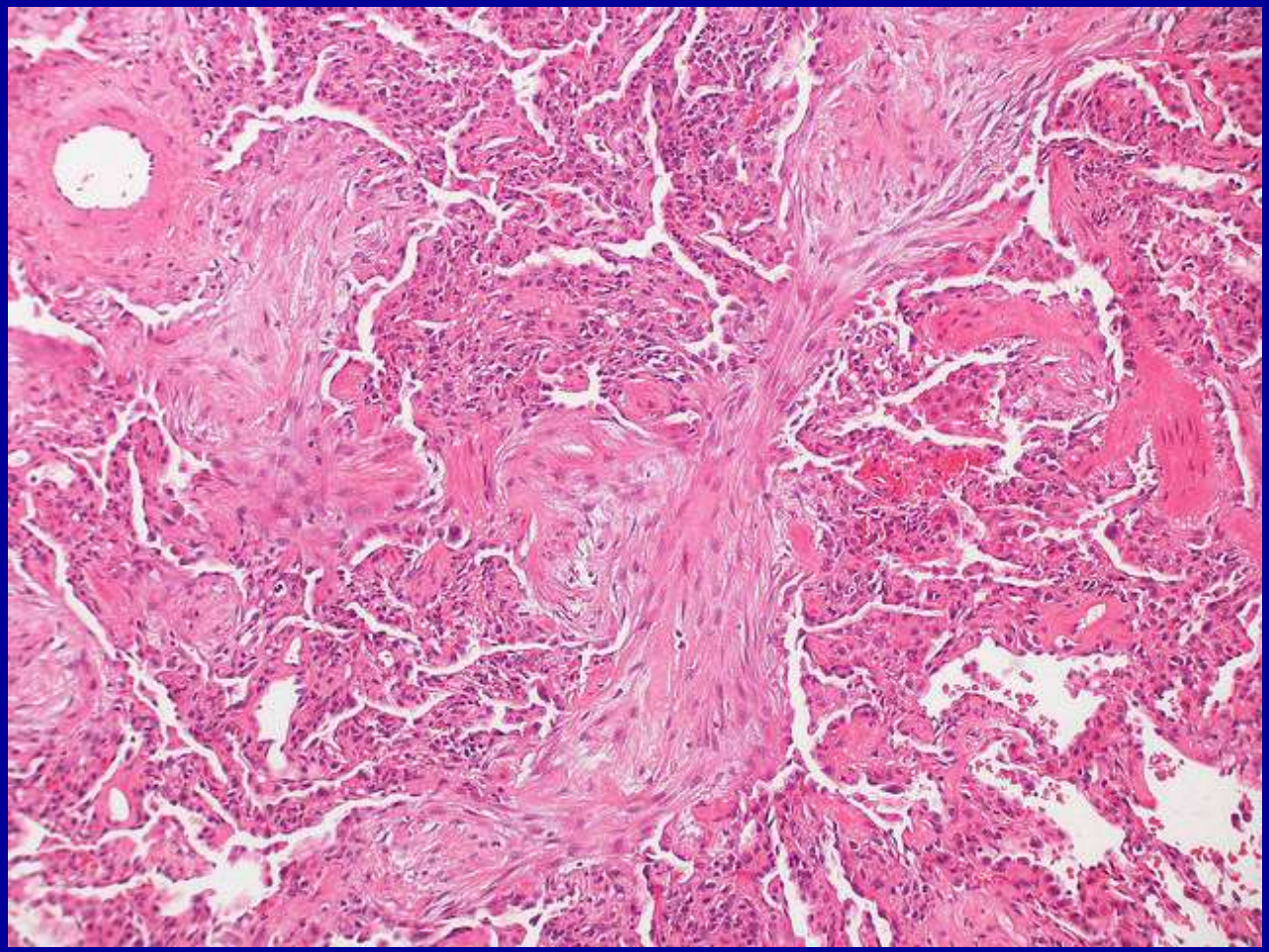
Case Presentation…

- What should we do next??

Case Presentation

- VATS wedge resection of LUL performed

Pathology



Case Presentation

- Diagnosis: Cryptogenic organizing pneumonia

- Outcome: Successful orthotropic liver transplantation

COP

Case 3

Case Presentation

- CC: “I have a little bit of everything”
- HPI:
  - 32 y/o female
  - No past medical history
  - Cold
  - Progressively worsening shortness of breath
  - Admitted to outside hospital

Case Presentation

- PMHx: denies any medical problems
- Surgical Hx: no prior surgeries
- Family History: no illnesses
- Social History:
  - Quit smoking 9 months ago
  - No alcohol or illicit drugs
  - No TB, pets, birds or other exposures

Case Presentation

- ROS
  - Fatigue
  - SOB
- Allergies: azithromycin (urticaria)
- Home medications: none

Case Presentation

- P/E:
  - V/S: 130/81, 109, 97.9, 18, 98% on 2LNC
  - Gen: obese, in no apparent distress
  - Lungs: bilateral basilar dry crackles

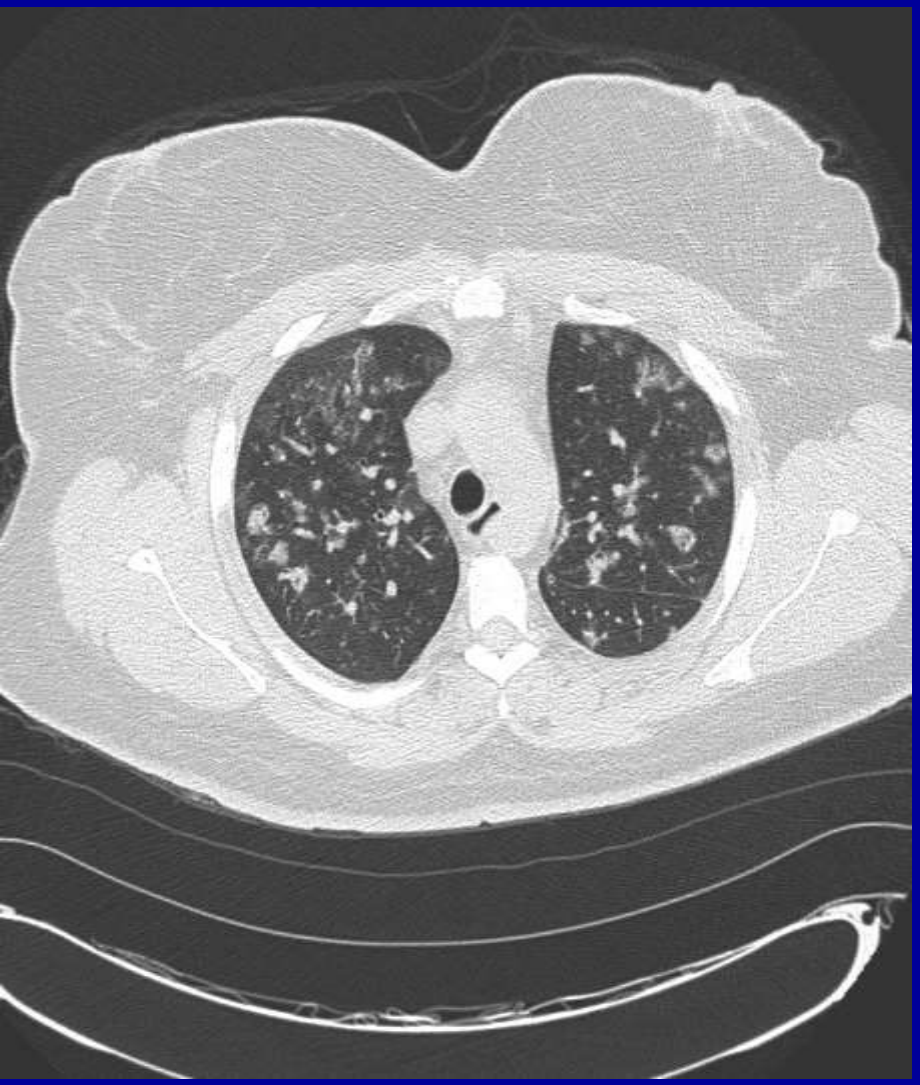
- Imaging studies

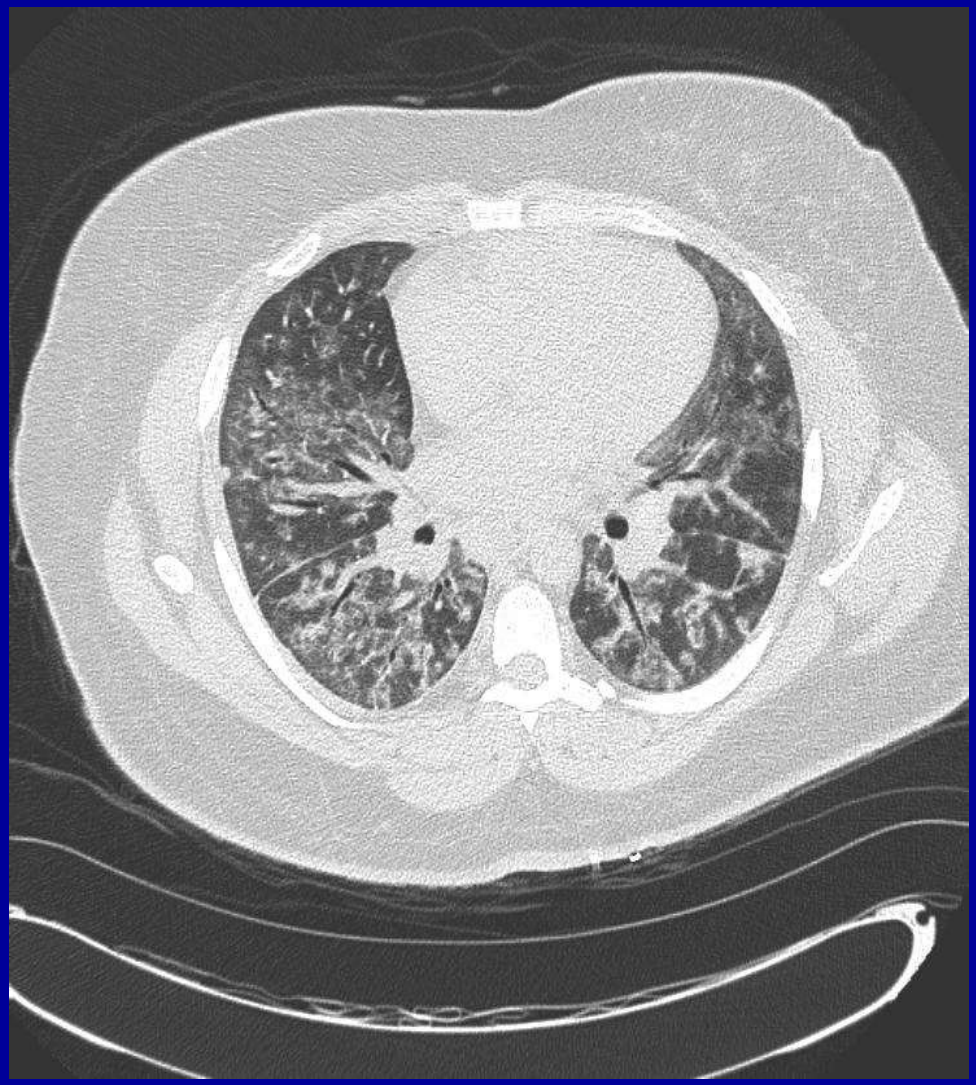
Case Presentation…

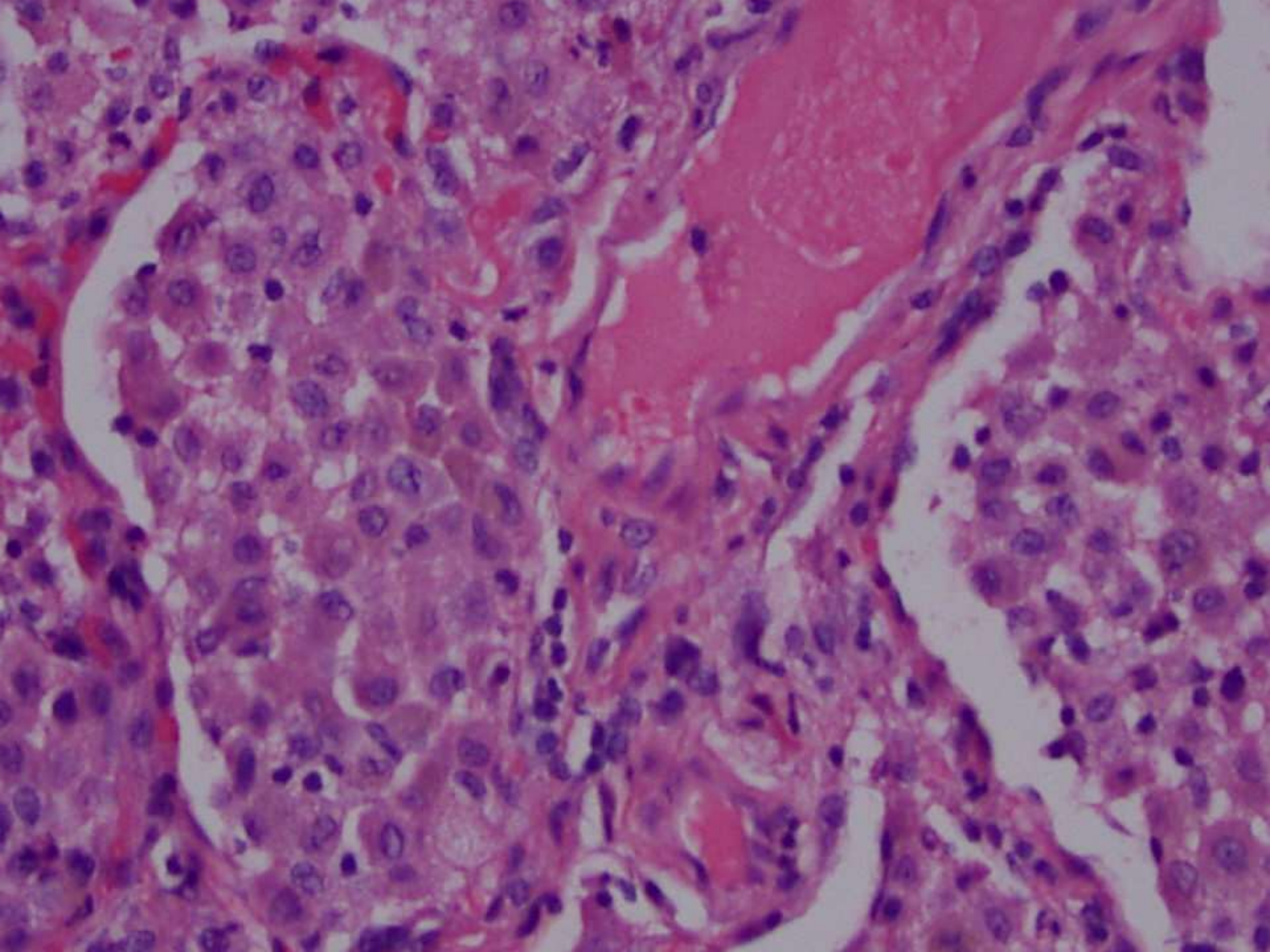
- PFTs:
  - FVC: 1.68 L (50%)
  - FEV1: 1.63 L (58%)
  - FEV1/FVC: 0.97
  - TLC 2.44 L (46%)
  - DLCO 8.1 (28%)

Case Presentation…

- Laboratory data:
  - ESR: 54 mm/h
  - PL-12 antibody: moderate positive (Mayo Medical Laboratories, Rochester MN)
  - JO-1 antibody: negative
  - PL-7 negative
  - Rest of myositis antibody panel: negative





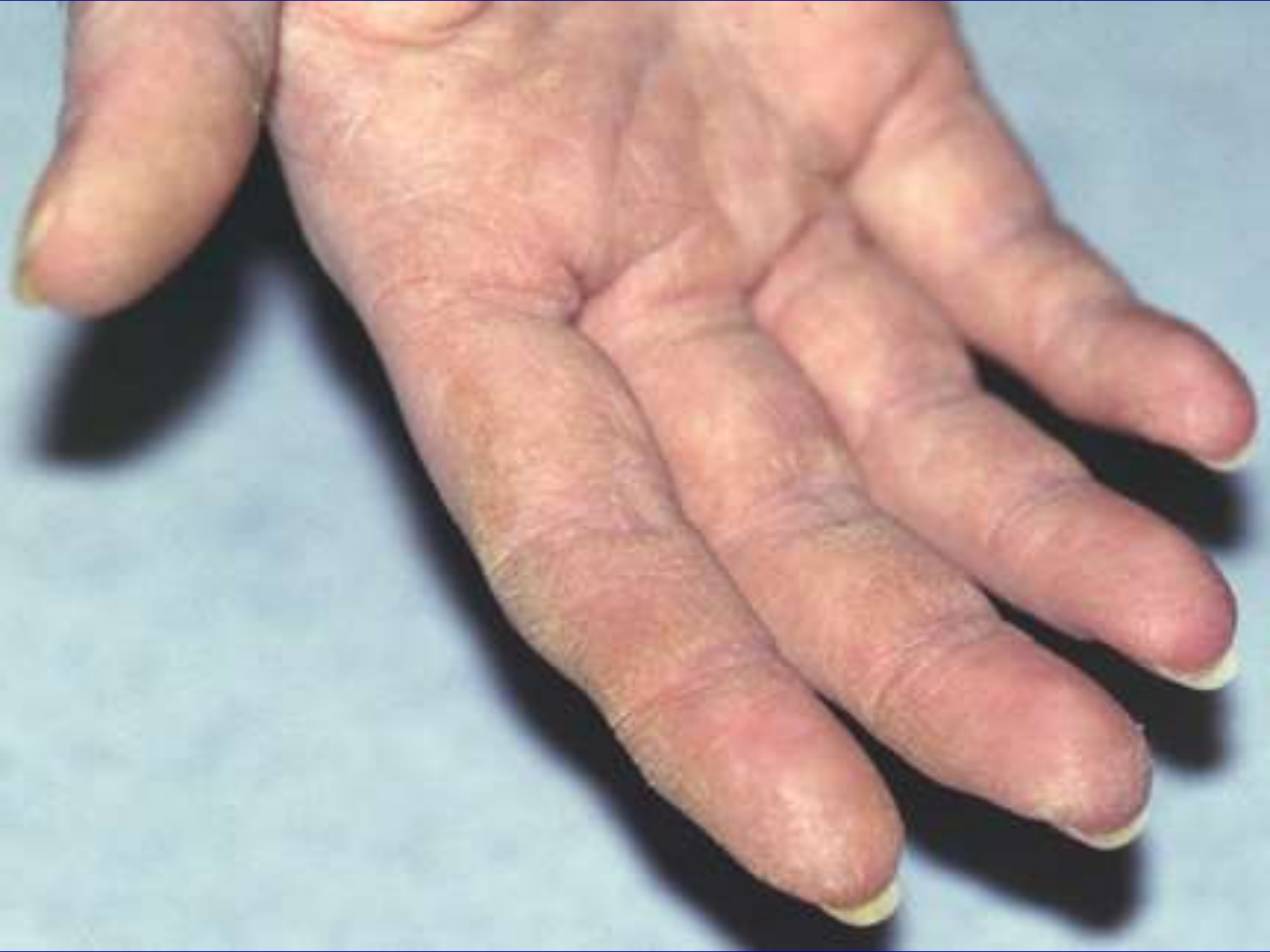


Anti-synthetase Syndrome Associated Capillaritis

Definition

- Autoimmune syndrome[1]
  - Inflammatory myopathy
  - Arthritis or arthralgias
  - Interstitial lung disease
  - Fever
  - Raynaud's phenomenon
  - Mechanic's hands
- Myositis specific antibodies[1]: aminoacyl-transfer RNA synthetases

1. Katzap E, Barilla-Barca ML, Marder G. Curr Rheumatol Rep 2011;13:175-181.



Proposed[2] Criteria

- An antisynthetase antibody plus two major criteria
- One major criteria plus two minor criteria
- Major criteria:
  1. ILD
  2. Polymyositis or dermatomyositis
- Minor criteria:
  1. Arthritis
  2. Raynaud's phenomenon
  3. Mechanic's hands

2. Solomon J, Swigris JJ, Brown KK. J Bras Pneumol 2011;37:100-109.

Antisynthetase antibodies[3]

- Myositis specific antibodies
- Aminoacyl-tRNA-synthetases
- 8 antibodies described
  - Anti-Jo-1
  - PL-7
  - PL-12 (alanyl-tRNA synthetase)
  - Others: EJ, OJ, KS, Zo and YRS

3. Labirua A and Lundberg IE. Current Opinion in Rheumatology 2010;22:633-638.

Table 1—*Prevalence of Antisynthetase Antibodies in PM/DM

MSAs (Anti-tRNA Synthetases)	Antigen	Prevalence
Jo-1	Histidyl-tRNA synthetase	15–20%
PL-7	Threonyl-tRNA synthetase	5–10%
PL-12	Alanyl-tRNA synthetase	< 5%
EJ	Glycyl-tRNA synthetase	5–10%
OJ	Isoleucyl-tRNA synthetase	5%
KS	Asparaginyl-tRNA synthetase	< 5%
Zo	Phenylalanyl-tRNA synthetase	< 1%
YRS	Tyrosyl-tRNA synthetase	< 1%

*Adapted from Table 1 in the article by Mimori et al.[1]

4. Kalluri M, Sahn SA, Oddis CV et al. CHEST 2009;135:1550-1556.

Clinical Features

Manifestation	Incidence, %	Subsets	Comments
ILD	70-90	NSIP, COP, DAD, UIP	May precede myositis
Myositis	78-91	Acute, subacute, late onset, subclinical	Jo-1: PM>DM PL-7 and PL-12: Amyopathic
Raynaud's	62	With or without Sclerodactyly	
Arthropathy	64-83	Arthralgia, polyarthritis	
Fever	20	No pattern	
Mechanic's hands	17-71	Hyperkeratosis	

5. Watanabe K, Handa T, Tanizawa K et al. Respiratory Medicine 2011;105:1238-1247.

Anti-PL12 Phenotype[4]

- Median age: 51 years (range 21 to 87)
- 81% females
- 52% African-American
- 90% with associated connective-tissue disorder
  - 32% PM
  - 19% DM
  - 16% Overlap syndromes

Anti-PL12 Phenotype

- 90% with ILD
- ILD preceded CTD diagnosis in 53% of cases
  - UIP 46%
  - COP 18%
  - NSIP 18%
- Less fever and mechanic's hands; variable Raynaud's

Anti-PL12 Phenotype…

- Radiographic findings:
  - Interlobular septal thickening (73%)
  - Traction bronchiectasis (59%)
  - Honeycombing (41%)
  - Ground-glass opacities (36%)
  - Lower-lobe predominance (82%)

1-3 Therapy

- Glucocorticoids (1 mg/kg)
- Other:
  - Cyclophosphamide
  - Azathioprine
  - Mycophenolate mofetil (MMF)
  - Cyclosporine, tacrolimus
  - Rituximab
  - Intravenous immunoglobulin (IVIG)

Unique features in this case

- Capillaritis and diffuse alveolar hemorrhage
- 3 case reports [6]
  - Paraneoplastic dermatomyositis [7]
  - 2 Polymyositis (one patient with anti-Jo1 Ab)
- No prior association between PL-12 antibodies and capillaritis

6. Do-Pham G, Pages C, Picard C et al. British Journal of Dermatology 2010;163:208-234.

7. Schwarz MI, Sutarik JM, Nick JA et al. Am J Respir Crit Care Med 1995;151:2037-2040.

Clinical Pearls

- Antisynthetase syndrome has a high prevalence of ILD
- Patients with ILD should be screened for the presence of antisynthetase antibodies
- ILD can precede the presence of myositis and other symptoms
- Anti-PL12 phenotype can be associated with capillaritis and diffuse alveolar hemorrhage

Case 4

Clinical Presentation

- 53 y/o AAF
- Progressive DOE for 1 week
- Fever
- hemoptysis

Clinical Presentation

- PMH:
  - CKI
  - SLE (pleuritis and pericarditis)
  - DVT/PE
  - A Fib
  - Hepatitis B
  - Sickle cell trait
- PSH
  - C-section
- SHX:
  - 1ppd smoker
  - + Crack cocaine

Clinical Presentation

- Medications:

  Colace
  Warfarin
  Oxycodone
  Lisinopril
  Plaquenil
  Prednsione (10mg daily)
  Carvedilol
  Ambien

Clinical Presentation

- PE:

  VS: T: 100.6; P: 92; RR: 20; BP: 155/89;

  O2 sats 96% on 4 LPM

  Lung: Bibasilar crackles (non-velcro)

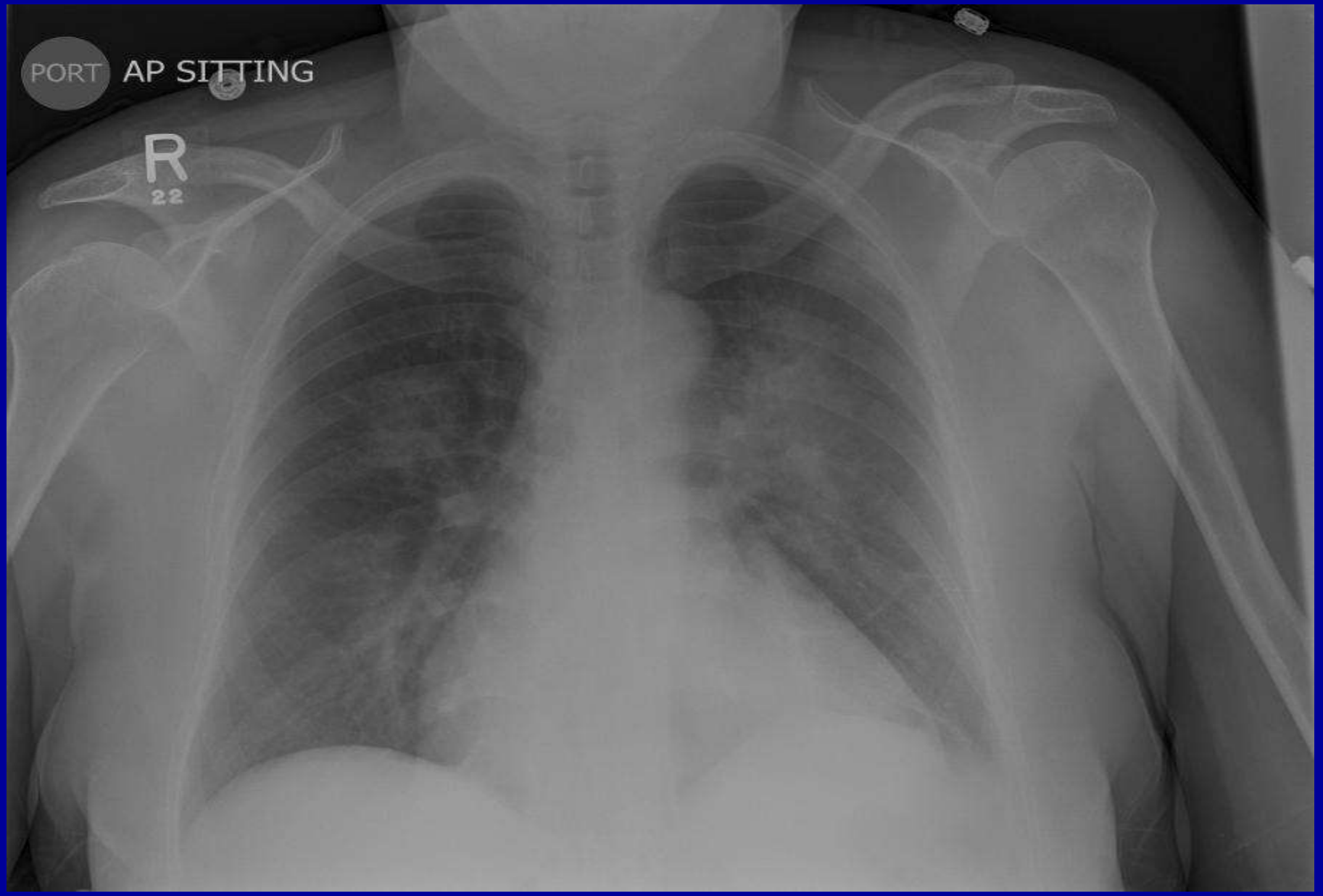
  Remainder of PE unremarkable

Clinical Presentation

- LABS
  - U/A: > 300 protein, 7 RBC
  - INR 0.93
  - WBC: 10
  - HgB: 9.7
  - PLTs: 609
  - Cr: 2.2

Clinical Presentation

- ANA: 1:1280 speckled
- Ds DNA 30.6 (<30)
- RO: 141.7 EU
- LA: 121.4 EU
- Anti- Smith: 0.9 EU
- C3: 155 mg/dl
- C4: 79.2 mg/dl
- Cryoglobulins: negative
- ANCA: 1:1280
  perinuclear staining
- + MPO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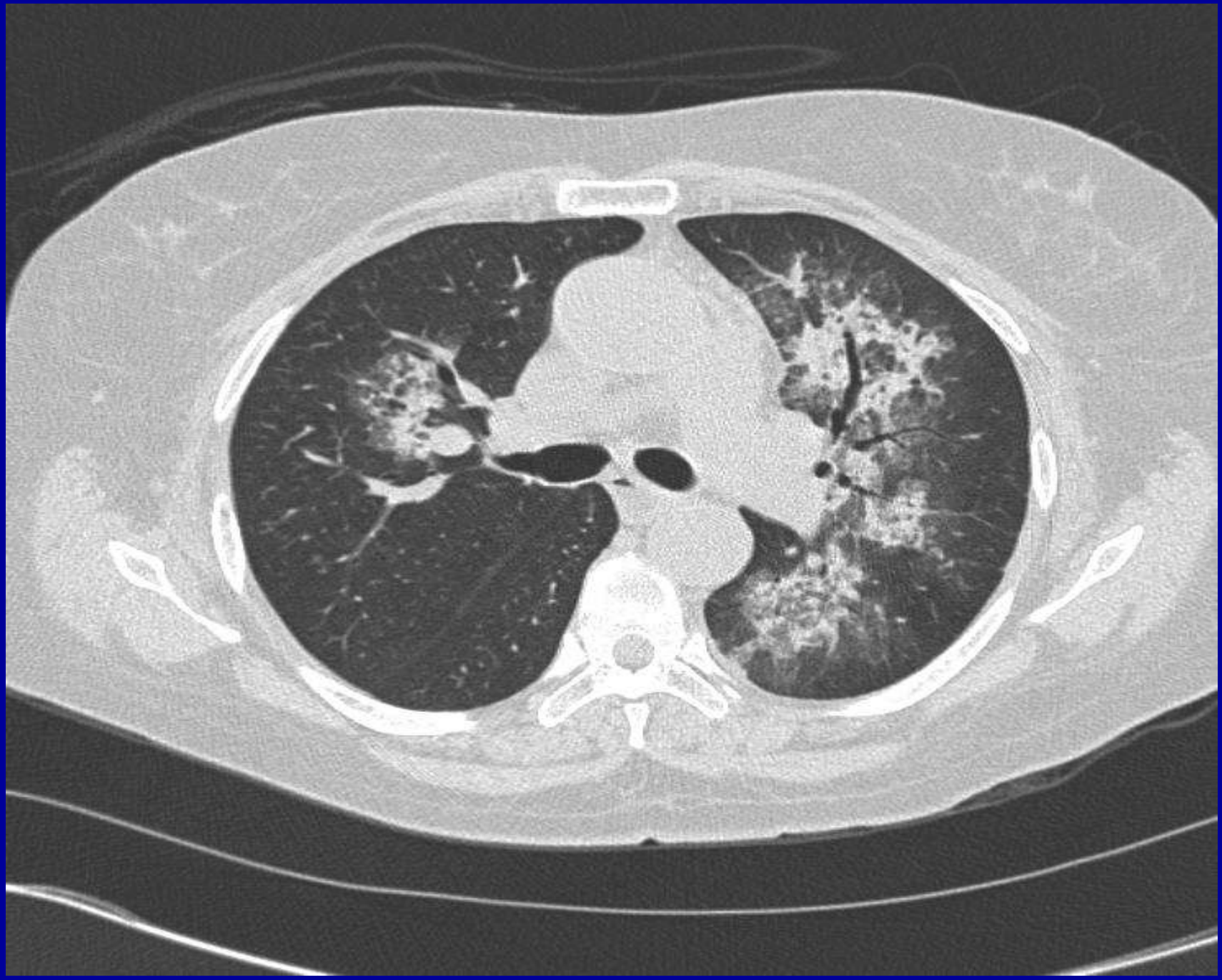


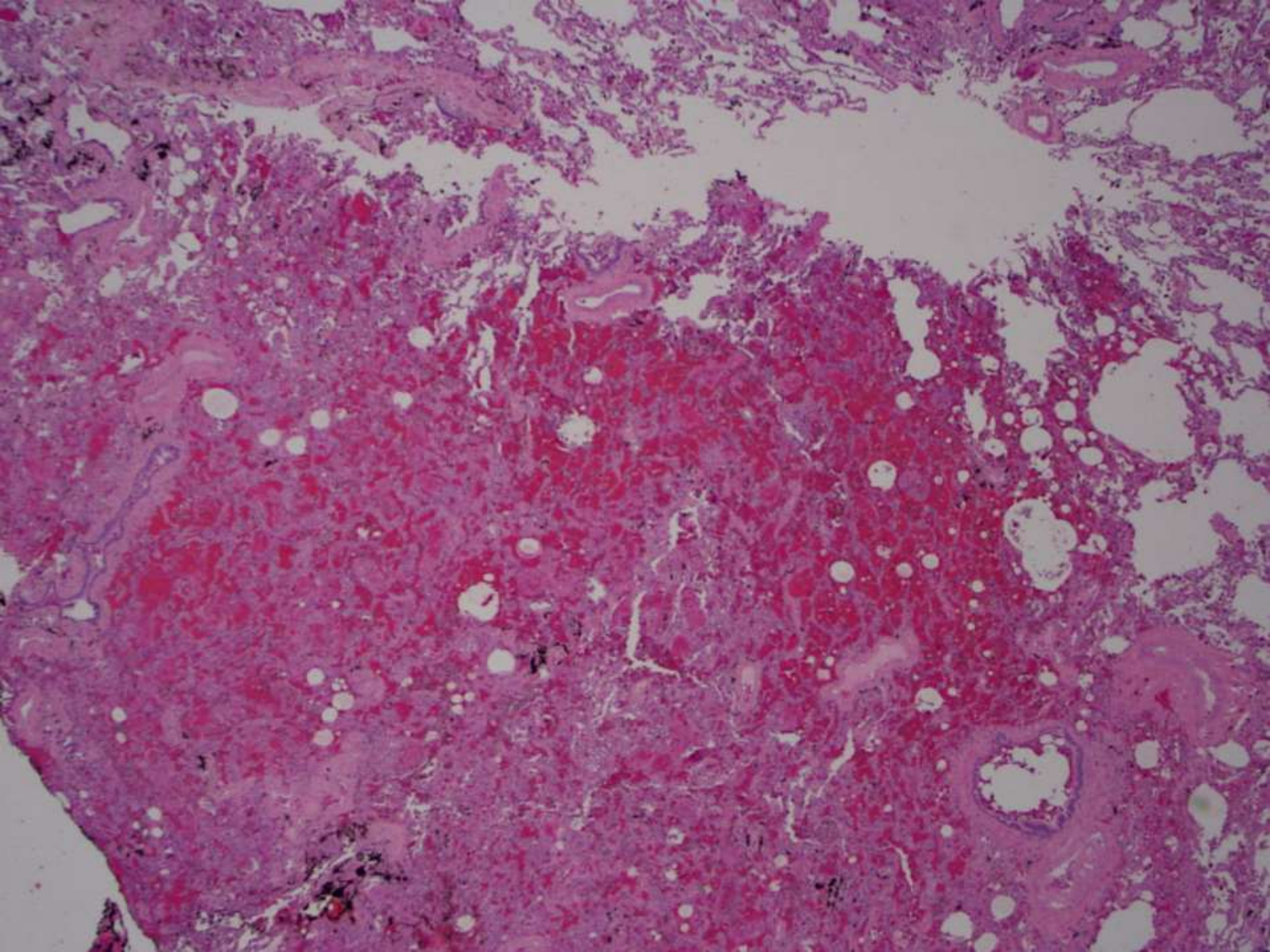
Clinical Presentation

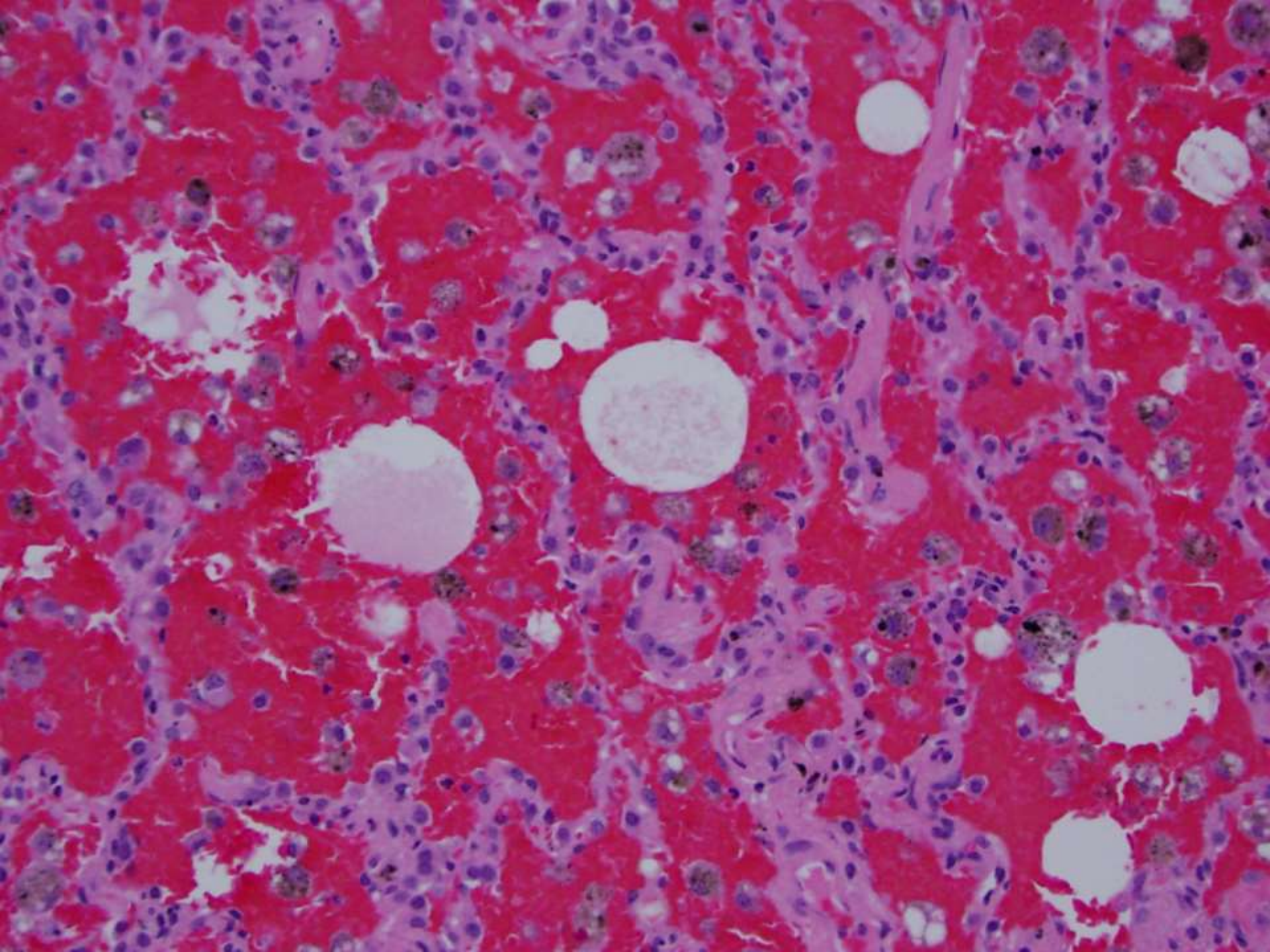
- Bronchoscopy with BAL

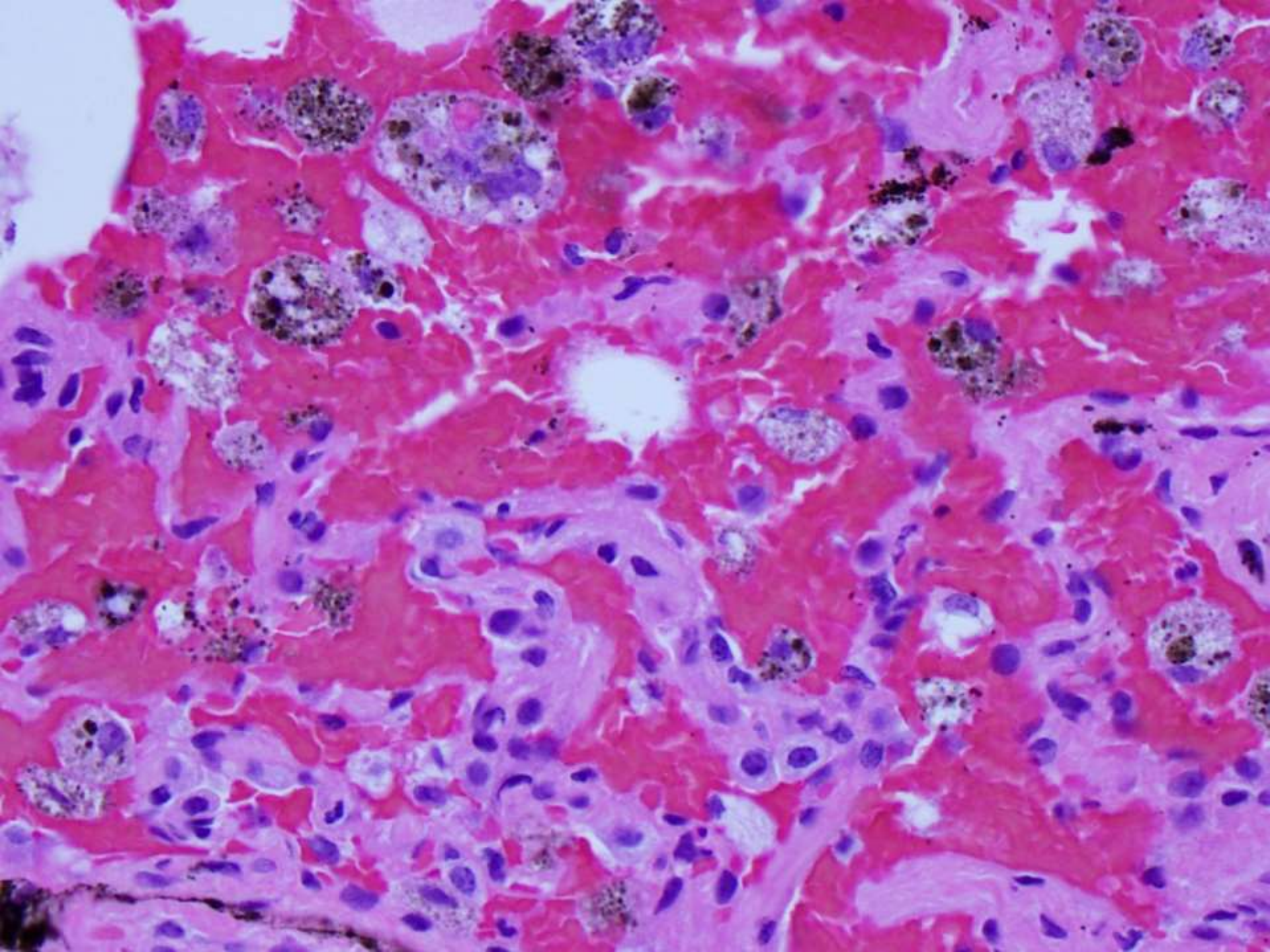
  - progressive bloody return

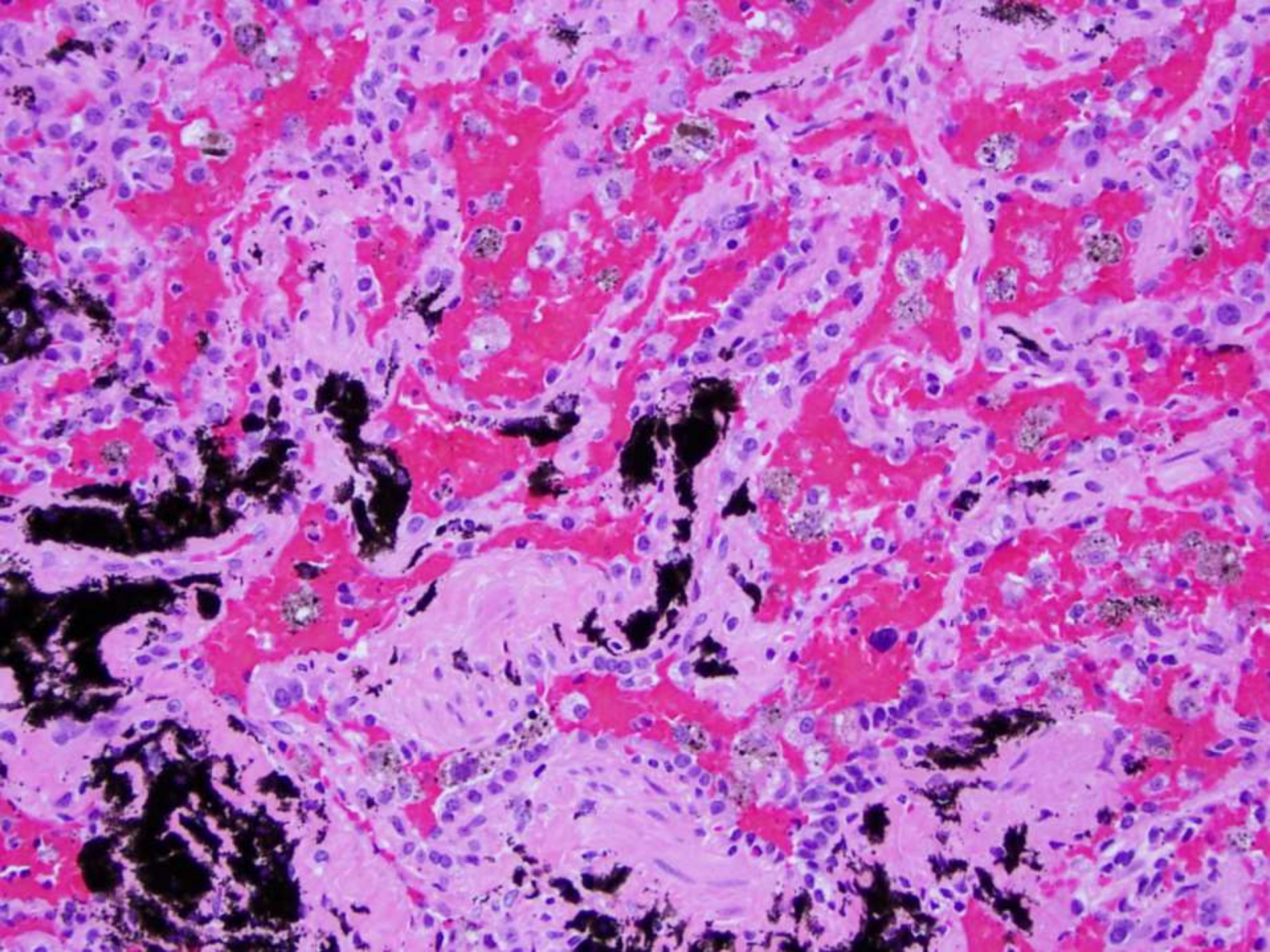
  - cytology: abundant hemosiderin laden macrophages

  - micro studies negative including respiratory viral PCR









Causes of Diffuse Alveolar Hemorrhage Syndromes (DAH)
Based on Histologic Appearance

Capillaritis

Systemic vasculitides
- Wegener's granulomatosis
- Microscopic polyangiitis
- Henoch-Schoenlein purpura
- Cryoglobulinemia
- Behcet's syndrome

Collagen vascular diseases
- Systemic lupus erythematosus*
- Polymyositis
- Rheumatoid arthritis
- Scleroderma
- Mixed connective tissue disease

Other
- Isolated pulmonary capillaritis
- Goodpasture's syndrome*
- Primary antiphospholipid syndrome
- Pauci-immune glomerulonephritis
- Autologus bone marrow transplant
- Lung transplant rejection
- Idiopathic pulmonary fibrosis
- Infective endocarditis
- Retinoic acid syndrome
- Propylthiouracil
- Phenytoin

Bland hemorrhage
- Goodpasture's syndrome*
- Systemic lupus erythematosus*
- Idiopathic pulmonary hemosiderosis
- Severe coagulopathies
- Mitral stenosis
- Trimellitic anhydride inhalation
- Penicillamine, nitrofurantoin, amiodarone
- Rapamycin

Diffuse alveolar damage
- ARDS (any cause)
- Cytotoxic drug toxicity
- Polymyositis
- Systemic lupus erythematosus
- Crack cocaine inhalation
- Infections in the immunocompromised host

Miscellaneous conditions
- Lymphangioleiomyomatosis
- Tuberous sclerosis
- Pulmonary veno-occlusive disease
- Pulmonary capillary hemangiomatosis
- Pulmonary infarction

*Both bland hemorrhage and DAH with capillaritis can be seen in these conditions.

Acute Lung Injury with Crack

- Typically develops within 1-48 hours
- 25% of users with develop respiratory symptoms including fever, cough, nonspecific chest pain, hemoptysis, back pain, hyperpnea, dyspnea, melanoptysis, wheezing
- Diffuse pulmonary infiltrates, eosinophilic pleural effusions, acute lung injury pattern
- Eosinophilia

Chronic Exposure to Crack

- Pulmonary fibrosis
- Diffuse alveolar hemorrhage
- Hemosiderosis
- Pulmonary infarction
- Eosinophilic interstitial lung disease
- Bullous emphysema
- Medial artery hypertrophy
- Noncardiogenic pulmonary edema
- Increased risk of pneumonia, multifactorial problem

Microenvironment and Cocaine

- Cocaine inhibits alveolar macrophages ability to kill most bacteria and tumor cells in vitro
- Cocaine users are unable to kill bacteria using nitric oxide as an antibacterial effector molecule
- These changes may predispose to increase pulmonary infections in these users
- Marijuana has similar adverse effects. Inhibits phagocytosis Staph aureus

Crack Pulmonary Injuries I

- Barotrauma, ischemia, provocation of inflammatory damage, and direct cellular toxicity
- Barotrauma is the result of Valsalva maneuver after inhalation and the forceful inhalation of air into partners. Pneumothoraces, pneumomediastinum, and pneumopericardium
- Ischemia is the result of the vasoconstrictive properties
- Severe bronchospasm in patients with preexisting asthma

Treatment of Crack Lung

- Need to make history of exposure
- Supportive
- Role of steroids unproven, helpful in those patients with bronchospasm
- Screen for HIV and concomitant drugs
- Drug treatment

ANCA in DAH Diagnosis

- Antineutrophilic cytoplasmic antibodies (ANCA)first described in 1982 in association with pauci-immune glomerulonephritis
- ANCA described in association with GPA in 1985
- Subsequently described in microscopic polyangitis (MPA) and limited renal vasculitis

Davies DJ, et al. Br Med J 1982; 285:606

Van der Woude FJ, et al. Lancet 1985; 1:425

Falk RJ, et al. N Engl J Med 1988; 318:1651

TABLE 22–1 Systemic Vasculitides*

Name	*Vasculitic Lung Involvement*	*ANCA Findings*
Large vessel vasculitis		
Giant cell (temporal) arteritis	Rare	No
Takayasu arteritis	Frequent	No
Medium-sized vessel vasculitis		
Polyarteritis nodosa	Rare	No
Kawasaki disease	No	No
Small vessel vasculitis		
WG	Frequent	PR3-ANCA
CSS	Frequent	MPO-ANCA or PR3-ANCA
MPA	Frequent	MPO-ANCA or PR3-ANCA
Henoch-Schönlein purpura	No	IgA-possible
Essential cryoglobulinemic vasculitis	No	No

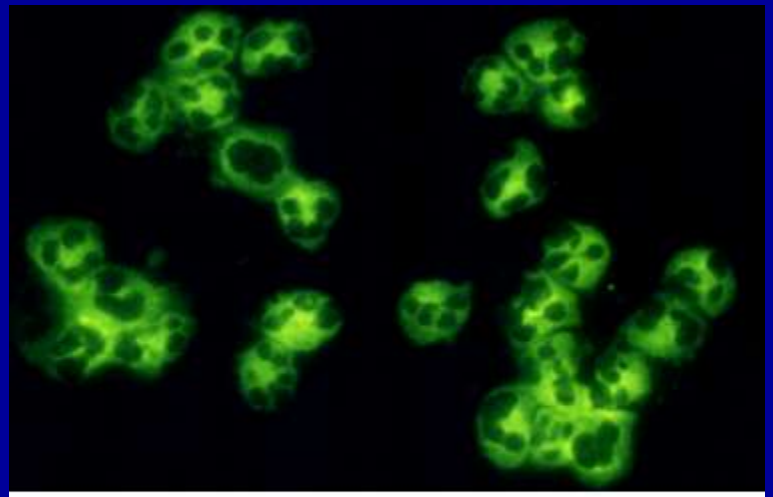
*Systemic vasculitides as defined by the 1992 Chapel Hill international consensus conference on the nomenclature of systemic vasculitis.[8] ANCA = antineutrophil cytoplasmic antibodies; CSS = Churg-Strauss syndrome; MPA = microscopic polyangiitis; MPO = myeloperoxidase; PR3 = proteinase 3; WG = Wegener's granulomatosis.

ANCA Testing

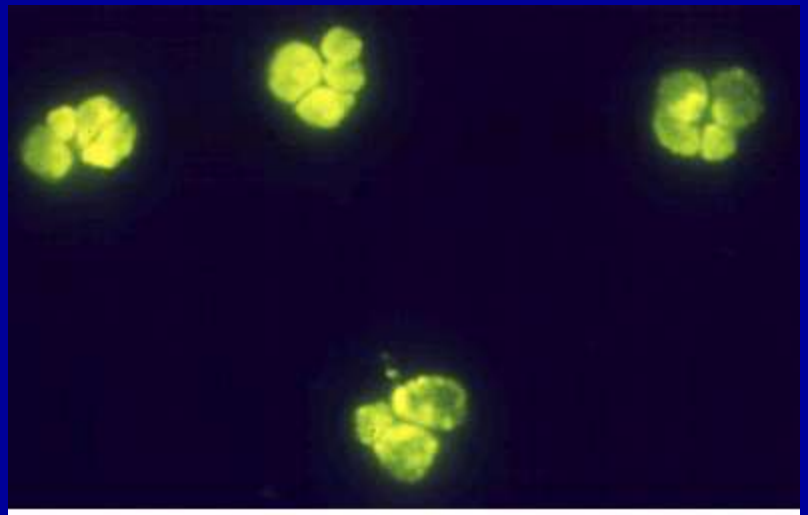
- Indirect immunofluorescence assay (IIA) is more sensitive
- Enzyme link immunosorbent assay (ELISA) is more specific
- Best used in conjunction with IIA for screening and ELISA for confirmation
- Two relative antigens in vasculitic diseases, proteinase 3 (PR3) and myeloperoxidase (MPO)
- Antigens are found in neutrophils and monocytes
- PR3-ANCA and MPO-ANCA

Immunofluorescence Patterns In **Vasculitis**

- Sera from patients with suspected ANCA related **vasculitis** are incubated in ethanol fixed neutrophils
- Two distinct patterns of fixation identified, c-ANCA with cytoplasmic pattern and p-ANCA with perinuclear pattern
- c-ANCA pattern is **typically** associated with antibodies against PR3
- p-ANCA is **typically** associated with antibodies against MPO



C-ANCA pattern Demonstration of **cytoplasmic** antineutrophil cytoplasmic antibodies (C-ANCA) by indirect immunofluorescence with normal neutrophils. There is heavy staining in the cytoplasm while the multilobulated nuclei (clear zones) are nonreactive. These antibodies are usually directed against proteinase 3 and most patients have Wegener's granulomatosis. Courtesy of Helmut Rennke, MD.



P-ANCA pattern Demonstration of **perinuclear** antineutrophil cytoplasmic antibodies (P-ANCA) by indirect immunofluorescence with normal neutrophils. Staining is limited to the perinuclear region and the cytoplasm is nonreactive. Among patients with vasculitis, the antibodies are usually directed against myeloperoxidase. However, a P-ANCA pattern can also be seen with autoantibodies against a number of other antigens including lactoferrin and elastase. Non-MPO P-ANCA can be seen in a variety of nonvasculitic disorders. Courtesy of Helmut Rennke, MD.

Cocaine and ANCA association

- >70% of illicit cocaine is cut levamisole
- Levamisole-contaminated cocaine is associated with ANCA vasculitis
- Largest series described (n=30)

  arthralgias (83%)
  skin lesions (61%)
  constitutional symptoms (72%)
  MPO-ANCA (100%)
  PR3-ANCA (50%)
  Leukopenia (28%)
  Abnormal U/A (27%)
  Pulmonary hemorrhage (7%)

McGarth MM, et al. Clin J Soc Nephrol 2011; 6:2799.

Case 5

Case Presentation

- 38 y/o AAF
- Chronic and progressive DOE
- DOE reproducible at 30 feet

Case presentation

- PMH:
  - BPAD
  - Endometriosis
  - +PPD s/p INH x 6 mos
  - CLB
  - Migraines
- PSH
  - Oophorectomy
- SHX:
  - 2ppd smoker

Case presentation

- Medications:

  Elavil
  Azithromycin
  Baclofen
  Citalopram
  Neurotonin
  Methocarbamol
  Topomax

Case presentation

- PE:

  VS: T: 98.3; P: 88; RR: 20; BP: 140/80;

  O2 sats 94% on 4 LPM

  Lung: Bibasilar crackles (non-velcro) with radiation to axilla

  Remainder of PE unremarkable

Case presentation

- PFTS:

  FVC 38% of pred.

  FEV1 34% of pred

  FEV1/FVC ratio 92% of pred

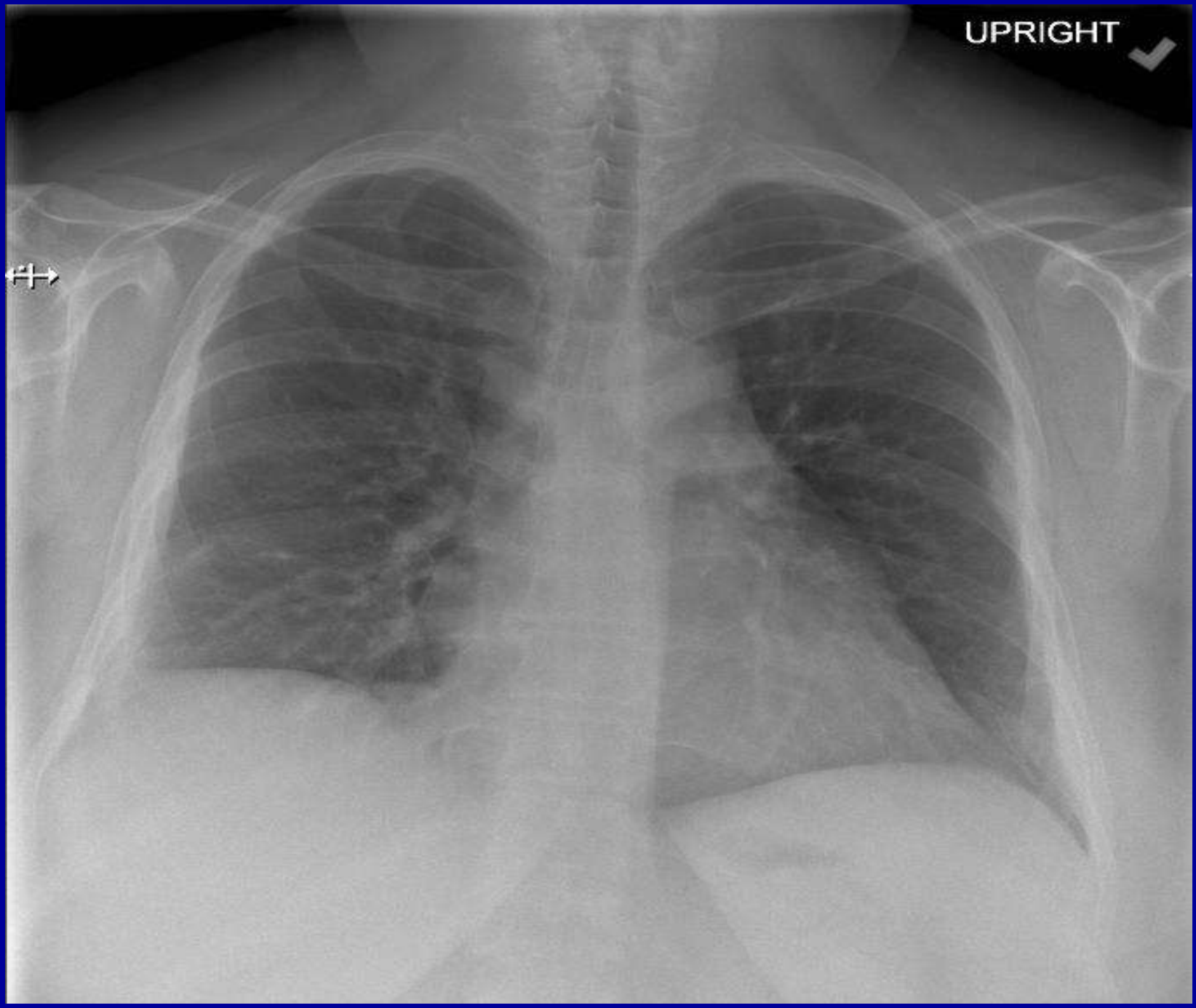
  DLCO 39% of pred

- 6 MW

  810 feet

  O2 desat to 84% on 4 LPM

  appropriate CV response to excercise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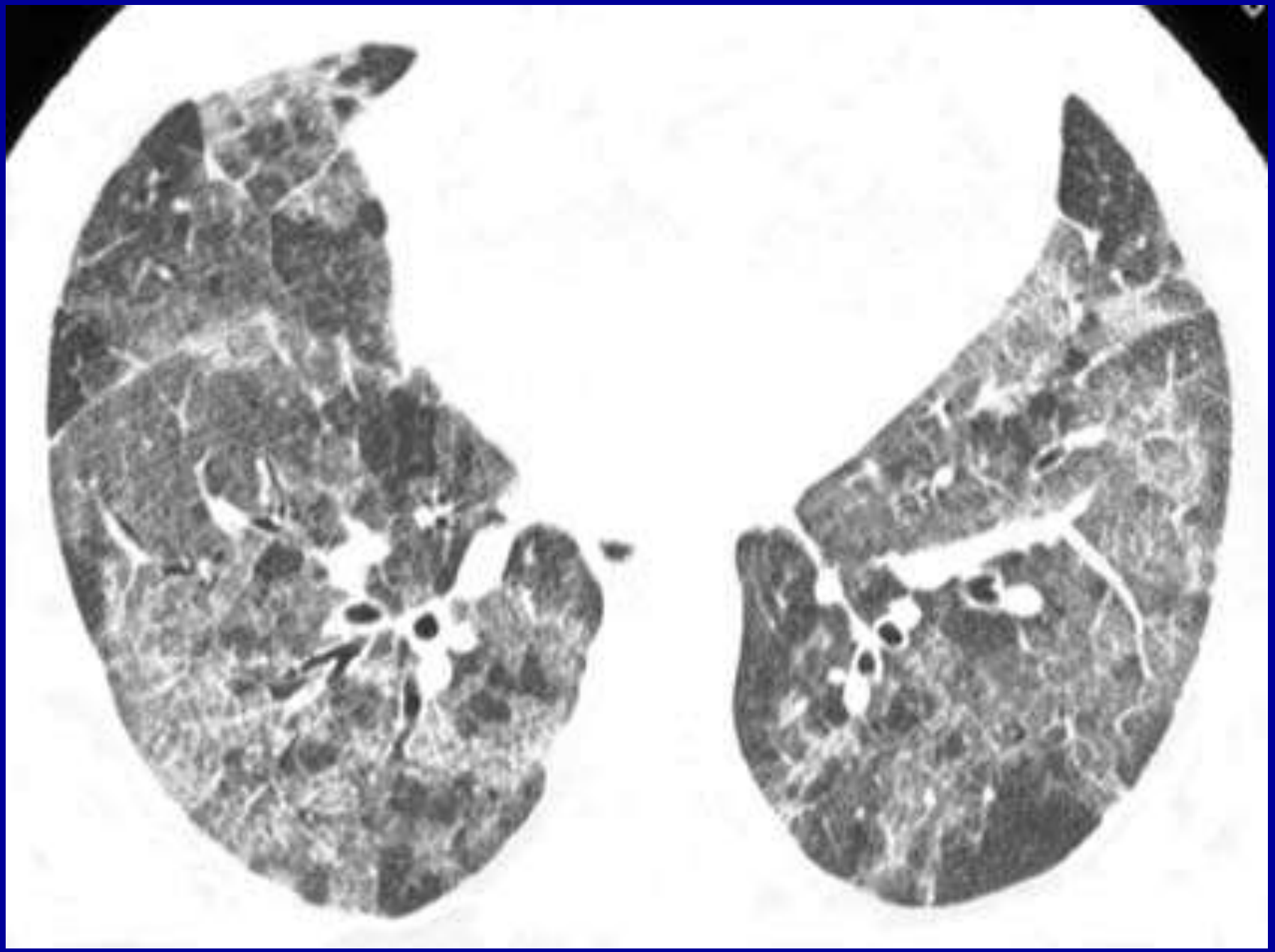

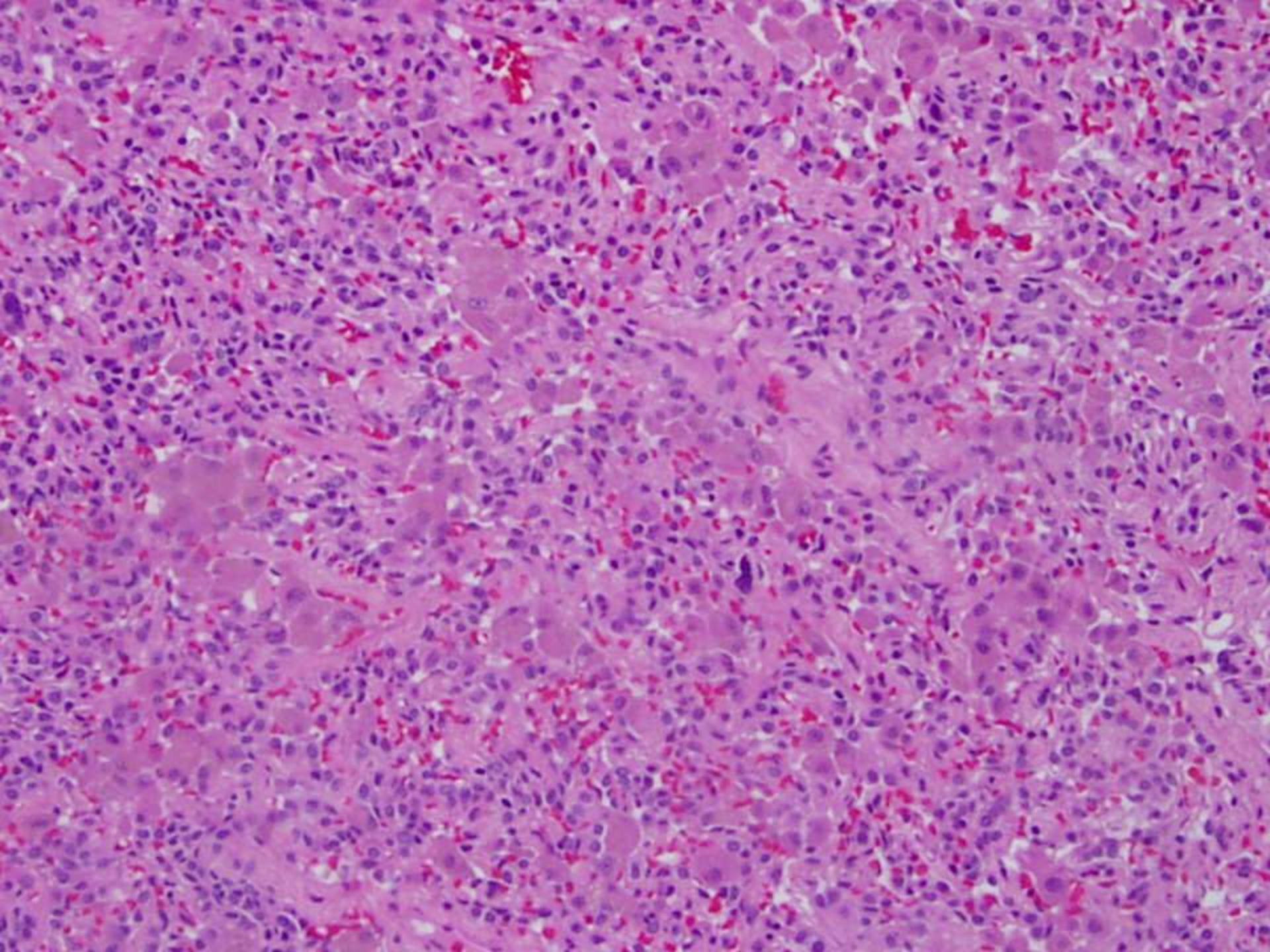

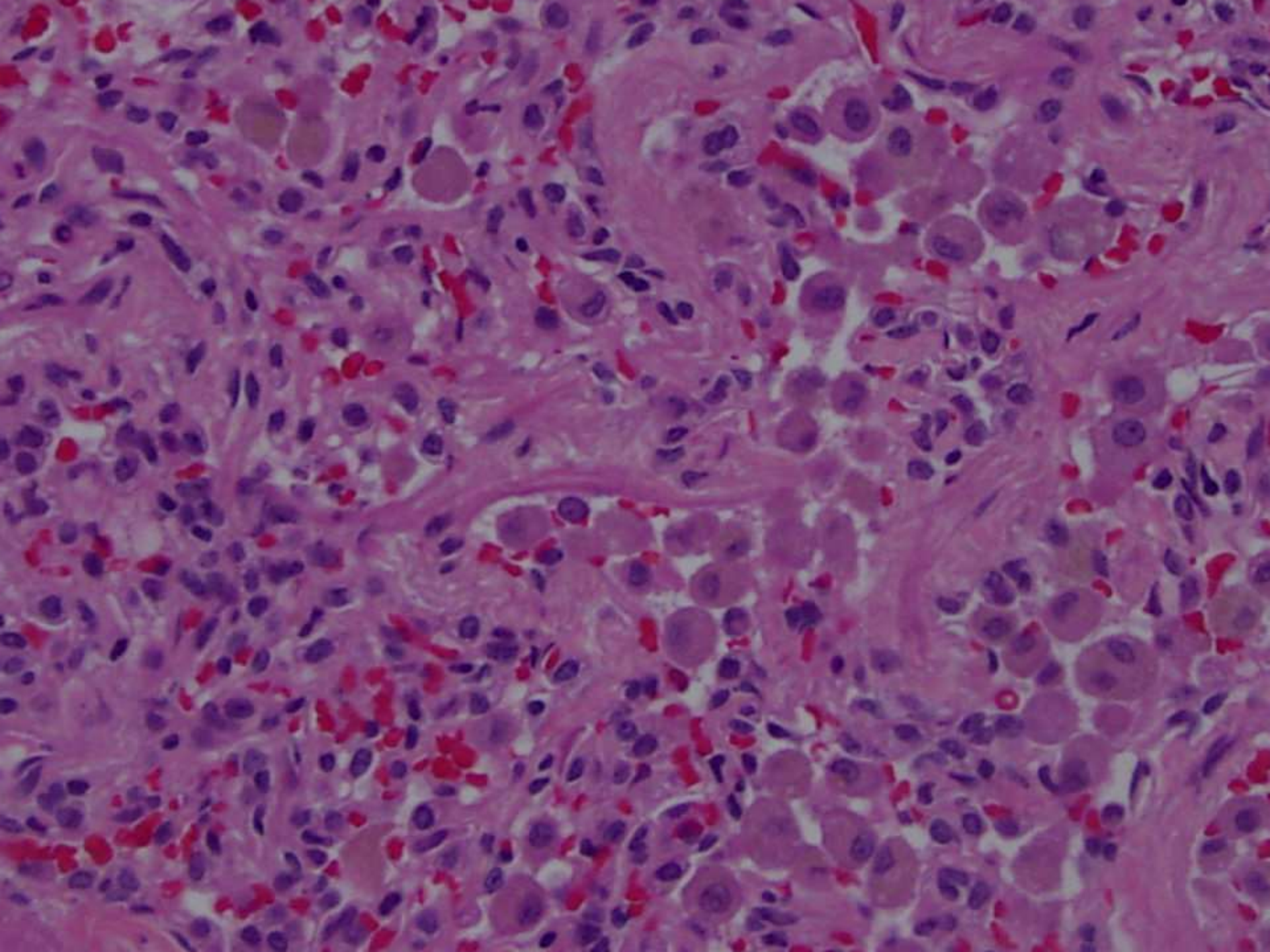

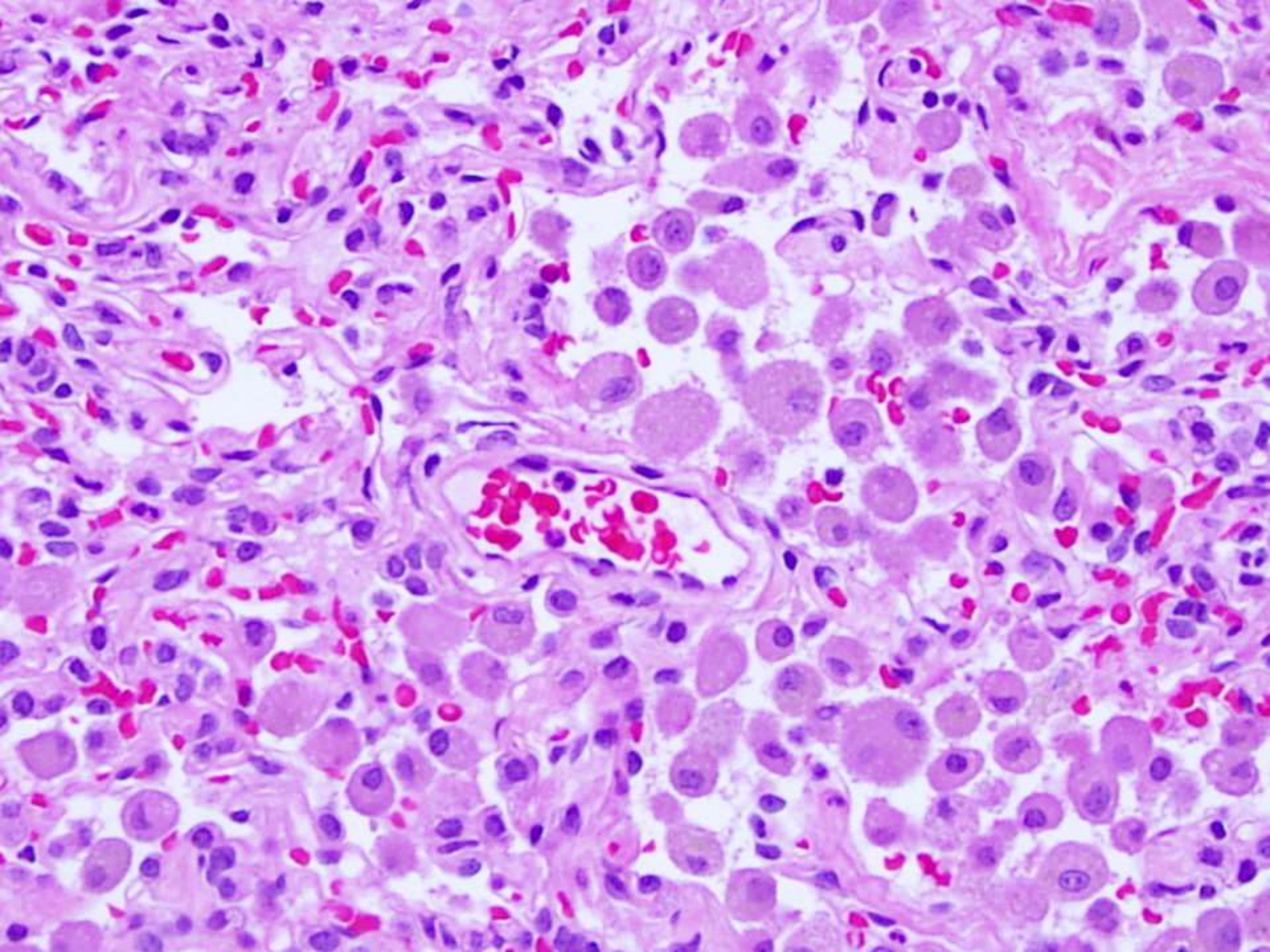

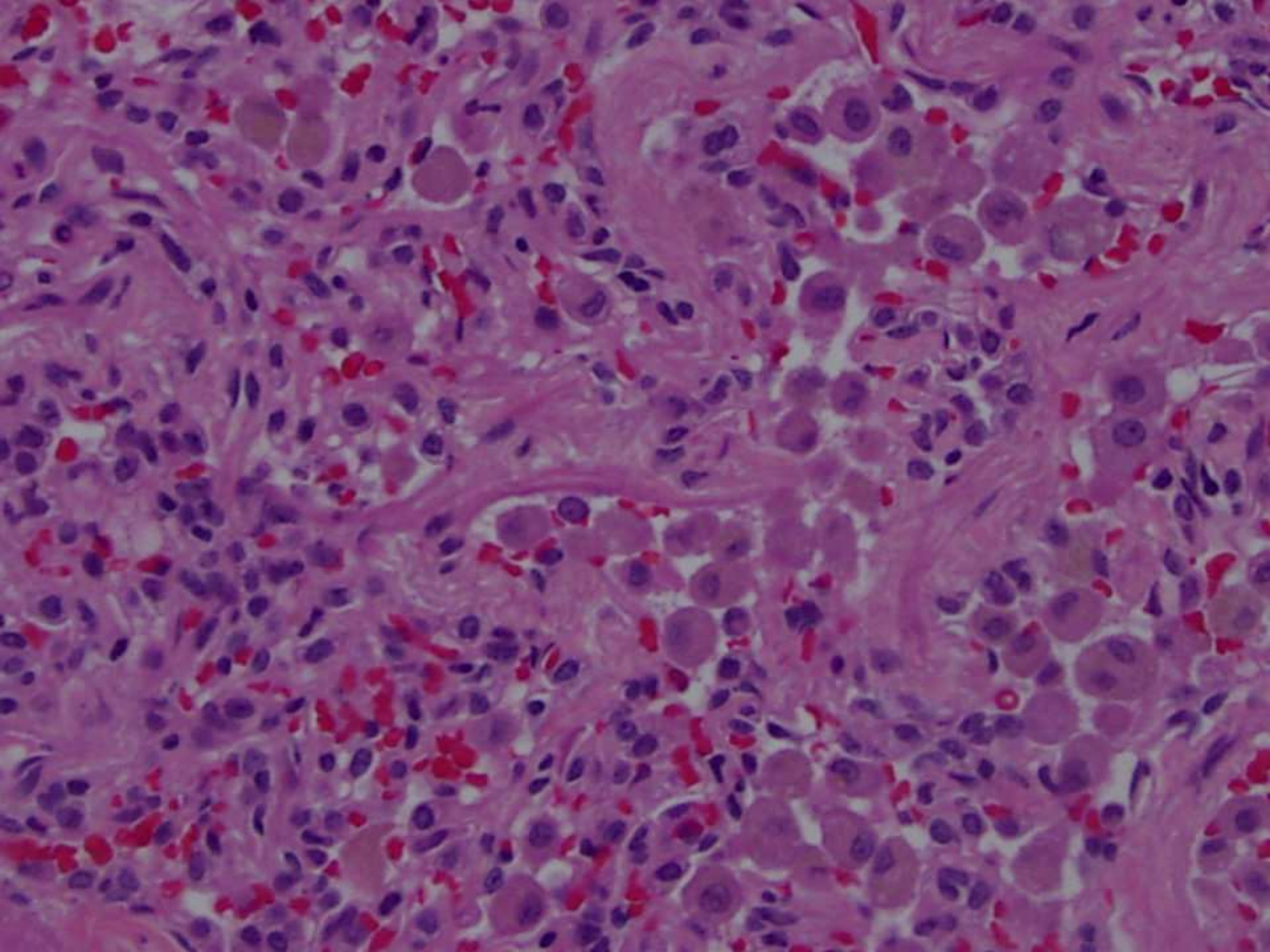

Desquamative Interstitial Pneumonia

	DIP	RBILD	PLCH	IPF
Smoking (%)	90	100	>90	41-83
Age	3rd-5th decades	3rd-5th decades	3rd-4th decades	Middle aged to older
Occurrence in children	Rare	NO	Rare	Rare
Onset	Insidious	Insidious	Insidious	Insidious
Presenting sxs	Dyspnea, cough	Dyspnea, cough	Dyspnea, cough	Dyspnea, cough
Crackles (%)	60	50	Usually absent	100
Clubbing (%)	Nearly 50	Rare	Rare	50-70
CXR	Interstitial, patchy ground-glass	Interstitial or normal	Interstitial/cystic or nodular, with basilar sparing	Interstitial, honeycombing basilar predominance
HRCT	Ground glass with lower lung predominance	Patchy ground glass	Nodules and cyst; basilar sparing	Subpleural honeycombing; basilar predominance
PFTs	Restrictive	Mixed defect to normal	Obstructive or restrictive	Restrictive
Treatment	Smoking cessation, steroids	Smoking cessation	Smoking cessation; ?steroids	None
Response of steroids	Good	Good	Fair	Poor
Prognosis	Good	Good	Fair	Poor
Complete recovery possible	Yes	Yes	Yes	No

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3. Lynch DA. *Radiol Clin* North Am 2002; 39:1153-1170
4. Ryu JH, et al *Eur Respir* J 2001; 17: 122-132
5. Ryu JH, et al. *Chest* 2005; 127:178-184
6. Katzenstein AA, Myers JL. *Am J Respir Crit* Care Med 1998; 157:1301-1315

Case 6

Case Presentation

- 53 year old male
- 6 months of progressive DOE and cough
- Dry cough
- 2 years treated for several bouts of “walking pneumonia” and bronchitis

Case presentation

- PMH:
  - hypothyroidism
  - hyperlipidemia
  - thrombocytopenia
  - NASH cirrhosis
- PSH:
  - chole
  - sinus surgery

Case presentation

- Medications:

  synthroid 100 mcg daily

  loratadaine 10 mg daily

  pravastatin 80 mg daily

  flonase prn

  naproxen prn

Social Hx:

life-long non-smoker; no ETOH or illicit drugs; no birds, asbestos, hot-tubs; humidifiers

Case presentation

- Family Hx:

  Sister deceased secondary to IPF and lung transplant complications

Case presentation

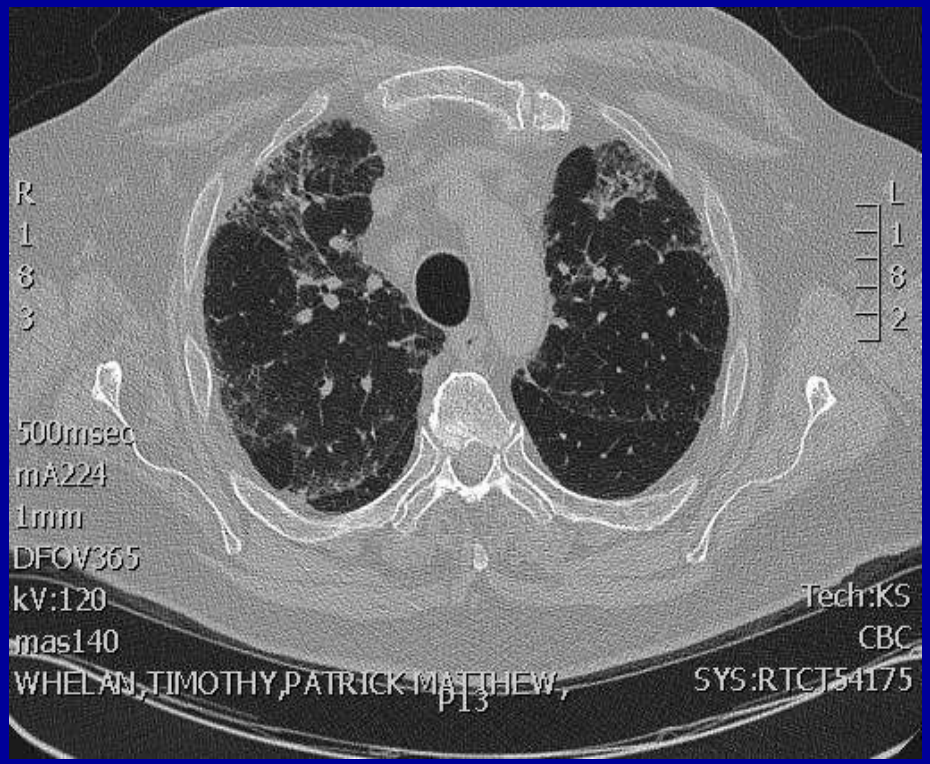
- PE: AF/ 76/ 18/ 140/79/ 94% on room air
- Pertinent positives:

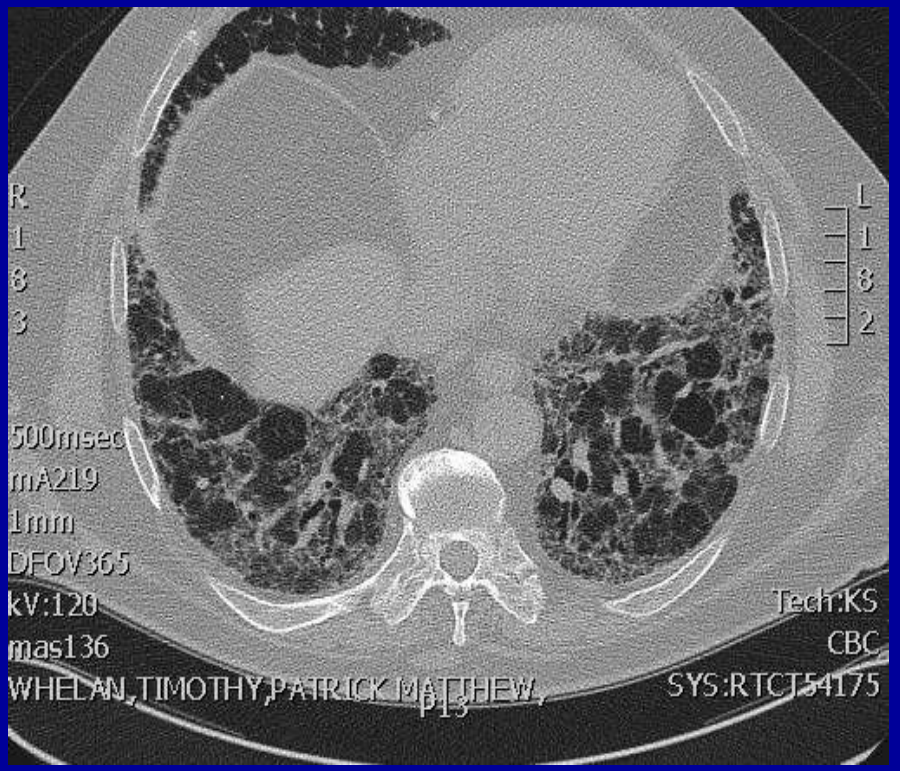
  bi-basilar crackles
- 6MWT on room air: O2 desaturation to 83%
- PFTS: FVC 58% of predicted; FEV1/FVC ratio 0.85; TLC 55% of predicted; DLCO 42%

Case presentation

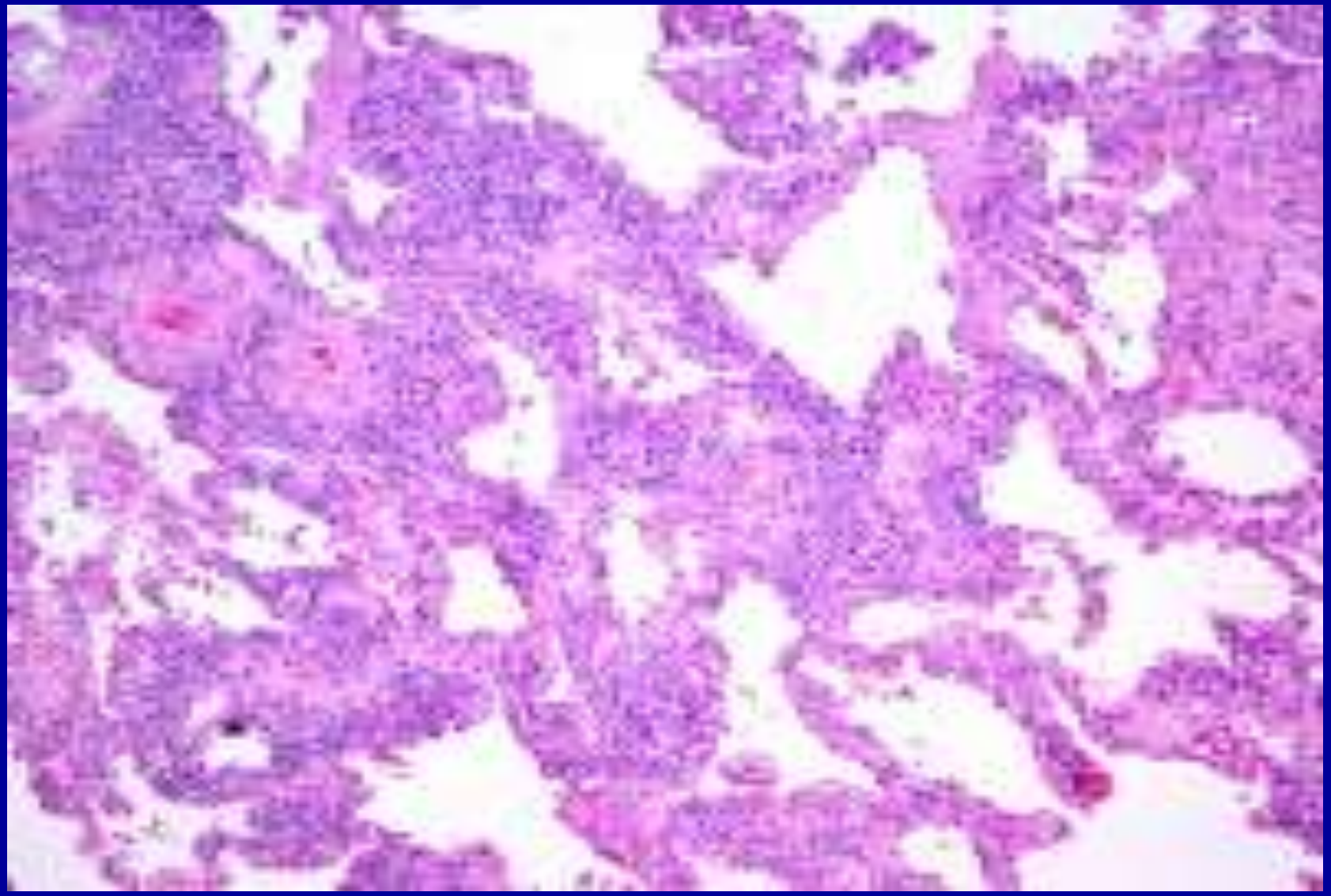
- ANA, RF, CCP negative

Case presentation





Case presentation



Case Presentation

- Diagnosis: Chronic Hypersensitivity pneumonitis
- More History: Prior home with water damage
- HP panel: positive for A flavus

Acknowledgment

- Ellen Riemer, MD (Dept. of Pathology, MSUC)