Interstitial lung diseases (AKA DPLDs)

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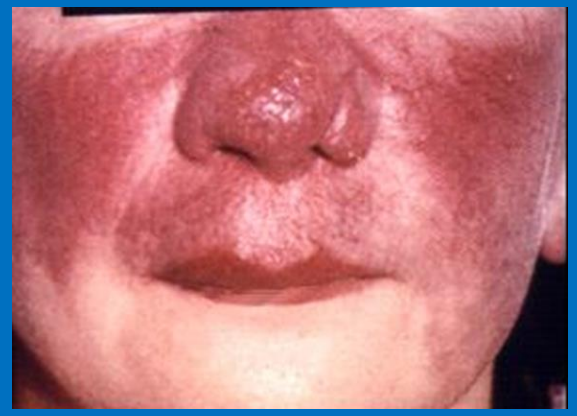
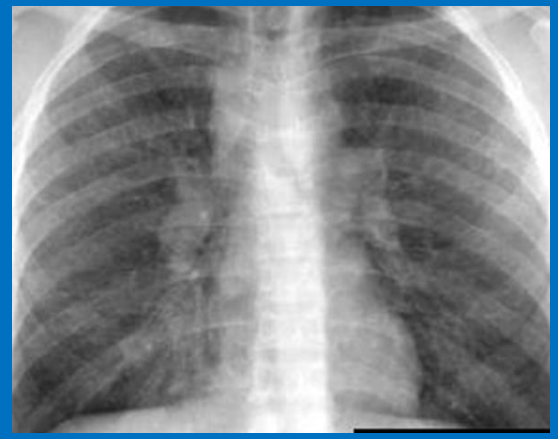
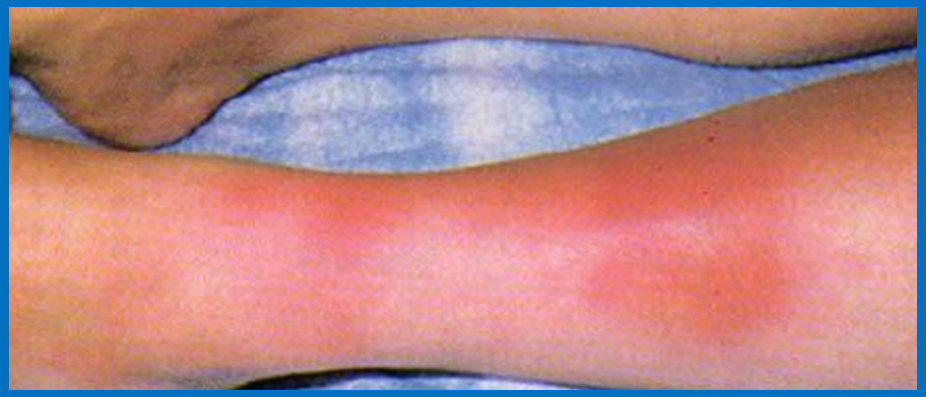
UA CoM-P

QUESTION 1.

30 yo female presents with cough of 6 weeks duration. She has also developed facial rash and skin lesions





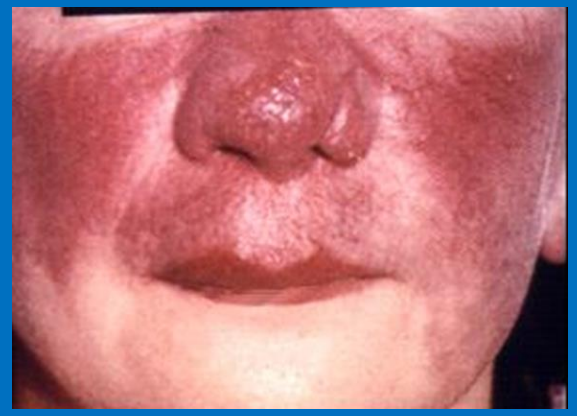


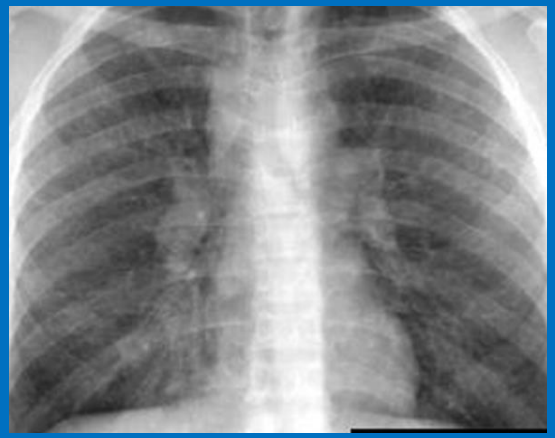
What is the most likely diagnosis?

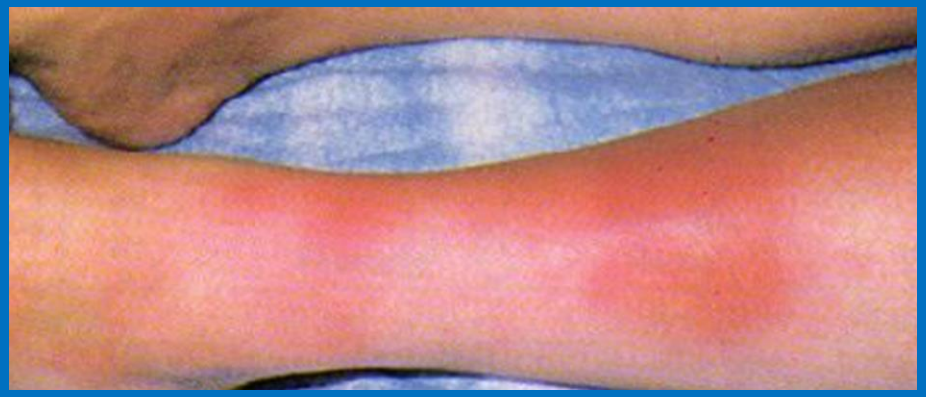
A. Lupus-related alveolar hemorrhage
B. Sarcoidosis with lung and skin involvement
C. Scleroderma related NSIP
D. Lymphoma with cutaneous manifestations
E. Valley fever complicating ILD

QUESTION 2.

You desire to have a biopsy supported diagnosis for this patient before starting therapy





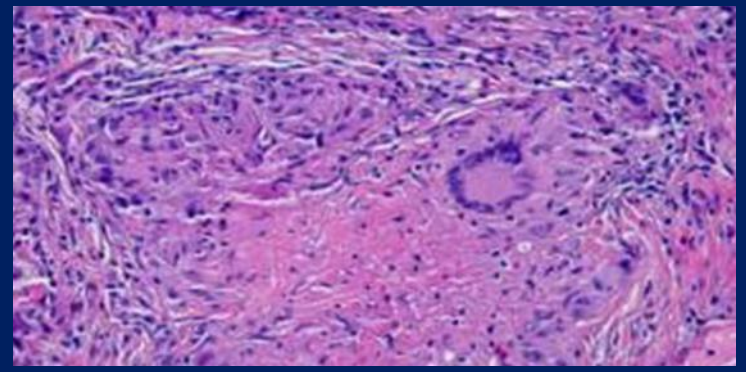


Which biopsy site is *least likely* to give you a diagnosis of sarcoid?

A. Facial rash
B. Lung parenchyma by bronchoscopic biopsy
C. Leg lesion
D. Mediastinal node by EBUS
E. Liver biopsy

Sarcoidosis

- Chronic inflammatory disorder
- Granulomatous inflammation
- Unknown etiology
- Diagnosis of exclusion
- Can affect any organ
- Steroids, MTX, AZA

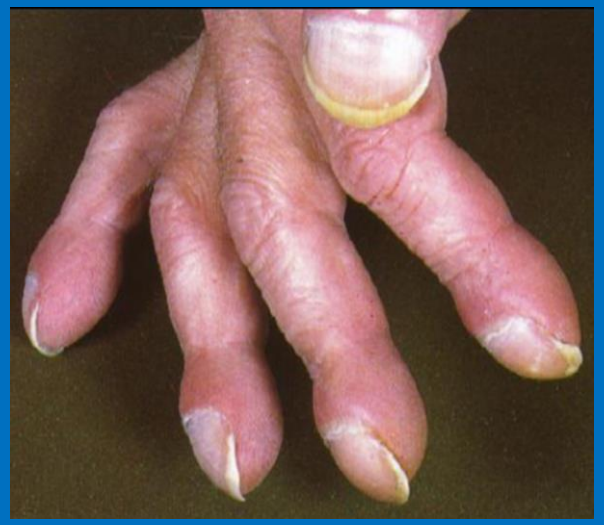


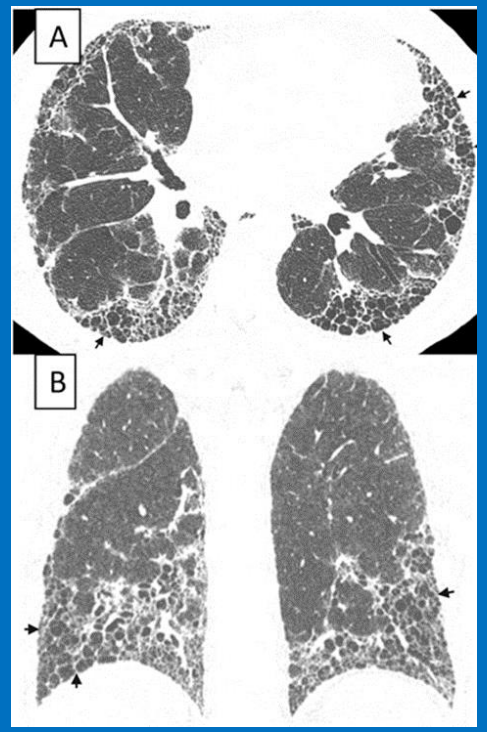
Infection	TB, Histoplasma, Coccy
Exposures	Beryllium exposure, Hypersensitivity pneumonitis
Systemic disease	Wegener's granulomatosis SLE, psoriatic rashes , CVID
Malignancy	Lymphangitic spread of primary Lymphoma

Eye	Uveitis, iritis, conjunctival
Cardiac	Dysrhythmia, CHF
Neurosarcoidosis	CNS – meningitis, Cranial nerves, peripheral neuropathy
Calcium metabolism	Hypercalcemia, Hypercalciuria
Lymphoreticular	Lymphadenopathy Hepato- and spleno-megaly

QUESTION 3.

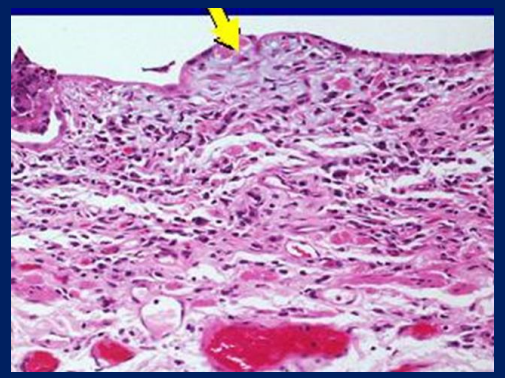
56 yo male presents with progressive dyspnea and decreasing exercise tolerance over a 3-year period. He denies occupational and environmental-related exposures. Meds- diltiazem, sildenafil, testosterone, aspirin. He has bibasilar crackles and nail changes





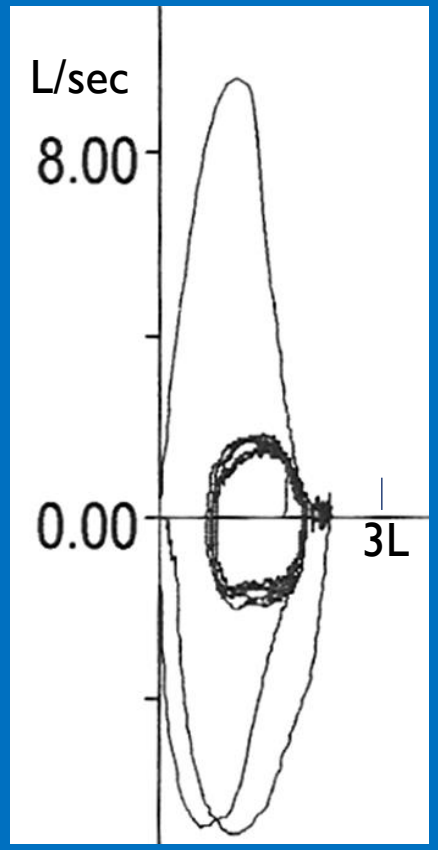
What is the most likely diagnosis?

A. Cystic fibrosis
B. Lupus-associated ILD
C. Hypersensitivity pneumonitis
D. Idiopathic pulmonary fibrosis
E. Idiopathic nonspecific interstitial pneumonia



QUESTION 4.

You get pulmonary function tests and a six minute walk. He requires 1 liter of oxygen while ambulating. You desire to treat your patient for IPF.



	Pre-Drug Measured	Pre-Drug % Predicted
Drug Name		
FVC	2.22	59
FEV1	1.98	67
FEV1/FVC	89.10	113
FEF25-75%	5.85	173
FEFmax	9.64	
FIFmax	6.94	
FEF50/FIF50	142.58	
TET	6.75	
	SINGLE-BREATH DIFFU	
Dsb	9.70	30
DsbHb	9.70	30
D/VAsb	3.22	65
D/VAsbHb	3.22	65
VAsb	3.01	54

What are the next steps?

A. He has severe *Restriction* and should be treated with steroids
B. He has severe *Obstruction* and should be treated with steroids
C. He has severe *Restriction* and should be treated with pirfenidone
D. He should only be treated with oxygen therapy at this time because his FVC is only mildly reduced

QUESTION 5.

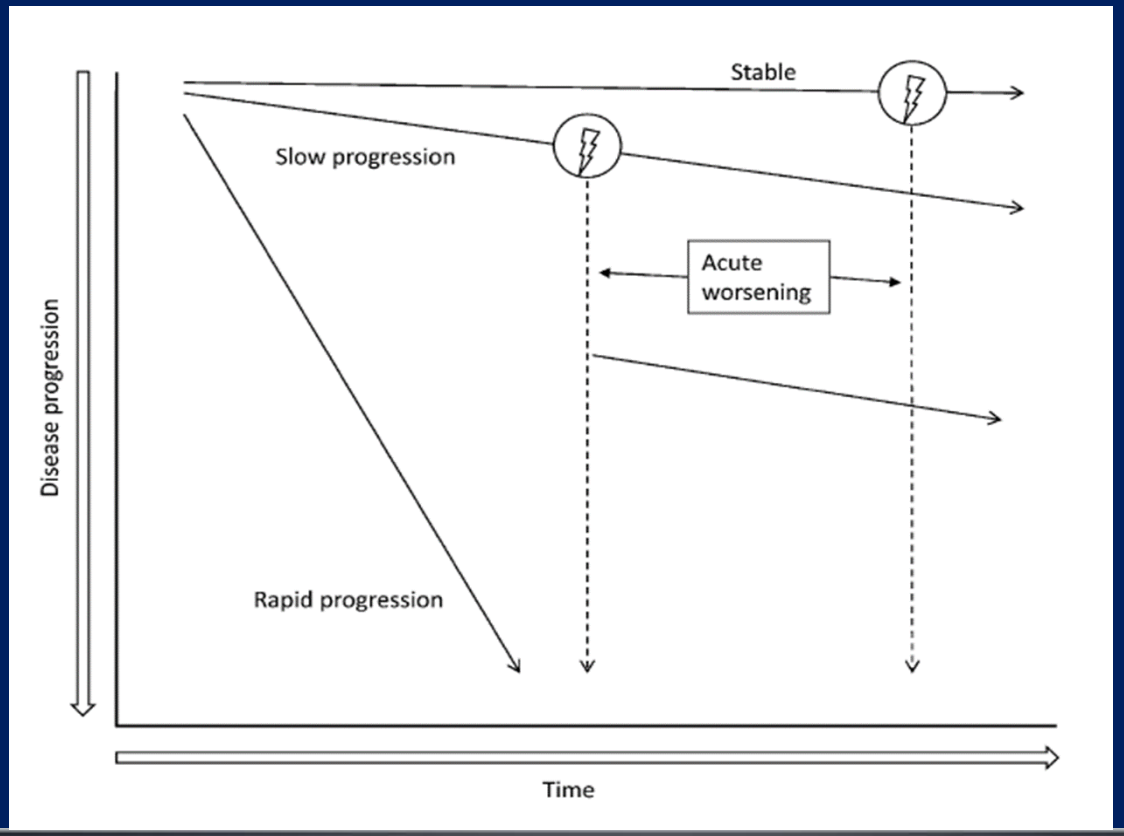
6 months of Pirfenidone therapy, he returns to the ED requiring 50% Oxygen by mask with saturation 89%. Crackles that are worse at the bases and some rhonchi anteriorly. No jugular venous distention. B-type natriuretic peptide is 20 pg/mL. High-resolution CT scan with contrast demonstrates new bilateral ground-glass opacities on a background of UIP. No evidence of pulmonary embolism. Bronchoalveolar lavage is negative for pathogens with 30% lymphocytes.

What are the next steps?

A. He has acute exacerbation IPF requiring steroids
B. He has acute exacerbation IPF requiring addition of nintedanib
C. He has streptococcal pneumonia needing antibiotics
D. He was misdiagnosed as IPF, necessitating a lung biopsy
E. He has heart failure and needs diuresis

IPF

- UIP pattern on CT scan
- Diagnosis of exclusion
- No other underlying causes
- Prognosis worse than iNSIP and decline unpredictable
- Oxygen therapy and Pulmonary Rehab
- Treat comorbidities
- Transplant
- Palliative care if needing intubation and not a transplant candidate



Referral

- UIP or fibrotic NSIP
- FVC < 80% or DLCO < 40%
- Dyspnea or functional limitation
- Oxygen requirement
- Failure to improve with a trial of medical therapy

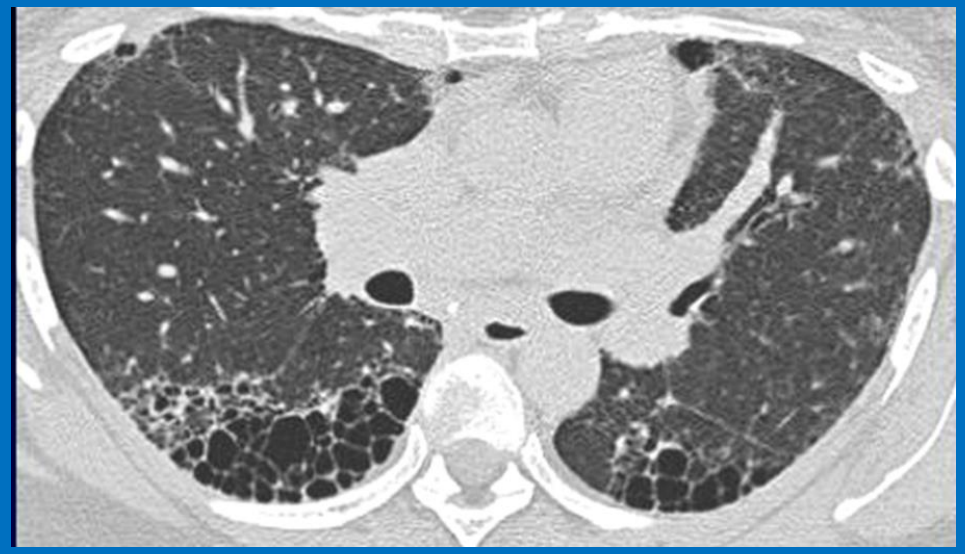
Listing

- 5-10% decline in FVC or 15% decline in DLCO over 6 months
- 6-minute-walk test
  - Desaturation < 88%
  - <250m
  - >50m decline over 6 months
- Pulmonary hypertension
- Hospitalization for respiratory decline or pneumothorax

Clinical	Image/Bronch	Therapy
IPF- Clubbing and crackles	Honeycombing,linear, traction bronchiectasis, UIP pattern Mod Neutrophils BAL	Nintedanib, pirfenidone
NSIP- clear, crackles	Peripheral linear, ground glass, honeycomb, traction Mod lymphs BAL	Prednisone Mycophenolate ? antifibrotics
COP- Inspiratory squeaks, crackles, rhonchi	Nodules, alvelolar infiltrates, bronchiolar involvement “Full house” inflammatory BAL	Prednisone Azathiorpine
AIP- Diffuse rales, bronchiolar breath sounds	ARDS-like 5 lobe infiltrates Intense neutrophilic BAL Diffuse alveolar damage on path	High dose steroids, maintenance therapy

QUESTION 6.

39-year old female presents with progressive dyspnea, pleuritic chest pain and cough. She denies occupational and environmental exposures. No medications. She has noted a rash on her arms and face and her fingers will change color when she is in cold environments. Lungs- crackles. ANA antibody **(+).** Rheumatology diagnosed her with scleroderma.



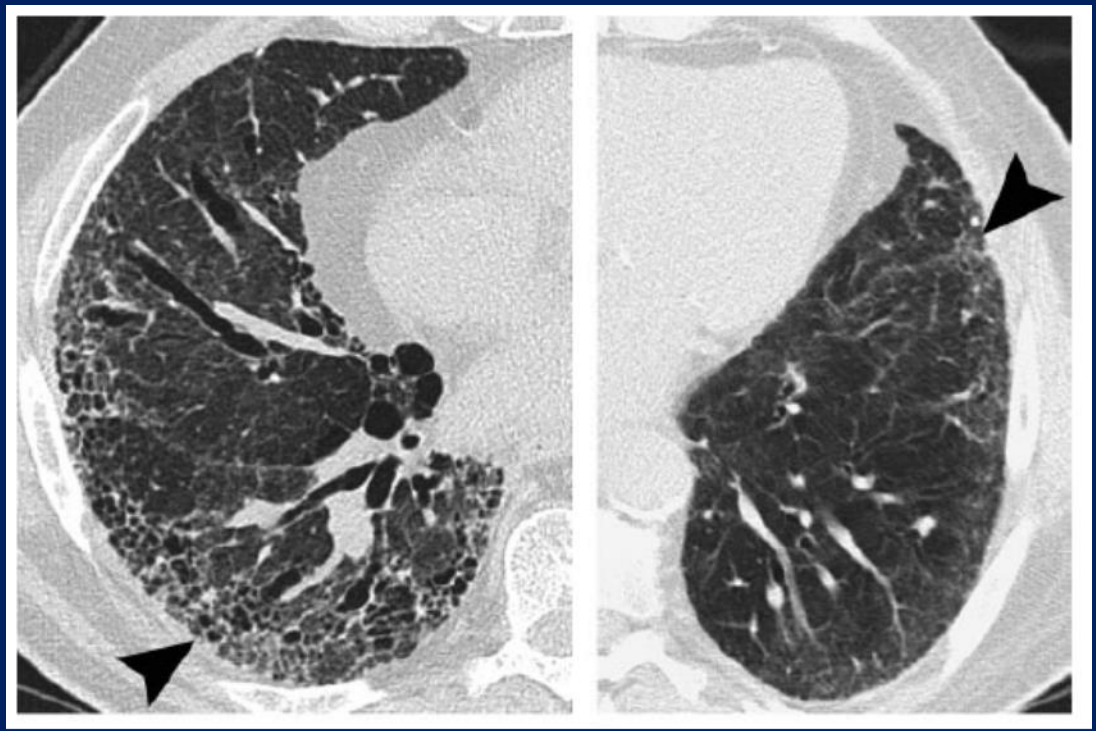
What is the most likely cause of her dyspnea?

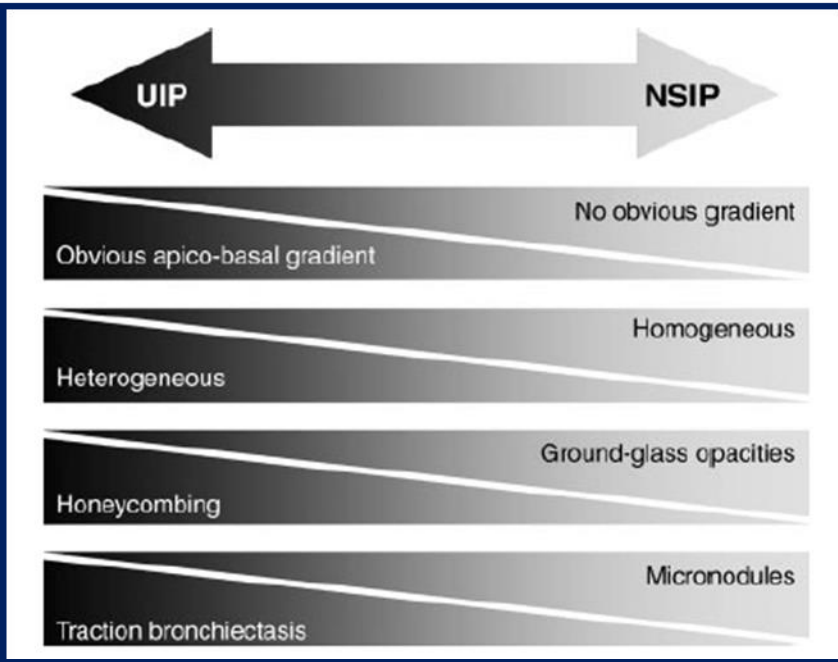
A. Aspiration pneumonia
B. Pulmonary hypertension
C. Connective tissue disease related ILD
D. Idiopathic usual interstitial pneumonia
E. Idiopathic nonspecific interstitial pneumonia

CTD-ILD

- Must have CTD diagnosis
- If CTD diagnosis, no biopsy
- NSIP pattern most common
- Prognosis better than iNSIP
- Lots of pulm complications

	ILD	ILD pattern ** other
Scleroderma	+++++	NSIP >>> UIP (aspiration) ** PAH
Rheumatoid	++	UIP > NSIP > OP = DAD ** airway(++++), Pleura(++++)
Polymyositis	++++	NSIP = OP > DAD > UIP ++ muscle
Sjogren's	+++	NSIP > LIP > OP = UIP = DAD ** airway(++++)
Lupus	+	NSIP > DAD = LIP = OP = UIP ** hemorrhage, Pleura(++++), PAH(++)

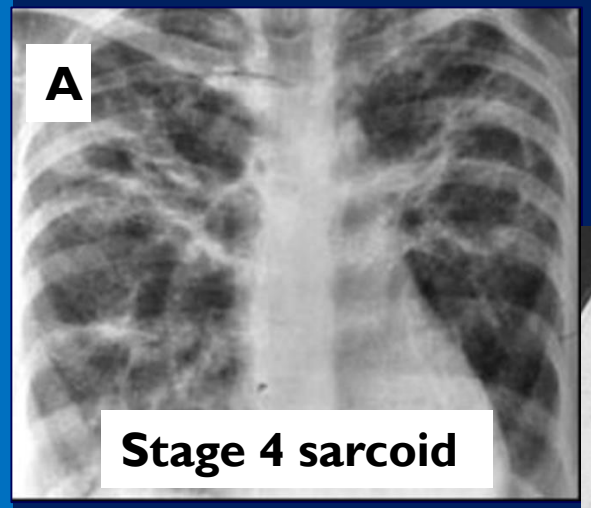


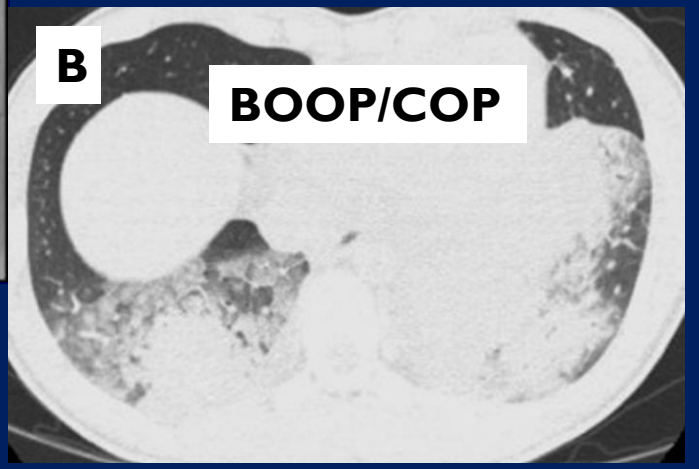


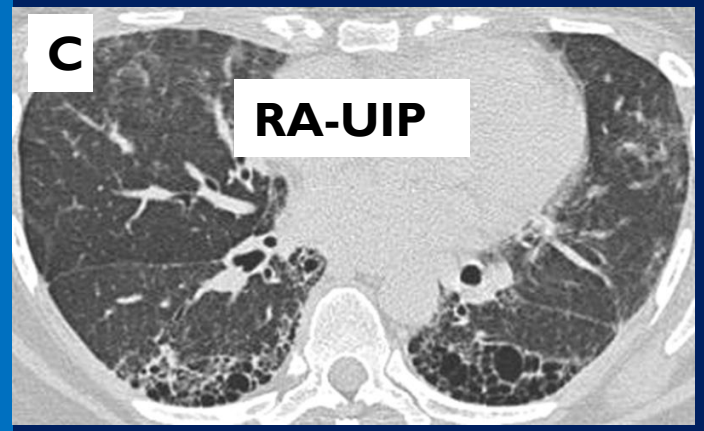
QUESTION 7.

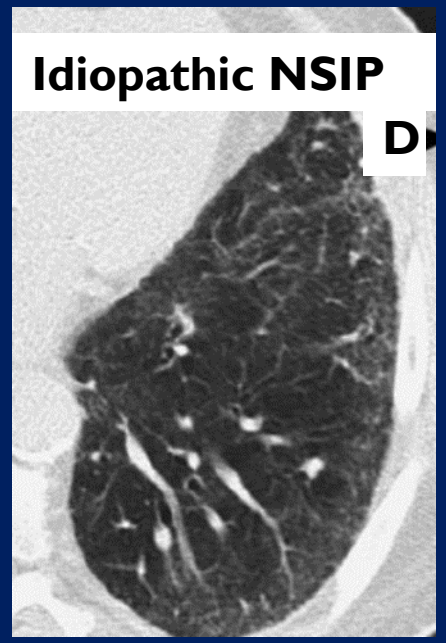
A 41-year-old woman is evaluated for dry cough and progressive dyspnea. She initially presented 8 weeks ago with cough, fever, sputum production, and dyspnea. A chest radiograph at that time revealed left-lower-lobe opacities; she was diagnosed with pneumonia and treated with azithromycin but had little improvement in her symptoms. Her fever and sputum production have resolved. She is a nonsmoker. Lungs have scant inspiratory squeaks and rhonchi.

Which image is most compatible with her diagnosis?









QUESTION 8.

A 50 year old female smoker on nitrofurantoin for chronic uti prophylaxis lives on a farm, owns 2 birds and grinds counter tops for a living.

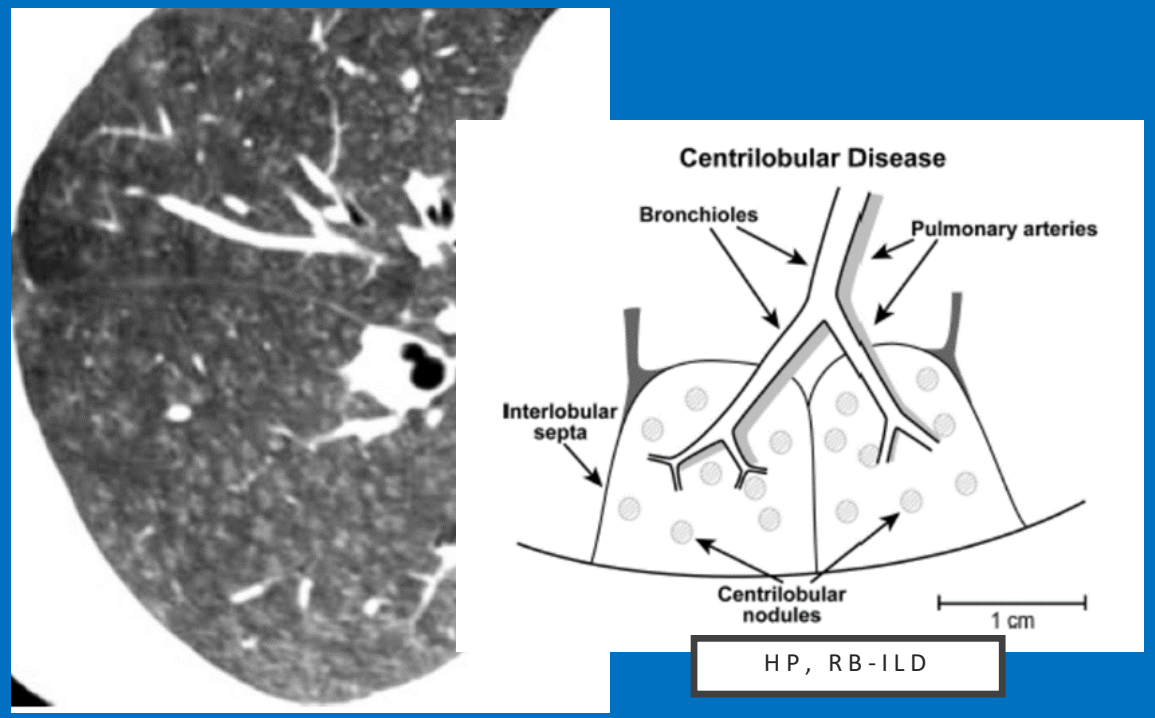
Drug-induced	Nitrofurantoin, Amiodarone, Bleomycin http://www.pneumotox.com
Occupational/environmental	Silicosis, Asbestosis, Farmer's and Bird Breeder's lung, HP
Smoking- related	DIP, RB-ILD, PLCH

What is a likely cause of her dyspnea?

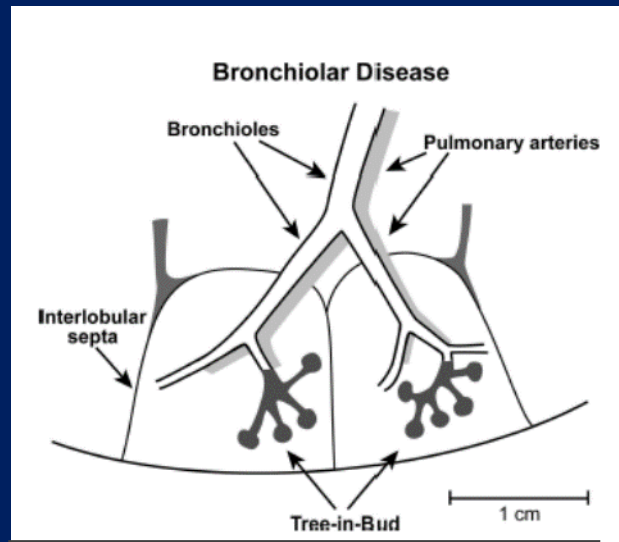
A. Hypersensitivity pneumonitis
B. Drug-induced ILD
C. Farmer's lung
D. Silicosis
E. RB-ILD/DIP
F. All of the above

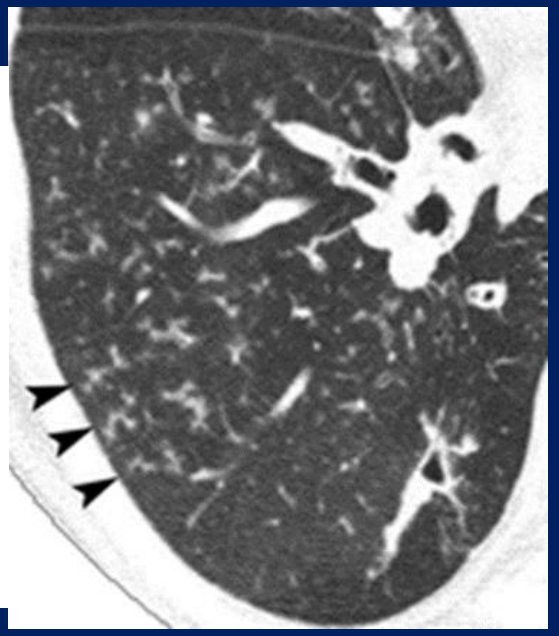
Exposure- ILD

- History is critical
- Acute- nodular, upper lobe
- Chronic- scarring, mimics IIPs
- Smoking- concomitant COPD

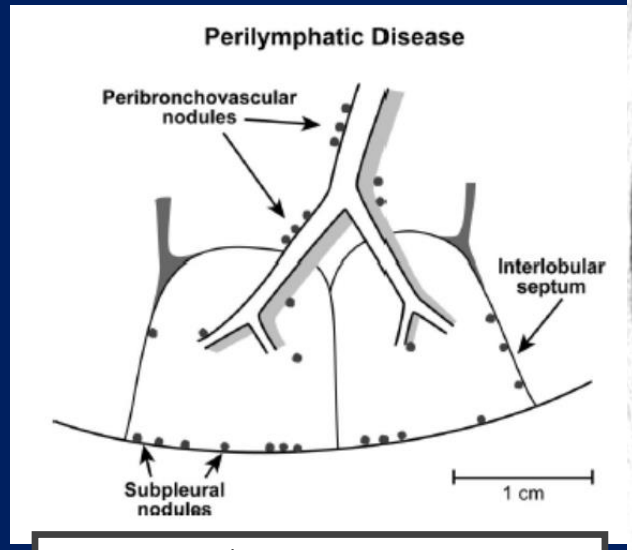


HP, RB-ILD

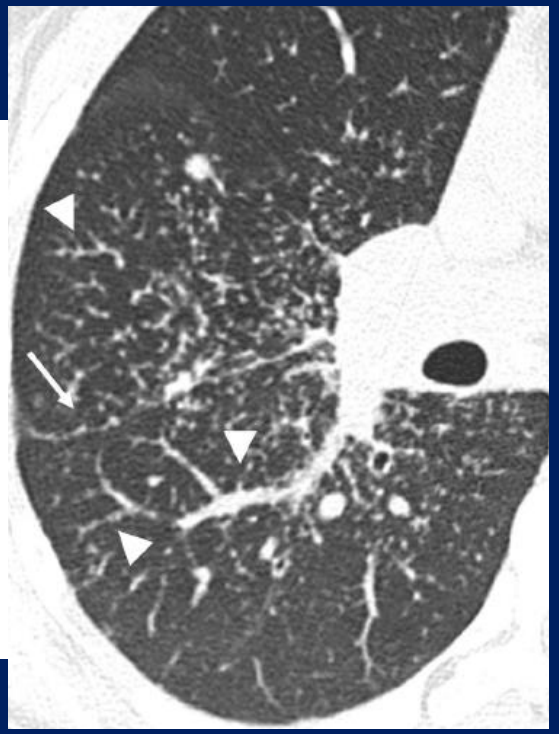




MAC, Bronchiectaisis, PLCH



Sarcoid, penumoconiosis



Sarcoid	ACE, Lysozyme
Granulomatosis with polyangitis (GPA)	c-ANCA, p-ANCA in related diseases (microscopic polyangitis) Anti-GBM
Rheumatoid	Rheumatoid factor, CCP
Scleroderma	SCL-70, anti-centromere
Sjogren's	SS-A, SS-B
Lupus	DS-DNA, anti-histone
Polymyositis	Anti Jo-1
Mixed CTDz	Anti-RNP

Disease	Antigen	Source
Farmer's lung	*Faeni rectivirgula*	Moldy hay, grain, silage
Humidifier lung	***Thermoactinomyces***	Contaminated water reservoirs
Bagassosis	*Thermoactinomyces vulgaris*	Moldy sugarcane
Pigeon breeder's lung	Avian droppings, feathers, serum	Parakeets, pigeons, chickens, turkeys
Woodworkers lung	*Alternaria sp.*, wood dust	Oak, cedar, mahogany dust, pine and spruce pulp

Agent	Clinical Syndrome
Bleomycin, Methotrexate	Pulmonary fibrosis and HP-like
Cyclophosphamide	Pulmonary fibrosis
Nitrofurantoin	Acute Infiltrates with Eos – 1mo Chronic – 6 mos. to 2 y
Amiodarone	Pulmonary fibrosis

Drugs	Nitrofurantoin, tetracyclines
Infections	Loffler's-ascaris, Strongyloides
ABPA (aspergillosis)	Asthma, skin test, IgE, precipitins
Churg-Strauss	Asthma, vasculitis, granulomas
Chronic eosinophilic pneumonia	Peripheral infiltrates "photographic negative of pulmonary edema"
Acute eosinophilic pneumonia	Does not usually recur after prednisone therapy
Hypereosinophilic syndrome	Systemic disease, can be fatal

References:
- Radiographics. 2007;27:595-615
- ATS/ERS Statement. AJRCCM 2011
- Terrence Demos Sarcoid website
- Slide courtesy of KO Leslie, MD. ATS Slide Set
- PATS. 2006;3:81-95
- ERJ. 2009;33:882-896
- Chronic Respiratory Disease. 2011;8:53