Progressive ILD in a patient with autoimmune disease

Radiology Elective Case Presentation

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MSTP 9th year
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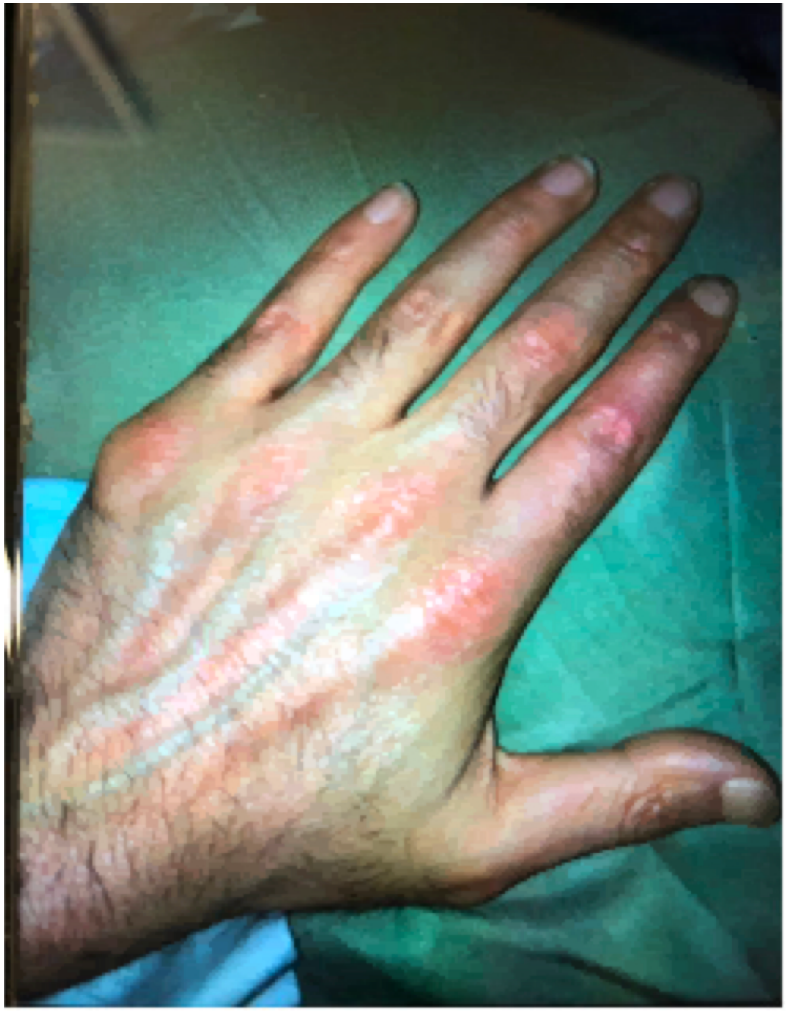
Patient history- 27 y.o. M “R.F.”

- Patient presented at age 15 with diaphragm and lung issues, unclear history with limited EPIC records
- Treated from 2007-2012 with prednisone and methotrexate
- Lost 100 lbs after prednisone taper, methotrexate was restarted in 2017 for progression of stiffness, back pain
- Methotrexate dose increased 3 weeks prior to recent visit, radiograph for worsening fatigability, worse exercise tolerance, and difficulty taking a deep breath
- No cough
- Decreased hand mobility, and chronic back pain treated with topical pain relievers
- PE: clear lungs, 4/5 hip abductor and adductor strength, and hand tenderness with skin findings:

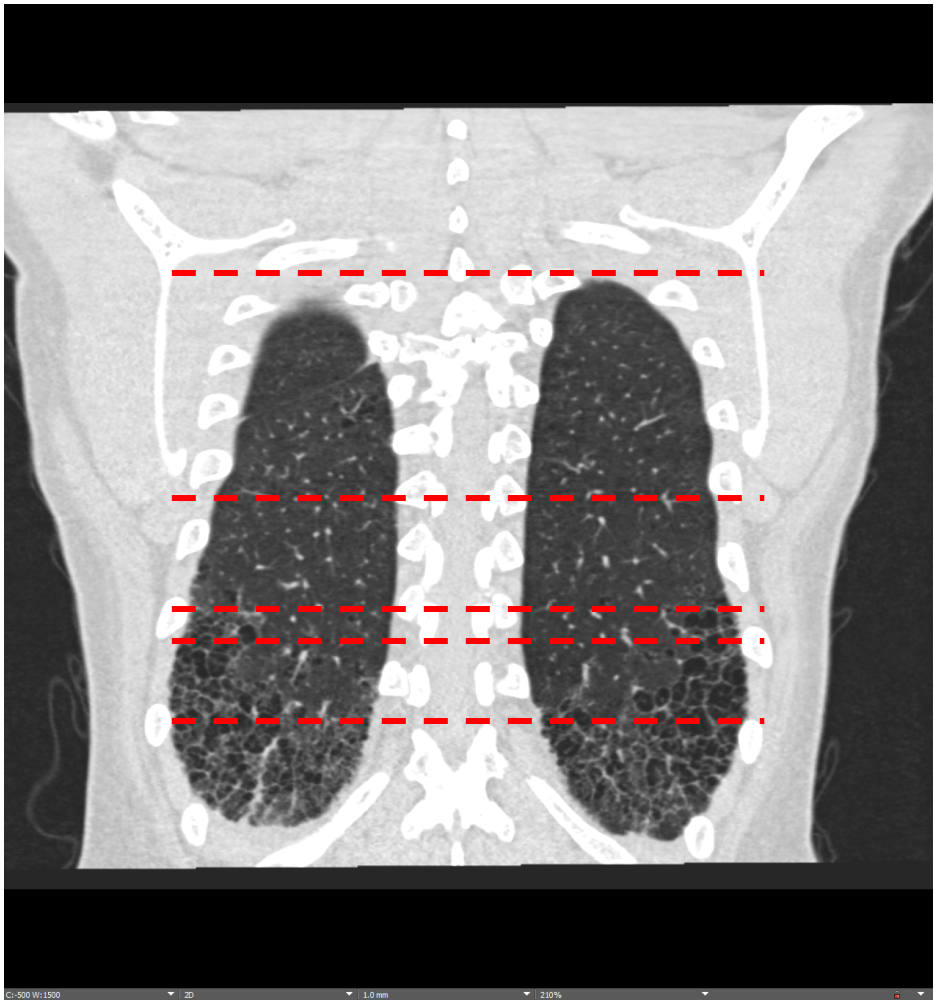


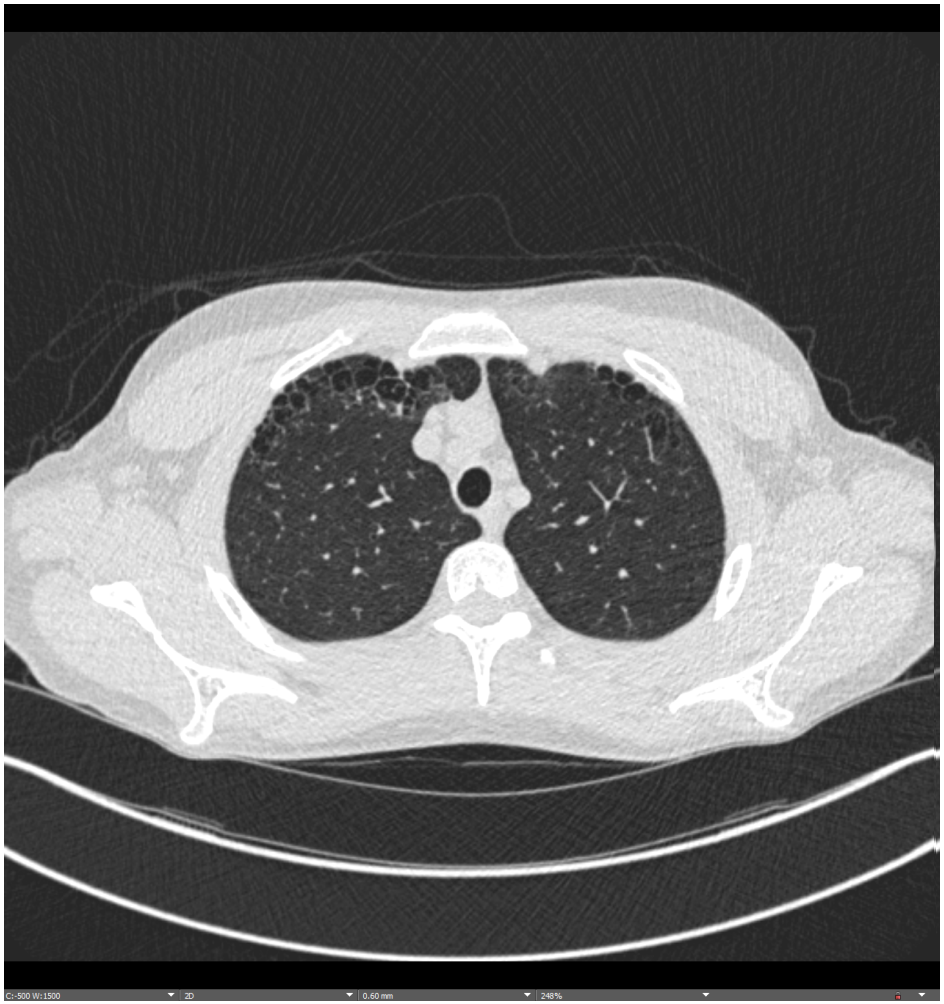
Labs and Diagnostics

- ANA 1:640 (since 2012)
- Negative (7/2019):
  - Lyme, anaplasma, Babesia
  - PM-Scl Ab; Scl70; RNA polymerase III
  - Smith/RNP
  - Histone ab
  - SPEP/IEP
  - RPR and HIV
  - Ds dna/ SS-A/ SS-B
  - Vitamin b12
  - Cortisol level, Pro BNP, TSH
  - ESR, CRP
  - Anti-Jo negative in 2016
- PFTs
  - DLCO of 84%
  - Was 105% in 1/2019
- Echo
  - RVSP of 25
  - EF 58%
- L-spine XR 7/2019:
  - "Incidental **reticular appearing opacity** along the posterior periphery of the lungs. This corresponds with **cystic changes present on CT abdomen July 1, 2017**, and is most suggestive of an interstitial lung disease."

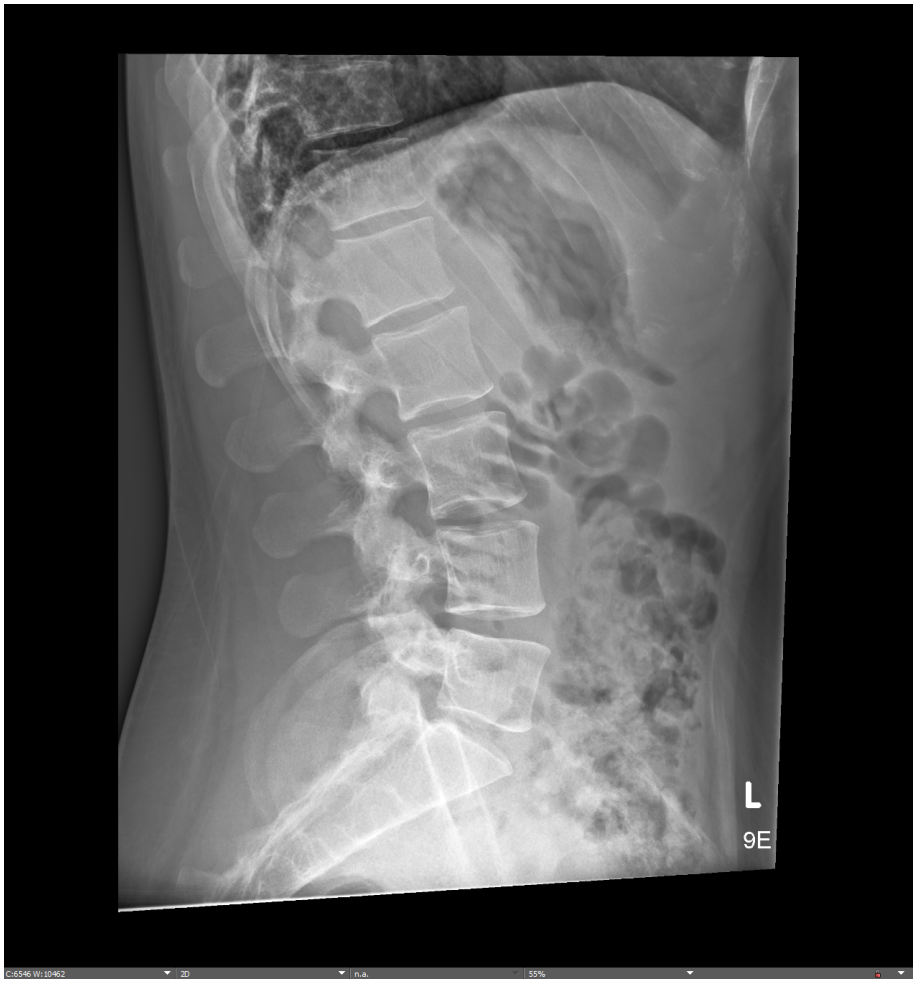
Overall, the patient carries a diagnosis of overlap juvenile dermatomyositis-scleroderma and has progression of lung symptoms since March

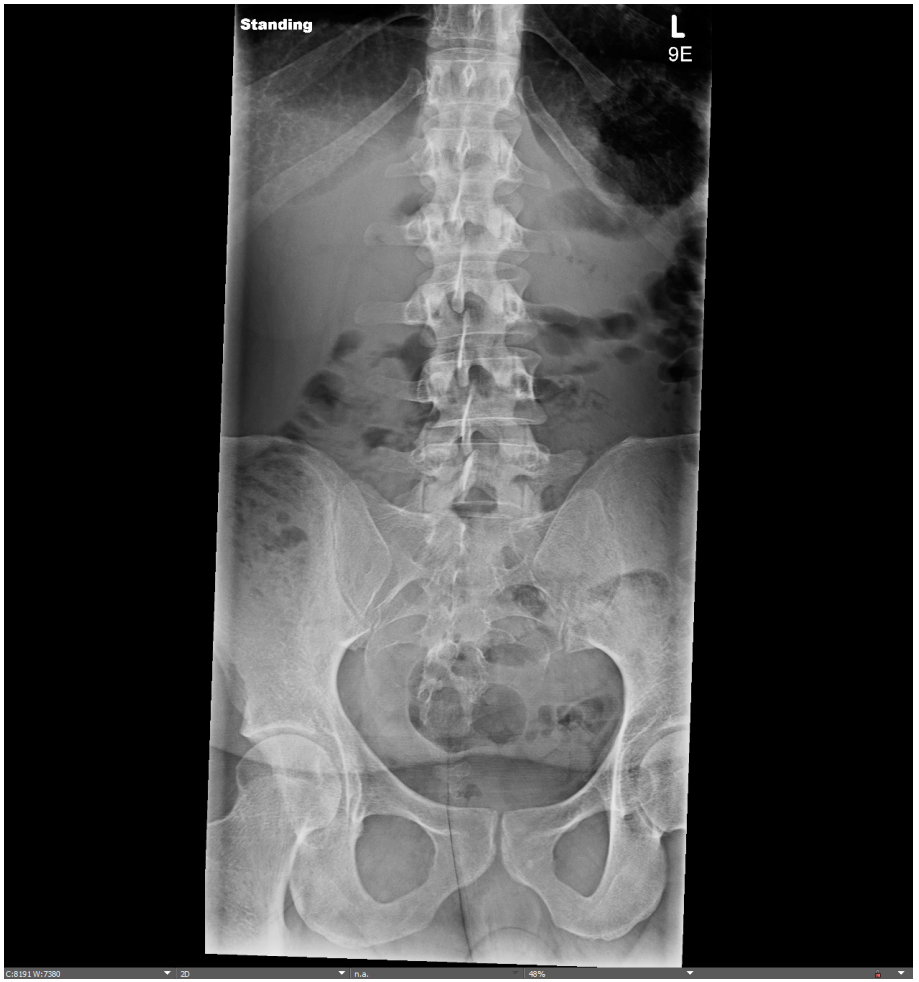
CT Chest w/o IV contrast High Resolution





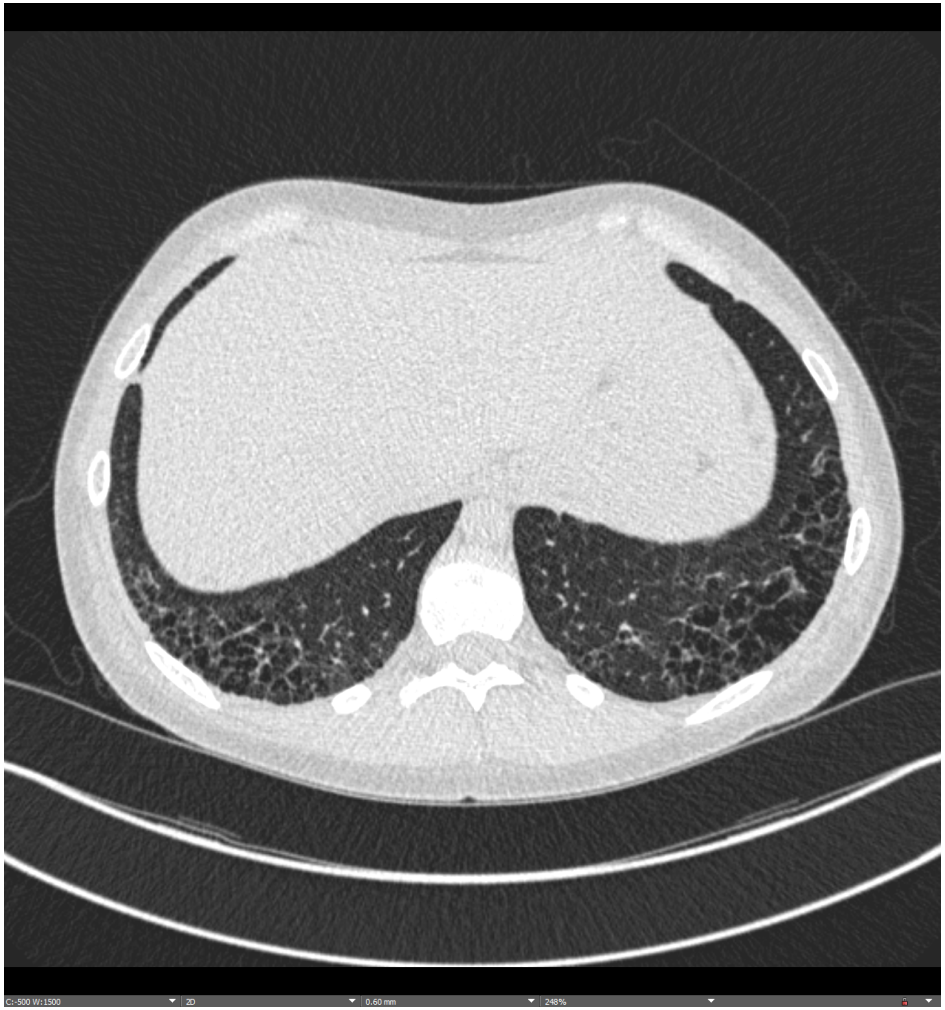
Lumbar Spine XR



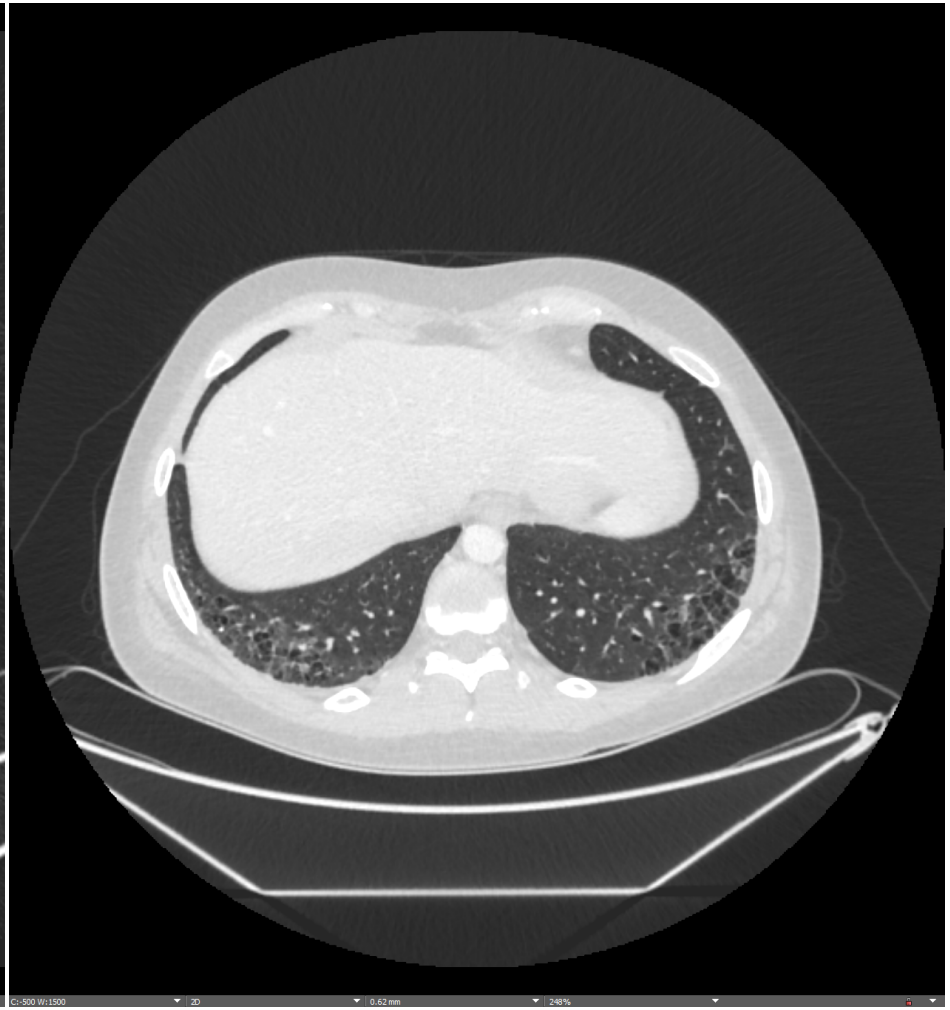


Comparison to CT Abdomen Pelvis 2017

2019



2017



Final Read

FINDINGS:
Lungs: There is a 6 mm nodule within the right lower lobe directly adjacent to the minor interlobar fissure, best seen on series 4 image 207, stable since 2017. Intrafissural lymph node is seen on series 4 image 156. Cluster **subpleural** cystic structures are seen throughout the posterior bilateral lower lobes, left slightly greater than right, **consistent with honeycombing**. More focal areas of similar-appearing honeycombing are seen in the **anterior bilateral upper lobes.** Lower lobe **honeycombing has worsened when compared to the abdominal CT from July 2017**. Mild bronchiectasis of the lower lobes without significant bronchial wall thickening. **No evidence of groundglass opacities.** Expiratory images demonstrate no evidence of air trapping.

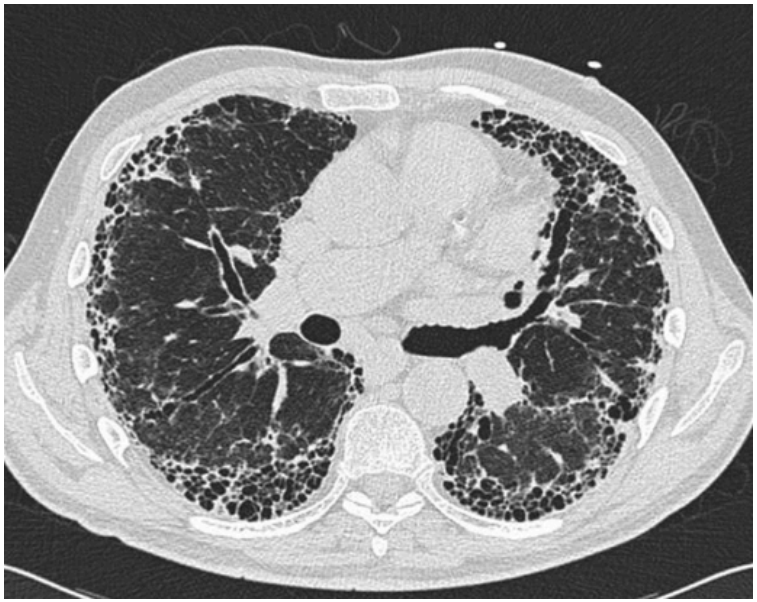
IMPRESSION:
1. Bilateral areas of honeycombing, most consistent with connective tissue disorder related ILD in this patient with JDM-scleroderma overlap.

Interstitial Lung Disease Differential Diagnosis

- Idiopathic interstitial pneumonias (IIPs)
  - Chronic fibrosing (IPF, INSIP)
  - Smoking-related
  - Acute or subacute (organizing pneumonia)
- Hypersensitivity pneumonitis
- Collagen vascular diseases
- Familial Interstitial pneumonia
- Other
  - Drug-associated
  - Vasculitis/GPA
  - Sarcoidosis
- Coexisting patterns

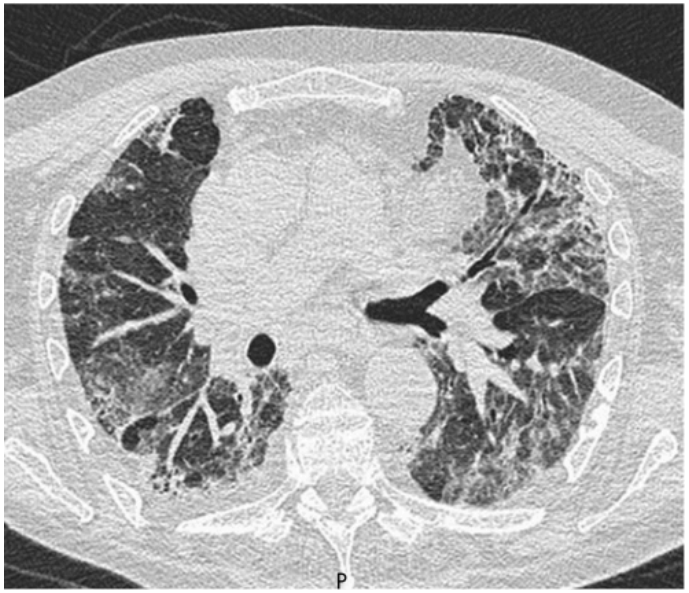
UIP vs. NSIP radiologic and histopathology

Usual Interstitial Pneumonia



Subplural, basal, bilateral, peripheral
Reticular, honeycombing
IPF- 20-30% 5 yr survival

Non-specific Interstitial Pneumonia



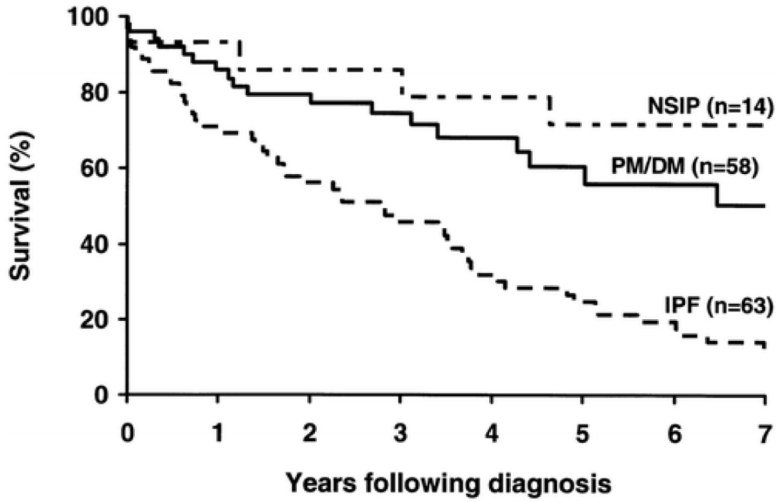
Non-basal, non-peripheral
Ground glass
INSIP- 60% 5 yr survival

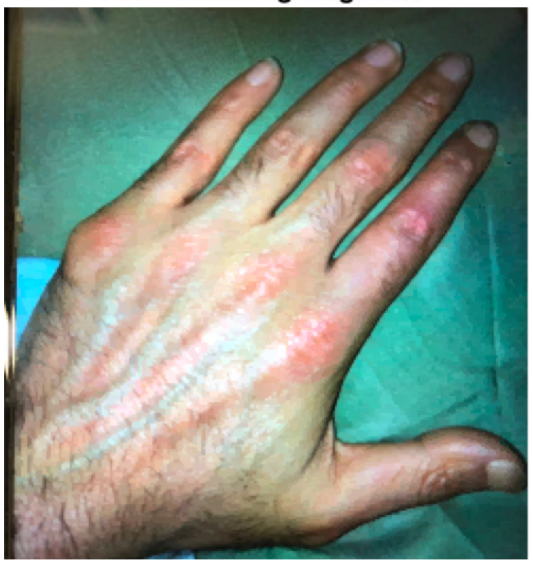
Connective tissue diseases can present with either pattern, with NSIP most common

Du Bois and King. Thorax (2007).

Dermatomyositis/scleroderma CTD and ILD

- ILD in 20-80% of DM patients, 86% of anti-Jo positive
- Anti-MDA5 antibody associated with rapid progression
- Consider effects of drugs, esp. methotrexate
- High resolution CT has efficacy in diagnosing ILD subtype, monitoring response/progression
- Survival not affected by anti-Jo positivity, improved with NSIP pattern of disease





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Resources

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