Progressive ILD in a patient with autoimmune disease

Radiology Elective Case Presentation

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MSTP 9th year
September 2019
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
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
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

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

Resources


