RADY 401 Case Presentation: Interstitial Lung Disease

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Focused patient history and workup

- 40-year-old female with PMHx of seropositive erosive rheumatoid arthritis with possible overlapping systemic sclerosis presented to UNC Rheumatology with worsening DOE x 3 months.
- She reports SOB after climbing more than 1 flight of stairs, walking more than 1 block, and with housework and yardwork; symptoms improve with rest
- No cough, hemoptysis, or morning cough
- Pt also endorses significant esophageal symptoms: worsening GERD, dysphagia/food getting stuck in throat, vomiting undigested food; no changes in appetite, no weight loss
- Smoking 1/4-1/2 ppd x 22 years as well as significant marijuana use x 15 years
- Exam: lungs CTAB, no crackles
- Initial work-up: CXR



Differential Diagnoses: Chronic Dyspnea on Exertion

- Presentation consistent with chronic dyspnea given worsening over several months
- Differential:
 - Cardiac: Heart Failure
 - Pulmonary: COPD, restrictive lung disease (ie ILD), pulmonary HTN
 - high concern for ILD or pulmonary HTN in pt with systemic rheumatic disease
 - GI: Recurrent aspiration



List of imaging studies

- Chest X-ray
 - Low Cost
 - Radiation: 0.1 mSv, comparable to 10 days background radiation
 - Sensitivity 80%, specificity 82% for detecting diffuse lung disease
- High-Resolution Chest CT without contrast
 - High Cost
 - Radiation: 6.1 mSv, comparable to 2 years background radiation
 - Sensitivity 95%, specificity close to 100%
 - Smaller slices: 0.6 mm (at UNC) vs 5mm standard thickness
 - Indications: useful for assessing diffuse lung disease
 - ie interstitial lung disease, cystic fibrosis, bronchiectasis, small airways disease, pulmonary micronodules



Imaging studies from PACS 1: Chest X-ray 2 views

Upright PA



Upright Lateral



Findings: what abnormalities do you see?



Imaging studies from PACS 1: Chest X-ray 2 views

Upright PA



Upright Lateral



Findings: subtly increased coarse interstitial markings near the lung bases. In setting of known connective tissue disease suspicion is raised for ILD. Recommend HRCT.



Side by Side

Normal



Our Patient



DDNC - Radiology

(Saripalli et al., 2020)

Chest Xray Discussion

Revised 2018

American College of Radiology ACR Appropriateness Criteria[®] Chronic Dyspnea-Noncardiovascular Origin

Variant 1:

Chronic dyspnea. Unclear etiology. Initial imaging.

| Procedure | Appropriateness Category | Relative Radiation Level |
|--|-----------------------------------|--------------------------|
| Radiography chest | Usually Appropriate | |
| CT chest without IV contrast | May Be Appropriate (Disagreement) | *** |
| CT chest with IV contrast | May Be Appropriate | *** |
| CT chest without and with IV contrast | Usually Not Appropriate | *** |
| FDG-PET/CT skull base to mid-thigh | Usually Not Appropriate | **** |
| MRI chest without and with IV contrast | Usually Not Appropriate | 0 |
| MRI chest without IV contrast | Usually Not Appropriate | 0 |
| US chest | Usually Not Appropriate | 0 |



Imaging studies from PACS 2: High-resolution Chest CT without contrast



Findings: what abnormalities do you see?



Imaging studies from PACS 2: High-resolution Chest CT without contrast



Additional HRCT Findings...



Findings: what abnormalities do you see?

Additional HRCT Findings...





bronchiectasis

traction bronchiectasis

HRCT Final Report

Findings:

-bibasilar predominant largely subpleural reticular and ground glass opacities associated with bronchiectasis/bronchiolectasis and cyst like changes -some of these cystic changes coalesce to form small regions of honeycombing -patchy/nodular areas of ground glass are present somewhat more diffusely in lung bases

Impression:

Pulmonary parenchymal changes characterized above are **consistent with interstitial lung disease**. Notably, there is clear evidence of fibrosis, including several small regions of honeycombing. The imaging pattern is considered "typical" with that of usual interstitial pneumonia (UIP).



Radiographic and histopathologic appearances of non-specific interstitial pneumonia (NSIP) and usual interstitial pneumonia (UIP)



A: NSIP pattern; HRCT scan demonstrating the characteristic radiographic appearance of NSIP with **bilateral, ground-glass opacities.**

C: UIP pattern; HRCT scan demonstrating the characteristic radiographic appearance of a UIP pattern with **bibasilar**, **reticular abnormalities**, **traction bronchiectasis**, **and honeycombing**.

Common findings associated with RA-ILD; considered diagnostic!

High-resolution Chest CT without contrast discussion

Variant 4:

Chronic dyspnea. Suspected interstitial lung disease. Initial imaging.

| Procedure | Appropriateness Category | Relative Radiation Level |
|--|-----------------------------------|--------------------------|
| CT chest without IV contrast | Usually Appropriate | *** |
| Radiography chest | Usually Appropriate | ÷ |
| CT chest with IV contrast | May Be Appropriate (Disagreement) | *** |
| MRI chest without and with IV contrast | Usually Not Appropriate | 0 |
| MRI chest without IV contrast | Usually Not Appropriate | 0 |
| US chest | Usually Not Appropriate | 0 |
| CT chest without and with IV contrast | Usually Not Appropriate | *** |
| FDG-PET/CT skull base to mid-thigh | Usually Not Appropriate | **** |



Interstitial Lung Disease

- Large subset of parenchymal lung diseases; all share same basic physiology:
 - Some type of injury/insult to lung parenchyma → aberrant wound healing → fibrosis → remodeling of interstitium
- Includes idiopathic causes such as idiopathic pulmonary fibrosis (IPF), connective tissue disease-associated ILD, hypersensitivity pneumonitis, sarcoidosis, environmental and occupational exposure induced ILD (ie silicosis, asbestosis), as well as other rare forms.
- Most common types are IPF, connective tissue disease associated ILD, chronic HP, and smoking related ILD



ILD Overview



In this patient, presentation was felt to be most consistent with RA/MCTD-ILD

(Hoffman-Vold et al., 2019)



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Patient treatment or outcome

- PFTs obtained which showed restrictive pattern
- Echo also obtained which showed normal EF, no wall thickening
- Patient diagnosed with interstitial lung disease as a complication of her rheumatoid arthritis
- Pulmonary Recommendations:
 - Quit smoking cigarettes and marijuana as soon as possible
 - Increase prednisone to 20 mg daily
 - Start Bactrim prophylaxis
 - Control GERD aggressively as uncontrolled reflux can worsen ILD
 - Start Cellcept 500 mg BID monitoring CBC in 4 weeks, goal to increase to 1 g BID
 - Consider Rituximab if pt does not tolerate Cellcept



UNC Top Three: Interstitial Lung Disease

- 1. HRCT is gold-standard imaging for suspected ILD; can be diagnostic and if so, avoids biopsy
- 2. Honeycombing and traction bronchiectasis are indicative of usual interstitial pneumonia (UIP) pattern on HRCT, typically thought to be interchangeable with idiopathic pulmonary fibrosis (IPF) but also diagnostic for RA-ILD



3. It is critical to test everyone diagnosed with ILD for rheumatic disease if not already diagnosed

CHOOL OF MEDICINE

Radiology

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