Dermatomyositis and interstitial lung disease

Asmin Tulpule, HMS III
November 16, 2009
Core Radiology Clerkship

Beth Israel Deaconess Medical Center
Agenda

• Presentation of our patient

• Menu of radiologic tests

• Basics of interstitial lung disease (ILD)

• Radiologic survey of ILD

• Application of radiologic findings to our patient’s clinical course
Ms. L is a 57 year old F with a recent diagnosis of dermatomyositis (DM) and ILD transferred to BIDMC for worsening dyspnea and hypoxia over the past 2-3 days.

Initial diagnosis of DM two months prior based on myalgias, Gottron’s papules, and heliotrope rash. Confirmed by skin biopsy.

No respiratory complaints at first and initial CT scan at OSH showed mild ILD. Started on 40 mg prednisone.
Note: DM is an idiopathic inflammatory myopathy with a strong association with ILD (30-40%) characterized by pulmonary fibrosis, as well as a predisposition to developing visceral malignancies (stomach, pancreas).
Our patient: Past Medical Hx

- Began developing dyspnea on exertion after a month. Thought to be progressing pulmonary fibrosis associated with DM. Receiving increasing prednisone (up to 60 mg PO qday) and started on intermittent infusions of Solumedrol. She was also tried on IVIG and azathioprine for ILD associated with DM. Her respiratory status did not improve.

- She was NOT placed on any antibiotic prophylaxis

- She reports night sweats for the past two months. She denies any fevers, chills, or symptoms of GI illness.

- No prior history of smoking. No recent international travel history.
Our patient: Initial Evaluation

- On the floor, she reports an acute 2-3 day worsening of respiratory status. She is no longer able to walk more than a few steps without developing acute shortness of breath.

Physical Exam:
Vital Signs: Tc 97.2 BP 117/79 HR 77
- RR 24 O2 sat: 93% on 6L and 70%FM
GEN: Ill appearing, speaking in full sentences but appears dyspneic with conversation, occasionally stopping to catch her breath
Lungs: clear anteriorly. Posterior fine crackles mid and lower lung fields. No wheezes or rhonchi.
Cardiac: RRR. No m/r/g. Prominent S2
Let’s now discuss the menu of radiologic tests that could help aid us in the diagnosis and management of our patient.
Agenda

- Presentation of our patient
- Menu of radiologic tests
- Basics of interstitial lung disease (ILD)
- Radiologic survey of ILD
- Application of radiologic findings to our patient’s clinical course
Menu of Radiologic Tests

1. Chest X-Ray (CXR), both PA and LATERAL

   PROS: - Excellent initial screening test
          - Fast and inexpensive
          - Sensitive for acute pulmonary processes
   CONS: Shows projection images – not sensitive for parenchymal detail or interstitial disease

2. Chest CT (non-contrast)

   PROS: The gold standard for evaluating interstitial disease. + Contrast for pulmonary nodules and masses
   CONS: High cost and radiation dose. Not useful as a screening tool
Menu of Radiologic Tests cont’d

Lesser Used Tests:

3. Chest MR
   - for patients with contrast allergy or renal failure
   - patients who want to minimize radiation exposure
     (pregnant women)

4. Specialized nuclear medicine testing
   - Gallium scan for inflammatory (sarcoidosis) or infectious processes
   - PET scan – monitoring of lymphoma, lung cancer
Our patient: Initial CXR w/ interstitial opacities

Always compare with prior CXR

Diffuse reticular (interstitial) opacities diffusely involving both lungs, most prominent at the bases.

Upright PA Chest X-ray
BIDMC, PACS
So, the CXR helps rule out a lot of common causes of dyspnea. We don’t see signs of a lobar pneumonia (like pneumococcal), pulmonary edema, pneumothorax, or congestive heart failure exacerbation.

We are dealing with Interstitial Lung Disease (aka Diffuse Parenchymal Lung Disease).

What imaging test would you order next?
Our patient: Evolution of ILD on Chest CT

Initial axial non-contrast chest CT on diagnosis

Axial non-contrast chest CT 1 month later

Mild ground glass opacities at L lung base

Bilateral ground glass opacities at the bases
Peripheral honeycombing
Summary of Imaging

1) CXR showed diffuse bilateral reticulo-nodular opacities
2) CT showed ground glass opacities and honeycombing, w/o focal consolidation

This is classic imaging of ILD (also known as diffuse parenchymal lung disease DPLD)
Summary of Imaging

Does this tell us what our patient has?

Could it be Pneumocystis carinii pneumonia (PCP) given the fact that our patient received high dose steroids without antibiotic prophylaxis?

What about progressive pulmonary fibrosis associated with DM?
Can we distinguish between the different causes of ILD?

Let’s review some lung anatomy…
Agenda

• Presentation of our patient

• Menu of radiologic tests

• Basics of interstitial lung disease (ILD)

• Radiologic survey of ILD

• Application of radiologic findings to our patient’s clinical course
Interstitial Lung Diseases: Anatomy

Alveolar unit

ILD = Any infiltration and/or fibrosis of the lung interstitium

What can do that?

Cells (Immune or Malignant)
Infection
Fluid
Fibrosis (reactive or primary)

p.a. – pulmonary artery
p.v. – pulmonary vein
a.s. – alveolar sac
l - lymphatics

http://en.wikivisual.com/images/c/ce/Alveoli.gif (Reprint from Gray’s Anatomy, 1918)
Interstitial Lung Disease: Differential Dx

Modified 2002 American Thoracic Society Guidelines for DPLD:
Acute vs Chronic:

Chronic:
1) Known causes
   a) Inhaled substances: Inorganic, Silicosis, Asbestosis, Berylliosis
   b) Connective tissue disorders: Systemic sclerosis, Polymyositis, dermatomyositis, lupus
   c) Drugs: Antibiotics, Chemotherapeutic, Anti-arrhythmic drugs

2) Granulomatous
   a) Sarcoidosis
   b) Hypersensitivity pneumonitis

3) Rare DPLDs
   a) Langerhans histiocytosis
   b) Eosinophilic pneumonia
**Interstitial Lung Disease: Differential Dx**

Modified 2002 ATS Guidelines for DPLD continued…

4) Idiopathic Interstitial Pneumonias (IIPs)
   * divided pathologically
   a) Usual Interstitial Pneumonia (UIP) – clinical correlate
      Idiopathic Pulmonary Fibrosis (IPF)
   b) Non-Specific Intersitial Pneumonia (NSIP)
   c) Cryptogenic organizing pneumonia

Acute:
5) “Known” Infections - mimics
   a) Atypical pneumonia
   b) Pneumocystis pneumonia (PCP)
   c) Viral pneumonias (RSV)

6) Pulmonary edema - CHF

And many others....
Can we differentiate the different causes of ILD on CXR or CT?

No. But, we can get clues as to more/less likely etiologies.

Examples:
- a) NSIP vs UIP – no honeycombing in NSIP
- b) Hilar adenopathy – sarcoid vs lymphoma vs tumor

www.breader.com/diagram-teaching-files/index.html
So, imaging can provide us with some insight into the different causes of ILD.

Let’s take a look at some companion patients with classic findings within the broader category of ILD.
Agenda

• Presentation of our patient
• Menu of radiologic tests
• Basics of interstitial lung disease (ILD)
• Radiologic survey of ILD
• Application of radiologic findings to our patient’s clinical course
Companion patient #1: Systemic Sclerosis on CXR

Diffuse reticulo-nodular (interstitial) opacities diffusely involving both lungs, perhaps R>L, most prominent at the bases.

Looks similar to our patient’s CXR, though less severe!
Companion patient #1: Systemic Sclerosis on CT

Ground glass opacities noted at the lung bases

Traction bronchiechstasis due to fibrous tissue tethering of the airways

Axial non-contrast chest CT
BIDMC, PACS
Companion patient #2: Idiopathic Interstitial Pneumonia on CXR

Lung bases show fine reticulation and small nodules.

Smaller number of tiny nodules are also suggested in the upper lobes.

Better evaluated with the chest CT to determine whether there is a fibrotic honeycombing and to assess nodules, if any.

Why?
Fine reticular on CXR = UIP or NSIP
Companion patient #2: Idiopathic Interstitial Pneumonia on CT

Diffuse ground-glass changes throughout the lower lobes bilaterally

No honeycombing.

Axial non-contrast chest CT
BIDMC, PACS
Honeycombing: Recall our patient

Axial non-contrast chest CT

Peripheral honeycombing
Companion patient #2: Idiopathic Interstitial Pneumonia on CT

The combination of fine reticular opacities on CXR, with ground glass opacities in the absence of honeycombing on CT, helps to narrow the differential.

Most likely due to fibrosing type NSIP
Companion patient #3: Sarcoidosis on CXR

Diffuse small discrete and confluent nodular opacities of varying sizes, involving both lungs.

Bilateral hilar and right paratracheal soft tissue prominence, corresponding to the known adenopathy.

Upright PA Chest X-ray
BIDMC, PACS
Companion patient #3: Sarcoidosis on CT

Extensive progression of parenchymal ground glass and micronodular opacities with a peribronchovascular and perilymphatic distribution consistent with worsening pulmonary sarcoidosis. No significant fibrosis.
We have taken a look at a number of different patients with classic findings of ILD that suggest a specific cause.

Now, let's apply what we have learned about ILD to our patient.
Agenda

• Presentation of our patient
• Menu of radiologic tests
• Basics of interstitial lung disease (ILD)
• Radiologic survey of ILD
• Application of radiologic findings to our patient’s clinical course
Recall our patient

Upright PA Chest X-ray

Axial non-contrast chest CT

Combining the radiologic findings and clinical history, can we narrow our differential of ILD causes?
Recall the Differential Dx for ILD

1) Known causes
   a) Inhaled substances: Inorganic, Silicosis, Asbestosis, Berylliosis
   b) Connective tissue disorders: Systemic sclerosis, Polymyositis, dermatomyositis, lupus
   c) Drugs: Antibiotics, Chemotherapeutic, Anti-arrhythmic drugs

2) Granulomatous
   a) Sarcoidosis
   b) Hypersensitivity pneumonitis

3) Rare DPLDs

4) Idiopathic Interstitial Pneumonias (IIPs) * divided pathologically
   a) UIP – clinical correlate IPF
   b) NSIP

5) “Known” Infections - mimics
   a) Atypical pneumonia
   b) Pneumocystis pneumonia (PCP)
   c) Viral pneumonias (RSV)
Basics of PCP pneumonia

Most common cause of interstitial pneumonia in immunocompromised patients (AIDS, high dose steroids, chemotherapy)

Imaging:
Normal CXR (10-40%)
Often starts out as bilateral reticular infiltrates
Rapidly continues to diffuse airspace disease

Classic CT finding:
- Exudative alveolitis w/ accumulation of fluid, organisms, fibrin, debris in alveolar spaces → ground glass opacity
- Patchy, mosaic appearance of diseased lung next to normal
Basics of PCP pneumonia

Diagnosis: Sputum or BAL – confirmed in our patient

Clinical Course:
Started on high dose bactrim.
Usually responds to bactrim therapy in 5-7 days. Very low levels of resistance.
Redistribution of infection to upper lobes

Complications:
Cystic lung disease
Spontaneous pneumothorax, frequently bilateral (6-7%)

Or.....
Our patient: Worsening CXR on Day 11

Findings:
- Pneumopericardium
- Pneumomediastinum
- Small R apical pneumothorax.

Slight increased opacity projecting over the mid right lung field may represent atelectasis versus early consolidation.

(1) Endotracheal tube
(2) PICC
(3) Nasogastric tube
Our Patient: Progression of ILD on CT

Diffuse central and peripheral ground-glass opacification, with new involvement of the basal segments of the left lower lobe with atelectasis.

Axial non-contrast chest CT
BIDMC, PACS
Our patient: Outcome

Very sadly, after being on the ventilator for 17 days, our patient passed away on 9/14/09.

The final conclusion is that the cause of death was diffuse alveolar damage associated with pneumocystis pneumonia.
1) When giving high dose steroids, always prophylax against opportunistic infections.

2) CXR is a great screening tool for identifying ILD, but does not provide many specific clues about etiology. Always consider the first key branchpoint: acute vs chronic process?

3) Hi-resolution CT scan provides parenchymal detail and can identify patterns to narrow the ILD differential (honeycombing, ground glass). However, pathology, microbiology and other studies are almost always needed to confirm a diagnosis.


Acknowledgements

Erine Yeh

David O’Donnell

Maria Levantakis

Gillian Lieberman