The Journey to a Diagnosis: A Multidisciplinary Approach

Seminar for Patients, Caregivers, and Families

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5/6/2019
Disclosures

• I have nothing to disclose
Why multidisciplinary teams?

• Making an accurate diagnosis of a specific form of interstitial lung disease is challenging

• Pulmonary clinicians, radiologist and pathologists are traditionally separated in a hospital setting

• Agreement between providers and diagnostic confidence improve as more data is available

• Dynamic discussions lead to changes in diagnosis and improved confidence
What is a multidisciplinary discussion?

- Expert ILD clinicians, radiologists, and pathologists integrate all available clinical data, laboratory results, high-resolution computed tomography [HRCT] findings, and lung biopsy [when performed].

- Goals:
  - Diagnosis
  - Therapeutic decisions
  - Discussion of prognosis

Case Presentation
History

• 67 year old man with slow development of dry cough and shortness of breath. Recently diagnosed with IPF
• Medical History: atrial fibrillation, diabetes mellitus
• Medications: apixaban, metformin, pirfenidone
• Allergies: None
• Family history:
  • No family history of ILD, cryptogenic cirrhosis, aplastic anemia, myelodysplastic syndrome, leukemia
  • No family history of early graying
History: Exposures

• **Smoking:**
  • Former smoker – smoked 1 pack per day for 30 years.
  • Quit 20 years ago

• **Environmental:**
  • Down comforter
  • Nightly humidifier use
  • Possible mold in work environment

• **Occupational:**
  • Retired general contractor
Review of Systems

- Constitutional: no weight loss, fevers, chills, night sweats, fatigue
- Head, Eyes, Ears, Nose, Throat: No dry eyes, dry mouth
- Respiratory: **cough, shortness of breath**; No pain with deep breaths, hemoptysis, or wheezing
- Cardiovascular: **palpitations**, no chest pain
- Gastrointestinal: no heartburn, no difficulty swallowing, no nausea, vomiting, diarrhea or constipation
- Musculoskeletal: **back pain, joint pain in hands and fingers, chronic knee pain**
- Skin: No rashes, **Raynaud’s**
Physical Exam

- Eyes: no redness/inflammation, normal vision
- Ears, Nose and Throat: normal salivary pool, good dentition
- Cardiovascular: Irregular heart rhythm, normal heart rate and sounds.
  No leg swelling
- Respiratory: normal effort, no wheezes, rales, or rhonchi, crackles at the bases of the lungs
- Gastrointestinal: abdomen soft, nontender, no masses
- Skin: no rashes, skin dryness or cracking, no changes to the nail bed, thickening or tightening of the skin. No nodules. No clubbing
- Neuro: normal muscle strength, normal reflexes
- Psych: normal mood and affect
- Musculoskeletal: Swelling of the finger joints, right hand and right knee. Tenderness of both wrists and right knee, No warmth or redness.
Diagnostic studies
Diagnostic studies
Diagnostic studies
Diagnostic studies
Information recap

• History:
  • 67 year old male, former smoker
  • Respiratory symptoms + joint pain
  • Down comforter use
  • No clear medication exposures
  • No family history or findings to suggest genetic predisposition

• Exam: crackles, joint tenderness and swelling

• PFTs: Normal lung volumes, reduced diffusion capacity

• HRCT: Definite UIP pattern

• TTE: Normal
Question for Multidisciplinary Conference

• What is the diagnosis?
• What are the next steps? Does he need a surgical lung biopsy?
• What is the appropriate therapeutic options? Should he continue pirfenidone?
What is the diagnosis?

- Idiopathic Pulmonary Fibrosis
- Connective tissue-interstitial lung disease
- *Chronic Hypersensitivity Pneumonitis*
Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
   - ANA - negative
   - RF - positive
   - C-Reactive Protein - elevated
   - Anti-CCP - elevated
Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
2. Hand films
Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
2. Hand films
3. Rheumatology referral – inflammatory arthritis

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Rheumatoid Arthritis
Next diagnostic steps?

1. Blood work to evaluate for an autoimmune disease
2. Hand films
3. Rheumatology referral – inflammatory arthritis
   \[ \rightarrow \]
   Rheumatoid Arthritis

4. Surgical lung biopsy???
Rheumatoid Arthritis - Interstitial Lung Disease

• Rheumatoid arthritis is an inflammatory and autoimmune disease ~1% of the population

• Up to 60% of patients with RA have interstitial lung disease, but a minority of those patients will develop clinically significant disease

• Diagnosis:
  • Various histologic patterns
  • Various pathologic patterns

• Variable response to treatment and clinical course

Olson et al. Am J Respir Crit Care Med. 2011
Therapeutic Options: Anti-inflammatory

• Pretreatment:
  • Baseline blood work
  • Evaluate for infectious risks: hepatitis B and C, latent tuberculosis
Therapeutic Options: Anti-inflammatory

• Pretreatment:

• Types of Therapies
  • Rapid acting: NSAIDs and glucocorticoids
  • Corticosteroid sparing agents: mycophenolate mofetil, azathioprine, cyclophosphamide
  • Disease-modifying anti-rheumatic drug (DMARD): hydroxychloroquine, leflunomide*, methotrexate*
  • Biologics:
    • Anti-TNF therapy: etanercept*, infliximab*, adalimumab*
    • Abatacept
    • Rituximab
Therapeutic Options: Additional Therapies

• Pirfenidone and nintedanib?
  • Under investigation as a therapeutic option for patients with RA-ILD
    (ClinicalTrials.gov identifier: NCT02808871)

• Smoking cessation
• Supplemental oxygen
• Pulmonary rehabilitation
• Lung transplantation
Take Home Points

• Interstitial lung diseases are challenging to diagnose
• Increased information (clinical, physical exam, blood work, diagnostic testing) improves agreement amongst providers
• Multidisciplinary approach improves the accuracy and confidence of a diagnosis
Thank you for involving us in your care

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