Multidisciplinary Diagnosis in Action: Challenging Case Presentations

Interstitial Lung Disease: Advances in Diagnosis and Management
UCSF CME
November 8, 2014

Case 1 – 69 yo M

- 3 year history of intermittent cough and exertional dyspnea
- Over prior 2 years, increased functional limitation, now SOB walking across room
- New exertional hypoxemia

- ROS: Negative, no reflux symptoms or symptoms of connective tissue disease
69 yo M

• PMHx
  – R-sided primary spontaneous pneumothorax in his 20’s, pleurectomy
  – GERD

• NKDA

• Medications
  – Pantoprazole 20 mg daily
  – ASA 81 mg daily
  – Losartan 50 mg daily

Environmental / Occupational History

• Down comforter, used in winter
• No birds
• No other mold exposures
• No occupational exposures
• Works in media and runs a non-profit organization
69 Yo M

• Social Hx
  – Lifetime non-smoker
  – No alcohol or illicit drug abuse

• FHx:
  – No known ILD
  – Mother had PMR

Physical Exam

• BP 120/56, HR 65, T 36.9, RR 18, Sats 96% on R/A
• Mild bibasilar velcro crackles R>L
• Otherwise unremarkable
• Normal cardiac exam
• No clubbing
• No signs of PH
• No signs of CTD
Physiology

- Oxygen saturation 97% on R/A at rest but decreased to 85% with 3 mins walking

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<tr>
<td>March 4, 2014</td>
<td>3.19 L (67%)</td>
<td>2.72 L (77%)</td>
<td>74%</td>
<td>4.90 L (66%)</td>
<td>11.97 (35%)</td>
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<tr>
<td>May 23, 2011</td>
<td>3.86 L (80%)</td>
<td>3.15 L (88%)</td>
<td>85%</td>
<td>5.24 L (71%)</td>
<td>15.48 (44%)</td>
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Labs

- June 2011
- ESR 31 mm/hr
- Negative - Scl-70, ds-DNA, C3, C4, D-dimer, Jo-1, anti-centromere Ab, ACE level, anti-CCP Ab
MDD discussion

• Dx – unclassifiable without additional information

• We recommended SLB

Surgical Lung Biopsy

• 3 surgical biopsies from the left lung
  – All lower lobe (basilar anterior x 2 and superior)
  – Discussed during surgery
Case 1 Diagnosis

- Interstitial fibrosis, usual interstitial pneumonia pattern, with increased peribronchiolar fibrosis (basilar).

- Interstitial fibrosis, predominantly bronchiolocentric, with small non-necrotizing granulomas (superior).

- Overlapping features that suggest chronic hypersensitivity pneumonia versus usual interstitial pneumonia.
Multidisciplinary Decision

- Based on clinical, radiologic and surgical lung biopsy, felt to be most consistent with hypersensitivity pneumonitis
# Multidisciplinary Management Plan

- Remove down comforter from house
- HP checklist
- Professional home cleaning
- Prednisone 50 mg daily x 4 week then taper by 5 mg/week until on 20 mg and return with PFTs after 3 months of treatment
- Rehab, vaccines
- Serial PFTs

# Final Thoughts from the Panel
Case 2 - 77 yo M

- Developed dry cough that started 5 years ago
- Denies dyspnea, although he has "slowed down" over the last several years.
- Denies constitutional symptoms
- No reflux
- Denies myalgias, arthralgia, sicca, Raynaud’s phenomena, rash, skin changes, joint stiffness or muscle weakness

77 yo M

- PMHx
  - BPH
- NKDA

- Medications
  - Finasteride 5 mg daily
  - Tamsulosin 0.4 mg daily
Environmental / Occupational History

- Retired factory (HVAC) and field worker
- Owned parakeets and cockatiels for past 2-3 years; kept outside away from home
- Changes the cage q 2 weeks; feeds them on alternating days. Does not handle the birds
- Woodworking as a hobby (builds furniture)

77 yo M

- Social Hx
  – Minimal smoking history
  – No alcohol or illicit drug use

- Family Hx
  – No ILD or CTD
Physical Exam

- BP 128/69, HR 58, T 36.5, RR 20, Sats 99% R/A
- Bilateral inspiratory crackles at both bases
- No clubbing
- Negative rheumatologic exam
- No signs of pulmonary hypertension

Case 2 - PFTs

- Office walk test: Room air saturation at rest 97%, after 3 minutes walking 96%

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<td>June 24, 2014</td>
<td>3.70 L (117%)</td>
<td>2.90 L (118%)</td>
<td>78%</td>
<td>5.86 L (92%)</td>
<td>17.16 (62%)</td>
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<td>July 12, 2013</td>
<td>3.81 L (119%)</td>
<td>3.18 L (128%)</td>
<td>84%</td>
<td>5.61 L (88%)</td>
<td>19.30 (70%)</td>
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Surgical Lung Biopsy

- Two biopsies, right upper and lower lobes.
  - 3 x 1 cm
  - “Similar wedge that is approximately 5 cm”
Case 2 - Diagnosis

- Interstitial fibrosis, usual interstitial pneumonia pattern.
- Minor component of chronic inflammation, but associated with the regions of fibrosis.
Initial Multidisciplinary Discussion

• Given clinical, radiology and surgical lung biopsy, felt the diagnosis is idiopathic pulmonary fibrosis

Multidisciplinary Management Plan

• Remove birds
• Pulmonary rehab
• Serial PFTs
• Vaccines
• Baseline serologies
Labs

- ANA 1:640 (speckled)
- RF 125
- SSA >150
- SSB 145
- Anti-CCP, Scl-70, SM antibody, Sm/RNP antibody, PM-Scl antibody negative
- Myositis panel negative
  - EJ, Jo-1, KU, MI-2, OJ, PL-12, PL-7, SRP autoantibodies
Multidisciplinary Management Plan

- Lip biopsy to r/o Sjogren’s
- Rheumatology consult

Final Thoughts from the Panel
Case 3 - 46 yo M

- Developed exertional dyspnea and non-productive cough in spring 2013
- Progression of dyspnea over past year
- Cardiac work up including angiogram was negative
- Endorses symptoms of reflux
- No CTD symptoms except dry mouth

- Presented to ER Feb 2014 with worsening SOB, treated with solumedrol, antibiotics. CT and SLB arranged.
- Discharged home on prednisone, PPI and NAC

46 yo Male

- PMHx
  - HTN
  - T2DM
  - Dyslipidemia
  - GERD
  - Renal cyst
  - Periodic limb movement disorder
  - B12 deficiency
  - AVN of hip with hip replacement
  - OSA

- Medications
  - Diltiazem 180 mg daily
  - Lovastatin 40 mg daily
  - Metformin 1000 mg BID
  - Omeprazole 20 mg daily
  - Prednisone 10 mg daily

- NKDA
Environmental / Occupational History

• Down pillow
• No birds
• Regular humidifier use
• Mold on central air filters
• Worked as a manager and engineer for construction companies on dusty work-sites

46 yo M

• Family Hx
  – Father - “black lung”
    • Died at 41
  – 2 paternal uncles, 1 aunt with ILD
    • Died at 46, 46, and 51
  – Grandmother – ILD
    • Died 72
  – History of early graying in 30s

• Social Hx
  – Life long non-smoker
  – No illicit drugs
  – 7 alcohol units weekly
Physical exam

- BP 137/88, HR 117, RR 18, T 36.4, Sats 96% R/A
- No clubbing
- No crackles, wheeze
- No signs of pulmonary hypertension
- No signs of connective tissue disease

Physiology

Room air saturation was 94% at rest. After 3 minutes of walking, oxygen saturation was 92%.

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<td>March 3, 2014</td>
<td>2.43L (53%)</td>
<td>2.16L (58%)</td>
<td>89%</td>
<td>3.55L (54%)</td>
<td>14.4 (46%)</td>
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<td>May 11, 2011</td>
<td>3.10 L (60%)</td>
<td>2.63L (64%)</td>
<td>85%</td>
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Labs

- WBC 13, Hb 12.6, Plt 165
- CPK normal
- ANA, RF, ANCA negative
- HP panel negative
- ACE normal
Surgical Lung Biopsy

- Left upper lobe surgical biopsy
  - 3.6 x 1.6 x 0.7 cm
Case 3 - Diagnosis

• Cellular and fibrosing nonspecific interstitial pneumonia with focal bronchiolocentric fibrosis and organizing pneumonia.

• Alveolar hemorrhage, likely procedure-related.

• Connective tissue disease, HP, drug reaction, or familial ILD (known at time of dx).
Multidisciplinary Discussion

- Unclassifiable ILD
- Differential includes
  - Chronic hypersensitivity pneumonitis (HP)
  - Fibrotic nonspecific interstitial pneumonia (NSIP)
- With a familial predisposition
Case 3 - Management

- Pulmonary rehab, Vaccines
- Mycophenolate (MMF) 1000 mg twice daily
- Wean prednisone once on full dose of MMF
- Continue PPI
- Weight loss
- Remove down and humidifier
- Serial PFTs
- Repeat serologies (Negative)

Final Thoughts from the Panel