Déjà vu all over again

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Disclosures

• None

HPI

• 49 y/o woman presents for lung transplant evaluation for Hypersensitivity Pneumonitis

  • Exposures: Open cheese whey pond behind home; pigeons

  • Serologies: +avian and +fungal precipitins

HPI continued

• Pond cleaned up, symptoms persist

• Pt moved to another town in same region

• Treated intermittently with prednisone and azathioprine over 18 years

• Periodic respiratory decompensation, unknown triggers

• At transplant eval, on 6LPM
Physiologic Data

**PFTs:**
- FEV\textsubscript{1} 0.74 L (27%)
- FVC 0.88 L (25%)
- TLC 2.39 L (44%)
- DLCO: could not obtain

Bilateral lung transplant: 3/2009

- Uneventful operative course
- Immunosuppression: Tacrolimus, prednisone, mycophenolate mofetil

CT: Airtrapping, Bronch: purulence, aspergillus+
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Needs reevaluation...
Ideas on what might be going on?

Approach to acute allograft dysfunction

• Goal: Identify reversible cause
  • Airway issue
  • Infection
  • Acute rejection
  • Drug reaction

• Identify reversible cause of allograft dysfunction
  • Airway issue
  • Infection
  • Acute rejection
  • Drug reaction
• Negative workup = Chronic Lung Allograft Dysfunction
  • Bronchiolitis obliterans syndrome (BOS)
  • Restrictive allograft syndrome (RAS)
  • Acute fibrinoid organizing pneumonia (AFOP)
Dynamic CT scan with air trapping

Transbronchial Biopsy

Diagnosis: Recurrent HP

- Pulsed with 40 mg of prednisone for a month followed by taper
- Upon direct questioning, patient noted pigeons roosting outside her window
- Son put chicken wire around window sill, ineffective at keeping away pigeons
Further Follow-up

- Patient moves...again
- All subsequent biopsies negative for granulomas
- Lung function stabilizes
- Last seen 3/2015- doing well with stable DOE

Disease recurrence in the allograft

- 1,394 lung transplant recipients
- Recurrence in 15 (1%)

<table>
<thead>
<tr>
<th>Disease</th>
<th>Number</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sarcoidosis</td>
<td>9/26</td>
<td>35%</td>
</tr>
<tr>
<td>LAM</td>
<td>2/21</td>
<td>10%</td>
</tr>
<tr>
<td>Pulmonary Langerhans</td>
<td>1/4</td>
<td>25%</td>
</tr>
<tr>
<td>Talc Granulomatosis</td>
<td>1/1</td>
<td>100%</td>
</tr>
<tr>
<td>Diffuse Panbronchiolitis</td>
<td>1/1</td>
<td>100%</td>
</tr>
<tr>
<td>Alveolar Proteinosis (PAP)</td>
<td>1/2</td>
<td>50%</td>
</tr>
</tbody>
</table>

Collins, Radiology 2001

Other published case reports

- Sarcoidosis
- Lymphangioleiomyomatosis (LAM)
- Pulmonary Langerhans cell histiocytosis (PLCH)
- Talc granulomatosis
- Diffuse panbronchiolitis
- Pulmonary alveolar proteinosis (PAP)
- Desquamative interstitial pneumonitis (DIP)
- Giant cell interstitial pneumonia (Hard metal lung/Cobalt)
- Alpha-1 antitrypsin deficiency related COPD
- Bronchoalveolar carcinoma

~30 studies reported recurrence of primary disease after transplant
Hypersensitivity Pneumonitis

- Extrinsic allergic alveolitis caused by inhaled organic particles
  - > 300 potential causal agents
- Antigen unknown ~40% of cases
  - Avoidance difficult if agent unknown
- HP = 1% of lung transplants performed in U.S.
  - No published reports on outcomes

Selman, AJRCCM 2012

Lung Transplantation for HP at UCSF

- Retrospective cohort study of LTx at UCSF between 2000-2013
- All LTx for ILD re-reviewed by multidisciplinary ILD approach
  - Clinical history, radiology, VATS and explant pathology

Kern RM. CHEST. 2014

Lung Transplantation for HP at UCSF

- Of 406 LTx, 31 were for HP (8%)
- Only 23 of 31 HP cases were diagnosed before LTx (74%)
- 2 definitive, 1 probable case of recurrence after LTx

Kern RM. CHEST. 2014
Survival after LT for HP is quite good

Conclusions

• HP is a surprisingly common indication for transplant
  • Hard to diagnose accurately
• HP can recur after lung transplant
  • Identifying antigen is important
  • Be aware that it is possible: immunosuppression is not protective

Thank you.