Pulmonary Infiltrates with Eosinophilia Presenting as Heart Failure

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TEACHING POINTS

- Identifying the underlying etiology of pulmonary infiltrates with eosinophilia (PIE) is critical to prompt institution of appropriate treatment.
- Reliance on histology and traditional serological markers may result in failure to establish a timely diagnosis.

CASE

HPI: A 76 year-old female presented with two months of dyspnea on exertion. She was admitted with non-ST elevation myocardial infarction, new onset heart failure and bilateral pulmonary infiltrates on chest X-ray.

PMH: Included severe asthma diagnosed at age 59 and requiring frequent intermittent steroids, recurrent acute sinusitis, allergic rhinitis, a prior stroke attributed to cerebral venous thrombosis, chronic right foot drop, and a 25 pound weight loss over 1 year.

EXAM: She was cachectic with temporal wasting. JVP was elevated to 10cm. Fine inspiratory bibasilar lung crackles were auscultated. There was 1/5 strength on right foot dorsiflexion, and 3/5 on the left. Sensation to pinprick was reduced in both feet.

LABS

Chemistry:

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Na</td>
<td>134</td>
<td>136-145</td>
</tr>
<tr>
<td>K</td>
<td>3.3</td>
<td>3.5-5.3</td>
</tr>
<tr>
<td>Cl</td>
<td>10.7</td>
<td>10.6-11.2</td>
</tr>
<tr>
<td>Ca</td>
<td>2.7</td>
<td>2.6-3.6</td>
</tr>
<tr>
<td>Mg</td>
<td>1.4</td>
<td>1.5-2.2</td>
</tr>
<tr>
<td>TP</td>
<td>12</td>
<td>6.5-8.5</td>
</tr>
<tr>
<td>ALP</td>
<td>207</td>
<td>30-150</td>
</tr>
<tr>
<td>AST</td>
<td>102</td>
<td>0-36</td>
</tr>
<tr>
<td>ALT</td>
<td>27</td>
<td>0-35</td>
</tr>
<tr>
<td>BUN</td>
<td>10.7</td>
<td>1.3-2.1</td>
</tr>
<tr>
<td>Cr</td>
<td>0.7</td>
<td>0.6-1.4</td>
</tr>
<tr>
<td>T Chol</td>
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<td>150-250</td>
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<tr>
<td>HDL Chol</td>
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<td>35-130</td>
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<tr>
<td>LDL Chol</td>
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<td>70-160</td>
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<tr>
<td>MCV</td>
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<td>75-100</td>
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<tr>
<td>ESR</td>
<td>0</td>
<td>0-150</td>
</tr>
<tr>
<td>CRP</td>
<td>0.2</td>
<td>0-0.8</td>
</tr>
</tbody>
</table>

Tryptase: 5.8 µg/L (normal), Vitamin B12: 402 pg/mL (normal)

Immunologic:

- Serum IgE: 187 IU/L
- ANA, c-ANCA, and p-ANCA: negative
- FISH for 4q13 anomalies (FIP1L1/PDGFRα, PDGFRβ): normal

Infectious:

- Stool examination for ova and parasites: negative
- Aspergillus spp. Antibody and Coccidioides serology: negative

Procedures:

- Cardiac catheterization confirmed dilated cardiomyopathy without significant coronary artery disease
- EMG confirmed mononeuritis multiplex
- Sural nerve biopsy showed no inflammatory or eosinophilic infiltrates
- Bronchoscopy showed normal mucosa and airways. BAL and transbronchial biopsy were normal
- Bone marrow biopsy & flow cytometry were negative for malignancy

In 2005, mediastinal windows were unremarkable. By 2008, there has been interval development of anterior pericardial thickening, posterior pericardial effusion, and bilateral pleural effusions.

IMAGES

CT CHEST 08/2005

CT CHEST 11/2008

HOSPITAL COURSE

After her initial admission, she was discharged with an outpatient referral to pulmonology for workup of PIE syndrome.

- Suspicion for Churg Strauss Syndrome (CSS) or Hypereosinophilic Syndrome (HES) was high
- Lung biopsy was planned, but this was precluded by a marked clinical deterioration due to her cardiomyopathy and rapidly progressive neuropathy
- She was readmitted to the hospital and started on systemic corticosteroid therapy
- Dramatic clinical improvement ensued with resolution of her heart failure and stabilization of her peripheral neuropathy
- On follow-up one year later, her eosinophil count remains less than 300/microlL

DISCUSSION

- Eosinophilic lung disorders comprise a heterogeneous group of diseases. These include both primary & secondary processes such as:
  - Helminthic and Nonhelminthic Infections
  - Medications and Toxins
  - Malignancy
  - Allergic Bronchopulmonary Aspergillosis
  - Acute and Chronic Eosinophilic Pneumonias
  - Churg Strauss Syndrome (CSS)
  - Hypereosinophilic Syndrome (HES)

- Multifactorial involvement should prompt consideration of CSS & HES
- Cardiac and neurologic involvement are major causes of morbidity and mortality in both HES and CSS
- This patient met the American College of Rheumatology criteria for a diagnosis of CSS
- CSS is uniformly fatal without treatment. 50% of whom will die within 3 months after the onset of vasculitis
- Studies suggest that ANCA negative CSS patients are more likely than ANCA positive patients to have cardiac involvement, and that vasculitis is less frequently captured histologically in ANCA negative patients
- This emphasizes the need for clinicians to maintain an awareness of this disease, despite its rarity, as relying on traditional serological markers may result in failure to identify and promptly treat these high risk CSS patients

REFERENCES