A Curious Case of Dyspnea

Maine Chapter ACP Conference
Sean Lena, PGY 3
September 22, 2018
76 y/o woman who presents to the ER with progressive shortness of breath
HISTORY

- Started 4 weeks prior
- SOB with exertion
- SOB when lying flat
- Uses 3 pillows to sleep
- Bilateral leg swelling
- History of DVT
HISTORY

- No fever, rigors
- No cough, sputum
- No sick contacts
- No Vomiting or Diarrhea
- No Dysuria
- No recent trauma
Medical History:
- Hypothyroid
- Polycythemia Vera, JAK2+
- On experimental chemotherapy at Dana Farber
- Osteoporosis
- Remote DVT—Tx Warfarin

Social:
- Non Smoker
- No alcohol or drug use
- Married
- Lives in Poland, Maine
- Two healthy adult children

Medications:
- Allopurinol
- Iron
- Folic Acid
- Levothyroxine
- Omeprazole
- Lasix
- Potassium
- X-25

Family History:
- Father: Renal Failure
- Mother: CHF
PHYSICAL EXAM

- Hypoxic (80% on RA)
- 3/6 systolic murmur, RUSB
- 12 cm JVD
- Bibasilar crackles
- Venous stasis changes
- +2 Pitting Edema
**LABS**

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<th>CMP</th>
<th>139</th>
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<th>44</th>
<th>110</th>
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<td></td>
<td>4.9</td>
<td>20</td>
<td>1.5</td>
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Alk Phos: 135  
AST: 13  
ALT: 7

NT Pro BNP: **11,621**  
TSH: 14.99  
D Dimer: 213

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Questions?
Differential Diagnosis
Flattened Septum

RV

LV

Posterior Pericardial Effusion
Right Heart Catheterization

- Elevated right atrial pressure 20mmHg
- Normal wedge pressure (14 MMHG)
- Elevated pulmonary artery pressures/resistance
- PVR (PA=60/27/39) PVR 460 ARU
- Normal Fick and Cardiac output
- Non responder to inhaled Nitric oxide
Questions?
Right Heart Failure Secondary to Pulmonary Hypertension
Dr. Abel Ayerza
1. Pulmonary Arterial Hypertension
   - Idiopathic
   - Heritable

2. PH Due to Left Heart Disease
   Systolic/Diastolic HF, Valvular disease

3. PH Due to lung disease/hypoxia
   COPD
   Interstitial Lung Disease
   Obstructive Sleep Apnea

4. Chronic Thromboembolic Pulmonary Hypertension

5. PH Due to Multifactorial/Unclear Mechanisms
   - Systemic Disorders
     Sarcoi, Neurofibromatosis, Vasculitis
   - Hematologic
     Chronic Hemolytic anemia
     Myeloproliferative disease

2. PH Due to Left Heart Disease
   Systolic/Diastolic HF, Valvular disease

Image: Pulmonary hypertension: diagnosis and management, BMJ 2013; 346 16 April 2013)
1. Pulmonary Arterial Hypertension
10%

2. PH Due to Left Heart Disease
60%

3. PH Due to lung disease/hypoxia
20%

4. Chronic Thromboembolic Pulmonary Hypertension
10%

5. PH Due to Multifactorial OR Unclear Mechanisms

10% Combination

Image: Pulmonary hypertension: diagnosis and management, BMJ 2013; 346 16 April 2013)
Polycythemia Vera and Pulmonary Hypertension

- Polycythemia results from clonal proliferation of myeloid cells, mostly RBC
- Mutation of JAK/STAT pathway
- Polycythemia Vera and other myeloproliferative disorders are associated with pulmonary hypertension
- Etiology is multifactorial and incompletely understood
- Incidence is unclear due to lack of data, some studies have estimated 30-50%
- Presence of PH is frequently associated with worse outcomes
Management

- Optimal treatment in these patients is not well defined, but depends on underlying etiology
- If resulting from CTEPH, pulmonary endarterectomy, anticoagulation
- If drug induced, stop the offending agent
- Treat underlying cause with cytoreductive therapy
- Not amenable to treatment with central vasodilators
- Symptomatic management
- Prognosis is poor, survival of 6-9 months,
Clinical Course:

- Negative VQ Scan Ruled out CTEPH--Not started on systemic anticoagulation
- Workup for Other Causes of PH: HIV, ANA, RF, Sjogrens, SCL 70—Negative
- Patient was discharged with follow up with her oncologist, chest medicine associates, home oxygen, and diuretics
- Opted for palliative management of her disease, died in March 2018
Takeaway Points

- Pulmonary hypertension is defined as a sustained elevated mean pulmonary artery pressure $\geq 25$ mmHg, or pulmonary vascular resistance $> 3$ Wood units by RHC.
- PH is classified into 5 groups, divided into shared pathophysiology and clinical features.
- Right heart catheterization is the gold standard for diagnosis of PH.
- Myeloproliferative disorders are associated with pulmonary hypertension.
- Prevalence is unknown, pathophysiology is multifactorial and poorly understood.
- Treatment is aimed at the underlying etiology and the prognosis is poor.
References


Thank You!