The Boy’s Not All Right

Aaron St-Laurent
Montreal Children’s Hospital - Pulmonology
Cross Canada Rounds
Case

- Adolecent patient presenting with massive hemoptysis
Differential Diagnosis

- Rheumatologic disorders:
  - Granulomatosis and polyangiitis, anti-GBM disease, SLE, microscopic polyangiitis, pulmonary capillaritis
- Ateriovenous malformation
- Oncologic / Hematologic
- Tuberculosis and infectious
- Inhalant abuse
- Idiopathic pulmonary hemosiderosis
- Trauma
- Foreign Body
- Bronchiectasis
- CPAM
- Pulmonary hypertension
- Pulmonary embolus
Systemic artery to pulmonary vein
arteriovenous malformations
(SP-AVM)
A case-based review of the literature
SP-AVM

• SP-AVMs extremely rare

• 3-4% of pulmonary AVMs
  – Dines\textsuperscript{1} - 3/101
  – Bosher\textsuperscript{2} - 12/350

• Systemic connections
  – Most frequently internal mammary arteries\textsuperscript{3}
  – Anomalous aortic branches, bronchial, intercostal, lateral thoracic, esophageal

“Typical” Pulmonary AVM

- Pulmonary artery to pulmonary vein
- Cyanotic
Systemic to Pulmonary AVM

- Normal systemic artery to pulmonary vein
- Aberrant aortic artery to pulmonary vein
- Acyanotic

http://www1.imperial.ac.uk/resources/52F91CDC-2A31-43C7-B644-C79BF10E162D/pavms.jpg
# Difference Between Pulmonary AVM and SP-AVM

<table>
<thead>
<tr>
<th>Pulmonary AVM</th>
<th>Systemic to pulmonary AVM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female &gt; Male</td>
<td>Male &gt; Female</td>
</tr>
<tr>
<td>Related to HHT</td>
<td>No relationship</td>
</tr>
<tr>
<td>Right-to-left shunt</td>
<td>Left-to-left shunt</td>
</tr>
<tr>
<td>Single or multiple</td>
<td>Single</td>
</tr>
<tr>
<td>Local or diffuse</td>
<td>Local</td>
</tr>
</tbody>
</table>

SP-AVM - Presentations

• Not associated with hereditary hemorrhagic telangiectasia
  – No telangiectasia

• Does not present with classic triad of PAVM
  – Cyanosis
  – Clubbing
  – Desaturation
SP-AVM - Presentations

• Physical exam
  – Continuous systolic murmur
  – Bounding pulses

• Asymptomatic
  – Incidental finding

• Symptomatic
  – Heart failure
  – Hemoptysis
SP-AVM - Presentations

Chest radiographs
• Typically abnormal, often non-specific
• Single lesion
• May involve more than one lobe
Presentation – Hemoptysis

- 21 y.o. ♂
- Hemoptysis - 600cc upon awakening
- On exam:
  - Crackles to right base
  - No murmur
- Bronchoscopy – clots in RLL
- Diagnosis – Bronchial artery to pulmonary vein
- Treatment – bilobectomy

Presentation – Asymptomatic

- 6 y.o. ♀
- Continuous murmur, misdiagnosed as PDA
- Admitted for cardiac catheterization
- Hypertensive 130/60 with bounding pulses
- ECG: Left ventricular hypertrophy
- Diagnosis – SP-AVM, abnormal aortic branch
- Treatment – Left lower lobectomy, ligation of artery

Increased density behind heart

Presentation – Heart Failure

- Term neonate
- APGARS 5 at 1 min, 3 at 5 min
- Complicated neonatal course
  - Congestive heart failure, sepsis, mechanical ventilation, seizures, cerebral edema
- At 6 weeks
  - Congestive heart failure
  - Biphasic stridor
  - Tachypnea, Tachycardia
- Bronchoscopy – extramural vascular compression
- Diagnosis - 2 aberrant aortic vessel from descending aorta to pulmonary vein

SP-AVM - Diagnosis

Traditionally – selective angiography of systemic artery

– Contrast or bubble contrast
– Return to pulmonary vein, left atrium

SP-AVM - Diagnosis

CT-angiography with 3D reconstruction

– Generally still confirmed with selective angiography

Fig. 2 – a–c: CT angiography with 3D volume rendering showing aberrant vessels originating from the left subclavian artery and abdominal aorta to the entire left lung which drained into the left atrium via normal pulmonary veins.

SP-AVM Management

- Historically: Surgical
  - Surgical ligation
  - Lobectomy, only if lung damaged\(^7\) or ligation not possible

- Interventional Radiology
  - Coil embolization
  - Vascular plug

SP-AVM Management

A review of 21 cases of SP-AVM

<table>
<thead>
<tr>
<th>Management</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lobectomy</td>
<td>1</td>
</tr>
<tr>
<td>Surgical Ligation</td>
<td>11</td>
</tr>
<tr>
<td>Lobectomy and Surgical Ligation</td>
<td>3</td>
</tr>
<tr>
<td>Interventional</td>
<td>4</td>
</tr>
<tr>
<td>Refused Treatment</td>
<td>2</td>
</tr>
</tbody>
</table>

Interventional Management

Coil Embolization\(^6\)

Vascular Plug & Coil\(^4\)


Risks of Embolization

• Treatment failure
  – Acute
  – Recurrence

• Migration of coil, material systemically → stroke
  – Higher risk than pulmonary AVMs
Summary - I

- Extremely rare, not associated with HHT
- Often asymptomatic, incidental finding
- May be hemodynamically significant SP-AVMs
- Not associated with cyanosis or increased pulmonary blood flow
- Theoretically increased risk of hemoptysis compared to other pulmonary AVMs
Summary - II

• Diagnosis: Selective angiography – passage of contrast to pulmonary vein and left atrium

• Management:
  – Arterial ligation
  – Lobectomy if lung damaged, ligation not possible
  – Interventional radiology used increasingly