5 Consecutive Cases of Pulmonary AVM

James R. Gossage, MD
Georgia Regents University
Augusta, Georgia
Case MA 1994

History:

- 42 yo white man
- Hospitalized for ARDS and found to have L lung nodule (?since 1983)
- PHx: GI bleeding from NSAIDS
- No hemoptysis, SOB, nosebleeds
- SHx: prior smoker, furniture refinishing
- Mom has GI AVM
Case MA 1994

Exam:

• Normal vital signs
• 10-15 telangiectasias on lips, tongue, and fingers
• Lungs clear
• Heart and abdomen normal
• No leg edema or finger clubbing
Case MA 1994
Pulmonary angiogram
Coil Embolization of PAVM
Case MA 1994

Follow up:

• Diagnosis: HHT complicated by PAVM
• No recurrence of lung AVM despite repeat CT scans over 20 years
• Developed AVM in multiple other organs
• Recurrent severe GI bleeds from AVM
• Died 2013 from GI bleeding
Pulmonary AVM “Lingo”

**Definition:**
- An abnormal communication between pulmonary arteries and pulmonary veins.

**Terminology:**
- Pulmonary AV fistulas.
- Pulmonary telangiectasias or spiders.
- Cavernous angiomas or hemangiomatas of the lung.
**Pathology of PAVM**

- Single in 64% of patients
- Unilateral in 75% of cases
- 53-70% located in lower lobes
- 81% involve the pleura surface
- 7-11% of patients have diffuse microvascular PAVM (especially in HHT)
- 95% of PAVM are supplied by PA branches
Case SS 2008
### Symptoms of PAVM

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Range</th>
<th>Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptomatic</td>
<td>44-91</td>
<td>76</td>
</tr>
<tr>
<td>Epistaxis (HHT)</td>
<td>29-79</td>
<td>55</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>32-71</td>
<td>53</td>
</tr>
<tr>
<td>Hemoptysis</td>
<td>5-15</td>
<td>12</td>
</tr>
</tbody>
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Gossage, *AJRCCM* 1998; 158:643
## Signs of PAVM

<table>
<thead>
<tr>
<th></th>
<th>Range</th>
<th>Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>Telangiectases</td>
<td>34-79</td>
<td>53</td>
</tr>
<tr>
<td>Lung bruits</td>
<td>29-67</td>
<td>43</td>
</tr>
<tr>
<td>Clubbing</td>
<td>13-54</td>
<td>27</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>16-29</td>
<td>24</td>
</tr>
</tbody>
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Gossage, *AJRCCM* 1998; 158:643
Contrast Echo

Procedure:

• Rapid injection of agitated saline into peripheral IV
• Look for 1 or more bubbles in the LV
• Early bubbles (≤1 beat) suggest PFO
• Delayed bubbles (after 3 beats) are 99% sensitive for presence of PAVM
• 6% of normal people have a low grade delayed shunt

van Gent, Chest 2010; 138:833
Contrast Echo

Results:

• Grade 1  <30 bubbles
  ▷ Treatable PAVM in <1%
  ▷ CT chest not usually recommended

• Grade 2  30-100 bubbles
  ▷ Treatable PAVM in 17%

• Grade 3  >100 bubbles
  ▷ Treatable PAVM in 53%

van Gent, Chest 2010; 138:833
COOK GUIDE + 5 Fr. COAXIAL

NESTER COIL DEPLOYING
Standard Push Coils

Cannot reposition once deployed
PAVM - Magnified View

Catheter in Pulm artery

Draining vein
PAVM - Coil Placed

Coil in feeding artery
PAVM - Flow Persists
PAVM – Final Occlusion!
Amplatzer Vascular Plug

- Used in large feeding arteries
- Can reposition before deploying
- Used alone or with coils
## Treatment of PAVM

<table>
<thead>
<tr>
<th></th>
<th>Stroke</th>
<th>Brain abscess</th>
<th>Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>Untreated</td>
<td>0-50%</td>
<td>8-50%</td>
<td>8-55%</td>
</tr>
<tr>
<td>Surgery</td>
<td>0-4%</td>
<td>0</td>
<td>0-9%</td>
</tr>
<tr>
<td>Embolization</td>
<td>1%</td>
<td>1%</td>
<td>0</td>
</tr>
</tbody>
</table>

Gossage, *AJRCCM* 1998; 158:643
Case PS 2008

History:

- 64 yo white woman
- Referred for incidental PAVM seen on abdominal CT after car accident; rib fractures but no penetrating trauma
- ROS: no nosebleeds, SOB, or bleeding
- FHx: son had ICH after getting kicked by horse; no one with nosebleeds
History:

- 64 yo white woman
- Referred for incidental PAVM seen on abdominal CT after car accident; rib fractures but no penetrating trauma
- ROS: no nosebleeds, SOB, or bleeding
- FHx: son had ICH after getting kicked by horse; one son with occasional nosebleeds
Exam:

- BP 150/71, otherwise normal vital signs
- 9 tiny telangiectasias on lips, tongue, and fingers
- Lungs clear
- Heart and abdomen normal
- 1+ leg edema; no finger clubbing
Case PS 2008

Labs:

- ABG on 100% oxygen: 5.9% shunt
- Echo w contrast: normal EF, grade 2/3 delayed shunt, normal RVSP
- MRI brain: several lesions suggestive of recent trauma; no AVM
Case PS 2008
Case PS 2008

Additional f/u:

• Exam of both sons showed typical telangiectasias and nosebleeds
• Son with ICH was kicked by horse 3 years earlier; could never get MRI
• She almost certainly has HHT
• AVM embolized successfully but patient and family lost to follow up
Epidemiology of PAVM

- 4.5 cases/year at Mayo clinic
- Twice as common in women
- 90-95% of patients with PAVM have hereditary hemorrhagic telangiectasia (HHT)
- Other causes: liver disease, trauma, actinomycosis, bronchiectasis, idiopathic
Hereditary Hemorrhagic Telangiectasia

• Aka Osler-Weber-Rendu
• Autosomal dominant inheritance; 5 genes
• US incidence 1 / 5,000
• Epistaxis initially in 90% of patients
• 50-80% with mucocutaneous telangiectasias
• 50% develop pulmonary AVM
• 50-80% develop other internal AVM including GI tract, liver, and brain
Diagnosis of HHT

Definite = 3 or more of the following:

- Spontaneous and recurrent nosebleeds
- Telangiectasias at characteristic sites
- Organ manifestations: pulmonary, gastrointestinal, cerebral, hepatic AVM
- Affected first degree relative

Case CH 2014

History:

• 36 yo white woman
• Hospitalized for ruptured PAVM with hemothorax 3 mo earlier while 26 weeks pregnant
• L thoracotomy with PAVM embolization
• Weekly nosebleeds since childhood
• Strong family history of HHT
Case CH 2014

Exam:

• Normal vital signs
• 6 telangiectasias on lips, tongue, and fingers
• Lungs clear
• Heart and abdomen normal
• No leg edema or finger clubbing
**Case CH 2014**

**Labs:**

- Echo w contrast: normal EF, grade 2/3 delayed shunt, normal RVSP
- Recent CT chest: closed LUL AVM, small LUL AVM, RUL AVM with 3mm feeding artery, probable liver AVM
- Pulse ox: 97% supine, 95-98 seated
- MRI brain: normal
Case CH 2014

Plan:

• Schedule embolization of RUL AVM
• Antibiotics before dental hygiene using AHA bacterial endocarditis guidelines
• Avoid air in IV lines or use inline filter
• Avoid scuba diving
• DNA screening of children for HHT
Common Presentations of PAVM

- Abnormal Chest X-ray or CT scan
  - Most are asymptomatic
- Occult stroke or brain abscess
- Unexplained hypoxemia
- Unexplained shortness of breath
- Hemoptyisis or hemothorax
- Clubbing and/or cyanosis
PAVM During Pregnancy

• Often enlarge during the 2nd and 3rd trimesters

• Hemoptysis in 1.1%, hemothorax 2.1%
  ‣ Almost always in the 3rd trimester

• Recommend screening HHT patients within 2 years before getting pregnant

• Embolization therapy safe after 16 wk

de Gussem, Obstet Gynecol 2014
Case SB 2013

History:

• 51 yo white woman
• Referred for possible PAVM seen on chest CT done for workup of SOB
• ROS: fatigue and DOE but no bleeding or nosebleeds
• PHx: negative
• FHx: son with nosebleeds from allergies; no other nosebleeds or AVM
Exam:

- BP 142/82, otherwise normal vital signs
- No telangiectasias on skin or mucus membranes
- Lungs clear
- Heart and abdomen normal
- No leg edema or finger clubbing
Labs:

- **Pulse ox**: 98% supine, 97-99% seated
- **Echo w contrast**: EF 59, grade 1/3 delayed shunt (10 bubbles), RVSP 20-25
- **CTA chest**: 1 cm enhancing nodule LLL with a 2.2 mm feeding artery but no definite draining vein
Case SB 2013 - CTA
Case SB 2013 – Pulm Angiogram
Case SB 2013

Follow-up:

- No macroscopic PAVM
- Echo is borderline positive for micro PAVM
- Consider prophylactic antibiotics
- She meets no criteria for HHT
- No further testing for HHT is recommended for she or her family
Large Complex PAVM
Multiple Small PAVM
Small PAVM
Enlarged Vessels into PAVM
Enlarged Vessels into PAVM
Case CH 2010

History:

- 39 yo white woman
- 12/2010: referred for exercise desaturation
- 2/2010: Dx with incidental PAVM
- 4/2010: Embolization of single R PAVM
- 9/2010: significant decrease in exercise tolerance with episodic SOB, SVT, CP
Case CH 2010

OSH evaluation:

- Contrast echo: delayed shunt, grade 1/3
- Pulmonary angiogram: normal
- Treadmill exercise test: desaturation to 83% after 8 min of exercise
- Right heart cath: normal, no shunt
- Referred to GRU for occult PAVM as a cause for desaturation and exercise intolerance
Case CH 2010

Other History:

- **PHx:** migraine HA, cholecystitis, GERD, GI bleeds from upper GI AVM
- **ROS:** nosebleeds since age 13
- **FHx:** dad, brother and nephew with recurrent nosebleeds; no other hx of AVM
- **SHx:** no alcohol or smoking
Case CH 2010

Exam:

- Normal vital signs
- 8-10 tiny telangiectasias on lips, face, and nasal mucosa
- Lungs clear
- Heart and abdomen normal
- No leg edema or finger clubbing
Cardiopulmonary exercise test:

- Exercised 10.5 min at 12 watts/ min
- Peak VO2 123% of predicted
- Peak HR 92% predicted
- Anaerobic threshold, breathing reserve, & gas exchange normal
- PaO2: rest, 101; peak exercise, 117
- Conclusion: no right to left shunt
Conclusions:

- She may have microscopic PAVM but these are not the cause of her symptoms.
- Prior episodes of desaturation likely due to poor tracking of the pulse oximeter.
- She has no significant cardiopulmonary disease.
Follow-up After PAVM Embol

Routine:

- Contrasted or non-contrasted CT at about 6 months
  - Reperfusion is seen in 10-15%
- Non-contrasted CT every 3-5 years
- Contrast echo usually not helpful because it remains positive in 90% due to persistent micro PAVM
Follow-up After PAVM Embol

For dyspnea or hypoxemia:

- Supine and seated pulse oximetry:
  - Usually <97% with significant PAVM
- 100% oxygen shunt test: >5% abnormal
- Exercise ABG: PaO2 should increase
- Invasive cardiopulmonary exercise test
- Contrast echo only helpful if negative
- Consider repeat CT if above positive
- Pulmonary angiogram rarely needed