RADY 401 Case Presentation

Pulmonary Arteriovenous Malformation (PAVM)

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6/14/21
Patient History and Workup

30yoF G3P2 at 24wks with PMH of asthma presents to the ED with cough and shortness of breath over the past two months. Increased fatigue with little activity, and blood in sputum with coughing. She has no recent illness, no COVID-19 exposures, no sick contacts, and no other alarming symptoms.

Physical Exam: Tachycardia to the 120s, otherwise unremarkable.

Notable Labs & Studies: WBC 20.1, ECG shows sinus tachycardia, new right axis deviation, and T waves present in inferior and lateral leads.
List of imaging studies

- Chest X-ray
  - Estimated cost: National average $420. ²
  - Radiation: ~0.1 mSv. ⁹
- CTA Chest with IV contrast
  - Estimated cost: National average $6200. ³
  - Radiation: ~10 mSv. ⁹
Imaging Studies: Chest X-ray Findings

2.5 cm lobulated soft tissue density structure projections over the right mid lung zone.
Imaging Studies: CTA Chest findings

Lateral RML shows an opacified serpiginous pulmonary artery distally with a vascular nidus measuring 8 x 7 x 7mm with dilated feeding pulmonary artery and draining pulmonary vein.
Pulmonary Arteriovenous Malformation

• An abnormal communication between a pulmonary artery and vein. These connections can create a high-flow right-to-left shunt.⁴

• They are commonly associated with Hereditary hemorrhagic telangiectasia (HHT) which can be characterized as abnormalities of vascular structures involving multiple organs.⁶

• Screening in HHT⁶:

![Diagram of screening process]

  - TTCE
  - Positive → Chest CT → Diagnostic PAVM
  - Negative
  - Not seen → Possible microscopic PAVM
Pulmonary Arteriovenous Malformation

• Epidemiology: uncommon, the Mayo Clinic suggests 194 cases of PAVMs over 45 years.  

• Symptoms: asymptomatic in ~50% of individuals; most common symptoms are dyspnea, hemoptysis, hypoxia, epistaxis, mucocutaneous telangiectases, or presence of a nodule with a history of cerebral infarct or abscess. 

• Diagnosis: CT pulmonary angiography is the gold standard test.  
  • Sensitivity – 68%; Specificity – 100%.  

• Treatment: Endovascular embolization.
## ACR Criteria

### American College of Radiology

**ACR Appropriateness Criteria**

**Clinical Condition:** Clinically Suspected Pulmonary Arteriovenous Malformation (PAVM)

<table>
<thead>
<tr>
<th>Radiologic Procedure</th>
<th>Rating</th>
<th>Comments</th>
<th>RRL*</th>
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<tbody>
<tr>
<td>US echocardiography transthoracic with IV contrast</td>
<td>8</td>
<td>This procedure is often used following positive TTE.</td>
<td>🌟🌟🌟🌟</td>
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<tr>
<td>CTA chest with IV contrast</td>
<td>8</td>
<td>This procedure is complementary to other examinations, such as TTE.</td>
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<td>X-ray chest</td>
<td>7</td>
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<tr>
<td>US echocardiography transesophageal with IV contrast</td>
<td>6</td>
<td>This procedure is the reference standard for detecting right-to-left shunts but is more invasive than TTE.</td>
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<tr>
<td>MRA chest without and with IV contrast</td>
<td>6</td>
<td></td>
<td>🌟🌟🌟🌟</td>
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<tr>
<td>CT chest without IV contrast</td>
<td>6</td>
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<td>🌟🌟🌟🌟</td>
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<tr>
<td>Arteriography pulmonary</td>
<td>5</td>
<td>Although this procedure is appropriate for preinterventional planning, it is usually not appropriate as an initial test.</td>
<td>🌟🌟🌟🌟</td>
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<tr>
<td>US transcranial with IV contrast</td>
<td>5</td>
<td>This procedure is an alternative to TTE, although it is less widely available.</td>
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<td>Perterchnetate albumin pulmonary scan</td>
<td>4</td>
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<tr>
<td>MRA chest without IV contrast</td>
<td>3</td>
<td></td>
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**Rating Scale:** 1, 2, 3 Usually not appropriate; 4, 5, 6 May be appropriate; 7, 8, 9 Usually appropriate

*Relative Radiation Level*
Patient Treatment

• Patient was admitted to the Maternal Fetal Medicine antepartum floor.

• Interventional Radiology was consulted to perform pulmonary arteriograms and possible embolization.

• The procedure was scheduled for the following day.
Imaging studies: Right Pulmonary Arteriogram
Imaging studies: Right Pulmonary Arteriogram
Treatment: Right Pulmonary Artery Embolization

Two MVP-5Q plugs and three Nester coils were deployed to diminish flow to the AVM.
Treatment: Right Pulmonary Artery Embolization

Follow up arteriogram showing no further filling of the PAVM.
Patient Outcome

• The patient’s pulmonary arteriovenous malformation was successfully sealed with the embolization procedure.

• The patient was later found to have a uterine AVM, and it remains undetermined as to whether the patient possibly had Hereditary hemorrhagic telangiectasia (HHT).
UNC Top Three

• Pulmonary arteriovenous malformations are commonly associated with Hereditary hemorrhagic telangiectasia.

• Screening is performed with transthoracic contrast echocardiography. Diagnosis is confirmed with CT pulmonary angiography.

• Treatment is endovascular embolization performed by interventional radiology.
References


