Radiologic Imaging and Treatment of Pulmonary AVMs

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Clinical Presentation of Pt A

• 34 yo female with acute onset of R sided hemiparesis and aphasia
Pt A: Head CT on Presentation

Head CT without contrast showed a hyperdense L MCA sign.
Pt A: Occlusion of L MCA and ICA

Decreased blood flow in area of L MCA

Completed occluded L ICA and L MCA

PACS, BIDMC
Pt A Percutaneous Clot Retrieval

Angio revealed clot extending from L ICA into L MCA

IA tPA administered, MERCI clot retriever deployed

Good flow achieved through L ICA and MCA

http://www.med.cornell.edu/science/2006/03_06/06/03_06-2.shtml
Etiology of Thrombotic Infarct

Most Likely < 40yo

• Carotid or Vertebral Dissection
• Underlying coagulopathy
  – Protein S or C Def, Antithrombin III def, Factor V Leiden, Antiphospholipid syndrome

Less Likely

• Congenital Heart Defect
  – PFO, ASD, PDA
• Prolonged immobility
• Neoplastic Process
• Collagen vascular disorders
  – SLE, RA, Sjogrens, FMD, Myositis
• Trauma
Menu of Tests For Pt Work-Up

• Most Common
  – CTA Neck w & w/out contrast
  – ECG
  – ECHO with agitated saline
  – MR Head w/o Contrast

• Less Common
  – Chest X-ray
  – Venous Duplex Lower Extremities Bilaterally
  – CT Chest, Abdomen, Pelvis w/ contrast
Pt A: Opacity in R Lung Base

- ET tube tip is 3 cm about carina
- Lungs w/ minimal atelectasis at the L base
- Rounded opacity in R cardiophrenic angle
  - 3 cm in diameter projecting mostly over R lung base
- Linear opacity proximal to R hilus
Pt A: AVM on Contrast CT

- Homogenously enhancing structure in R middle lobe adjacent to pericardial fat
- Large feeding vessel arising from pulmonary artery approaches lesion
- Findings consistent with large pulmonary arteriovenous malformation (AVM)
- AVM enhances homogenously w/out evidence of thrombus
Pt A: Pulmonary Arteriovenous Malformation (PAVM)

- Aberrant connection between pulmonary artery and venous circulation that bypasses capillary system
- High flow low resistance
- 50-70% of PAVMs are located in the lower lobes
Normal Anatomy vs PAVM

- Pulmonary angiogram demonstrated substantial aneurysmal dilation of interlobar branch of with finding consistent with a large AVM
Complications from PAVM

- Hemorrhage: Hemoptysis or Hemothorax

- Massive R to L shunting: Hypoxemia, Dyspnea, Clubbing, Cyanosis, Polycythemia

- **Paradoxical Emboli**: cerebral abscess, embolic stroke, TIA’s
  - Serious neurological complications occur in up to 35% of pt w/ PAVM*

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Treatment

- AVM > 3 mm require tx
- Transcatheter Embolotherapy (TCE) is treatment of choice
  - avoids major surgery and general anesthesia
  - loss of lung parenchyma

- TCE Procedure: Coils are threaded through a catheter into the pulmonary artery and placed both distally in the draining vein and proximally in the feeding artery of the AVM
Treatment Complications

• Most Common:
  – Pleuritic chest pain
  – Paradoxic embolization
  – Air emboli
  – Recanalization

• Less Common
  – Infection
  – Pulmonary infarction
  – Pulmonary Hypertension
  – Deep venous thrombosis
  – Dislocation of coils
  – Bleeding

Companion Patient #1
Pt A: Treatment

- Substantial dilation of interlobar branch appreciated on angiogram
- Feeding artery unsuitable for secure placement of balloons or coils due to size and high flow
- 10mm Amplatz occluder device deployed
- No filling or arterial phase opacification of the AV on follow-up angiogram
DDx: Pulmonary AVM

• Most Common:
  – Hereditary Hemorrhagic Telangiectasia (HHT) (80%)

• Less Common:
  – Chronic Inflammatory Conditions
  – Hepatic Cirrhosis
  – Mitral Stenosis
  – Schistosomiasis
  – Actinomycosis
  – Trauma
  – Fanconi’s Syndrome
  – Metastatic Thyroid Carcinoma
Hereditary Hemorrhagic Telangiectasia

• Autosomal Dominant:
  – Incidence 1/10,000
  – Varying penetrance
  – Mutations in genes encoding Endoglin or ALK-1, both membrane receptors of TGF-β

• Diagnostic Criteria:
  1. Spontaneous, recurrent epistaxis
  2. Multiple mucocutaneous telangiectasias
  3. Visceral involvement
  4. 1° relative with HHT

Clinical Features of HHT

- Recurrent Epistaxis (95%)
- Multiple mucocutaneous telangiectasias (75%)
  - lips, hands, face
- GI bleeding (20-25%)
- AVMs
  - Pulmonary (30-50%)
  - Cerebral (5-20%)
  - Hepatic (up to 30%)*
- Fe deficiency anemia

PAVMs: Summary

• Morbidity and mortality
  – embolic stroke, brain abscess, and intracranial hemorrhage

• HHT is the most common etiology for PAVM
  – Patients and 1º relatives should be screened

• Embolotherapy or surgery is required for:
  – Symptomatic patients
  – PAVM with feeding arteries > 3 mm in diameter
References

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