Chronic Thromboembolic Pulmonary Hypertension (CTEPH): A Primer

H. Page McAdams, MD
Duke University Medical Center
Durham, NC 27710
page.mcadams@duke.edu
Which of the following imaging tests is LEAST helpful for distinguishing CTEPH from other causes of pulmonary hypertension?

a. CT angiography
b. MR angiography
c. Chest radiography
d. Ventilation-Perfusion scintigraphy (V/Q)
e. Thin-section CT
Pre-Capillary Pulm HTN

Capillaries

Pulmonary artery

Pulmonary vein

RV

LA
Pre-Capillary Pulm HTN

Primary Pulmonary HTN

Eisenmenger (ASD/VSD/PDA)

Chronic Emboli (CTEPH)
CTEPH: Pathogenesis

- Normal fibrinolytic system
  - lysis
  - fragmentation
  - more peripheral emboli
  - organization
  - recanalization
CTEPH: Pathogenesis

- Normal fibrinolytic system
- Occasionally fails
  - chronic scarring
  - secondary pulmonary hypertension (CTEPH)
CTEPH: Pathogenesis

Acute
CTEPH: Pathogenesis

Acute

Chronic
CTEPH: Pathogenesis

- Normal fibrinolytic system
- Occasionally fails
- PAH develops
  - enough small vessels occluded
  - CTEPH develops $\approx 5\%$ acute PE
CTEPH: Pathogenesis

- Normal fibrinolytic system
- Occasionally fails
- PAH develops
- Up to 50% have no history of previous embolic events
CTEPH

• Radiologic evaluation
  • CXR
  • V/Q scan
  • Chest CT/CTA

all patients
CTEPH

• Radiologic evaluation
  • CXR
  • V/Q scan
  • Chest CT/CTA
  • PAogram, RH cath
  • MR imaging

selected patients
CTEPH: CT (MR) Findings

- Enlarged PAs, right heart
- Peripheral, mural thrombus
- Webs, bands
- Small, occluded vessels
- Mosaic attenuation
- Collateral vessels
CTEPH: CT Findings

- Mural thrombus
  - peripheral
  - obtuse angles with vessel wall
  - calcification uncommon
CTEPH: CT Findings

- Webs
  - thin, central filling defects
  - not acute emboli
CTEPH: CT Findings

- Webs
  - thin, central filling defects
  - not acute emboli
  - MPRs to distinguish from bifurcation
CTEPH: CT Findings

- Bands
  - circumferential constriction
  - best seen on MPRs
CTEPH: CT Findings

- Small, occluded vessels
CTEPH: CT Findings

- Mosaic attenuation
CTEPH: CT Findings

- Collateral vessels
CTEPH: CT Findings

- Collateral vessels
CTEPH: Treatment

- Anticoagulation
- IVC filter
- Thromboendarterectomy
  - substantially improve PAH, RH function
  - CTA modality of choice for presurgical evaluation
CTEPH: Good Surgical Candidate

- Organized thrombi
  - proximal to segmental arteries
- Clinical
  - younger patients
  - thrombophilic disorders
Severe PAH: ? CTEPH
Severe PAH: ? CTEPH
Severe PAH: ? CTEPH
Severe PAH: CTEPH
Good Surgical Candidate
Severe PAH: ?CTEPH
Severe PAH: ?CTEPH
Severe IV Contrast Allergy
Severe PAH: CTEPH
Good Surgical Candidate
CTEPH: Poor Surgical Candidate

- Doesn’t have CTEPH
- Distal clot only
- Clinical
  - older patients
  - splenectomy, malignancy
  - less likely to remember acute PE episode
Severe PAH: ?CTEPH
Severe PAH: ?CTEPH
Severe PAH: ?CTEPH
Poor Surgical Candidate
CTEPH: Summary

- Important, treatable cause of PAH
- Many have no prior history of PE
- Imaging evaluation key
  - CTA modality of choice
  - findings subtle, easily overlooked
  - need to see most/all findings
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