Emerging techniques in imaging of Chronic Thromboembolic Pulmonary Hypertension (CTEPH)

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Teaching points

- To understand the pathophysiology of CTEPH
- To understand the goals of imaging in diagnosis and clinical decision making
- To understand the role of newer imaging techniques (such as Dual-energy CT and SPECT-V/Q imaging) that allow combined anatomic and functional imaging.

Pathophysiology of CTEPH

- Natural history of acute PE is complete resolution with no hemodynamic consequence.
- Still unclear why certain patients develop chronic thromboembolic disease (CTE).
- Role of procoagulant states such as anti-phospholipid antibodies postulated.
- Subsequent development of PH is multifactorial.

Introduction

- Chronic thromboembolic disease is one of the increasingly recognized causes of pulmonary hypertension (PH) that follows a single or recurrent event of acute pulmonary embolism (PE).
- Occurs after approximately 0.5% to 3.8% of acute pulmonary embolic events and in up to 10% of patients with recurrent PE.
- Pulmonary thromboendarterectomy (PEA) is the procedure of choice, with favorable short-term and long-term outcomes.

Layering of eccentric thrombus
- Weblike filling defects
- Stenosis and retraction of vessels
- Shear stress and release of cytokines
Work-up of CTEPH

- Nonspecific symptoms and clinical picture: High index of suspicion necessary.
- Usual first step is Doppler echocardiography that gives a rough estimate of presence of PH.

What is the role of imaging?
- To diagnose the presence of CTE and PH.
- Is the PH attributable to CTE (CTEPH)?
- To assess the extent of chronic thromboembolic disease.
- To assess surgical candidacy.

STEP 1

First question after PH is diagnosed: is PH due to chronic thromboembolic disease?

Preferred tests:
- V/Q scanning
- CTA
- Pulmonary angiogram
- MRI

V/Q scanning

Most clinicians use V/Q scanning as initial screening test of choice to establish diagnosis of CTEPH.
- Very easy to interpret.

Computed tomography (CTA)

Layered eccentric thrombus
- Atretic poorly opacified artery
- Web-like defects
- Stenosis

CTA—although extremely detailed, can be difficult sometimes to interpret and may need trained radiologists.
64/F with history of acute PE in November 2013 presents for evaluation of CTEPH in April 2014.

CTA-almost normal with possible attenuation of caliber of RLL segmental branch.

V/Q scanning shows multiple mismatched perfusion defects, predominantly in the right lung.

HIGH PROBABILITY SCAN

Second question-how much is the disease and is it operable?

Extent of disease
Pulmonary vascular resistance (PVR)

Decision for surgical intervention

Extent of disease

Preferred tests
CTA Pulmonary angiogram
MRI-alternative to CTA
V/Q-No role

Pulmonary vascular resistance

Test of choice
Right heart catheterization

MRI Echocardiography V/Q-No role

Typical >600-1200 dynes-sec/cm5
>300 dynes-sec/cm5

CTEPH

V/Q scanning
Initial screen Perfusion defects
CTA
Extent of disease
Pulmonary angiogram
Extent of disease Perfusion defects***
Echocardiography MRI
RV function
Why search for newer tools?

- One-stop shop imaging test to reduce imaging costs
- Better assessment of lung perfusion and microvasculature—better selection of patients

Spectral CT

Attenuation of photons is energy dependent and each substance has a specific attenuation curve.

SPECTRAL CT

- Use of 2 tubes that operate at different kVp levels
- Use of rapid kVp switching producing spectra of lower and higher energies
- Energy sensitive detectors: top layer absorbs lower energy photons while lower layer absorbs higher energy ones

Dual-energy CT (DECT)

- Technique: Using dual source CT, scans are obtained at two different kilo-voltages (80/100 and 140 kVps).
- Iodine attenuates X-ray spectra differently at lower kVp settings from higher ones, which is used to generate an iodine map.

53 year old male with CTEPH. a) Perfusion blood volume images show perfusion defects in the right upper and lower lobes, which match those seen on the corresponding pulmonary angiogram.
Study number | Conclusion
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Simultaneous information on the presence of endoluminal thrombus and lung perfusion impairment is possible with Dual-energy MDCT.

Dual-energy CTA of pulmonary embolism is feasible and allows the assessment of perfusion defects caused by pulmonary embolism.

Abnormal pulmonary blood distribution, as shown at dual-source CT, improves detection of acute PE in rabbits.

Dournes G, Verdier D, Montaudon M, Bullier E, Rivière A, Dromer C, Picard F, Billes MA, Corneloup O, Laurent F, Lederlin M. Dual-energy CT perfusion and angiography in chronic thromboembolic pulmonary hypertension: diagnostic accuracy and concordance with radionuclide scintigraphy. Eur Radiol. 2014;24(1):42-51. | Fourteen patients were diagnosed with CTEPH and 26 with other aetiologies. DECT perfusion and angiography correctly diagnosed CTEPH in 14 patients (100% sensitivity, 96% specificity) and in 26 patients with other aetiologies (100% sensitivity, 96% specificity). DECT perfusion showed moderate agreement (κ = 0.44) with scintigraphy. Agreement between CT angiography and scintigraphy ranged from fair (κ = 0.44) to substantial (κ = 0.67) depending on whether completely or partially occlusive patterns were considered, respectively.


Applications in CTEPH
- Anatomic quantification of disease
- Assessment of lung perfusion
- Assessment of surgical candidacy
- Post-procedural functional improvement

SPECT and SPECT-CT V/Q imaging
- Data on SPECT V/Q scanning published as early as 1987.
- Several studies have since documented superiority of SPECT V/Q imaging over planar imaging.
- Some of the key advantages include:
  - Lesser number of indeterminate/non-diagnostic studies.
  - Greater accuracy and specificity.

Planar V/Q
- As discussed, remains the initial screening test of choice.
- Easy to perform and interpret and has been extensively validated.
- Some limitations of planar V/Q imaging include:
  - Segmental overlap
  - Shine-through from underlying normal lung segments
  - Difficulty in visualization of all segments

Findings

Studies comparing DECT with V/Q scanning

Applications in CTEPH

Planar V/Q
Normal co-registered perfusion (top) and ventilation (bottom) SPECT-CT V/Q images

Abnormal co-registered SPECT-CT V/Q images: Multiple mismatched perfusion defects in both lungs (arrows)

Value of SPECT-CT V/Q scanning in follow-up of patients. Bottom image (b) shows decreased perfusion in the left upper lobe in this patient with left superior pulmonary vein stenosis. Following stenting of the stenosed left superior pulmonary vein, the perfusion improved, as seen in the top image (a).

Magnetic resonance imaging (MRI)

MRI is generally accepted as a second-choice to CTA in assessment of anatomic extent of the disease*. Better spatial resolution, faster scan times and detailed assessment of both lung parenchyma and vasculature are some of the advantages of CTA over MRI.

Strengths of MRI

MRI is the current gold standard for assessment of right ventricular volumes.

MRI based measurements of mean pulmonary arterial pressure (mPAP) and PVR have been encouraging.

Assessment of RV hemodynamics can be used to monitor response to therapy (1,2).

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Summary

- Imaging plays a critical role in diagnosis and preoperative planning of patients with CTEPH.
- Newer imaging tools such as Dual-energy CT, SPECT-CT V/Q imaging and MRI are expected to play increasing role in the evaluation of CTEPH.

Thank you!!