Pulmonary Vascular Diseases

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KEYWORDS

• Acute pulmonary embolism • Chronic thromboembolic pulmonary hypertension
• Pulmonary arteriovenous malformation • Hereditary hemorrhagic telangiectasia
• Granulomatosis with polyangitis • Microscopic polyangitis

KEY POINTS

- Multidetector CT (MDCT) is the study of choice for the evaluation of acute pulmonary embolism (PE), simultaneously detecting thromboemboli and, in its absence, providing alternative explanations for presentation.
- MDCT plays a role in the detection and distribution characterization of disease in patients with chronic thromboembolic pulmonary hypertension (CTEPH), especially in cases of prior emboli or abnormal ventilation-perfusion (V/Q) scan.
- In patients with suspected pulmonary arteriovenous malformation (PAVM) or HHT who have abnormal bubble echocardiography, MDCT is the study of choice.
- Vasculitides can affect the large central pulmonary vessels or more commonly the small vessels of the lung, resulting in various parenchymal manifestations.

INTRODUCTION

Pulmonary vascular diseases encompass a large and diverse group of underlying pathologies ranging from venous thromboembolism to congenital malformations to inflammatory vasculitides. As a result, patients can present either acutely with dyspnea and chest pain or chronically with dyspnea on exertion, hypoxia, and right heart failure. Imaging, particularly with MDCT, plays a key role in the evaluation and management of patients with suspected pulmonary vascular disease.

ACUTE PULMONARY EMBOLISM

Venous thromboembolism is reportedly the third most common vascular disease after myocardial infarction and stroke, affecting approximately 600,000 people each year.[1] Because acute PE can be a challenging clinical diagnosis to make given the variability in patient presentation, CT pulmonary angiography (CTPA) is one of the most commonly ordered imaging studies performed for pulmonary vascular disease, especially in the emergency department.

The vast majority of PE arises from the deep venous system of the lower extremities, frequently in the setting of risk factors, such as recent surgery, prolonged immobilization, hypercoagulable disorders, and malignancy. No predisposing factor, however, is identified in up to 30% of cases,[2] and 50% of patients studied with deep venous thrombosis are found to have clinically silent PE.[3] For these reasons, clinical prediction criteria, such as the Wells score, and laboratory assays, like the D-dimer, are used to risk stratify for acute PE. The combination of the 2 can safely exclude the risk of acute PE.[4]

MDCT has emerged as the imaging study of choice for those patients with intermediate or high

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clinical risk for acute PE and those with abnormal D-dimer assays. In PIOPED II (Prospective Investigation of Pulmonary Embolism Diagnosis), which studied the accuracy of CTPA with V/Q and conventional pulmonary angiography, CTPA in patients with a high pretest probability was shown to have a sensitivity of 83% and specificity of 96% for the diagnosis.[5] The rapid evolution of MDCT technology has, however, likely resulted in greatly improved accuracy in comparison to the 4-, 8-, and 16-row scanners used in the PIOPED II study. In addition to its widespread availability and rapid assessment, another major advantage of MDCT over V/Q and conventional angiography is the ability to detect alternative causes for clinical symptoms in up to a third of cases, which is important given the widespread low positivity rates for acute PE, in many cases as low as 10%.[6]

Although techniques vary slightly among institutions and vendors, generally CTPA is performed with 80 to 100 mL of nonionic iodinated intravenous contrast administered at a rate of 4 mL/s from an upper extremity antecubital vein cannulated with an 18- or 20-gauge intravenous (IV). In the era of widespread advanced and even ultrafast MDCT, automated bolus tracking and test bolus techniques are widely used to time optimal contrast delivery to the pulmonary system during the time of imaging. Test bolus technique is useful in patients with very poor cardiac function who frequently have delayed opacification of the pulmonary arteries. Although caudal to cranial scanning is often used to reduce effect of respiratory motion at the lung bases where more emboli are seen and more diaphragmatic motion occurs, the availability of ultrafast MDCT usually obviates this modification. Thin collimation allowing for isovolumetric 1-mm slice reconstruction allows for detailed analysis and multiplanar reconstruction.

The hallmark finding of acute PE on MDCT is a low-attenuation filling defect in a contrast-filled pulmonary artery. MDCT findings of acute PE include (**Fig. 1**)[7,8]

- Complete arterial occlusion (vessel cutoff) without luminal enhancement with or without vessel dilatation
- Partial, central filling defect surrounded by contrast (rim or polo mint sign when viewed perpendicular to the vessel axis or railway sign when viewed longitudinal to the vessel axis)
- Peripheral filling defects forming acute angles with the vessel walls or at vessel branch points

To prevent obscuration of small segmental emboli by dense iodinated contrast, window and level values that enable visualization of right heart structures, such as trabeculations, or the pulmonary valve leaflets should be chosen.

Parenchymal findings that can be seen with acute PE include (**Fig. 2**)

- Pulmonary hemorrhage (ground-glass opacification)
- Infarct (peripheral consolidation with central ground-glass)
- Oligemia (vascular mosaic attenuation)
- Atelectasis
- Pleural effusion

The prognosis for patients with acute PE has been shown to correlate with circulatory effects and right ventricular dysfunction. Clinically, the term, *massive PE*, is applied to patients with evidence of hemodynamic compromise (systemic hypotension), whereas those patients with hemodynamic stability are labeled *submassive*. Cardiac findings that can be seen in the setting of acute PE include (**Fig. 3**)

- Right ventricle–to–left ventricle (RV:LV) diameter ratio greater than 1 (measured on axial image)
- Leftward interventricular septal bowing
- Inferior vena cava and/or azygous vein dilatation
- Reflux of contrast material into the inferior vena cava

Of the findings listed, RV:LV ratio best correlates with echocardiographic evidence of right heart strain with reported sensitivities of 80% or greater for RV:LV ratio greater than 1 but variable specificity.[9] CT findings of right heart dysfunction should be reported, because they may triage a patient to a more closely monitored nursing division. The CT findings of right heart dysfunction, however, are not used for determining whether thrombolysis is warranted. That decision is based on systemic symptoms and echocardiographic findings.

Artifacts that can limit assessment for acute PE include

- Flow artifacts (transient interruption of the contrast column)
- Respiratory or patient motion
- Image noise
- Partial volume averaging with adjacent structures

It is important to interrogate the pulmonary arteries on all contrast-enhanced CT scans, especially in oncologic populations, because PEs