A trainee's guide to imaging pulmonary arterial hypertension

Poster No.: C-1049  
Congress: ECR 2013  
Type: Educational Exhibit  
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Keywords: Cardiovascular system, Arteries / Aorta, Respiratory system, CT, CT-Angiography, eLearning, Education and training, Embolism / Thrombosis  
DOI: 10.1594/ecr2013/C-1049

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Learning objectives

CT has a central role in the diagnosis and assessment of pulmonary hypertension. This poster is designed to illustrate the various CT signs that suggest pulmonary hypertension and findings that help identify the aetiology, in particular, chronic thromboembolic pulmonary hypertension.
Background

Raised pulmonary artery pressure can be an idiopathic disorder or, more commonly, the consequence of many disparate clinical conditions. Although there is no cure for pulmonary arterial hypertension, several different therapies have been developed that have led to improvements in symptoms, exercise tolerance, and in some cases survival.\(^1\) This has emphasized the importance of early and accurate detection of increased pulmonary arterial pressure and its underlying aetiology. Given the very non-specific nature of the symptoms of pulmonary arterial hypertension it may be the radiologist who is first able to suggest this diagnosis.

Right heart catheterization remains the most accurate method of measuring pulmonary artery pressure\(^2\) but it is an invasive and relatively expensive test. CT is widely available, non-invasive and it is possible to image the main and peripheral pulmonary arteries, heart and lung parenchyma in one examination.

The role of CT scanning is to confirm the diagnosis of pulmonary hypertension and to help establish the aetiology. There are several CT findings in patients with pulmonary hypertension that taken in isolation have limited value due to lack specificity. However, when taken in combination it is usually possible to make the diagnosis.

It is particularly important to recognize the CT findings in chronic thromboembolic pulmonary hypertension (CTEPH) because this may be amenable to surgery (pulmonary endarterectomy).

This poster will review the CT findings of raised pulmonary arterial pressure and in particular, the signs that suggest CTEPH as the likely aetiology.
Imaging findings OR Procedure details

Diagnosing pulmonary hypertension (raised pulmonary artery pressure of any cause):

1. The most extensively investigated CT sign of pulmonary hypertension (PH) is the diameter of the main pulmonary artery (mPA). A diameter of >29mm is suggestive of PH. However, its size is dependent on factors other than mean pulmonary arterial pressure (mPAP), e.g. body surface area. It is reasonable to conclude that these other factors affect the ascending aorta (AA) to the same extent as the mPA. This has led to the concept of the ratio of the mPA:AA. A ratio of >1 is highly specific for PH and correlates strongly with mPAP.\(^3,4\) (Although no more so than mPA diameter >29mm alone). Fig. 1 on page 6

2. Enlargement and hypertrophy of the right ventricle and atrium are common in patients with pulmonary hypertension, along with flattening or reversal of the interventricular septum. The left ventricle is usually compressed and displaced posteriorly unless the pulmonary hypertension has developed secondary to left sided cardiac dysfunction; in which case, left atrial and ventricular dilatation may be present. Fig. 2 on page 6

3. Reflux of contrast from the right atrium into the IVC during the first pass is another useful sign of pulmonary hypertension on CT. It occurs because of secondary tricuspid regurgitation in patients with pulmonary hypertension and the degree of reflux correlates with mPAP at right heart catheterisation\(^5\). Caution is required when interpreting this sign as IVC reflux may be seen in normal individuals when contrast injection rates exceed 3 mls/second. However, in routine practice, it is unusual to see reflux into the hepatic veins in patients without significant right heart dysfunction or tricuspid regurgitation. Fig. 3 on page 7

4. Pericardial effusions are not an infrequent finding in pulmonary hypertension\(^6\). Their pathogenesis is unclear but they are usually refractory to diuretics and larger effusions are associated with evidence of heart failure, impaired exercise tolerance and poor prognosis. Fig. 4 on page 8

5. Within the lung, the diameter of the pulmonary arteries should be approximately the same as the accompanying bronchi. In patients with pulmonary hypertension or increased blood flow due to left to right shunts, there may be significant dilatation of intrapulmonary arteries relative to their accompanying bronchi. Fig. 5 on page 9
The following signs are not specific to CTEPH. However, they are more commonly seen in CTEPH compared to other causes of PH. Their presence should therefore increase the suspicion of CTEPH as the likely aetiology:

1. One of the most conspicuous findings in CTEPH is eccentric laminated thrombus, resulting in a crescentic filling defect adjacent to the vessel wall. It is worth noting that this peripheral laminated filling defect in the central PA's is not specific for CTEPH. There are a number of conditions which may give rise to a similar appearance on CT, thus mimicking CTEPH. In Eisenmenger's syndrome, due to very slow flow and giant pulmonary arteries, laminated thrombus may develop in situ in the proximal pulmonary arteries. Other mimics of CTEPH include large vessel vasculitis and pulmonary artery sarcoma. It is also worth noting that the absence of an eccentric filling defect does not exclude CTEPH.

2. The bronchial arteries usually arise from the proximal descending aorta and supply the walls of the central bronchial tree and proximal pulmonary arteries. Hypertrophy of the bronchial arteries has been defined as curvilinear mediastinal vessels > 1.5 mm in diameter, seen along the course of the proximal bronchial tree. In practice, given that the bronchial arteries are inconspicuous on CT in normal individuals, hypertrophied arteries are usually easily identified, especially on maximum intensity projection images. Bronchial artery hypertrophy occurs in approximately half the patients with CTEPH and only rarely in pulmonary arterial hypertension (PAH). However, bronchial artery hypertrophy does occur with sufficient frequency in other causes of pulmonary hypertension, such as PAH and Eisenmenger's Syndrome, as to make it an unreliable sign on its own for the diagnosis of CTEPH.

3. A mosaic pattern refers to regional differences in density of lung parenchyma, and in CTEPH is caused by obliteration of parts of the vascular bed. This results in hypoperfusion with arteries of diminished size in some areas, and normal or increased perfusion with enlarged arteries in others; "mosaic oligemia". In the context of pulmonary hypertension, this finding used to be considered virtually pathognomonic of CTEPH. However, some studies have been published, albeit with small population sizes, which suggest that a mosaic pattern is not as uncommon in other causes of pulmonary hypertension as was previously thought.

4. Peripheral lung parenchymal opacities are a common finding in CTEPH. They represent pulmonary infarcts due to occlusion of segmental and smaller pulmonary arteries.
Fig. 1: Axial CT demonstrating an enlarged main pulmonary artery diameter (i.e. >29mm) and a ratio of main pulmonary artery diameter: ascending aorta of >1. This is indicative of raised pulmonary artery pressure.

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**Fig. 2:** Axial contrast enhanced CT of the thorax demonstrating enlargement of the right atrium and ventricle, flattening of the interventricular septum (arrow) and a displaced and compressed left ventricle.

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Fig. 3: Contrast enhanced axial CT at the level of the hepatic veins demonstrating reflux of contrast from the right side of heart into the IVC and hepatic veins.

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Fig. 4: Axial contrast enhanced CT thorax demonstrating a pericardial effusion (arrow) and dilated right sided heart chambers in a patient with pulmonary hypertension.

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Fig. 5: Axial CT image. In the right lung there is an increase in the segmental pulmonary artery: bronchus ratio (arrow). There is also regional variation in lung density (mosaic pattern).

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**Fig. 6:** Axial contrast enhanced CT in a patient with CTEPH demonstrating peripheral laminated thrombus in the right main pulmonary artery (arrow).

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Fig. 7: Contrast enhanced axial CT demonstrating an enlarged bronchial artery originating from the descending aorta (arrow).

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Fig. 8: Peripheral opacities representing pulmonary infarcts in a patient with CTEPH (arrows).

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Conclusion

After reading through this poster we hope that you will recognise the more common CT signs found in pulmonary hypertension.

It must be stressed that these signs, when taken in isolation are of limited value due to lack of sensitivity and specificity. However, when taken in combination, in the appropriate clinical setting, a confident diagnosis can usually be made.

It is particularly important to recognise CTEPH as a cause of PH, because it is unique in that it may be amenable to surgical treatment.
References