Imaging findings in pulmonary hypertension and its underlying etiological mechanisms.

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Learning objectives

The purpose of our educational exhibit is to:

1. Describe imaging findings in the diagnosis and management of pulmonary hypertension (PH).
2. List the main causes of pulmonary hypertension with reference to the underlying pathophysiologic mechanisms.
3. Identify conditions that have suggestive findings at chest radiography and multidetector computed tomography.
Background

Pulmonary hypertension is defined as an abnormal elevation of pressure in pulmonary circulation, with a mean pulmonary arterial pressure higher than 25 mmHg.

This condition represents a spectrum of idiopathic and associated diseases that result in vascular remodeling: intimal thickening, medial hypertrophy and extension of media into acinar arterioles, all of which may be secondary reaction to localized fibrosis or inflammation of any etiology.

A large component of the morbidity of PH of any cause is the secondary right heart failure; there is progressive hypertrophy with dilatation occurring as right heart failure ensues.

PH may primarily affect either the arterial (precapillary) or the venous (postcapillary) pulmonary circulation.

Pulmonary arterial hypertension (PAH) may be idiopathic or arise in association with pulmonary thromboembolism; parasitic or foreign material; parenchymal lung disease; liver disease; vasculitis; human immunodeficiency virus infection or a longstanding cardiac left-to-right shunt due to congenital anomaly.

Pulmonary venous hypertension (PVH) may result from pulmonary veno-occlusive disease, pulmonary venous compression by extrinsic lesions, left-sided cardiac disease, or pulmonary vein stenosis. Evidence of coexistent pulmonary arterial hypertension due to retrograde transmission of elevated venous pressures across the capillary bed also is commonly seen at imaging and histopathologic analysis.
Findings and procedure details

The following chest radiographic findings may indicate the presence of pulmonary hypertension, but the sensitivity and specificity are not high enough for a definitive diagnosis: enlargement of the right and left main pulmonary arteries (hilar enlargement) (fig. 1); tapering of peripheral pulmonary arteries; enlargement of the right interlobar artery (greater than 15-mm diameter on a posteroanterior frontal radiograph) (fig. 2); right atrial and right ventricular enlargement; and areas of oligemia, which appear as increased lucency and decreased vascularity (fig. 3).

Features of pulmonary arterial hypertension that may be seen at computed tomography (CT) are: dilatation of the pulmonary artery trunk (29 mm or more), the diameter of which frequently exceeds that of the ascending aorta (greater than or equal to 1) (fig. 4); dilatation of the right and left main pulmonary arteries (left PA: >28 mm; right PA: >24.3 mm); abrupt narrowing and tapering of the peripheral pulmonary vessels; right ventricular hypertrophy; and right ventricular and atrial enlargement with inversion of the interventricular septum and dilatation of the tricuspid valve annulus.

The characteristic CT features of pulmonary venous hypertension are interlobular septal thickening, pleural effusion, and, occasionally, airspace opacities.

Idiopathic pulmonary arterial hypertension:

Characteristic vascular features of idiopathic PAH depicted at CT are central pulmonary artery dilatation, usually in the absence of detectable intraluminal thrombi (fig. 4); small tortuous peripheral vessels representing plexogenic arteriopathy; and an abrupt decrease in the caliber of segmental and subsegmental arteries. Additional CT findings may include right heart enlargement, pericardial effusion, (fig. 5) and a mosaic pattern of attenuation in lung parenchyma. However, there are no distinct radiologic features to distinguish idiopathic pulmonary hypertension from other causes of PAH, it is a clinical and radiological diagnosis of exclusion.

Congenital heart disease:

Without correction, a ventricular septal defect, an atrial septal defect, a patent ductus arteriosus, or a truncus arteriosus may relentlessly assault the pulmonary arterial circuit with high flow volume and systemic arterial pressure.
On CT and chest radiograph, the central pulmonary arteries may be outlined by linear calcifications representing atheromatous plaque. Dilated central pulmonary arteries may develop chronic thrombus due to sustained turbulent flow. The central pulmonary arteries may become so large that they produce airway compromise (fig. 6). Bronchial arterial collateral vessels and peripheral nodular "neovascularity" are occasionally evident as angiocentric pulmonary nodules.

**Thromboembolic vaso-oclusion, chronic thromboembolic pulmonary hypertension (CTEPH):**

The CT features of CTEPH can be classified as vascular or parenchymal changes. Directly visualized intraluminal thrombi in the pulmonary arteries is the vascular CT finding with the highest specificity for a diagnosis of CTEPH.

Vascular changes include central pulmonary artery dilatation, right heart chamber enlargement, atherosclerotic plaques, complete or partial thromboembolic obstruction (fig. 7), bands or webs in the pulmonary arteries, and a systemic collateral supply.

Parenchymal changes include mosaic lung perfusion and peripheral parenchymal opacities. Mosaic lung perfusion is characterized by sharply demarcated regions of hypoattenuation with reduced vessel size and without air trapping, interspersed with adjacent areas of normal attenuation or relative hyperattenuation (fig. 8). Findings of disparity in the size of segmental vessels and mosaic lung attenuation reliably distinguish CTEPH from nonthromboembolic pulmonary arterial hypertension.

**Pulmonary interstitial disease and hypoxic states:**

Lung disease is the most common cause of pulmonary hypertension, and the presence of pulmonary hypertension in this setting is an unfavorable prognostic sign. The most common lung diseases that are associated with PH include chronic obstructive pulmonary disease, interstitial lung disease, and other diffuse pulmonary diseases such as connective tissue disease, sarcoidosis, and pulmonary Langerhans cell histiocytosis.

The pathophysiologic mechanisms include acute hypoxic vasoconstriction, vascular remodeling due to sustained alveolar hypoxia, loss of cross-sectional lung area caused by destruction of the alveolar capillary septa, compression of the alveolar vessels by increased intraalveolar pressures (chronic obstructive pulmonary disease), and fibrotic compression or obliteration of the pulmonary vessels (interstitial lung disease). At CT, the characteristic features of pulmonary arterial hypertension are seen in combination with pathologic changes caused by the underlying lung disease (fig. 9). In patients with
advanced pulmonary fibrosis, the pulmonary artery diameter often does not correlate with the presence or degree of pulmonary hypertension. In these patients, the ratio of the pulmonary artery diameter to that of the ascending aorta is a more reliable indicator of pulmonary hypertension than the absolute main pulmonary artery diameter.

**Associated systemic disorders (HIV):**

The mechanism leading to the development of pulmonary arterial hypertension in the setting of HIV infection is unknown. Indirect action of the virus through second messengers such as cytokines and growth factors and genetic predisposition may be involved.

Because of considerable overlap between HIV-associated and idiopathic pulmonary arterial hypertension with regard to their clinical, pathologic, and radiologic manifestations, HIV testing should be considered in patients with CT evidence of pulmonary arterial hypertension without another identifiable cause.

**Pulmonary Veno-occlusive Disease**

Pulmonary veno-occlusive disease is histologically characterized by organized and recanalized thrombi and eccentric intimal fibrosis in the pulmonary veins and venules.

At CT, the combination of features of pulmonary arterial hypertension with interstitial and alveolar edema is virtually diagnostic of pulmonary veno-occlusive disease. CT scans show markedly small central pulmonary veins, interlobular septal thickening, and patchy centrilobular ground-glass opacities representing interstitial and alveolar edema.

Additional CT findings include dilatation of the central pulmonary arteries, right ventricular enlargement, a normal-sized left atrium, pleural effusion, and mediastinal lymphadenopathy.

**Pulmonary Venous Hypertension Associated with Cardiac Disease**

PVH is most frequently caused by left-sided cardiac disorders such as left ventricular failure, its most common cause both in adults and in children. The distinguishing feature is left atrial enlargement (fig. 10), which is frequently seen in left-sided cardiac disease but not in pulmonary veno-occlusive disease.
Extravascular constriction

CT facilitates direct visualization and extent of vessel encasement as well as redirected blood flow. With direct venous obstruction, lung parenchymal changes on CT include dilated pulmonary veins, thickened interlobular septa, prominent bronchial walls, ground-glass opacities and paraseptal venous infarcts, which all reflect significant pulmonary venous congestion.
Fig. 1: Posteroanterior (PA) chest radiograph of a patient with pulmonary artery hypertension shows moderate cardiomegaly, marked enlargement of the pulmonary trunk (left arrow) and right pulmonary artery (right arrow). Note the tapering of the peripheral pulmonary arteries.

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Fig. 2: Left: Axial multidetector CT angiogram shows dilatation of the main pulmonary arteries. Right: chest radiograph correlation. Notice the marked enlargement of the right interlobar artery.

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Fig. 3: Posteroanterior chest radiograph and axial multidetector CT obtained in a 77 years-old man with CTEPH show hilar enlargement, tapering of peripheral pulmonary arteries and areas of oligemia (best seen on CT).

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**Fig. 4:** Idiopathic PH. Axial contrast-enhanced CT scan shows dilatation of the pulmonary artery trunk (42 mm) and the right main pulmonary artery.

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**Fig. 5:** Idiopathic PH. Notice the right heart enlargement, with straightening of the interventricular septum and pericardial effusion.

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Fig. 6: Congenital heart disease. Axial contrast-enhanced CT scan obtained in a 51 years-old man diagnosed with atrial septal defect shows marked enlargement of the pulmonary arteries and dilatation of the right ventricle. Notice the abnormal opening in the atrial septum allowing communication between the right and left atria.

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**Fig. 7:** CTEPH. Axial pulmonary CT Angiogram shows an eccentric filling defect in the left main pulmonary artery and right interlobar artery consistent with a chronic thromboembolus. Note that the thrombus is muraly adherent and not surrounded by contrast material. The main pulmonary artery and right heart are enlarged.

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![CT Angiogram](image)

**Fig. 8:** Axial CECT of a patient with longstanding PAH shows alternating geographic regions and increased and decreased lung attenuation. These findings are consistent with mosaic perfusion of the lungs in chronic PAH.

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Fig. 9: Chronic obstructive pulmonary disease. Axial multidetector CT images show enlarged central pulmonary arteries. Parenchimal findings include diffuse bronchial wall thickening and focal areas of emphysema and discrete bullae.

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**Fig. 10:** Left-sided cardiac disease. Axial CECT obtained in a 85 years-old woman with left heart failure shows bilateral pleural and pericardial effusions, marked dilation of the left ventricle and left atrium. Right ventricular overload demonstrates enlarged pulmonary arteries and reflux of contrast into the hepatic veins.

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Conclusion

Radiologic imaging provides an essential tool for the comprehensive analysis of the pulmonary vasculature, lung parenchyma and cardiac morphology and function in pulmonary hypertension.

Chest radiography and CT with standard axial and angiographic acquisitions help to localize the primary site of disease as either precapillary or postcapillary, and further may provide insight into the underlying etiology and differentiating among the various causes of secondary pulmonary hypertension.
References


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