ASK THE EXPERT

Radiographic Signs and Patterns of Pulmonary Hypertension: A Pictorial Essay

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INTRODUCTION
Pulmonary hypertension (PH) is a disease characterized by elevated mean pulmonary arterial pressure and/or elevated pulmonary vascular resistance, as measured by right heart catheterization.1–4 Individuals often present with dyspnea and decreased functional exertion. Imaging features of PH may be present prior to clinical diagnosis. Certain radiographic signs can be useful in identifying the etiology of the PH.5 We present a variety of imaging features that are aimed at making these etiologies apparent to clinicians, with a focus on those etiologies that do not otherwise usually have distinguishing clinical manifestations.

EVIDENCE OF PH ON PLAIN CHEST RADIOGRAPHY
There are a variety of signs that are present in advanced PH regardless of etiology. The most obvious begin with the changes in the main pulmonary artery (PA) itself. On plain radiography, enlargement of the main PA typically obscures the expected concavity between the aortic arch and the main PA silhouette, the so-called “aortopulmonary window” (Figure 1). Additionally, dilated descending left and right PAs can suggest PH (Figures 2 and 3). Right atrial enlargement and right ventricle (RV) enlargement (Figure 2) may also be evident.

Key Words—pulmonary hypertension, pulmonary arterial hypertension, radiographic signs, CT signs

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Figure 1: Frontal chest radiograph of a 25-year-old woman with Group 1 pulmonary arterial hypertension secondary to systemic lupus erythematosus demonstrating loss of the aortopulmonary window (thick black arrow), now replaced by an outward convexity, due to main pulmonary artery enlargement. Additionally, the individual has bilateral dilated descending pulmonary arteries (thin black arrow [right] and white arrow [left]).

Figure 2: Lateral chest radiograph of a 33-year-old woman with pulmonary arterial hypertension and plexogenic arteriopathy demonstrating right ventricular enlargement as evidenced by obscuration of the expected retrosternal clear space (white arrow).
EVIDENCE OF PH ON CT

The main PA diameter (mPAD) should be measured on computed tomography (CT) 1 cm from the bifurcation of the right and left PAs, or at least at the widest portion within 3 cm of the bifurcation\(^6\) (Figure 4). An mPAD $\geq 29$ mm has a sensitivity of 87%, specificity of 89%, and positive predictive value (PPV) of 0.97 for PH.\(^6\) Another common measurement compares mPAD to aorta diameter, abbreviated here as PA-Ao ratio. A PA-Ao ratio $> 1$ has a sensitivity of 57% and a specificity of 81% for PH.\(^7\) While mPAD dilation can suggest the presence of PH, it does not correlate with PH severity, but rather to the length of time that PH has been present.\(^8\)

In addition to static measurements of the great vessels, Scelsi et al.\(^7\) have shown that being able to visualize the main PA at the same level as the most caudal aspect of the aortic arch, the so called “egg-and-banana” sign (Figure 5), is also associated with the presence of PH. Particularly, they noted that its presence had a similar PPV (85%) to the PA-Ao ratio $> 1$ (PPV 87%) and that the combination increased the PPV to 90%.

Left ventricle (LV) and RV characteristics can also point to the presence of PH (Figure 6). An RV free wall thick-
Figure 5: Computed tomographic angiogram of the chest at the level of the caudal aspect of the aortic arch (yellow) in a 32-year-old man with Wiskott-Aldrich syndrome complicated with pulmonary hypertension. Note that the main pulmonary artery (pink) is visible at this level, the so-called "egg-and-banana" sign.

Figure 6: Computed tomography of the chest with contrast, demonstrating an increased right ventricle-to-left ventricle ratio, as well as interventricular septal flattening (black arrow) in an individual with pulmonary hypertension, suggesting right ventricular pressure overload.
ness \( \geq 6 \text{ mm} \) on CT has an odds ratio (OR) of 30.5 for PH, and an RV-to-LV luminal diameter ratio of \( \geq 1.28 \) carries an OR of 28.8. Interventricular septal flattening and reflux of contrast material down the inferior vena cava also suggest RV pressure overload\(^9\) (Figure 7).

**ETIOLOGIES OF PH**

While many of the above findings may still be nonspecific to PH, if PH is strongly suspected or confirmed clinically, the following findings might provide strong clues as to the etiology of the PH. There are a few diseases that have very distinct imaging findings and usually have little extrapulmonary clinical manifestations. We have decided to group these findings in order by the proposed World Symposium on Pulmonary Hypertension clinical group classifications for PH.\(^3,11,12\)

**Group 1**

This group is made up of a diverse set of precapillary and postcapillary diseases that lead to pulmonary arterial hypertrophy.

Figure 7: Computed tomographic angiogram of the chest demonstrating reflux at the level of the inferior vena cava (IVC; black arrow) due to elevated pulmonary artery pressures and elevated pulmonary vascular resistance.

Figure 8: Computed tomographic angiogram of the chest in a 33-year-old woman with pulmonary arterial hypertension and plexogenic arteriopathy. Images show tortuous, corkscrew-like peripheral pulmonary arteries (black arrow) and faint centrilobular nodules (thin black lines).
Plexogenic pulmonary arteriopathy is the pathognomonic histopathologic lesion of PAH, with its presence signifying the ongoing angiogenic pathology of PAH. The pulmonary vasculature undergoes a progression of changes from arterial medial hypertrophy to subsequent intimal proliferation and eventually compensatory plexus. Tortuous hypertrophic arterioles may be seen peripherally as a result (Figure 8). Poorly defined ground glass attenuation centrilobular nodules can be seen on CT (Figure 9), which might represent either the plexogenic lesions or cholesterol deposits.

Partial anomalous pulmonary venous return (PAPVR) is an anomalous venous connection in which some pulmonary venous return drains back to the right atrium, either via direct connection or through an atrial septal defect (ASD; Figure 10), but does not cause total cyanosis as might a total anomalous connection. Left-to-right shunting is present which might eventually lead to PH. PAPVR is overall a rare finding, but its presence should prompt search for ASD, especially a sinus venous type of ASD if the location of PAPVR is in the right upper lobe (Figure 11), as these are highly associated.

Pulmonary veno-occlusive disease (PVOD) is a postcapillary disease with
characteristic findings of interlobular septal thickening, resembling the parenchymal imaging findings of left-sided heart failure without clinical evidence of left-sided congestion (Figure 12).

Pulmonary capillary hemangiomatosis (PCH) may be on the spectrum with PVOD as one disease.20 PCH tends to present with diffuse, random ground glass attenuation nodules (Figure 13) corresponding to foci of capillary proliferation in the secondary pulmonary lobules. Characteristically, PCH is a postcapillary contributor to PH, as is PVOD. Notably, PCH and PVOD do not respond to pulmonary vasodilators, and there is an increase in risk of pulmonary edema and respiratory failure21 with these advanced vasodilator therapies.

Figure 11: T2 black blood sequence in the same individual as in Figure 10 confirms the presence of a sinus venosus type of atrial-septal defect (white arrow). Right atrium (RA), left atrium (LA), aorta (A), right ventricle (RV), and left ventricle (LV) are labeled on this image.

Figure 12: 42-year-old woman with pulmonary veno-occlusive disease (PVOD). Computed tomography of the chest demonstrating interlobular septal thickening (thin black arrow) and mediastinal and hilar lymphadenopathy (thick black arrow). These features are common to both PVOD and left-sided heart failure.

Figure 13: 17-year-old man with pulmonary capillary hemangiomatosis (PCH). Computed tomography of the chest demonstrates multiple randomly distributed ground glass attenuation nodules characteristic of PCH.
**Group 2**
Chest radiography can show LV hypertrophy, as well as left atrial and ventricular dilation, all to suggest left-sided cardiomyopathy that is associated with Group 2 disease.

**Group 3**
The main causes of Group 3 disease are the chronic hypoxemic respiratory diseases such as interstitial lung disease (ILD) and chronic obstructive pulmonary disease. The role for chest CT in making a noninvasive diagnosis for this group of diseases, particularly ILD, is well established.

**Group 4**
Chronic thromboembolic PH (CTEPH) is an underdiagnosed etiology and is often diagnosed at the same time that an individual presents with manifestations of their PH. The diagnosis is increasingly important given advances in treatment. CTEPH has several characteristic imaging features, including imaging findings of acute and chronic PA thrombi (Figures 14–16), bronchial artery hypertrophy (Figures 17 and 18), and mosaic attenuation (Figures 19–21).

**Group 5**
Group 5 etiologies represent a variety of diseases for which the mechanism in association with PH is not yet well understood. These include sarcoidosis and hematologic malignancies, among others.

![Figure 14: 26-year-old woman with chronic thromboembolic pulmonary hypertension. Computed tomographic angiogram of the chest shows an acute pulmonary embolism (arrow) in the right main pulmonary artery, seen as a central near-completely occlusive filling defect.](image1)

![Figure 15: 26-year-old woman with chronic thromboembolic pulmonary hypertension (same individual as in Figure 14). Computed tomographic angiogram of the chest shows an acute pulmonary embolism in the right main pulmonary artery, seen as a central near-completely occlusive filling defect, superimposed on chronic pulmonary embolism (arrow), seen as peripheral filling defects in the basal trunk of the right lower lobe artery.](image2)
Figure 16: 26-year-old woman with chronic thromboembolic pulmonary hypertension (same individual as in Figures 14 and 15). Computed tomographic angiogram of the chest shows an acute pulmonary embolism in the right main pulmonary artery, seen as a central near-completely occlusive filling defect, superimposed on chronic pulmonary embolism, seen as peripheral filling defects in the basal trunk of the right lower lobe artery as well as the right lateral and posterior basal (arrow) segmental arteries.

Figure 17: 57-year-old man with chronic thromboembolic pulmonary hypertension. Computed tomographic angiogram of the chest demonstrates bronchial artery hypertrophy (white arrows) and diminished peripheral lung vascularity.
Figure 18: Same individual as in Figure 17. Computed tomographic angiogram of the chest in coronal projection demonstrates pulmonary artery narrowing (white arrow).

Figure 19: 29-year-old man with chronic thromboembolic pulmonary hypertension (CTEPH). Computed tomography of the chest demonstrating mosaic attenuation in the left lower lobe (black arrow). In individuals with pulmonary hypertension, mosaic attenuation of the lung is most commonly seen in individuals with CTEPH.\(^9\)

Figure 20: Same individual as in Figure 19. Left panel: Computed tomographic angiogram of the chest demonstrates a chronic pulmonary embolism in the basal trunk of the left lower lobe lobar artery. Middle panel: Single photon emission computed tomography perfusion image shows decreased perfusion (white arrow) in the corresponding left lower lobe artery distribution. Right panel: Conventional pulmonary angiogram shows both occlusion of the left lower lobe pulmonary artery and corresponding loss of contrast enhancement (black arrow).
CONCLUSIONS
Awareness of early and late radiographic and cross-sectional imaging features of PH may be helpful in early and accurate diagnosis. In many conditions that cause PH, imaging alone may suggest a specific etiology.

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