Cardiothoracic Interplay: When the clue is in the Heart and Vessels

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No relevant financial disclosures
Introduction

The pulmonary and cardiac systems compose a one functional unit where the pathology of one frequently results in dysfunction of the other.

“Cor pulmonale”, refers to altered function of the right ventricle secondary to abnormal respiratory function and is commonly secondary to chronic obstructive pulmonary diseases, interstitial lung diseases and other hypoventilatory disorders that may lead to abnormal gas exchange and pulmonary hypertension.
Primary cardiac pump dysfunction, pathologies of the cardiac valves, as well as primary and acquired pulmonary arterial and venous abnormalities, may all be the cause of pulmonary opacities in a patient presenting clinically with respiratory dysfunction or hemoptysis, and are less easily recognized during the initial diagnostic approach to the patient.
Learning objectives:

- Understand the intimate interaction between the cardiac and pulmonary physiology and how many cardiac disorders manifest first with abnormalities in the pulmonary parenchyma.

- Identify the specific main pulmonary and cardiovascular imaging findings of these entities that cue the diagnosis.

- Relate the imaging manifestations and the physiopathology of these entities.
Cardiovascular pathology that can first manifest as pulmonary opacities

Heart
- Congestive heart failure
- Cardiac valves pathology
  - Mitral valve disease
  - Endocarditis
- Amiodarone pulmonary toxicity
- Univentricular heart & PAVMs/VVs

Pulmonary veins
- PVOD/ PCH
- Cor triatriatum
- Venous stenosis
  - Fibrosing mediastinitis
  - Post ablation
  - Post lobectomy
  - Lung transplantation
Cardiovascular pathology that can first manifest as pulmonary opacities

- **Pulmonary Arteries**
  - Pulmonary Arterial Hypertension
  - Pulmonary Embolism

- **Systemic Arteries**
  - Pulmonary sequestration
  - Systemic vasculitis
- Congestive heart failure
- Cardiac valves pathology
  - Mitral valve disease
  - Endocarditis
- Amiodarone pulmonary toxicity
- Univentricular heart & PAVMs/VVs
Cardiogenic (Hydrostatic) Pulmonary Edema

Interstitial edema is manifested on chest radiograph or CT by interlobular septal thickening (Kerley's A or B lines), thickening of the fissures, peribronchial cuffing, poor definition of pulmonary vessels, “perihilar haze,” and ground-glass opacities.

As the process progresses and the hydrostatic pressure worsens, fluid overflows the interstitium and fills the alveoli. Findings of air-space edema include ill-defined or confluent alveolar opacities and “acinar” or airspace nodules. Air bronchogram may be seen in some patients.
Mitral valve disease

The hallmark of mitral valve disease is pulmonary venous hypertension which frequently leads to left ventricular failure. Early detection is paramount since medical and surgical treatment are especially successful in the early course of the disease. The morphologic changes in the left atrium are well known. Valvular function is usually assessed by echocardiography.

Parenchymal manifestations of mitral valve disease are usually less well known and are the result of pulmonary venous hypertension in mitral stenosis or regurgitant flow in mitral insufficiency.

**Stenosis**
- Pulmonary edema
- Diffuse alveolar hemorrhage
- Pulmonary hemosiderosis
- Pulmonary ossifications

**Insufficiency**
- Pulmonary edema
Mitral stenosis

Mitral stenosis is usually acquired and nearly always caused by rheumatic fever. While it is a pressure overload lesion that causes little increase in overall heart size during the early phase, it does result in characteristic enlargement of the left atrium (double density signs, splaying of the carina, dorsal displacement of left main bronchus on lateral view) and the left atrial appendage (convexity on the upper left cardiac border). Signs of pulmonary venous hypertension are characteristic.

Parenchymal abnormalities are the result of:
- **pulmonary edema**: interstitial or alveolar. In chronic mitral stenosis, septal lines may represent interstitial fibrosis and deposition of hemosiderin-laden macrophages.
- **diffuse alveolar hemorrhage**: confluent acinar or ground-glass areas of increased opacity, characteristic subpleural sparing.
- **pulmonary hemosiderosis**: small, ill-defined nodules or coarse reticular opacities.
- **pulmonary ossifications**: densely calcified micronodules.
Mitral insufficiency

Mitral insufficiency can be the result of infective endocarditis, myxomatous degeneration of the mitral valve, or rupture of the chordae tendineae or papillary muscle.

Imaging findings depend on the acuteness of the disease, with variable degrees of PVH, cardiomegaly, left atrial enlargement, and LV enlargement and sometimes signs of right-sided chamber enlargement.

Symmetric interstitial and alveolar pulmonary edema is the most frequent parenchymal manifestation.

A less common but pathognomonic manifestation of acute MI is asymmetric right upper lobe edema (arrows) due to the anatomic disposition of the mitral valve and the regurgitant jet (arrow) towards the right upper pulmonary vein.
Hematogenous dissemination ("septic embolization") of microorganisms to the lungs is usually encountered in patients with history intravenous drug abuse and bacterial tricuspid valve endocarditis.

Septic embolism typically presents as multiple pulmonary nodules that cavitate over a period of several days. These nodules are usually peripherally located and predominate in the lower lobes and are closely associated with pulmonary vessels: "feeding vessel sign". Wedge-shaped areas of peripheral consolidation, representing septic pulmonary infarction, are commonly seen.
Amiodarone pulmonary toxicity

Amiodarone is a triiodinated drug that has been commonly used to treat atrial fibrillation as well as supraventricular and ventricular tachycardia. Amiodarone causes numerous side effects, the most serious of which is pulmonary toxicity that occurs in approximately 5-10% of patients.

The most common manifestation is non-specific interstitial pneumonia characterized by scattered or diffuse areas of ground-glass attenuation usually associated with pleural effusion. Basilar predominant fibrosis can be seen at a later stage. Organizing pneumonia is less common and almost always appears associated to NSIP.

Focal, peripheral, homogenous consolidations with characteristic high-density are a distinguishing feature.

The concomitant increased liver and spleen attenuation together with frequent cardiomegaly with left atrial enlargement supports the diagnosis.
Extra-cardiac complications are frequent in patients with Glenn or Fontan correction of univentricular congenital heart diseases. Right to left shunts may occur due to the development of pulmonary arteriovenous fistulae (PAVFs) and the formation of systemic-pulmonary veno-venous shunts (VVS). The lack of pulsatility in the pulmonary arteries and the absence or asymmetrical distribution of the hepatoenteric inflow in the lungs are considered potential etiological factors. Veno-venous collateral development with connection between the systemic and pulmonary circulation, systemic-pulmonary VVS (*), is a consequence of an elevated central venous pressure.

Maximal intensity projection (MIP) reconstructions can help identifying PAVF and VVs on CTA. When clusters of small innumerable PAVFs occur, multifocal groundglass opacities can be seen on CT (*); additional regional early venous drainage is also frequently seen.
- Pulmonary Arterial Hypertension
- Pulmonary Embolism:
  - Acute
  - Chronic
Pulmonary Arterial Hypertension

Pulmonary hypertension is defined as an abnormal elevation of pressure in pulmonary circulation, with a mean pulmonary arterial pressure higher than 25 mmHg. The term pulmonary arterial hypertension (PAH) is restricted to cases with a hemodynamic profile in which high pulmonary pressure is a result of elevated precapillary pulmonary resistance and normal pulmonary venous pressure (shared by groups 3, 4, and 5 in the Dana Point Classification).

Chest radiography findings include enlargement of the pulmonary trunk (₁) and pruning of peripheral branches.

Enlargement of the pulmonary arteries characterized by a pulmonary trunk greater than 29 mm (₂) together with centrilobular nodules (₃) are characteristic CT findings. Pleural and pericardial effusions, and mediastinal lymphadenopathy are frequently seen.

The centrilobular nodules (especially common in patients with idiopathic PAH) represent cholesterol granulomas, which are caused by ingestion of red blood cells by pulmonary macrophages, a result of repeated episodes of pulmonary hemorrhage.
Focal parenchymal abnormalities, particularly atelectasis are the most common chest radiographic abnormalities in patients with PE.

Focal air-space consolidation may occur in patients with PE and may represent pulmonary hemorrhage without infarction or true pulmonary infarction with ischemic necrosis of lung tissue.

Infarcts often are multiple and occur most frequently in the subpleural regions of the lower lobes, usually within 12 to 24 hours of the onset of symptoms. Wedge shape peripheral consolidations with internal lucencies ( ) or air-bronchogram are characteristic.
Chronic emboli are eccentric in location and usually appear as a smooth or sometimes nodular thickening of the vessel wall on CTPA studies. It may calcify, and the main pulmonary arteries may be dilated because of associated pulmonary hypertension. Additionally, small linear filling defects, or “webs” are indicative of chronic PE.

Geographic regions of mosaic perfusion (oligemia) also may be seen either with or without central findings of chronic PE. Often pulmonary vessels appear smaller in the regions of hypoattenuation, a finding that aids in suggesting a vascular cause for inhomogeneous lung opacity over an airway etiology.
- PVOD/ PCH
- Cor triatriatum
- Venous stenosis
  - Fibrosing mediastinitis
  - Post ablation
  - Post lobectomy
  - Lung transplantation
Pulmonary veins pathology

- A fine balance between the two primary vascular networks of the lung, the pulmonary and bronchial systems, is maintained to support the physiology of the lung parenchyma.

- The pulmonary venous system is responsible for the post capillary pressure of the pulmonary system.

- Pathologic processes involving the pulmonary veins lead to vein stenosis and may result in pulmonary venous hypertension (PVH).
Pulmonary vein pathology

**Congenital**
- PVOD/ PCH
- Cor triatriatum

**Acquired**
- Venous stenosis
  - Neoplasms
  - Fibrosing mediastinitis
  - Post ablation
  - Post lobectomy
  - Lung transplantation
PVOD / PCH

Diffuse bilateral ground glass acinar and centrilobular nodules, smooth interlobular septal thickening and occasionally pleural effusions are the classic imaging findings in these patients.

The enlargement of the main pulmonary artery (> 29 mm) reflects the presence of PAH and cues the diagnosis when associated with the parenchymal findings, in a typically young patient with otherwise unexplained cause of PAH.
Pulmonary vein stenosis
Fibrosing mediastinitis

The parenchymal findings secondary to venous stenosis are variable according to the acuity of the occlusion and the number of veins involved, although reflect poor venous drainage and lobar pulmonary edema (arrows point thickening of interlobular septa). In chronic processes such as in this case, the occlusion results in parenchymal and pleural scarring ( ).
The fibrotic tissue (white and yellow arrows) infiltrates the mediastinum and characteristically encases the bronchi, vessels and airways. The right inferior pulmonary vein in this case is encased and occluded ( ). Long term venous and arterial occlusion are associated with development of collateral pathways of circulation through bronchial arteries ( ).
Right middle lobe vein accidental occlusion during right upper lobectomy

AP CXR 48 hours after surgery demonstrates consolidative opacities in the residual right lung, which have progressively worsened over time. Coronal CT reformat demonstrates surgical clip occluding the RML vein ( ), with consequent hemorrhagic infarction of the RML ( ).

Including anatomic variants of the pulmonary veins in the radiologic report of patients undergoing lung resection is essential to decrease the incidence of iatrogenic vascular complications.
Post surgical pulmonary edema and areas of parenchymal contusion after partial lung resection are expected to resolve during the first week after the procedure. The progressive consolidation of the residual lung with bulging of the fissure ( ) are findings secondary to torsion and hemorrhagic infarction of the RML, a known complication of RUL lobectomy.
Axial image and coronal reformats (left and middle images) of the same patient demonstrate consolidation of the RML ( ), representing hemorrhagic ischemia due to venous occlusion.

Oblique MIP reformat (far right) demonstrates swirling of the hilar vessels ( → ), characteristic of lobar torsion. Although the pulmonary arteries are patent, the pulmonary vein is compressed and occluded, leading to venous congestion and lobar infarction.
Systemic arteries

- Pulmonary sequestration
- Systemic vasculitis
Systemic Arteries pathology

- The causes of pulmonary opacities secondary to pathology of the systemic arteries are overall less common due to the relative low incidence of this group of entities.

- Since no morphologic abnormalities are usually depicted in the systemic arteries, primary and secondary pulmonary vasculitis are not discussed in this review. These may manifest primarily as parenchymal opacities, pulmonary nodules, cavities or alveolar hemorrhage.
The imaging manifestations of pulmonary sequestration vary according to location, volume of parenchyma involved and the presence of complications, more frequently being superimposed infection.

In this case, frequent episodes of infection in the left lower lobe in a young patient lead to a multiloculated pulmonary abscess (*) and multilobar bronchopneumonia ( )
Pulmonary sequestration

- Pulmonary sequestrations are classified as:
  - Intralobar: most frequent type, draining via pulmonary veins
  - Extralobar: venous drainage through systemic veins.
- The identification of the systemic arterial supply is the key to diagnosis
- Contrast enhanced CT is the imaging method of choice for diagnosis & treatment planning
- Assessment of the involved lung parenchyma and related complications such as superimposed infection, cavitation and pulmonary abscess formation, are also possible by CT.

Coronal CT reformat depicts the systemic artery arising form the descending thoracic aorta, supplying the sequestration in the LLL.
Conclusions

- Primary cardiac and vascular pathologies may lead to pulmonary abnormalities that are not infrequently the first clinical and radiologic manifestations of the disease.

- Recognizing diagnostic clues in the cardiovascular system helps orienting the diagnosis; optimizes and guides further imaging work-up and treatment, and impacts patient outcome.
References
