Beyond Pulmonary Emboli: Acquired Abnormalities of the Pulmonary Arteries on CTA

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Learning objectives

- Review common imaging features of bland pulmonary artery embolus

- How to identify tumor thrombus (primary or metastatic) on computed tomography angiogram (CTA)

- Discuss less common acquired pulmonary artery abnormalities, including pulmonary artery aneurysm and pulmonary artery vasculitis (i.e. Behçet disease)

- Brief discussion of chronic pulmonary emboli

- Identify common mimics of pulmonary embolism on CTA, including motion artifact, contrast opacification artifacts, bronchial obstruction, and pulmonary vein thrombus
Background

- CT angiograms of the thorax are a frequently performed study in the emergency department

- Incidence of pulmonary embolisms is estimated to be 60 to 70 per 100,000 people

- Pulmonary embolism ranks third among the most common types of cardiovascular disease

- Due to this large volume of studies, the general radiologist is likely to encounter even relatively rare abnormalities of the pulmonary arteries in their practice
Modalities for evaluation of the pulmonary arteries

- **CT angiography** of the pulmonary artery is the current standard of care for diagnosing and following pulmonary artery abnormalities

- Additional options for evaluation include: Nuclear medicine ventilation/perfusion imaging, MR angiography, echocardiography and catheter angiography
CT angiography – Technique

- Goal of the examination is to acquire images of the pulmonary vasculature during maximal enhancement (majority of contrast is in the pulmonary arteries)

- At our institution:

- **70 cc contrast** (iodine based with a density of 300-350 mg/ml) bolus is injected at a **fast rate (4 cc/sec)** through a 18 or 20g IV, followed by a **50 cc saline flush** to decrease streak artifact from bolus.

- **Imaging triggered with bolus tracking method**: Sequential low dose scans over the main pulmonary artery with an ROI are used to “track” the bolus until a threshold attenuation is reached (~150 HU). Imaging then begins after a **short delay (~6 seconds)**.

- **Important**: Prior to imaging the patient must be coached to stop breathing (not take in a breath and hold it) when the threshold trigger is reached. This helps to prevent transient interruption of the contrast bolus.

- Goal: **>200 HU** in the main pulmonary artery and minimal standard deviation
Case 1

49 year old female presents to the ED with acute shortness of breath, hypotension, tachycardia and lethargy.
Bland Pulmonary Embolism

What is it?
- Embolic thrombus usually from lower extremity venous vasculature

Clinical presentation
- Acute shortness of breath with or without chest pain
- Tachycardia
- Tachypnea
- New onset right heart failure

Imaging characteristics
- Filling defect without enhancement
- Tends to get “caught” at branching points of vessels (like the saddle embolism seen in this case)
- Pulmonary infarcts can also be seen in severe cases (wedge shaped areas of parenchymal opacity)

Treatment and Prognosis
- Initial treatment depends on hemodynamic stability
- In hemodynamically unstable patients embolectomy and thrombolytic therapy are considered
- Anticoagulation – duration depends on whether or not there is an underlying etiology

Axial CT images demonstrating a saddle pulmonary embolism at the branch point of the main pulmonary artery and a wedge shaped pulmonary infarct.
Bland Pulmonary Embolism – CT Signs of Increased Severity

Assess for evidence of right ventricular strain: Flattening of the interventricular septum and compression of the left heart which correlates with D sign on echocardiography. Right ventricular enlargement and reflux of contrast into distended IVC and hepatic veins. Mottled appearance of the liver parenchyma secondary to hepatic congestion can be seen on later phase abdominal imaging.
52 year old female presents with worsening shortness of breath and syncope. Recently diagnosed with pulmonary embolism and has been on Lovenox for 6 weeks.
Pulmonary artery sarcoma

**Subtle areas of linear contrast enhancement** within the hypodense filling defect of the main pulmonary artery, consistent with vascularity. The lesion extends through the **expected region of the pulmonary valve and into the right ventricular outflow tract**. Note how the lesion nearly fills the entirety of the **enlarged main pulmonary artery**, with greater than 90% occlusion. Importantly, this was increased in size when compared with prior imaging despite lovenox therapy.

Additional clinical history: patient presented with chronic increasing, not acute, shortness of breath.
Pulmonary artery sarcoma

What is it?
- Rare primary tumor of the pulmonary artery
- Originates from the intimal mesenchymal cells

Clinical presentation
- Worsening dyspnea or chest/back pain, possibly with cough or hemoptysis
- Nonspecific and may explain why these are frequently initially misdiagnosed as bland emboli
- Occasionally, more specific symptoms such as weight loss can be identified

Imaging characteristics
- Destruction of vascular structures (such as pulmonary valve in this case)
- Subtle areas of enhancement
- Persistence and growth on follow up imaging

Treatment and Prognosis
- Median survival of 1.5 months
- Treatment is surgical with or without chemotherapy.
  Different surgical approach compared with thrombectomy and preoperative diagnosis can be critical

Note clumps of small round blue cells within a highly cellular mass inside of the vessel lumen. Pulmonary artery wall of smooth muscle cells is not clearly invaded on this image
Case 3

59 year old man with PMHx notable for metastatic squamous cell carcinoma presents for restaging CT of the chest.
Pulmonary artery aneurysm/pseudoaneurysm

What is it?
- Focal dilatation of the pulmonary artery

Clinical presentation
- Related to the underlying etiology of the aneurysm/pseudoaneurysm formation
- Can cause massive hemoptysis
- Differential diagnosis is large (both congenital and acquired)
- Leading considerations in an adult patient should include infectious etiologies, vasculitis, and sequelae of malignancy
- Iatrogenic considerations include formation related to malpositioned Swan-Ganz Catheter

Imaging characteristics
- Focal dilatation or outpouching from a branch of the pulmonary arteries
- May be partially calcified (rim calcifications) or thrombosed

Treatment and prognosis
- Related to underlying etiology
- May be at risk for rupture depending on size and etiology (especially mycotic aneurysms) and may require surgical resection or endovascular treatment in certain cases

Eccentric outpouching of a right upper lobe pulmonary arterial branch vessel concerning for aneurysm/pseudoaneurysm formation. Surrounding soft tissue density associated with the lesion. Note separate left upper lobe lesion found to be metastatic squamous cell carcinoma. Pleural metastatic disease is also present.
Pulmonary artery aneurysm/pseudoaneurysm

Additional example of isolated pulmonary artery aneurysm/pseudoaneurysm. This case started as aneurysmal dilation proximal to a pulmonary embolism in a subsegmental branch vessel. Coronal MIP image shows connection to the pulmonary artery.
Case 4

20 year old woman with past medical history notable for known vasculitis and pulmonary artery pseudoaneurysm presents for evaluation of new hemoptysis.
Behçet’s disease

What is it?
- Chronic multisystem inflammatory disorder with vasculitis

Clinical presentation
- Generally a clinical diagnosis
- Recurrent oral ulcerations with two of the following:
  - Recurrent genital ulcerations
  - Eye lesions including uveitis and retinal vasculitis
  - Skin lesions (folliculitis, erythema nodosum)
  - Positive skin pathergy test

Imaging characteristics
- Common cause of pulmonary artery aneurysm/pseudoaneurysm, can undergo in situ thrombosis and form fistulae to bronchial tree, as suggested by small foci of air in this case
- Similar to other vasculitis, can be seen as thickening of vessel walls
- Can also manifest as pulmonary infarcts, hemorrhage, recurrent pneumonia and fibrosis

Treatment and prognosis
- Symptomatic treatment for manifestations of disease
- This case required both coil embolization and surgical resection
- Pulmonary artery aneurysm is associated with poor prognosis (30% of patients die within 2 years after diagnosis)
Images from interventional radiology coil embolization demonstrate conventional angiographic appearance of this pulmonary artery aneurysm. Note the altered blood flow to the right lower lobe with delayed parenchymal perfusion on the first image. Post procedure CT demonstrates complete coiling of the aneurysm.
34 year old female presents with embolic stroke on head CT. 4 small arteriovenous malformations identified on CT angiogram of the head and neck. CT chest imaging was preformed after right lower lung mass was seen on plain film imaging.
Osler-Weber-Rendu

What is it?
- Also known as Hereditary Hemorrhagic Telangiectasia (HHT)

Clinical presentation
- Combination of epistaxis, GI bleeding and iron deficiency anemia in patients with characteristic telangiectasia of the lips, oral mucosa and finger tips
- Estimated that greater than 50% of patients also have pulmonary involvement
- Approximately 10% have cerebral involvement
- Can present with embolic strokes due to right to left shunting, as is the likely scenario in our case

Imaging characteristics
- Arteriovenous malformations and arteriovenous shunts
- Generally thin walled (compared to vasculitis) and without surrounding soft tissue density
- Note lobulated appearance representing torturous course of multiple vessels

Treatment and prognosis
- Coil embolization and/or surgical resection depending on size and clinical significance
- Patients with clinical HHT should be screened for pulmonary AVMs given high incidence and associated morbidity. Screening options include CT and contrast enhanced echocardiography

Large AVM connecting the right middle lobar pulmonary artery to the superior right pulmonary vein.
Osler-Weber-Rendu – Cerebral Manifestations

Images from a noncontrast CT head and CT angiogram of the head from the same patient showing subacute infarct (star), subtle cutoff of the left superior cerebellar artery and cerebral arteriovenous malformations.
Case 6

45 year old man with hemoptysis preceded by infectious symptoms. Abdominal CT demonstrates ill-defined retroperitoneal mass-like region adjacent to the aorta.
Fibrosing Mediastinitis

What is it?
- Also known as sclerosing mediastinitis and mediastinal fibrosis
- Extensive fibrotic reaction in the mediastinum

Clinical presentation
- Thought to result from excessive host response to prior infection, generally involving mediastinal lymph nodes
- Majority of cases thought to be sequelae of Histoplasmosis (although still very rare with less than 1% of patients with histoplasmosis developing fibrosing mediastinitis
- Can also be idiopathic (possibly autoimmune) and associated with fibrosis in other sites, including retroperitoneal fibrosis

Imaging characteristics
- Mediastinal soft tissue density with or without calcifications
- Surrounds and narrows adjacent structures, such as the proximal pulmonary arteries and bronchi
- Two types – focal (generally calcified and related to prior histoplasmosis, more common) and diffuse (generally idiopathic)

Treatment and prognosis
- Symptomatic – endobronchial and endovascular stent placement for airway and vessel narrowing
- Systemic antifungals and/or corticosteroids if concerned for infectious etiology

Long segment narrowing of the right main pulmonary artery due to ill-defined soft tissue density of the mediastinum. Note associated calcification.
Axial CT images from a different patient with known diffuse fibrosing mediastinitis demonstrates appropriate positioning of pulmonary artery stents which are patent.
65 year old man with history notable for prior pulmonary embolism (seen on axial CTA image on the right). Presents for follow up imaging for resolution of ground glass opacities seen on prior study.
Chronic Pulmonary Embolism

What is it?
- Persistent calcified thrombus and/or change of the vessel walls after prior pulmonary embolism
- Calcifications within the pulmonary arteries

Clinical presentation
- Prior pulmonary embolism
- Usually asymptomatic but can progress to pulmonary hypertension

Imaging characteristics
- Calcifications within pulmonary embolism
- Eccentric wall thickening with narrowing of the vessel rather than expansion seen with acute PE

Treatment and prognosis
- Symptomatic

Axial CT image demonstrating a thin linear filling defect in this separate case. Finding is compatible with a web related to prior pulmonary embolism.
Pulmonary Hypertension

What is it?
- Elevated pressures in the pulmonary arteries
- Mean >25 mm Hg at rest or >30 mm Hg during exercise
- Primary (rare) and secondary (more common) causes
- Typically occurs secondary to underlying pulmonary or cardiac disease

Clinical presentation
- Longstanding chronic pulmonary embolism
- Chronic shortness of breath

Imaging characteristics
- Enlarged pulmonary arteries (main pulmonary artery) >2.9 cm in diameter or larger than adjacent aorta
- Peripheral pruning of the pulmonary arteries
- Pulmonary parenchymal changes – mosaic attenuation related to differential perfusion
- Enlargement of right atria and ventricle with signs of right heart failure

Treatment and prognosis
- Symptomatic – oxygen, diuretics and in advanced cases
- Calcium channel antagonists, nitric oxide, endothelin antagonists, prostanoids, phosphodiesterase inhibitors

Axial CT image demonstrating an enlarged main pulmonary artery (larger than aorta) measuring up to 3.8 cm.
Case 8

51 year old woman with PMHx notable for stage IV lung cancer presents for evaluation of worsening SOB and clinical suspicion for pulmonary embolism. **Filling defect** identified in the left superior pulmonary vein.
Related condition: Pulmonary vein thrombosis

What is it?
- Occlusion of the pulmonary vein either from bland or tumor thrombus
- Rare

Clinical presentation
- Nonspecific, may be asymptomatic
- Shortness of breath, hemoptysis, chest pain, fever

Imaging characteristics
- Low density filling defect in the pulmonary vein
- Look for causes, including tumor, extrinsic compression
- May cause pulmonary infarction or segmental pulmonary edema

Treatment and prognosis
- Anticoagulation may be used to prevent progression of thrombus into the left atrium and may lead to recanalization of the pulmonary vein
- Address underlying cause

Companion case: Occluded left superior pulmonary vein. Note adjacent well opacified and patent pulmonary artery.
Mimic: Transient interruption of the contrast bolus

Decreased expected opacification of PA leads to suboptimal eval. Why did this happen: ROI in correct place, threshold met; Bolus of unopacified blood from abdomen due to patient taking a deep breath just before imaging. Note that the SVC is well opacified, indicating contrast was still flowing in. This can be fixed by coaching patient not to take a deep breath.
Mimic: Streak artifact

Beam hardening artifact from contrast bolus positioned in the SVC. Can attempt to avoid by using saline flush to “push” the contrast through. Generally easy to distinguish from pulmonary embolism as it is easily tracked outside of the pulmonary artery (such as in this case where it is visible in the aorta as well)
Heterogeneous enhancement in the right pulmonary artery distally which may be free thrombus or more likely mixing artifact given ill defined appearance and overall higher attenuation. Commonly seen in patients with parenchymal abnormalities causing flow alteration, such as the tumor involving the right lung in this patient. Repeat imaging could be performed if clinically necessary to exclude pulmonary embolism.
CT images demonstrating **apparent tubular filling defect/low attenuation in a left lower lobe structure**. Switching imaging to lung windows and tracking the defect towards the hila show this correlates with a mucous impacted bronchus. Note **fleck** of air within this tubular structure.
References


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