Unusual Pulmonary Arterial Filling Defect caused by Systemic to Pulmonary Shunt in the Setting of Chronic Lung Disease Demonstrated by Dynamic 4D CTA

Kianoush Ansari-Gilani[1*], Robert C Gilkeson[1], Edward M Hsiao[2], Prabhakar Rajiah[1]

1. Department of Radiology, University Hospitals of Cleveland, Case Western Reserve University, Cleveland, Ohio, USA

2. Department of Radiology, Macquarie University Hospital, Macquarie University, Australia

* **Correspondence:** Kianoush Ansari-Gilani, MD, Department of Radiology, University Hospitals of Cleveland, Case Western Reserve University, Cleveland, OH 44106, USA
(*kianoush.ansari@gmail.com*)

Radiology Case. 2015 Nov; 9(11):17-23 :: DOI: 10.3941/jrcr.v9i11.2480

ABSTRACT

Even though pulmonary embolism is by far the most common cause of filling defect in the pulmonary arterial system, other less common etiologies should be considered especially in the setting of atypical clinical scenario or unusual imaging findings. Unusual pattern of filling defect in the pulmonary artery in the setting of chronic inflammatory/fibrotic parenchymal lung disease should raise the concern for systemic to pulmonary artery shunt. This diagnosis is typically made by conventional angiography. Dynamic 4D CT angiography however can be a safe, noninvasive and effective alternative tool for making such a diagnosis. It has the added value of multiplanar reconstruction capabilities and providing detailed anatomy which can be vital for interventional radiologists when planning their approach for possible intervention. We present 2 cases of such shunts, and illustrate the demonstration of these shunts by using dynamic 4D CT angiography.

CASE REPORT

CASE REPORT

In this manuscript we describe 2 cases that show unusual pulmonary artery filling defect raising the question of abnormal systemic to pulmonary artery shunt as the etiology and demonstrate how modern imaging technology can help in elucidating the etiology.

Case 1

An 81-year-old man with history of coronary artery disease, and amiodarone treatment for the past 3 years presented with chest pain and hemoptysis. Chest radiograph (Figure 1) showed increase in interstitial markings with distortion of the underlying parenchymal architecture consistent with changes of amiodarone lung toxicity. CT angiogram (Figure 2) showed a non-opacified left upper lobe segmental pulmonary artery with associated small intraluminal filling defect protruding more proximally into the left main pulmonary artery. Lung window confirmed the presence of interlobular septal thickening and reticulation, consistent with chronic changes of amiodarone lung toxicity and fibrosis (Figure 3). This unusual configuration of the protrusion of the filling defect to the more proximal portion of the pulmonary artery raised the possibility of systemic artery to pulmonary artery shunt, which was subsequently confirmed by using dynamic CT angiography (CTA) of the chest (see interactive image stack 1). Maximum-intensity projection (MIP) image (Figure 4) provided a better depiction of this abnormal communication.

Case 2

A 29-year-old woman with cystic fibrosis (CF) presented with hemoptysis and shortness of breath. Her chest radiograph (Figure 5) showed hyperinflation and upper lobe predominant

bronchiectasis in keeping with patient's underlying cystic fibrosis. CT angiogram (Figure 6) showed a filling defect in the bilateral pulmonary arteries with protrusion of the filling defects to the more proximal portion of the pulmonary arterial system and curvilinear areas of non-opacification raising the possibility of systemic artery to pulmonary artery shunt. Lung windows (Figure 7) also showed bronchial wall and peribronchial interstitial thickening with upper lobe predominant bronchiectasis, in keeping with underlying cystic fibrosis. The presence of systemic to pulmonary artery shunt was subsequently confirmed by dynamic CTA (see interactive image stack 2).

DISCUSSION

Etiology & Demographics:

Pulmonary embolism (PE) is the most common etiology for the presence of filling defect in the pulmonary artery and its branches [1]. PE is the third most common acute cardiovascular disease after myocardial infarction and stroke and results in thousands of deaths each year [2, 3]. However there are multiple other possible causes for pulmonary arterial filling defects other than PE, which should be considered in the setting of atypical clinical presentation or imaging findings.

Abnormal communication (shunt) between the systemic and pulmonary circulation presenting as a filling defect in the pulmonary artery is rare [4]. It can be caused by chronic inflammation of the lung parenchyma in the setting of chronic lung disease [4]. In chronic inflammation pulmonary circulation is reduced or occluded at the level of the pulmonary arterioles because of hypoxic vasoconstriction, intravascular thrombosis, and vasculitis. This causes proliferation of the bronchial and adjacent systemic arteries trying to replace the pulmonary circulation. Abnormal communication (shunt) can happen during this process [4].

In our patients, the filling defects were due to shunt between systemic and pulmonary circulation. The shunt was caused by chronic inflammatory processes, amiodarone toxicity in the first patient and cystic fibrosis in the second patient, which probably resulted in erosion of pulmonary artery and systemic artery, resulting in a communication between the two. Other causes of chronic inflammatory diseases reported to cause such shunts include tuberculosis and idiopathic pulmonary fibrosis [5]. Congenital and acquired etiologies such as surgery (Coronary artery bypass graft surgery), trauma and neoplasia [6] are other causes of shunts.

Clinical & Imaging findings:

Patients with systemic to pulmonary artery shunt are usually asymptomatic. Symptoms however can occur due to associated complications such as infection, hemorrhage, pulmonary hypertension and congestive heart failure [7].

Conventional angiography has been used for definitive diagnosis but it is relatively expensive, invasive and time-consuming. It is also associated with higher incidence of complications such as access site hematoma or embolic events [8]. Although the presence of systemic to pulmonary artery shunts has been reported in the past using conventional angiography, the recent advances in CT scan technology enable demonstration of these shunts and confirming the diagnosis. A 4D CTA (time-resolved CTA, fourth dimension being time) is now possible and can illustrate the temporal information of cardiovascular structures. In addition to generating cross-sectional images, 4D CTA, enables visualization of blood flow dynamics in the vessels. This has been useful in cardiovascular and cerebrovascular pathologies [9, 10]. 4D dynamic CTA, provide enough spatial resolution and temporal resolution to answer many clinically important questions that were previously addressed only by conventional angiography [11]. Generating cross-sectional images is an additional plus associated with these new techniques. MRA can also be used for the diagnosis of systemic to pulmonary artery shunt, but is more expensive, time consuming, prone to artifacts and technically inadequate in 25% of the times [12].

As demonstrated in case 1, this new technique directly demonstrates the flow from a systemic artery to pulmonary artery in systole, which manifests as a filling defect in a static CT image. Thus in cases with suspicion for this anomaly, the high temporal resolution of this technique enables confirmation of the abnormal communication (shunt) between systemic and pulmonary arteries.

Treatment & Prognosis:

Although the natural course of systemic to pulmonary artery shunt is not well known and definite indications for treatment have not been established [13], due to potential complications, some have recommended that intervention is necessary for asymptomatic patients because of future risk of endocarditis, angina pectoris, and congestive heart failure [14].

Selecting the appropriate therapeutic option is controversial. In the past, surgical intervention was the mainstay of treatment and the preferred procedure was lobectomy [13]. However, recently, embolization has become the preferred method since major surgery and general anesthesia are unnecessary and the loss of lung parenchyma is minimal [13]. Glue (N-butyl-2-cyanoacrylate) and gelfoam sponge particles can be used for those cases that shunts are very small. If the shunts are larger, coils can be used [13]. In addition, in cases with multiple shunts, embolization is more useful. Also embolization can be the primary choice for immediate control of hemoptysis [13].

Differential Diagnoses:

Acute and chronic PE are the most common causes of filling defect in the pulmonary artery. CT findings in acute PE include intraluminal filling defects, acute angles of the filling defect with the vessel wall, total cutoff of vascular enhancement and enlargement of an occluded vessel [15]. In chronic PE, the filling defect is usually in the periphery of the pulmonary artery lumen or its branches and may show calcification in the chronic thrombus. Other imaging findings in chronic PE include pulmonary arterial bands/pulmonary arterial webs. Signs related to pulmonary hypertension can also be seen which includes pulmonary artery dilation and/or calcification, narrowing of the peripheral pulmonary arteries, and right ventricular hypertrophy/enlargement [16].

There are multiple less common causes of filling defect in the pulmonary artery including systemic to pulmonary shunt, respiratory motion artifact, flow related artifact, partial volume artifact, presence of pulmonary artery catheter and perivascular edema. Primary pulmonary artery sarcoma or secondary tumor thrombus can also be confused with pulmonary embolism. Although there is overlap in the imaging findings of these entities, some clues can be helpful in narrowing the differential diagnosis (table 1).

When the pattern of filling defect in the pulmonary artery is unusual, meaning that the filling defect extends more proximally into the pulmonary artery and there is chronic inflammatory/fibrotic parenchymal lung disease in the background, one should raise the concern for systemic to pulmonary artery shunt as could be seen in our two cases. Dynamic CT angiography is a non-invasive and effective tool in diagnosing this condition. Its ability to provide detailed anatomy with multiplanar reconstruction could aid interventional radiologist in treatment planning.

TEACHING POINT

Filling defects in the pulmonary artery have many differential diagnoses other than pulmonary embolism. If the filling defect in the pulmonary artery has unusual appearance (extending to the more proximal left pulmonary artery or curvilinear appearance), and especially if this is happening in the setting of underlying chronic parenchymal lung disease, possibility of systemic to pulmonary artery shunt should be raised. 4D CTA (time-resolved CTA) can be used as a non-invasive and effective tool for reaching to this diagnosis. Its ability to provide detailed anatomy and multiplanar reconstruction could aid interventional radiologist in treatment planning.

REFERENCES

1. Wittram C, Maher MM, Yoo AJ, Kalra MK, Shepard JA, McLoud TC. CT angiography of pulmonary embolism: diagnostic criteria and causes of misdiagnosis. Radiographics. 2004 Sep-Oct; 24(5):1219-38. PMID: 15371604.

2. Anderson FA Jr, Wheeler B, Goldberg RJ, Hosmer DW, Patwardhan NA, Jovanovic B et al. A population-based perspective of the hospital incidence and case fatality rates of deep vein thrombosis and pulmonary embolism: the Worcester DVT study. Arch Intern Med 1991; 151:933-938. PMID: 2025141.

3. Giuntini C, Ricco GD, Marini C, Melillo E, Palla A. Pulmonary embolism: epidemiology. Chest 1995; 107(suppl): 3S-9S. PMID: 7813326.

4. Yoon W, Kim JK, Kim YH, Chung TW, Kang HK. Bronchial and nonbronchial systemic artery embolization for life-threatening hemoptysis: a comprehensive review. Radiographics 2002; 22:1395-409

5. Riehl G, Chaffanjon P, Frey G, Sessa C, Brichon PY. Postoperative systemic artery to pulmonary vessel fistula: analysis of three cases. Ann Thorac Surg. 2003 Dec; 76(6):1873-7. PMID: 14667603.

6. Jamshidi P, Ghaffari S, Mahmoodi K. Transpulmonary closing of left internal mammary artery to pulmonary artery fistula with polytetrafluoroethylene covered stent: a case report and review of literature.Cardiol J. 2009; 16(5):469-72. PMID: 19753528.

7. Saito T, Matsuda M, Yamaguchi T, Matsuzaki T, Nakagawa S. A case of a traumatic systemic-pulmonary arteriovenous fistula. Jpn Heart J 1975;16:196-203.

8. Bendszus M, Koltzenburg M, Burger R, Warmuth-Metz M, Hofmann E, Solymosi L. Silent embolism in diagnostic cerebral angiography and neurointerventional procedures: a prospective study. Lancet. 1999; 354:1594-1597. PMID: 10560674.

9. Klingebiel R, Siebert E, Diekmann S, Wiener E, Masuhr F, Wagner M, et al. 4-D imaging in cerebrovascular disorders by using 320-slice CT: feasibility and preliminary clinical experience. Acad Radiol 2009; 16:123-9. PMID: 19124096.

10. Siebert E, Bohner G, Dewey M, Masuhr F, Hoffmann KT, Mews J, et al. 320-slice CT neuroimaging: initial clinical experience and image quality evaluation. Br J Radiol 2009; 82:561-70. PMID: 19221186.

11. Willems PW, Taeshineetanakul P, Schenk B, Brouwer PA, Terbrugge KG, Krings T. The use of 4D-CTA in the diagnostic work-up of brain arteriovenous malformations. Neuroradiology. 2012; 54:123-31. PMID: 21465177.

12. Stein PD, Chenevert TL, Fowler SE, Goodman LR, Gottschalk A, Hales CA, Hull RD, Jablonski KA, Leeper KV Jr, Naidich DP, Sak DJ, Sostman HD, Tapson VF, Weg JG, Woodard PK; PIOPED III (Prospective Investigation of Pulmonary Embolism Diagnosis III) Investigators. Gadolinium-enhanced magnetic resonance angiography for pulmonary embolism: a multicenter prospective study (PIOPED III). Ann Intern Med. 2010 6;152:434-43

13. Lee JK1, Park JH, Kim J, Kim SJ, Lee AR, Lee CH, So YH. Embolization of multiple systemic artery to pulmonary artery fistula with recurrent hemoptysis. Tuberc Respir Dis. 2013;75:120-4.

14. Yamada Y, Imamura H, Amamoto Y, Ochi M, Nagano K, Ito M. Congenital internal mammary artery-to-pulmonary artery fistulas: a case report. Heart Vessels 1987;3:47-9.

15. Han D, Lee KS, Franquet T, Müller NL, Kim TS, Kim H, Kwon OJ, Byun HS. Thrombotic and nonthrombotic pulmonary arterial embolism: spectrum of imaging findings. Radiographics. 2003 ; 23:1521-39.

16. Castañer E, Gallardo X, Ballesteros E, Andreu M, Pallardó Y, Mata JM, Riera L. CT diagnosis of chronic pulmonary thromboembolism. Radiographics. 2009;29:31-50.

FIGURES

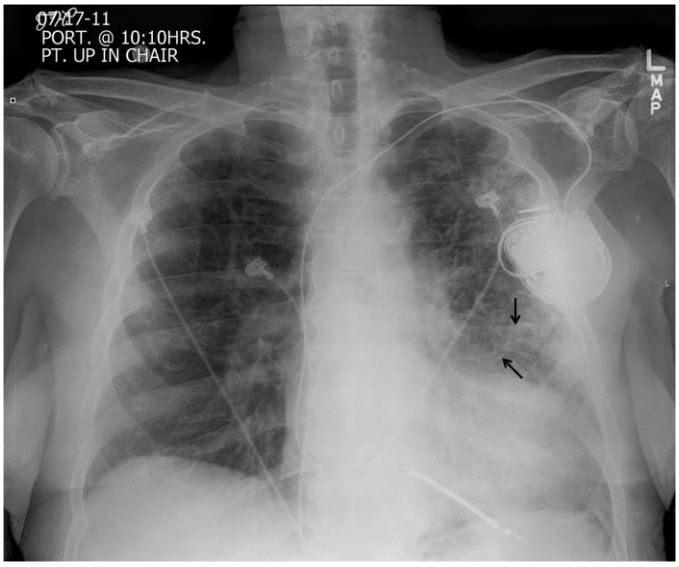

Figure 1: Portable frontal chest radiograph of an 81 year old male with systemic to pulmonary artery shunt and history of amiodarone lung toxicity shows increase in interstitial markings with distortion of the underlying parenchymal architecture, more in the left lung base (arrows), consistent with pulmonary fibrosis. There is also cardiomegaly and a defibrillator in place, in keeping with patient's known history of cardiac dysfunction.

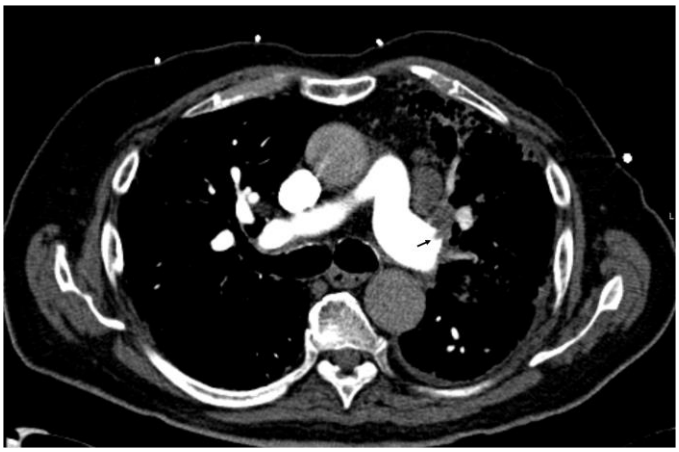

Figure 2: 81 year old male with systemic to pulmonary artery shunt. Axial contrast enhanced CT angiogram of the chest in the mediastinal window shows a non-opacified left upper lobe segmental pulmonary artery with associated small intraluminal filling defect is seen. The extension of the filling defect (arrow) into the more proximal left pulmonary artery is unusual for pulmonary embolism and therefore raised the possibility of an alternative diagnosis (shunt).
The CT is acquired on a 64-slice Siemens Somatom Force scanner using pulmonary angiogram protocol (contrast enhanced acquisition at peak pulmonary opacification with 1.25 mm overlapping collimation using 90 cc of Optiray 350 contrast at 120 kVp with tube current modulation and adaptive statistical iterative reconstruction.

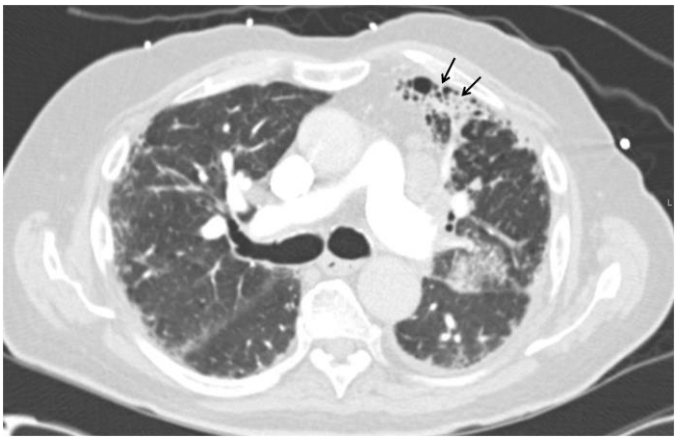

Figure 3: 81 year old male with systemic to pulmonary artery shunt. Axial contrast enhanced CT angiogram of the chest in the lung window shows coarse interlobular septal thickening and reticulation (arrows) in the lungs, more on the left, consistent with chronic changes of amiodarone induced lung toxicity and fibrosis.
The CT is acquired on a 64-slice Siemens Somatom Force scanner using pulmonary angiogram protocol (contrast enhanced acquisition at peak pulmonary opacification with 1.25 mm overlapping collimation using 90 cc of Optiray 350 contrast at 120 kVp with tube current modulation and adaptive statistical iterative reconstruction.

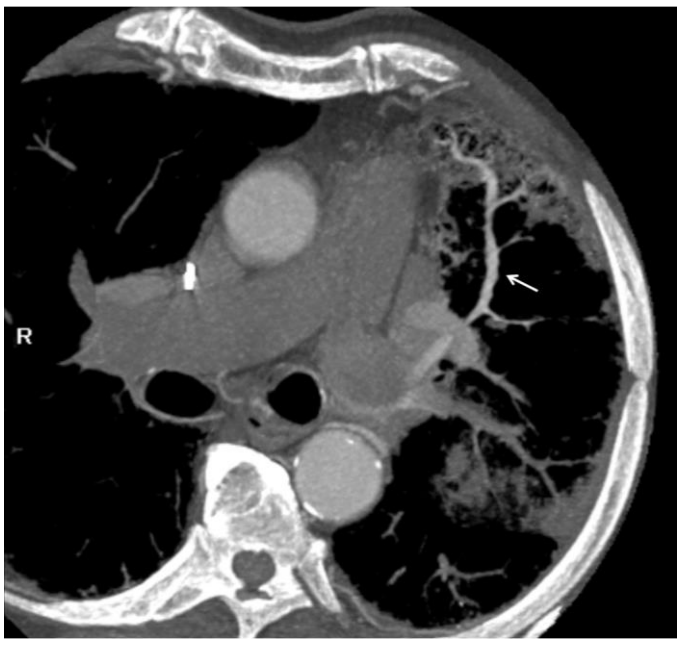

Figure 4: 81 year old male with systemic to pulmonary artery shunt. Axial contrast enhanced CT angiogram of the chest using maximum-intensity projection (MIP) provides a clear depiction (arrow) of abnormal communication between systemic and pulmonary artery.
The dynamic CTA is acquired on a 64-slice Siemens Somatom Force scanner, using a spiral 4D mode at 100 kVp, 100 mAs, with 60 mL contrast.

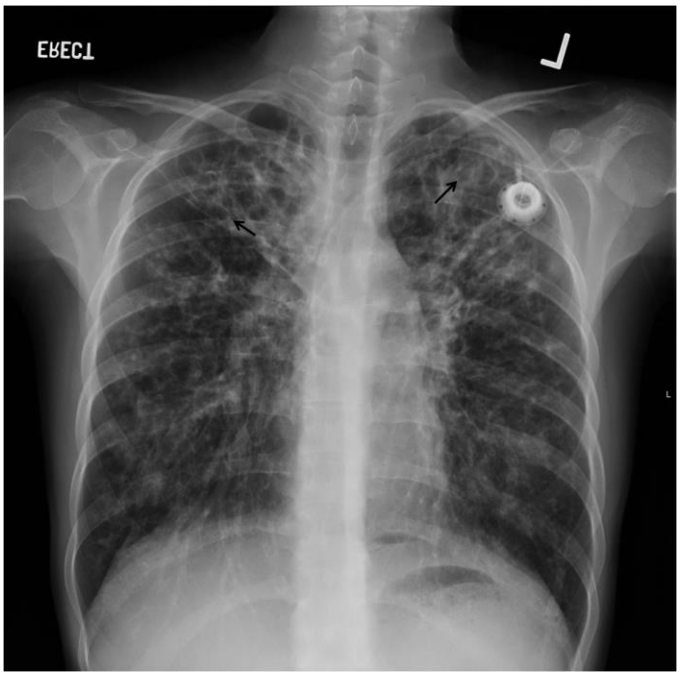

Figure 5: Posteroanterior frontal chest radiograph of a 29 year old female with systemic to pulmonary artery shunt in the setting of cystic fibrosis, shows hyperinflation and upper lobe predominant bronchiectasis (arrows) in keeping with patient's underlying cystic fibrosis.

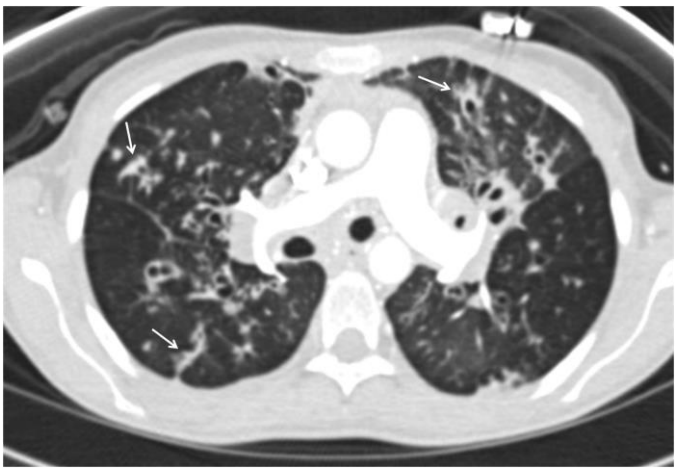

Figure 7: 29 year old female with systemic to pulmonary artery shunt in the setting of cystic fibrosis. Axial contrast enhanced CT angiogram of the chest in the lung window shows bronchial wall and peribronchial interstitial thickening with upper lobe predominant bronchiectasis (arrows) typical of Cystic Fibrosis.
CT is acquired on a 64-slice Siemens Somatom Force scanner. The CT is acquired using pulmonary angiogram protocol (contrast enhanced acquisition at peak pulmonary opacification with 1.25 mm overlapping collimation using 90 cc of Optiray 350 contrast at 100 kVp with tube current modulation and adaptive statistical iterative reconstruction.

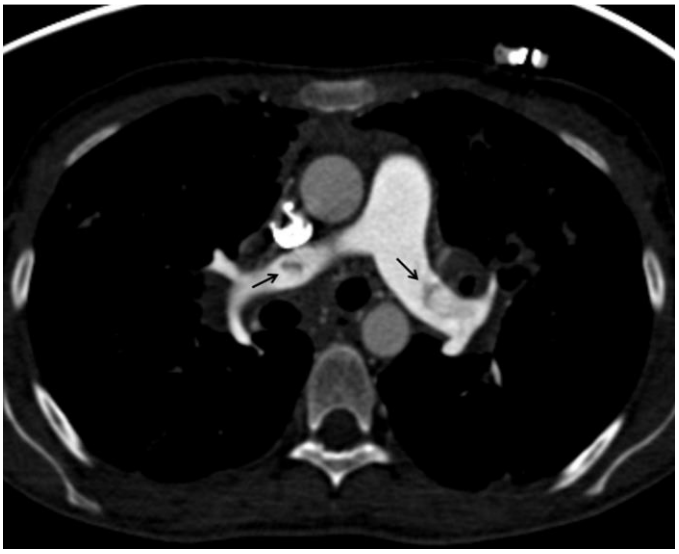

Figure 6 (left): 29 year old female with systemic to pulmonary artery shunt in the setting of cystic fibrosis. Axial contrast enhanced CT angiogram of the chest in the mediastinal window shows amourphous and curvilinear areas of non-opacification and filling defects (arrows) in the bilateral pulmonary arteries. As in the first case, this is unusual for pulmonary embolism which typically presents as a tubular filling defect starting from the more proximal portion of the pulmonary artery and extends to its more distal branches. Therefore possibility of an alternative diagnosis (shunt) was raised.
CT is acquired on a 64-slice Siemens Somatom Force scanner. The CT is acquired using pulmonary angiogram protocol (contrast enhanced acquisition at peak pulmonary opacification with 1.25 mm overlapping collimation using 90 cc of Optiray 350 contrast at 100 kVp with tube current modulation and adaptive statistical iterative reconstruction.

Differential Diagnosis	CT Angiography	4D CTA	Conventional angiography or other imaging modalities
Acute pulmonary embolism	Filling defect in the center of the pulmonary artery lumen or its branches.	The same findings as CTA. No abnormal communication between the systemic and pulmonary artery.	Conventional angiography is gold standard and shows filling defect in the center of the pulmonary artery or its branches.
Chronic pulmonary embolism	Filling defect in the periphery of the pulmonary artery lumen or its branches. Can show calcification in the chronic thrombus. May show pulmonary arterial bands/pulmonary arterial webs. Signs related to pulmonary hypertension can be seen (such as pulmonary artery dilation and/or calcification, narrowing of the peripheral pulmonary arteries, and right ventricular hypertrophy/enlargement)	The same findings as CTA. No abnormal communication between the systemic or pulmonary artery.	Conventional angiography shows filling defect in the periphery of the pulmonary artery or its branches.
Systemic to pulmonary artery shunt	Filling defect in the pulmonary artery with extension of the defect to the more proximal portion of the pulmonary artery.	Abnormal communication of the pulmonary artery to systemic artery or their branches.	Conventional angiography shows the abnormal communication of the pulmonary artery to systemic artery.
Respiratory motion artifact	Can mimic a filling defect, but respiratory motion artifact should be visible and provide clue to the diagnosis.	N/A	N/A
Flow related artifact (caused by inadequate mixing of contrast in the bloodstream).	Can mimic a filling defect but has a swirling appearance which can be helpful in differentiating it from a real defect.	If done, shows no abnormal communication between the systemic or pulmonary artery.	If done, conventional angiography shows no filling defect in the pulmonary artery. Also no abnormal communication between the systemic or pulmonary artery.
Partial volume artifact	Can mimic a filling defect on thicker cuts. Thinner cuts show no such defect.	N/A	N/A
Presence of pulmonary artery catheter	Can produce a filling defect. Appropriate windowing should show the catheter as the cause of defect.	N/A	Catheter can be seen on plain radiograph
Perivascular edema	Can mimic a filling defect on thicker cuts. Thinner cuts show no such defect. Circumferentially surrounding the vessel. Also is generalized and seen in both lungs.	N/A	N/A
Primary pulmonary artery sarcoma or secondary tumor thrombus	Large tumor emboli in the main, lobar, and segmental pulmonary arteries cause filling defects that mimic acute pulmonary thromboembolism. Small tumor emboli that affect subsegmental arteries produce multifocal dilatation or beading of vessels. Reconstruction in coronal and sagittal formats will be useful in showing the tumoral nature of the filling defect. The filling defect may show enhancement. In the case of primary pulmonary artery sarcoma, the border of the tumor may be irregular. There may be also extension of the tumor beyond the vascular margin. May show enhancement.	If done, shows no abnormal communication between the systemic or pulmonary artery.	If done, conventional angiography shows no abnormal communication between the systemic or pulmonary artery.

Table 1: Differential diagnosis of filling defect in the pulmonary artery.

Etiology	Abnormal communication of the systemic to pulmonary artery.
Incidence	Much less common than pulmonary embolism which is the most common cause of filling defect in the pulmonary artery and its branches.
Gender Ratio	No specific predilection.
Age predilection	Probably more common in older patients, as it is seen more in patients with underlying chronic inflammatory lung disease.
Risk factors	Chronic inflammatory diseases of the lung such as tuberculosis and idiopathic pulmonary fibrosis. Congenital and acquired etiologies such as surgery (Coronary artery bypass graft surgery), trauma and neoplasia.
Treatment	Surgical intervention such as lobectomy. Endovascular embolization.
Prognosis	Patients are usually asymptomatic, but even asymptomatic patients probably should be treated because of future risk of endocarditis, angina pectoris, and congestive heart failure. Surgical intervention such as lobectomy and embolization are definitive treatment.
Findings on Imaging	CTA: Unusual pattern of filling defect in the pulmonary artery with extension of the filling defect to the more proximal portion of the pulmonary artery. There is also a background of underlying chronic lung disease seen on lung window. Conventional angiography: Abnormal communication of the pulmonary artery to systemic artery. 4D CTA: Temporal resolution of this technique helps to show the abnormal communication of the pulmonary artery to systemic artery.

Table 2: Characteristics of systemic to pulmonary artery shunt.

ABBREVIATIONS

4D CTA: 4 dimensional computed tomography angiography
CABG: Coronary artery bypass graft
CF: Cystic Fibrosis
CT: computed tomography
MIP: Maximum-intensity projection
MRA: Magnetic Resonance Angiography
PE: Pulmonary Embolism

KEYWORDS

Shunt; pulmonary embolism; chronic lung disease; dynamic CT; angiography

Online access

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/2480

Peer discussion

Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad

www.EduRad.org