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Case Report

Differentiating systemic artery-to-pulmonary artery fistula from pulmonary arteriovenous malformation: A case report ☆

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ABSTRACT

This report describes a rare case of a systemic artery-to-pulmonary artery fistula initially misdiagnosed as a pulmonary arteriovenous malformation in a 64-year-old male with severe emphysema. While a suspected pulmonary arteriovenous malformation was identified in the left lung on plain computed tomography, contrast-enhanced imaging revealed an aneurysmal nodule connected to the left inferior phrenic and internal mammary arteries, suggesting a systemic artery-to-pulmonary artery fistula. Selective angiography confirmed the diagnosis. The absence of pulmonary vein dilation on computed tomography is the key to differentiating a systemic artery-to-pulmonary artery fistula from a pulmonary arteriovenous malformation. Additionally, using contrast-enhanced computed tomography with both pulmonary artery and late phases helps prevent misdiagnosis of pulmonary embolism and may raise the suspicion of a systemic artery-to-pulmonary artery fistula.

Introduction

Systemic artery-to-pulmonary artery fistula (SA-PAF) is a rare condition that can result in severe complications including hemoptysis and pulmonary hypertension [1]. SA-PAF may be congenital or acquired, with acquired cases often arising from chest trauma, surgery, malignancy, or infection [2]. It is frequently misdiagnosed as a pulmonary arteriovenous malformation (PAVM) or pulmonary embolism (PE) on computed

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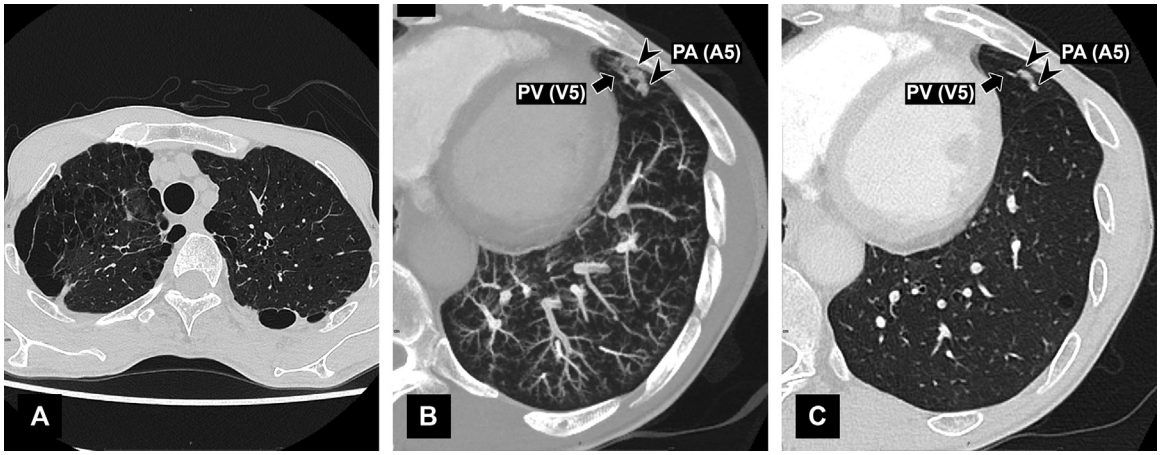

Fig. 1 – Plain computed tomography (CT) findings. Plain CT imaging reveals severe emphysema (A). The maximum intensity projection highlights a dilated and tortuous pulmonary artery (PA) A5 segment (arrowheads) in the lingula of the left lung leading to a bulbous, aneurysmal nodule with a well-defined border, adjacent to a nearby pulmonary vein (PV) V5 segment (arrow) (B). It is uncommon for pulmonary arteriovenous malformations to present with a dilated PA (arrowheads) while the PV (arrow) remains unaffected (C).

tomography (CT), leading to inappropriate treatment [3–5]. This report describes a case initially diagnosed as PAVM but was eventually suspected to be SA-PAF based on CT findings and subsequently confirmed by angiography.

Case report

A 64-year-old man with a suspected PAVM in the lingula on plain chest CT was referred for endovascular treatment. Although there was no family history or comorbidities, the patient had a smoking history of 30 pack-years and was a current smoker. The SpO2 level was 98% on room air, which was within the normal range. He was asymptomatic, and physical examination revealed no abnormalities, including signs of nail deformities. He had severe emphysema due to long-term smoking (Fig. 1A) but had no history of trauma or surgery. CT scan (Aquilion Prime SP TSX-303B, Canon Medical Systems Corporation, Tochigi, Japan) were set to 120 kVp, 160 reference mAs tube current, 80 × 6.5 mm collimation, 0.813 pitch, and standard filtered back projection with a slice thickness of 3 mm and 1 mm. CT scan revealed a dilated and tortuous pulmonary artery (A5 segment) with a nearby pulmonary vein in the lingula of the left lung. These vessels were connected through a bulbous aneurysmal formation, creating a rounded, mass-like nodule with well-defined borders (Fig. 1B). The peripheral pulmonary vein, which was suspected to serve as the drainage vein in the PAVM, was not dilated and appeared atypical (Fig. 1C).

Contrast-enhanced CT was performed to further evaluate the feeding artery and drainage vein. Similar to the findings on plain CT, an aneurysmal nodule with a similar density as and contiguous with the dilated and tortuous pulmonary artery was identified in the lingula, suggesting the presence of a vascular malformation (Fig. 2A). Additionally, the nodule was suspected to be contiguous with the dilated and meandering left inferior phrenic artery (lt-IPA) and left internal mammary artery (lt-IMA) (Fig. 2B, C). Another small aneurysmal nodule, suspected to be continuous with the lt-IMA, was likewise observed in the lingula (Fig. 2D). SA-PAF was strongly suspected, with the lt-IPA and lt-IMA presumed to be the feeding arteries and pulmonary artery segments A5 and A4 likely serving as the drainage pathways. During the pulmonary artery phase of the CT obtained 15 s after the start of contrast injection, a contrast defect was observed on the central side of the pulmonary artery A5 segment, whereas the late phase obtained 40 s after the start of contrast injection showed a uniform contrast enhancement (Fig. 2E, F). Retrograde blood flow in the pulmonary artery A5 segment, likely caused by systemic arterial supply, was suspected and considered, supporting the diagnosis of SA-PAF. The 3-dimensional reconstruction images using Synapse Vincent (v6.8, Fujifilm; Tokyo, Japan) clearly illustrated the relationships between the vessels (Fig. 3). Selective angiography was subsequently performed to confirm diagnosis.

Selective angiography of the lt-IPA revealed that it ascended in a dilated and tortuous manner, forming a fistula above the diaphragm, followed by retrograde depiction of the pulmonary artery A5 segment. The peripheral branches of the lt-IMA also exhibited slight retrograde flow (Fig. 4A). Selective angiography of the lt-IMA resulted in retrograde appearance of the pulmonary artery A5 segment, which was contiguous with the fistula identified in the lt-IPA. Additionally, the other small fistula that had previously been identified on CT was observed, along with a retrograde depiction of the pulmonary artery A4 segment (Fig. 4B). Pulmonary arteriography did not reveal a PAVM and failed to depict the fistula identified on selective angiography of the lt-IPA and lt-IMA (Fig. 4C, D).

Selective angiography confirmed the diagnosis of SA-PAF. Pulmonary hypertension was not present and no hemoptysis occurred. The patient remains to be on careful follow-up.

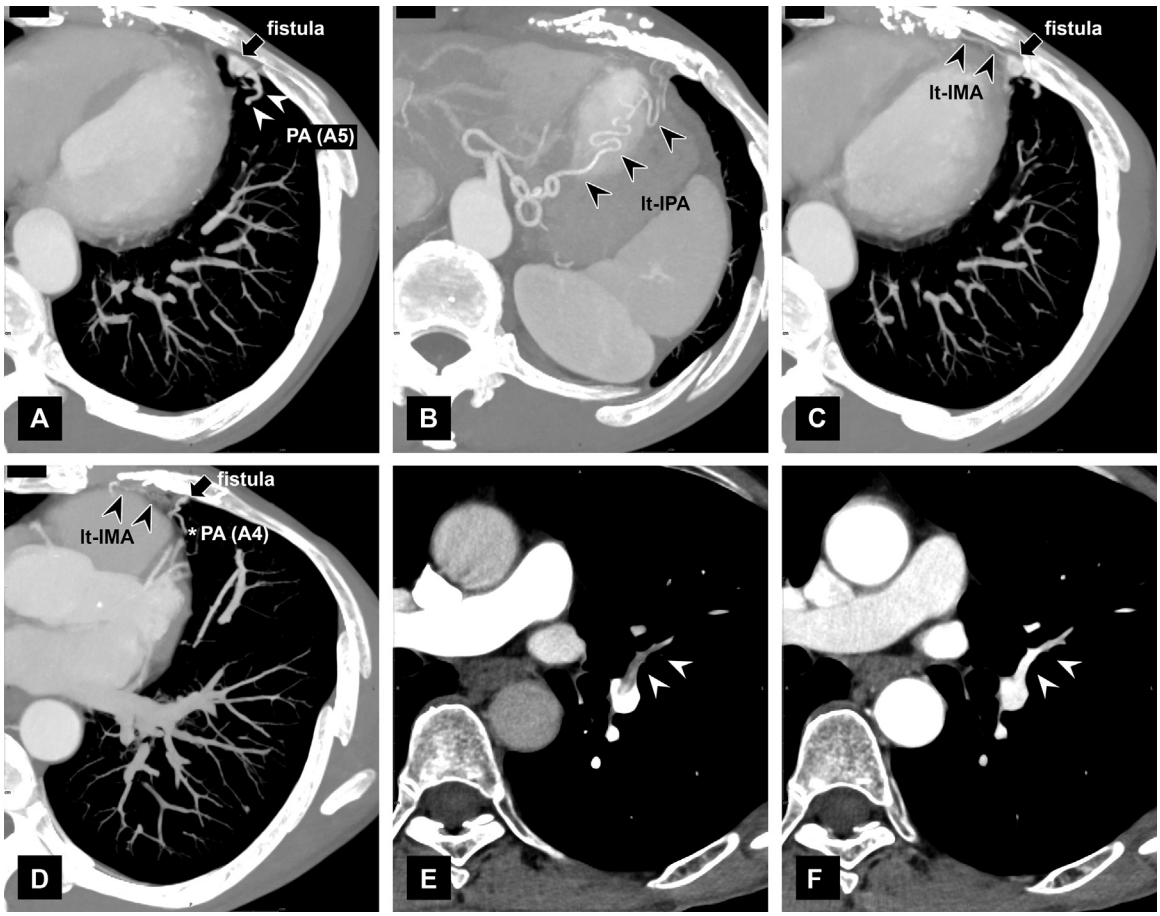

Fig. 2 – Contrast-enhanced computed tomography (CT) findings. Contrast-enhanced CT maximum intensity projection highlights a dilated and tortuous pulmonary artery (PA) A5 segment (arrowheads) forming a fistula (arrow) with a bulbous, aneurysmal dilation in the lingula of the left lung (A). The ascending left inferior phrenic artery (lt-IPA, arrowheads) extends and meanders toward the fistula (B). The left internal mammary artery (lt-IMA, arrowheads) runs near the fistula (arrow) and is suspected to be continuous with it (C). The lingula of the left lung shows another small fistula (arrow) contiguous with the PA A4 segment (asterisk), with the left internal mammary artery (lt-IMA, arrowheads) running nearby (D). In the pulmonary artery phase, contrast loss is observed on the central side of the PA A5 segment (E), while the late phase shows uniform contrast enhancement (F).

He began the process of quitting smoking because the inflammation caused by long-term smoking might have been a contributing factor.

Discussion

SA-PAF is often misdiagnosed as a PAVM or PE on CT, which can result in inappropriate treatment [3–5]. We will discuss the key imaging findings that differentiate these conditions.

SA-PAF is frequently misdiagnosed as a PAVM [3,4], mainly because the bulbous and aneurysmal morphology characteristic of PAVMs is also observed in SA-PAF, where the pulmonary artery serves as the drainage channel. Treatment options for SA-PAF include transcatheter embolization and surgical resection, although the indications for these procedures remain controversial [6,7]. Angiography, although slightly invasive, is preferred for definitive diagnosis.

In this case, a PAVM was suspected based on plain CT images; however, the absence of dilation in the pulmonary vein, presumed to be the draining vein (Fig. 1C), was atypical. In PAVMs, the draining vein is typically 1–2 mm larger than the feeding artery. This dilation is caused by the pressure gradient created by the direct anastomosis between the pulmonary artery and vein, bypassing the capillaries [8,9]. Plain CT is sufficient to assess dilation of the pulmonary vein, which is presumed to be the draining vein. We propose that the presence or absence of pulmonary vein dilation on plain CT is the key finding to differentiating SA-PAF from PAVM.

Similarly, SA-PAF is commonly misdiagnosed as PE [3–5]. This misdiagnosis occurs because of retrograde flow in the pulmonary artery caused by the pressure gradient difference between the systemic and pulmonary arteries. A similar reversal and reduction in blood flow can be induced by long-term inflammation during bronchial artery-pulmonary artery anastomosis, which is also observed in normal lungs [3]. During the pulmonary artery phase of contrast-enhanced CT, the pul-

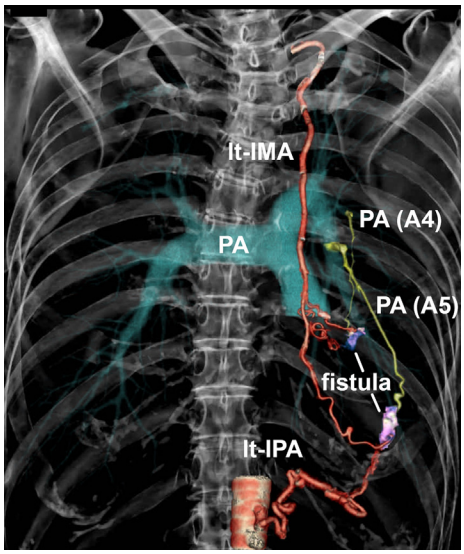

Fig. 3 – Three-dimensional (3D) reconstruction. The 3D reconstruction images using Synapse Vincent (v6.8, Fujifilm; Tokyo, Japan) clearly demonstrate the formation of fistulae between the left inferior phrenic artery (lt-IPA) and the left internal mammary artery (lt-IMA), as well as the connections between the pulmonary artery (PA) segments A5 and A4.

monary artery, which serves as the drainage route for SA-PAFs, is often misidentified as a pulmonary embolus. This misinterpretation occurs because the target pulmonary artery receives retrograde blood flow from the systemic arteries, and the contrast agent administered through the peripheral vein has not yet arrived in the pulmonary artery. However, in the delayed phase, when the contrast medium reaches the systemic artery, uniform contrast enhancement is observed, which is clearly distinguishable from PE (Fig. 2E, F). Therefore, imaging the delayed phase, in addition to the pulmonary artery phase, during contrast-enhanced CT is recommended. This characteristic finding, which reflects retrograde blood flow in the pulmonary artery, is instrumental in the diagnosis of SA-PAF.

Reports on SA-PAF with multiple feeders are scarce [10]. Previous studies have noted the involvement of various systemic arteries, including the bronchial [11], inferior phrenic [3,6], internal mammary [6], and coronary arteries [12]. In the present case, 2 feeding arteries, the lt-IPA and lt-IMA, were identified. The presence of multiple feeders and drainage veins complicates hemodynamics. Three-dimensional reconstruction images are particularly useful in identifying abnormal vessels [7]. In this case, these images were valuable for identifying the feeding artery and clarifying its relationship with other vessels prior to angiography (Fig. 3).

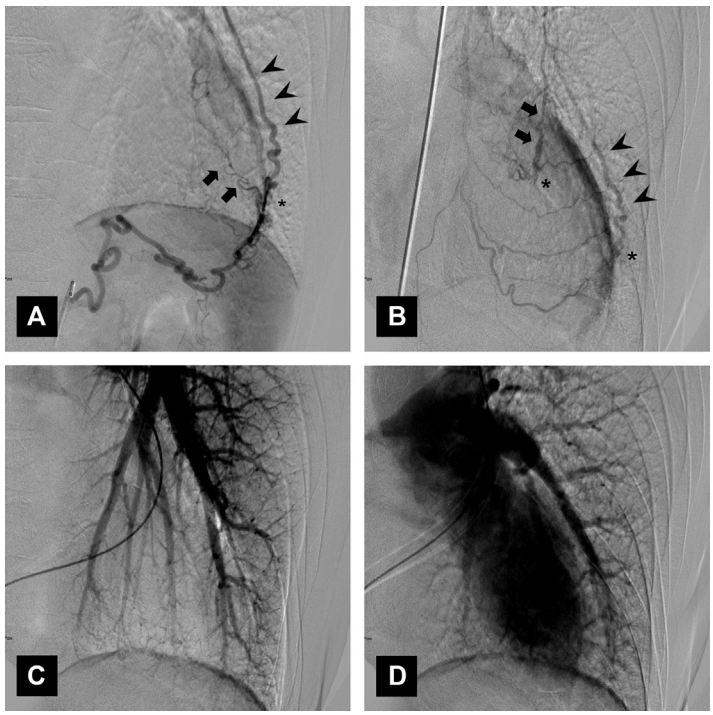

Fig. 4 – Selective angiography findings. Selective angiography of the left inferior phrenic artery shows retrograde visualization of the pulmonary artery A5 segment (arrowheads) and the left internal mammary artery (arrows) after forming a fistula (asterisk) (A). Selective angiography of the left internal mammary artery shows retrograde visualization of the pulmonary artery A5 (arrowheads) and A4 (arrows) segments after forming fistulae (asterisks) (B). Angiography of the main left pulmonary artery does not reveal any arteriovenous malformations or fistulae during the pulmonary artery (C) and venous phases (D).

Conclusion

SA-PAF is frequently misdiagnosed as a PAVM; however, the presence or absence of pulmonary vein dilation on CT can be a key distinguishing feature. Contrast-enhanced CT can reduce the risk of misdiagnosing SA-PAF as PE by imaging both the pulmonary artery and the late phases. Three-dimensional reconstructed images are valuable for identifying the nutrient arteries and their relationships with other vessels before angiography.

Patient consent

Informed written consent was obtained from the patient for the publication of this report and any accompanying images.

REFERENCES

[1] Woo JH, Cha YK, Lee SY, Kim JH, Kim JG, Son Y, et al. CT evaluation of systemic artery to pulmonary artery fistula: an underdiagnosed disease in patients with hemoptysis. J Thorac Dis 2023;15:5952–60. doi:10.21037/jtd-23-861.

[2] Livingston D, Grove M, Grage R, McKinney JM. Systemic artery-to-pulmonary artery fistula mimics pulmonary embolus. J Clin Imaging Sci 2019;9:41. doi:10.25259/JCIS_54_2019.

[3] Alsafi A, Shovlin CL, Jackson JE. Acquired transpleural systemic artery-to-pulmonary artery communication mimicking a pulmonary arteriovenous malformation and causing a false-positive diagnosis of a pulmonary embolus. J Vasc Interv Radiol 2018;29:1313–15. doi:10.1016/j.jvir.2017.12.016.

[4] Castañer E, Gallardo X, Rimola J, Pallardó Y, Mata JM, Perendreu J, et al. Congenital and acquired pulmonary artery anomalies in the adult: radiologic overview. RadioGraphics 2006;26:349–71. doi:10.1148/rg.262055092.

[5] Ansari-Gilani K, Gilkeson RC, Hsiao EM, Rajiah P. Unusual Pulmonary arterial Filling Defect caused by Systemic to Pulmonary Shunt in the Setting of Chronic Lung Disease Demonstrated by Dynamic 4D CTA. J Radiol Case Rep 2015;9:17–23. doi:10.3941/jrcr.v9i11.2480.

[6] Geyik S, Yavuz K, Keller FS. Unusual systemic artery to pulmonary artery malformation without evidence of systemic disease, trauma or surgery. Cardiovasc Intervent Radiol 2006;29:897–901. doi:10.1007/s00270-004-0289-9.

[7] Kuo CL, Lin KH, Ko KH, Huang TW. The aberrant systemic-pulmonary artery communication: three-dimensional image simulation. J Cardiothorac Surg 2019;14:35. doi:10.1186/s13019-019-0855-5.

[8] Saboo SS, Chamarthy M, Bhalla S, Park H, Sutphin P, Kay F, et al. Pulmonary arteriovenous malformations: diagnosis. Cardiovasc Diagn Ther 2018;8:325–37. doi:10.21037/cdt.2018.06.01.

[9] Tellapuri S, Park HS, Kalva SP. Pulmonary arteriovenous malformations. Int J Cardiovasc Imaging 2019;35:1421–8. doi:10.1007/s10554-018-1479-x.

[10] Lee JK, Park JH, Kim J, Kim SJ, Lee AR, Lee CH, et al. Embolization of multiple systemic artery to pulmonary artery fistula with recurrent hemoptysis. Tuberc Respir Dis (Seoul) 2013;75:120–4. doi:10.4046/trd.2013.75.3.120.

[11] Yon JR, Ravenel JG. Congenital bronchial artery-pulmonary artery fistula in an adult. J Comput Assist Tomogr 2010;34:418–20. doi:10.1097/RCT.0b013e3181d1e96e.

[12] Ohkura K, Yamashita K, Terada H, Washiyama N, Akuzawa S. Congenital systemic and coronary-to-pulmonary artery fistulas. Ann Thorac Cardiovasc Surg 2010;16:203–6.