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A Rare Congenital Multiple Systemic Artery-to-Pulmonary Artery Shunt Mimicking Pulmonary Arteriovenous Malformation

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1. Abstract

1.1. Background

Pulmonary arteriovenous malformations (PAVMs) are typically diagnosed using contrast-enhanced computed tomography (CT). However, certain vascular anomalies can mimic PAVMs. Systemic artery-to-pulmonary artery (SA–PA) shunts are rare mimickers, and congenital multiple SA–PA (mSA–PA) shunts resembling PAVM have not been previously reported.

1.2. Case Presentation

A 41-year-old man underwent chest CT during evaluation for tonsillitis, which incidentally revealed a vascular anomaly in the left lower lobe. Contrast-enhanced CT and digital subtraction angiography (DSA) demonstrated tortuous vessels supplied by two systemic arteries: a branch of the descending thoracic aorta and the inferior phrenic artery. These vessels drained into the posterior basal segmental pulmonary artery (A10), with retrograde flow into the left inferior pulmonary artery. Balloon-occluded A10 angiography showed antegrade pulmonary arterial flow with relatively early pulmonary venous visualization, raising suspicion for PAVM; however, no discrete arteriovenous nidus was identified. Intraoperatively, aberrant systemic arteries entered the lung via the diaphragm and mediastinum. Wedge resection was performed. Histopathological examination revealed dilated pulmonary arteries without evidence of an arteriovenous nidus.

1.3. Conclusions

We report a rare congenital mSA–PA shunt that closely mimicked PAVM on imaging. Unlike PAVMs, which involve direct arteriovenous connections, SA–PA shunts are characterized by systemic arterial inflow into the pulmonary arterial system without a nidus. Careful angiographic evaluation is essential for accurate diagnosis and appropriate management.

2. Case Presentation

A 41-year-old man underwent chest computed tomography (CT) as part of a workup for tonsillitis, which incidentally revealed a suspected pulmonary arteriovenous malformation (PAVM) in the left lower lobe. He had no history of thoracic trauma, surgery, or hereditary vascular disorders.

2.1. Image Findings

Contrast-enhanced CT showed dilated, tortuous vessels in the basal segment of the left lower lobe (Figure1). The vascular complex was supplied by two systemic arteries: one from the descending thoracic aorta running along the esophagus, and another from the left inferior phrenic artery. Three-dimensional CT angiography revealed direct communication between these arteries and the posterior basal segmental pulmonary artery (A10) (Figure 2). Digital subtraction angiography (DSA) confirmed retrograde flow from both systemic arteries into the A10, with subsequent drainage into the left inferior pulmonary vein (Figure 3). The higher pressure of the systemic arteries resulted in poor enhancement of A10 on pulmonary artery angiography (Figure 4-1). However, balloon-occluded A10 angiography demonstrated antegrade visualization of the segmental pulmonary artery, followed by relatively early visualization of the pulmonary vein, without identification of a definite arteriovenous nidus (Figure 4-2). Based on these findings, a systemic artery–to–pulmonary artery shunt with a possible complex-type PAVM was initially

suspected at the time of the DSA. The lesion did not meet the criteria for anomalous systemic arterial supply to the basal segment (ABLL), as the pulmonary artery was intact.

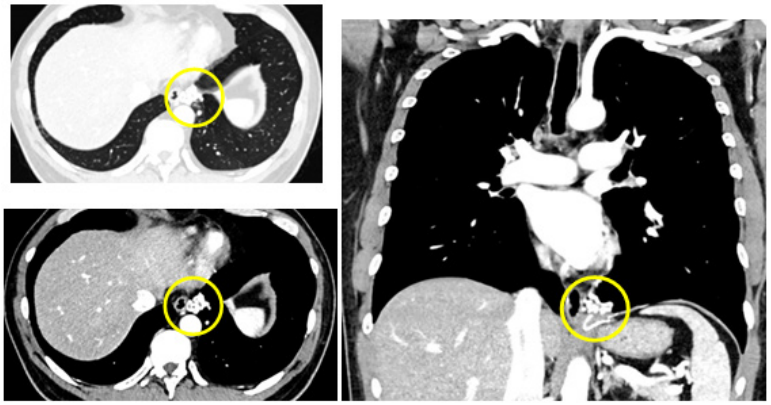

Figure 1: Contrast-enhanced CT shows dilated, tortuous vessels in the basal segment of the left lower lobe (yellow circle).

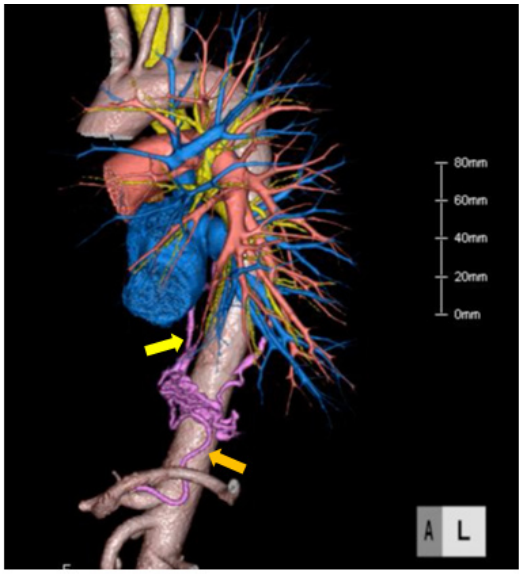

Figure 2: Three-dimensional CT angiography identifies two systemic feeding arteries arising from the descending thoracic aorta (yellow arrow) and the left inferior phrenic artery (orange arrow).

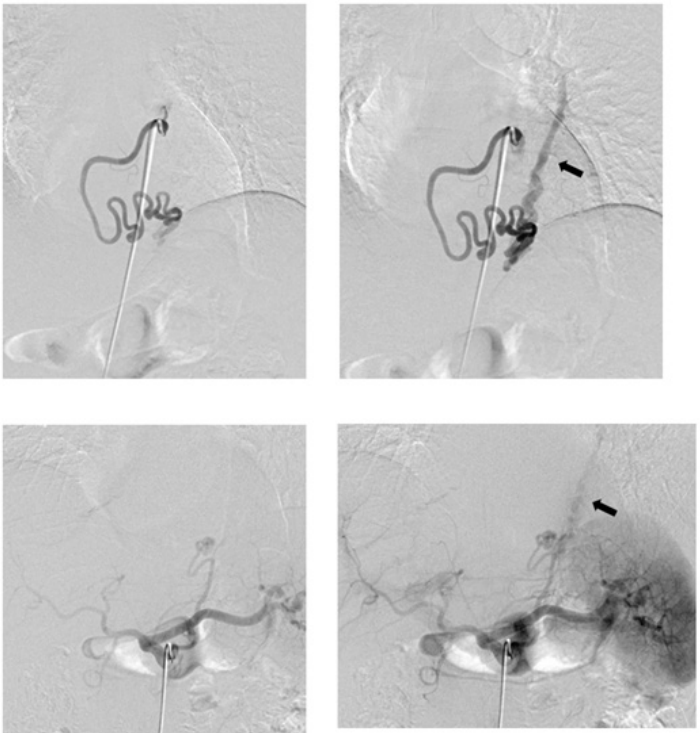

Figure 3: Digital subtraction angiography confirmed retrograde flow from descending aortic and left inferior phrenic branches into A10 (arrow).

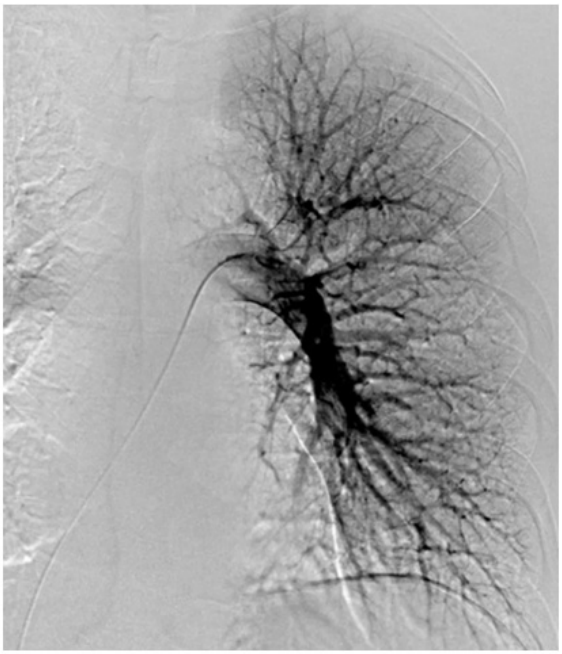

Figure 4-1: Right pulmonary artery angiography demonstrated no opacification of A10 (Figure 4-1), whereas selective A10 angiography revealed antegrade flow with drainage into the pulmonary vein.

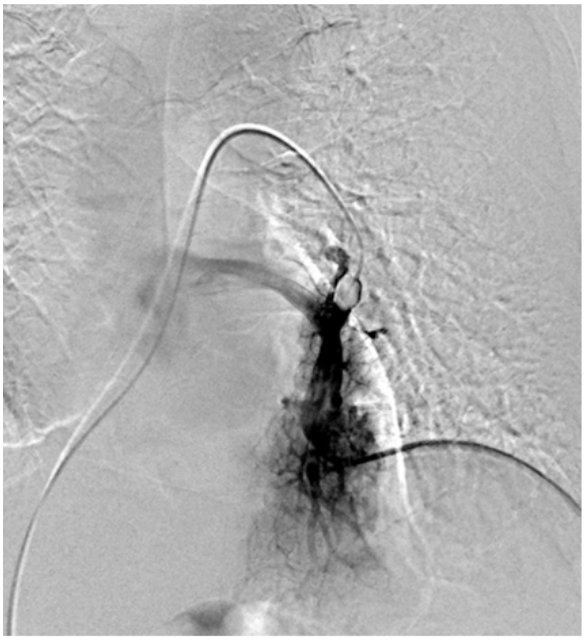

Figure 4-2: Mimicking the angiographic appearance of a pulmonary arteriovenous malformation (PAVM).

2.2. Operating Findings

Video-assisted thoracoscopic wedge resection was performed. Intraoperatively, tortuous vessels were found extending from the diaphragm and mediastinum into the lung. The aberrant arteries from the inferior phrenic artery and the descending thoracic aorta were carefully ligated and divided (Figure 5). The vascular anomaly was completely mobilized, and lung wedge resection was completed using a stapler device.

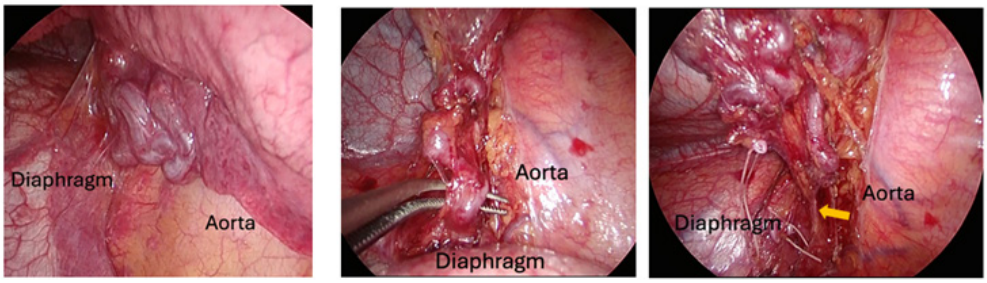

Figure 5: Tortuous vessels extended from diaphragm; inferior phrenic (orange arrow) and descending aorta (yellow arrow) ligated.

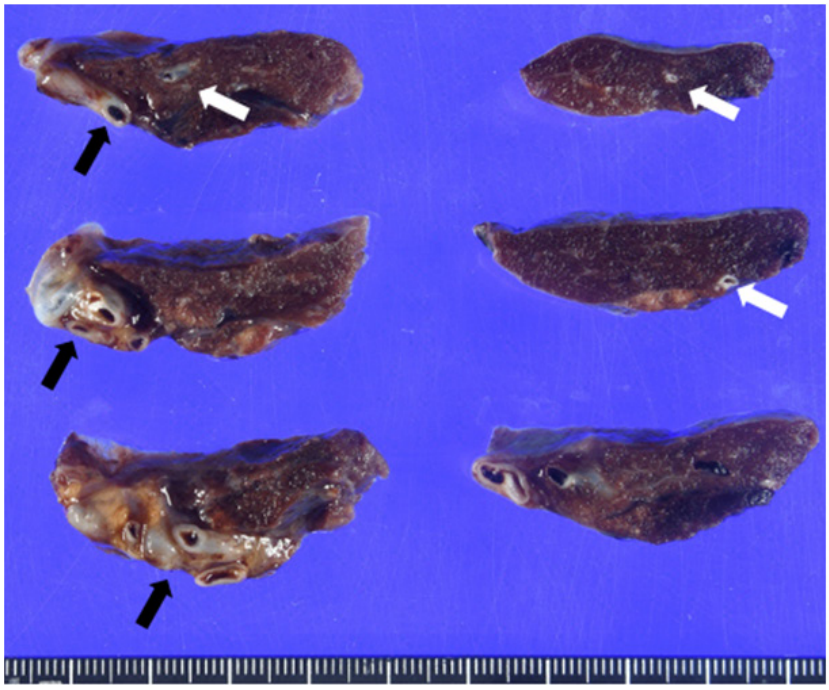

Figure 6: Tortuous vessels crowded on the surface (black arrows) and dilated A10 branches (white arrows).

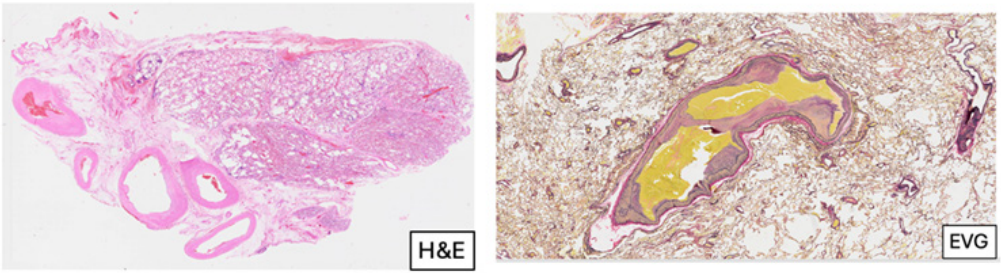

Figure 7: H&E and EVG staining showed tortuous pleural vessels and dilated A10 without arteriovenous nidus formation.

2.3. Pathology Findings

The specimen demonstrated clusters of blood vessels on the lung surface and the cut surface revieled tortuous vessels near the pleura along with dilated A10 branches in the lung parenchyma. Histopathological evaluation with hematoxylin and eosin (H&E) and Elastica van Gieson (EVG) staining demonstrated tortuous vessels densely distributed in the pleura and enlarged pulmonary arteries adjacent to a bronchus, without evidence of an arteriovenous nidus. The dilation of A10 was likely secondary to high-pressure inflow from the systemic arteries. These findings were consistent with a systemic artery–to–pulmonary artery shunt.

3. Discussion

Systemic artery–to–pulmonary artery (SA-PA) shunt is rare vascular anomalies characterized by direct communication between systemic arteries and pulmonary arteries, bypassing the normal pulmonary capillary network. These fistulas may be acquired or congenital. Acquired SA-PA shunt typically arises secondary to chronic inflammation, trauma, surgery, or infection, with neovascularization reported in conditions such as tuberculosis or pleuritis [1–3]. In contrast, congenital SA-PA shunt is uncommon and is often detected incidentally in patients without prior thoracic pathology. In the present case, the absence of thoracic trauma, surgery, or infection, together with dual systemic arterial inflow, strongly suggests a congenital origin. To our knowledge, this is the first reported case of a congenital multiple systemic artery–to–pulmonary artery (mSA-PA) shunt with dual systemic arterial supply from both the descending thoracic aorta and the inferior phrenic artery, confirmed histopathologically.

The lesion was initially suspected to represent an mSA-PA shunt based on contrast-enhanced CT findings. However, balloon-occluded A10 angiography demonstrated antegrade opacification of the segmental pulmonary artery, followed by relatively early filling of the pulmonary vein, without identification of a typical AVM nidus. These findings suggested the possibility of diffuse pulmonary micro-shunting, resembling Yakes type IV AVM [4]. Therefore, the preoperative differential diagnosis included mSA-PA shunt with a potential Yakes type IV AVM component.

Considering both entities is clinically important, as their pathophysiology and management differ substantially. PAVMs consist of direct communications between pulmonary arteries and pulmonary veins, are frequently associated with hereditary hemorrhagic telangiectasia, and are generally treated by transcatheter embolization of the pulmonary arterial feeders [5,6]. In contrast, SA-PA shunts involve systemic arterial inflow into the pulmonary circulation and typically require embolization of the systemic feeding arteries or surgical resection [7–9].

In the present case, a mixed vascular anomaly combining mSA-PA shunt and diffuse PAVM could not be excluded preoperatively based on angiographic findings. The early pulmonary venous filling raised concern for micro-arteriovenous shunting consistent with Yakes type IV AVM. However, definitive histopathological examination revealed no nidus formation or microfistulous arteriovenous communications, thereby excluding PAVM and establishing the diagnosis of congenital mSA-PA shunt.

The presence of a patent pulmonary artery supplying the affected segment excluded alternative diagnoses such as pulmonary sequestration or anomalous systemic arterial supply to the basal segment (ABLL), including Pryce type I [10,11]. Pulmonary sequestration is typically characterized by the absence of normal pulmonary arterial supply and nonfunctional lung parenchyma, features that were not observed in this case.

Involvement of the inferior phrenic artery in SA-PA shunt is particularly rare. Zhang et al. described a case in which the phrenic artery communicated with both the pulmonary artery and vein, although histological confirmation was lacking [12]. Notsuda et al. reported a surgically treated SA-PA shunt with histopathological confirmation, but involving only a single systemic feeder [13]. Serdar et al. described a case managed with transcatheter arterial embolization, also without histological evidence [14]. In contrast, the present case uniquely demonstrates dual systemic arterial inflow with definitive histopathological confirmation.

Although the patient was asymptomatic, the presence of dual systemic arterial feeders carries a potential risk of hemoptysis, pulmonary hypertension, or cardiac overload over time [7,14]. Given these considerations, surgical resection was selected, providing both definitive treatment and diagnostic confirmation. While transcatheter embolization may be appropriate in selected cases, particularly in acquired SA-PA shunt or when vascular anatomy is favorable [8,15], surgical resection remains preferable when multiple systemic feeders are present or when histological evaluation is required.

This case highlights the diagnostic challenges associated with rare pulmonary vascular anomalies and emphasizes the importance of multidisciplinary assessment. Clinicians should consider SA-PA shunt in the differential diagnosis of PAVM-like

lesions, particularly when systemic arterial supply is present without venous drainage. Early recognition of these congenital anomalies is essential for accurate diagnosis, appropriate risk stratification, and optimal treatment planning.

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