



## Isolated arterial pulmonary malinosculature without sequestration in an adult: A case report and literature review

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### ABSTRACT

Abnormal connections between systemic and pulmonary vascular systems are rare conditions and have been mostly documented in the pediatric population. We report a case of type B isolated arterial pulmonary malinosculature in an adult. The patient's chief complaint was intermittent hemoptysis during physical exertion. He had a dual arterial supply from the anomalous systemic artery and the pulmonary artery to the left lower lobe and the venous drainage was through the pulmonary vein. The bronchial connection appeared normal. The fistula was identified on chest computed tomography and was treated endovascularly. The patient remains asymptomatic at 1-year follow-up.

### 1. Introduction

Anomalous connections between the systemic and pulmonary vascular system, either congenital or acquired, are rare conditions [1,2]. These anomalies can be categorized as (1) anomalous systemic arterial supply to normal lung, (2) bronchopulmonary sequestration, (3) systemic artery to pulmonary vessel fistula, and (4) systemic artery to pulmonary vascular anastomoses secondary to chronic inflammatory diseases. In cases of intrapulmonary arteriopulmonary venous fistula, three types of vessels connecting to the pulmonary vein have been recognized including (1) a branch of the pulmonary artery, (2) a normal systemic artery such as the bronchial artery (most common), the intercostal artery, the internal mammary artery or some other branches of the aorta, (3) and an aberrant branch originating from the descending thoracic or proximal abdominal aorta [3]. The last type is extremely rare and has been sporadically reported in the infant population. In this type, the affected lobe is either normally developed or sequestered (not connected with the rest of the bronchial and pulmonary tree) [3]. Here, we report a case of an anomalous fistula between the descending aorta and the left inferior pulmonary vein without sequestration, termed as type B isolated arterial pulmonary malinosculature, in an adult. The anomaly was identified on chest computed tomography and confirmed on digital subtraction angiography. The patient was treated endovascularly and

remains asymptomatic at 1-year follow-up.

### 2. Case presentation

Based on our institutional policy, the Institutional Review Board is waived for case reports. A 46-year-old male patient presented with an acute episode of hemoptysis. He admitted mild-to-moderate volume of intermittent hemoptysis during physical exertion, but denied any history of smoking, chest trauma, or previous lung diseases. All laboratory tests were within the normal ranges. A chest X-ray revealed a homogeneous tubular opacity at the left lower lobe, otherwise unremarkable (Fig. 1). Bronchoscopy was unremarkable, except for the evidence of hemoptysis. Contrast-enhanced computed tomography demonstrated a high flow fistula between the descending aorta and the left lower pulmonary vein (Fig. 2). The afferent artery was dilated (maximum diameter was 11 mm) with wall calcification. Parenchymal ground-glass opacity was also evident. The affected lobe had a normal connection with the bronchial tree and pulmonary artery; thus, pulmonary sequestration was excluded. The fistula was confirmed on digital subtraction angiography and was embolized with multiple detachable coils (Interlock, Boston Scientific, Natick, MA, USA) (Fig. 3). The patient was discharged uneventfully and reported no recurrent hemoptysis during the 1-year follow-up.

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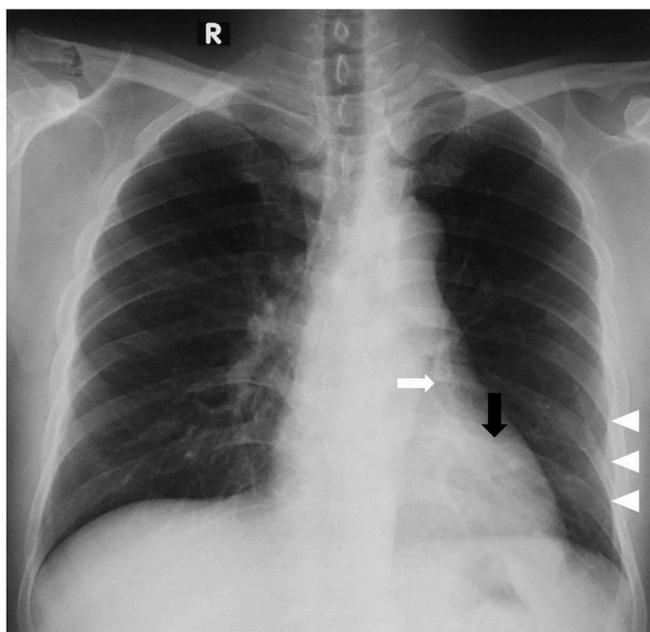
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**Fig. 1.** A plain chest radiograph reveals a homogenous, tubular retrocardiac/left lower lung opacity (black arrow). The normal left lower lobar pulmonary artery is still visible in the left infrahilar region (white arrow). Note increased interstitial markings in the peripheral left lower lung zone (arrowhead) compared to the normal right side.

### 3. Discussion

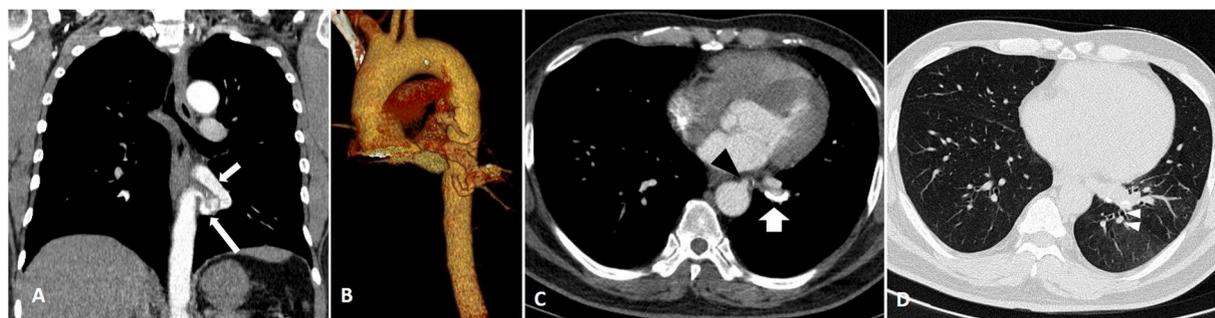
Congenital bronchopulmonary vascular malformations refer to a broad spectrum of disorders involving abnormalities in one or more of the three main components of the lung: the airways and lung parenchyma, arteries, and veins [1]. Anomalous systemic arterial supply to the basal segments without sequestration is a very rare congenital anomaly and has been sporadically reported in the English literature [2, 4]. Our patient had a dual arterial supply from an anomalous systemic artery and the pulmonary artery, whereas the bronchial tree and the venous drainage appeared normal. Neither structural abnormalities nor sequestration was found in the lung parenchyma, except for several ground-glass opacities presumably due to alveolar hemorrhage. This entity, therefore, can be classified as isolated arterial pulmonary malinosculation (type B) according to Lee's classification [5]. In this group, the arterial supply is abnormal, known as either interrupted pulmonary artery or anomalous systemic arterial supply or dual arterial supply to the normal lung [1,5].

In the present case, other potential differential diagnoses include other pulmonary vascular anomalies such as pulmonary arteriovenous malformations (PAVM), sequestration, or scimitar syndrome. PAVM refers to an abnormal communication between the pulmonary artery and pulmonary vein without an intervening capillary communication. As the afferent artery originated from the aorta, PAVM was excluded. Conversely, pulmonary sequestration was also excluded on the basis of a normal connection with the tracheobronchial tree and normal pulmonary arterial supply. Classic scimitar syndrome consists of hypoplasia of the right lung, hypoplasia of the right pulmonary artery, anomalous arterial supply from the abdominal aorta to the right lower lobe, and anomalous venous drainage of the right lung [6]. In our case, scimitar syndrome was effectively ruled out owing to normal pulmonary venous drainage and absent right lung hypoplasia [6,7].

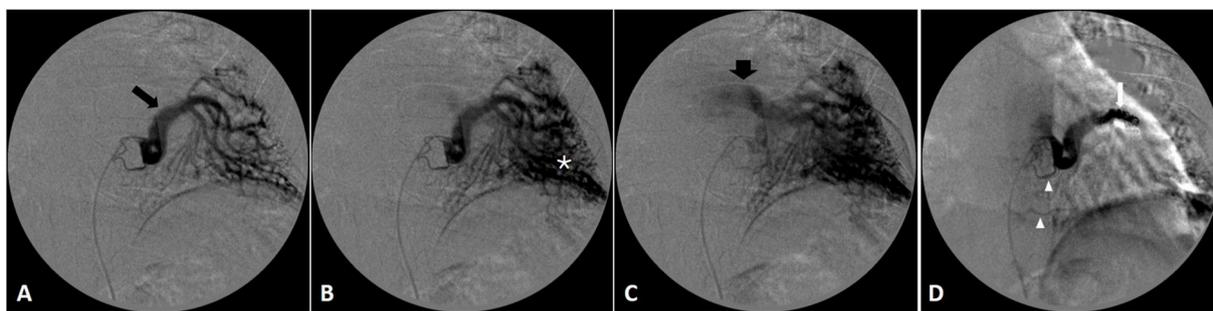
It is hypothesized that an abnormal persistence of embryonic aortic postbranchial arches supplying the lung buds before the development of the main pulmonary artery leads to this anomaly, though the exact cause remains unknown [8,9]. Wu discovered that 92% of the reported cases are Asian [10]. In a review, Miller found that most adult patients presented with symptoms such as hemoptysis (63%), pain (5%), cough (9%), and dyspnea on exertion or shortness of breath in (8%), whereas 26% were diagnosed incidentally on imaging [11]. The main clinical manifestations in children are heart murmur, respiratory distress, cardiomegaly, or heart failure [2]. The diagnosis should be suspected when a patient suffers from recurrent hemoptysis and cough, along with continuous murmur, cardiomegaly, and persistent shadow on chest radiographs [2].

On imaging, parenchymal changes can be appreciated in the affected lobe. Parenchymal changes include ground-glass opacities (GGO), volume loss, dilatation of the peripheral pulmonary vasculature, and possible arterial wall calcifications. GGO represents a mild degree of pulmonary congestion with or without associated alveolar hemorrhage induced by high systemic arterial pressure [12]. These lesions have been confirmed by pathological examination of the resected specimens [13]. Diffuse dilatation of peripheral pulmonary vasculature and areas of ground-glass attenuation seen on CT explain the increased interstitial markings on chest radiographs [12]. Thrombosis and premature atherosclerotic changes with wall calcification of the anomalous systemic artery have also been documented, suggesting a long-standing exposure of these vessels to high systemic arterial pressure [12,13]. Mild volume loss in the affected lobe, which was documented in all published cases, represents developmental hypoplasia [12].

Treatment is recommended even for asymptomatic patients because a left-to-left shunt may progress to heart failure and respiratory distress [9]. Established surgical treatments for this disease include anastomosis of the divided anomalous systemic artery to the pulmonary artery, simple ligation of the anomalous artery, and lower lobectomy or basal



**Fig. 2.** Coronal contrast-enhanced CT chest on mediastinal window (A) and 3D reconstruction (B) images demonstrate a tortuous, dilated anomalous artery (long arrow) originating from left antero-lateral descending thoracic aorta and entering the basal segments of the left lower lobe. Venous return (short arrow) is through the left lower pulmonary vein, indicating a left-to-left shunt. The left lower pulmonary vein is enlarged compared to its contralateral counterpart suggesting a high-flow fistula. Axial contrast-enhanced CT chest on mediastinal window (C) clearly depicts the aortic origin of the anomalous artery (black arrowhead) with wall calcification (thick arrow). Axial image on lung window (D) reveals a normal bronchial tree and accompanying pulmonary arteries (arrowhead). Note also a slight decrease in lung volume and subtle ground-glass opacity of the left lower lobe.



**Fig. 3.** Selective serial digital subtraction angiographic images of the anomalous artery shows (A) a tortuous and dilated afferent artery (black arrow), (B) an abnormal parenchymal blush and prominent vascular staining (asterisk) in the region of the left lower lobe, and (C) a normal venous return to the left pulmonary vein (thick arrow). There is no direct communication between the abnormal systemic artery and the veins of the involved lung tissue. These findings are compatible with a high-flow isolated arterial pulmonary malinosculature. Completion angiogram (D) confirms a total occlusion of the afferent artery after embolizing with multiple detachable coils (white arrow). Note the regurgitation of contrast media into the aorta and opacification of intercostal arteries (arrowheads). Bilateral bronchial arteries and other intercostal arteries have normal angiographic appearances and have no supply to the affected lobe (not shown).

segmentectomy with division of the anomalous artery [9]. Transcatheter embolization of the afferent artery has been validated as a safe and effective alternative treatment to traditional surgery [14–16]. Coil is the most preferable embolic material to achieve a permanent proximal occlusion of the afferent artery [17].

#### 4. Conclusion

Isolated arterial pulmonary malinosculature is a rare entity. Accurate interpretation of CT scan is important in making the correct diagnosis and treatment planning. Transarterial coil embolization is a safe and effective treatment modality for this anomaly.

#### Declaration of competing interest

No.

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