


Aberrant systemic arterial supply to normal lung arising from the proper hepatic artery discovered during transarterial chemoembolization

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ABSTRACT

We report a rare case of dual arterial supply to an otherwise normal lung discovered incidentally during initial angiography performed with the intent of chemoembolization of hepatocellular carcinoma. In addition to normal hepatic arterial supply, the proper hepatic artery provided systemic arterial supply to the lower lobe of the left lung. Subsequent chest computed tomography angiography demonstrated a normal tracheobronchial tree and normal pulmonary arterial supply to the lung. Although other anatomic variants have been reported, there are no other reported cases of systemic arterial supply from the proper hepatic artery to the lung. Identifying systemic arterial supply to the lung during angiography is important while performing transcatheter chemoembolization or radioembolization in the liver in order to minimize non-target embolization of the lung.

CASE REPORT

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A 53-year-old Cambodian-American man with hepatitis B and multifocal hepatocellular carcinoma presented for chemoembolization of right hepatic lobe lesions. The patient was asymptomatic. He denied hemoptysis or symptoms of heart failure. He also denied any history respiratory infections, chest surgery or trauma. Clinical examination, including that of the heart and lungs, was unremarkable. Laboratory evaluation was significant for elevated alpha-fetoprotein of 314,500 ng/mL (reference range: < 6 ng/mL), aspartate aminotransferase of 106 U/L (reference range: 10-44 U/L), alanine aminotransferase of 56 U/L (reference range: 7-54

U/L), and alkaline phosphatase of 195 U/L (reference range: 45-115 U/L).

Prior to embolization of liver lesions involving the right lobe, angiography of the common hepatic artery demonstrated an ascending branch from the proper hepatic artery that communicated with the left lower lobe pulmonary artery via a network of tortuous vessels. The pulmonary artery branch appeared otherwise normal, supplied normal-appearing lung parenchyma, and drained into the left inferior pulmonary vein (Figure 1). Computed tomography angiography (CTA) of the chest and abdomen also demonstrated the branch arising from the proper hepatic artery just distal to the origin of the

gastroduodenal artery. The vessel traversed the left hepatic lobe, entered the thorax through the esophageal hiatus, and then assumed a tortuous course before entering the left posterior basal segment (Figure 2 and Figure 3). The direction of flow in the systemic arterial vessel was definitively visualized as toward the pulmonary circulation from the proper hepatic artery. Pulmonary arterial supply to the left lower lobe was also present with normal branching patterns.

Chemoembolization proceeded routinely, with the microcatheter in distal branches of the right hepatic artery, well beyond the anomalous vessel. The patient recovered uneventfully aside from mild post-embolization syndrome, and three months later returned for a repeat chemoembolization with no change in the appearance of the anomalous vessel.

DISCUSSION

Etiology & Demographics:

Although different from classic intralobar bronchopulmonary sequestration, dual systemic and pulmonary vascular supply to normal lung likely represents an entity within the intralobar bronchopulmonary sequestration spectrum. Since Pryce et al. [1] originally described systemic artery supply to the lung as a type of bronchopulmonary sequestration, there has been ongoing controversy as to the etiology and classification of these anomalies [2,3]. Most authors believe the etiology of this entity to be the result of failure in regression of a remnant of systemic artery that supplied the embryonic lung. However, systemic arterial supply to the lung may also be acquired, particularly in the setting of congenital heart or lung disease, regional trauma, or infection [4-6]. The demographics of dual systemic and pulmonary arterial supply to normal lung are unknown since they are very rare and frequently asymptomatic. While the systemic arterial component of this vascular malformation likely represents a previously described inferior phrenic artery anatomic variant arising from the proper hepatic artery [7], dual vascular supply to the lung from this particular variant has not been previously described.

Clinical & Imaging Findings:

Patients with systemic vascular supply to normal lung are often asymptomatic but can present with recurrent hemoptysis. Less common complications may stem from systemic blood supply to the lungs over time and include infection, pleural effusion, and heart failure. Treatment may be warranted in the presence of these complications, but no treatment is required in asymptomatic cases [8, 9].

Chest radiography may show the anomalous systemic artery as a tubular or nodular opacity most frequently in the medial aspect of the left lower lobe. CT angiography is the most useful modality for evaluating cases of suspected sequestration or systemic arterial supply to the lungs, as it clearly shows both the bronchial and vascular anatomy of the lung [5,7,9]. Symptomatic patients with isolated systemic arterial supply to the right lower lobe [2,10], dual arterial supply from the celiac trunk to the left lower lobe [5,8], and

inferior phrenic artery arising from the aorta supplying right middle lobe [7] have been previously described.

Treatment & Prognosis:

When asymptomatic, no treatment is required for incidentally discovered dual arterial supply to normal lung. In symptomatic cases, embolization of the systemic artery is considered the first line of treatment. If this fails, surgical resection of the involved lung may be necessary. Due to the rarity of this condition, prognostic data are not available [5,7].

When performing intra-arterial embolization therapies, systemic arterial supply to the lung arising from the hepatic circulation must be recognized to avoid non-target embolization. In this case, placing the catheter proximal to the origin of the anomalous artery could have resulted in embolic particles entering the pulmonary vein, left heart, and subsequently allowing potentially disastrous embolization of end-organs via the systemic circulation. Super selective catheterization beyond the anomalous artery and careful administration of the embolic material should minimize this risk, but if such measures are impossible, then protective coil embolization of the anomalous artery should be considered. In this case, selective microcatheterization well beyond the aberrant arterial branch was performed prior to embolization. Therefore, it was unnecessary to perform protective coil embolization of the anomalous vessel.

Differential Diagnosis:

Once systemic arterial supply to the lung has been identified, a spectrum of diagnoses related to bronchopulmonary sequestration should be considered [4,8]. The differential diagnostic possibilities include the “classic” form of intralobar pulmonary sequestration, “variants” in the spectrum of intralobar pulmonary sequestration such as isolated systemic supply to normal lung and dual arterial supply to normal lung, and extralobar pulmonary sequestration. A definitive diagnosis may be achieved by characterizing the venous return of the involved lung, the presence or absence of pulmonary arterial supply to the involved lung, and the presence or absence of a normal bronchial tree and aeration of the involved lung [5-10]. Although many imaging modalities can evaluate components of these, CT angiography is the best single modality for evaluation of all of the components [5,7,9].

“Classic” Intralobar Pulmonary Sequestration

“Classic” intralobar pulmonary sequestration involves isolated systemic arterial supply to an abnormally aerated lung. The lung may be consolidated or cystic. The bronchial connections are incomplete. Venous return is typically via the pulmonary veins. This entity typically presents clinically in late childhood or early adulthood as recurrent pulmonary infections [5-10].

“Variant” forms of Intralobar Pulmonary Sequestration: Isolated Systemic Arterial Supply or Dual Arterial Supply to Normal Lung “Variant” forms in the spectrum of intralobar pulmonary sequestration may include dual systemic and pulmonary arterial supply to normal lung (as in this case) and isolated systemic supply to normal lung. In these entities, the

lung demonstrates normal bronchial connections and aeration and venous return is via the pulmonary veins. Although patients with these are usually asymptomatic, hemoptysis is the most common symptom [5-10].

Extralobar Pulmonary Sequestration

Extralobar pulmonary sequestration involves systemic arterial supply to consolidated, mass-like lung. In this condition, the sequestered lung has a separate pleural covering, lacks bronchial connections, and has systemic venous drainage [4-6]. Patients with extralobar sequestration usually experience respiratory distress in infancy.

TEACHING POINT

When performing embolization procedures, identification of variant vascular anatomy is critical for the prevention of non-target embolization. Aberrant systemic arteries supplying the lungs are rare, but may arise from several locations including hepatic arterial branches.

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FIGURES

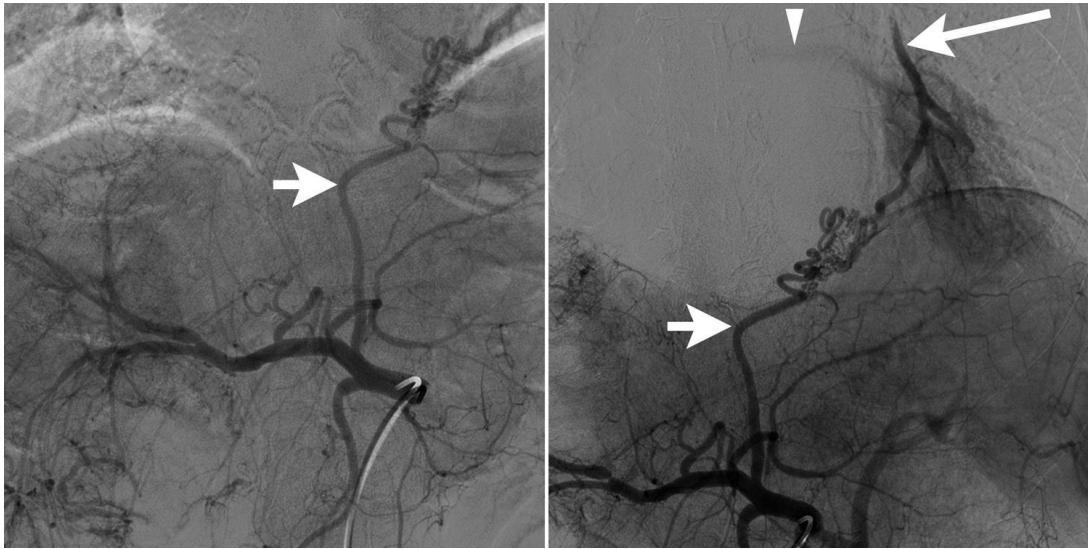


Figure 1: 53 year-old male with aberrant systemic arterial supply to normal lung

Findings: Digital subtraction angiography (DSA) of the common hepatic artery prior to chemoembolization. An aberrant systemic artery arising from the proper hepatic artery (short arrow) communicates with the left lower lobe posterior basal segmental artery of the lung (long arrow). The dual arterial supply supplies normal-appearing lung parenchyma and subsequently drains into the left inferior pulmonary vein (arrowhead).

Technique: Phillips Allura Xper, Angiogram with a catheter placed in the common hepatic artery, mAs 280, kVp 60. Iodixanol (Visipaque, GE Healthcare) injected at 4 ml/sec for a total of 16 ml

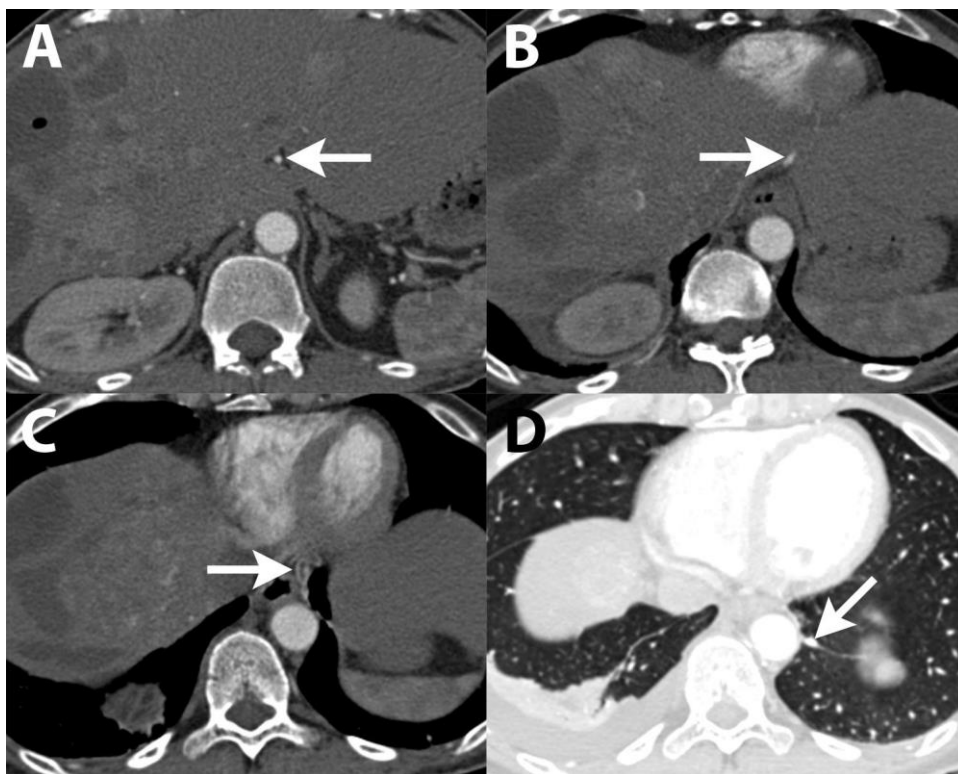


Figure 2: 53 year-old male with aberrant systemic arterial supply to normal lung

Findings: Axial computed tomography angiography demonstrating an aberrant systemic artery (arrows) passing superiorly from the proper hepatic artery within the medial aspect of the fissure of the ligamentum venosum (A), then anteriorly over the left lateral segment, (B) and dividing into a network of smaller vessels at the posteroinferior aspect of the pericardium (C). This network of vessels consolidates back into a single vessel and passes into the left lower lobe at the inferior pulmonary ligament (D).
Technique: Siemens Sensation 16 slice, 1 mm thickness, mAs 262, kVp 100. Iohexol (Omnipaque 350, GE Healthcare) at 4 ml/sec for a total of 60 ml

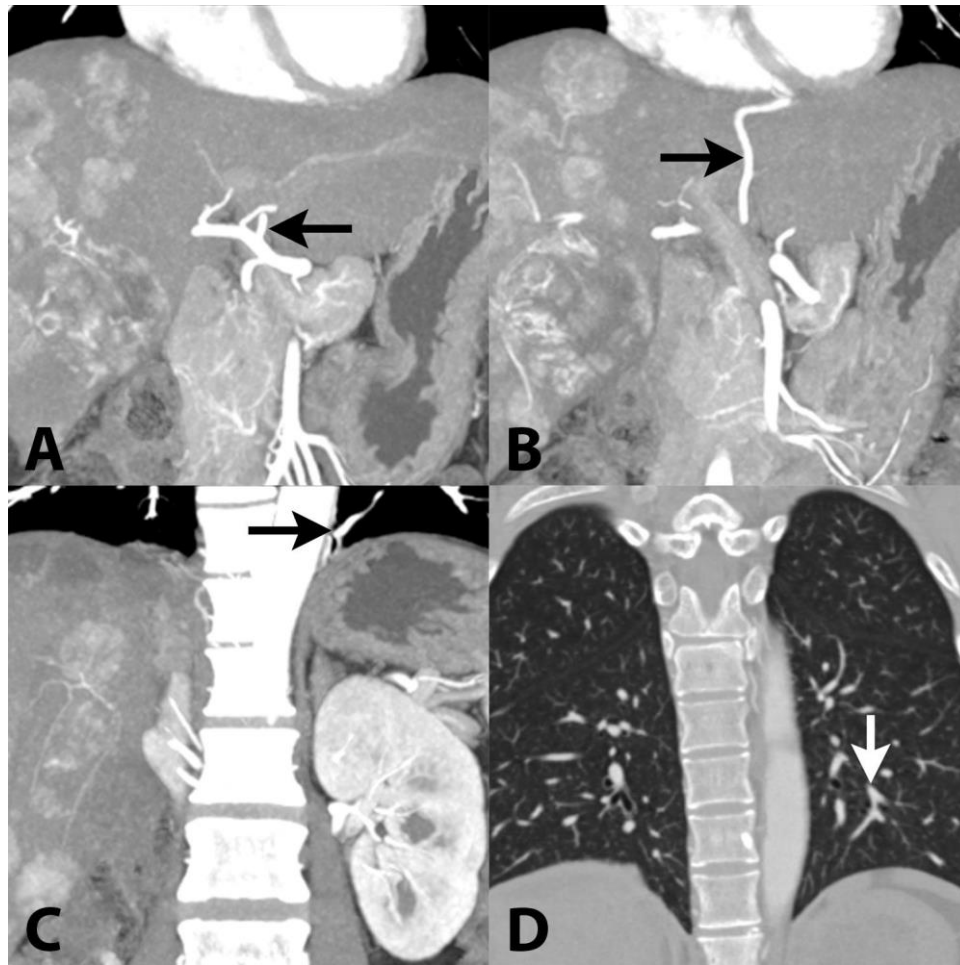


Figure 3: 53 year-old male with aberrant systemic arterial supply to normal lung

Findings: Coronal reconstructed maximal intensity projection (MIP) computed tomography angiography images demonstrating an aberrant systemic artery (arrow) originating from the proper hepatic artery (A), passing superiorly to the underside of the pericardium (B), and entering the left lower lobe (C) and joining the left lower lobe posterior basilar pulmonary artery (D).

Technique: Siemens Sensation 16 slice, MIP reconstructed at 10 mm slab thickness, mAs 262, kVp 100, Iohexol (Omnipaque 350, GE Healthcare) at 4 ml/sec for a total of 80 ml.

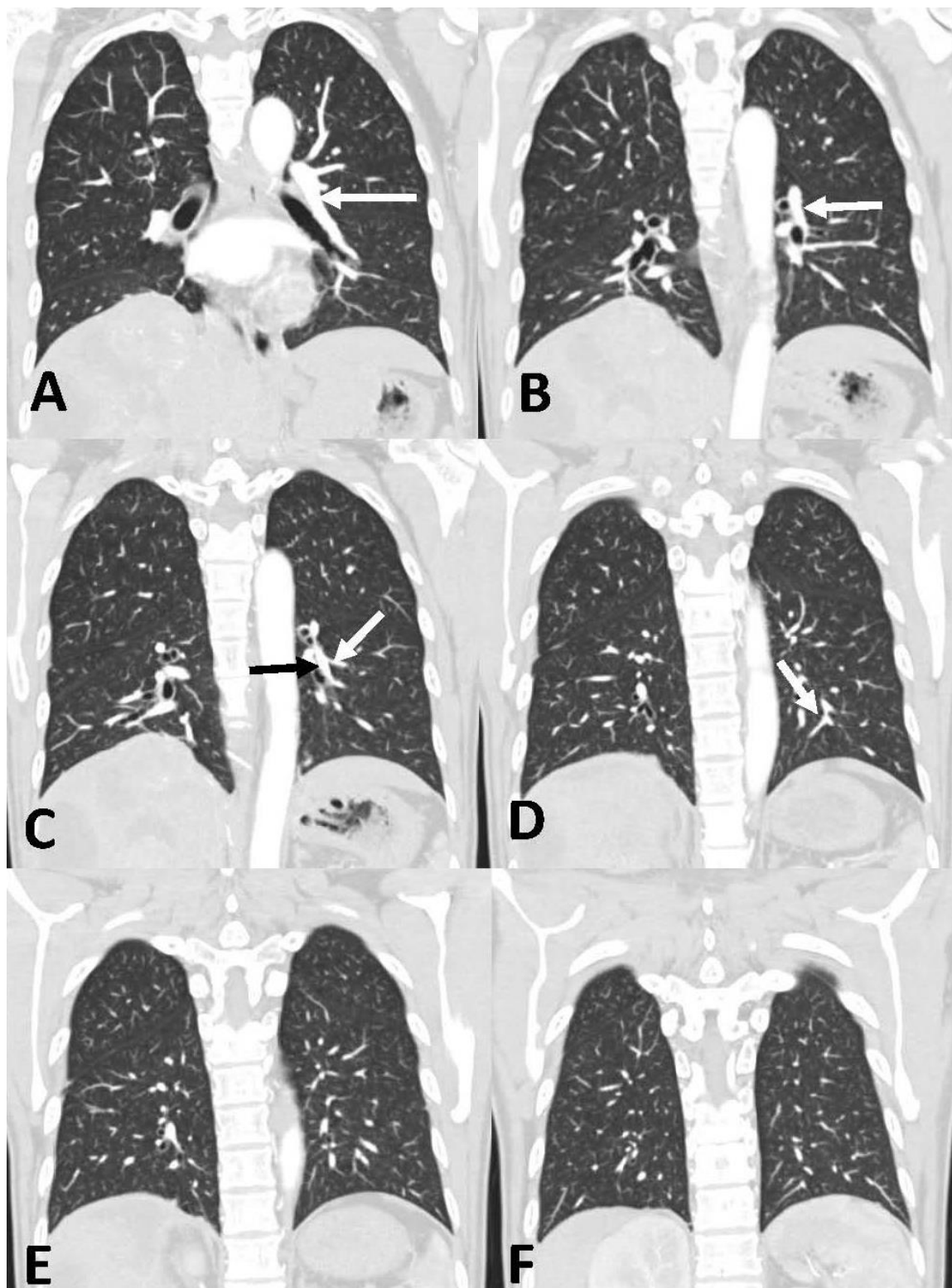


Figure 4: 53 year-old male with aberrant systemic arterial supply to normal lung

Findings: Coronal reconstructed computed tomography pulmonary angiography images demonstrating normal pulmonary arterial branching to the involved left lower lobe from the interlobar pulmonary artery (arrow) (A), to the basal trunk (arrow) (B), at the junction of the lateral basilar (white arrow) and posterior basilar (black arrow) pulmonary arteries (C), to the junction of the left lower lobe posterior basilar pulmonary artery with the aberrant systemic artery (arrow) (D), to the terminal branches of the posterior basilar pulmonary artery (E and F).

Technique: Siemens Sensation 16 slice, 1 mm thickness, mAs 130, kVp 120. Iohexol (Omnipaque 350, GE Healthcare) at 4 ml/sec for a total of 80 ml

Definition	Dual arterial supply from the pulmonary artery and a systemic artery in normal, aerated lung with a normal tracheobronchial tree. Venous drainage is via the pulmonary vein.
Etiology	Usually considered congenital from a failure of regression of a systemic artery that supplied the embryonic lung. Less commonly, this could be acquired after infection or trauma.
Incidence	Rare. Incidence is unknown as there are only case reports. This anomaly is frequently asymptomatic, so frequently goes undiagnosed.
Gender ratio	Uncertain due to rarity. Probably 1:1
Age predilection	Unknown. However, since this entity is usually congenital, there is likely no age predilection. When these are clinically symptomatic in children, they warrant evaluation for congenital cardiac
Anatomic Location	Lung bases are most common. The left is more common than the right. Systemic artery most frequently arises from the aorta.
Risk factors	Unknown for most. For the less common acquired form, prior lung infection or trauma.
Clinical Presentation	Usually asymptomatic. Most common symptom is hemoptysis. Other symptoms may include heart failure, pleural effusion, and infection.
Treatment	None if asymptomatic. If symptomatic, treat with embolization or surgery.
Prognosis	Data are lacking, but prognosis seems good based on reported cases.

Table 1: Summary table of dual systemic and pulmonary arterial supply to normal lung

Differential Diagnosis	Clinical Features	Features on Radiography	Features on CT	Features on Angiography
Dual arterial supply to normal lung	<ul style="list-style-type: none"> Usually asymptomatic. Most common presentation is hemoptysis. Less commonly: heart failure, pleural effusion, infection. 	<ul style="list-style-type: none"> Tubular or nodular opacity usually at the lung base (left > right). 	<ul style="list-style-type: none"> Systemic and pulmonary arterial supply to normally aerated lung with an intact bronchial tree. Venous drainage via the pulmonary veins. 	<ul style="list-style-type: none"> Systemic and pulmonary arterial supply to a portion of normal lung. Venous drainage via the pulmonary veins.
Isolated systemic arterial supply to normal lung	<ul style="list-style-type: none"> Usually asymptomatic. Most common presentation is hemoptysis. Less commonly: heart failure, pleural effusion or infection. 	<ul style="list-style-type: none"> Tubular or nodular opacity usually at the lung base (left > right). 	<ul style="list-style-type: none"> Systemic arterial supply only to normally aerated lung with an intact bronchial tree. Venous drainage via the pulmonary veins. 	<ul style="list-style-type: none"> Systemic arterial supply to a portion of normal lung. Venous drainage via the pulmonary veins.
“Classic” intralobar pulmonary sequestration	<ul style="list-style-type: none"> Usually present in later childhood or early adulthood with recurrent lung infections. Male: Female = 1:1 Rarely associated with other developmental disorders 	<ul style="list-style-type: none"> Mass-like opacity or cystic lesions usually at the lung base (left > right). 	<ul style="list-style-type: none"> Systemic arterial supply only to consolidated or abnormally cystic lung. Incomplete bronchial connections to the involved lung. Venous drainage is classically via the pulmonary veins, but systemic drainage has been described. 	<ul style="list-style-type: none"> Systemic arterial supply to a portion of abnormal lung. Venous drainage classically via the pulmonary veins, but systemic drainage has been described.
Extralobar pulmonary sequestration	<ul style="list-style-type: none"> Usually present in infancy with respiratory distress. Male: Female= 4:1 High association with other congenital abnormalities 	<ul style="list-style-type: none"> Mass-like opacity at the lung base (left>>right). 	<ul style="list-style-type: none"> Systemic arterial supply only to consolidated, mass-like lung. Venous drainage is via systemic veins. No bronchial connections. Involved portion of lung has separate visceral pleura covering. Sequestered lung may be subdiaphragmatic. 	<ul style="list-style-type: none"> Systemic arterial supply to a portion of abnormal lung. Venous drainage is via systemic veins.

Table 2: Differential diagnosis table for dual systemic and pulmonary arterial supply to normal lung

ABBREVIATIONS

ALT = alanine aminotransferase
AST = aspartate aminotransferase
CT = computed tomography

KEYWORDS

pulmonary-systemic vascular malformation;
bronchopulmonary sequestration; dual systemic-pulmonary
arterial supply to lung; aberrant arterial supply to lung;
systemic arterial supply to lung; chemoembolization

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