CASE REPORT

Anomalous Systemic Arterial Supply to the Normal Basal Segments of the Left Lower Lobe of the Lung

JJK Ip, PKT Hui, SHY Lam, SCW Cheung, WWM Lam, MT Chau

Department of Radiology, Queen Mary Hospital, Pokfulam, Hong Kong

ABSTRACT

Anomalous systemic arterial supply to the normal basal segments of the lower lobe of the lung without sequestration is a rare congenital abnormality. Whether the condition belongs to the broad spectrum of sequestration disorders remains controversial. This report is of a patient with anomalous systemic arterial supply to the normal basal segments of the left lower lobe, together with an attenuated pulmonary artery and normal tracheobronchial tree.

Key Words: Arteries; Arteriovenous malformations; Bronchopulmonary sequestration; Hemoptysis; Pulmonary artery

INTRODUCTION

Anomalous systemic arterial supply to the normal basal segments of the lower lobe of the lung without sequestration is a rare congenital abnormality. Whether the condition belongs to the broad spectrum of sequestration disorders remains controversial. This is a recognised congenital anomaly, which is not related to pulmonary arteriovenous malformation (AVM). Most patients with anomalous systemic arterial supply are asymptomatic, although occasionally haemoptysis may occur. This report is of a patient with anomalous systemic arterial supply to the basal segments of the left lower lobe, together with an attenuated pulmonary artery and normal tracheobronchial tree. The relationship of this anomaly with bronchopulmonary sequestration and how to differentiate these two similar conditions is also discussed.

CASE REPORT

A 30-year-old man, who did not smoke and was in good health, was noted to have a subtle abnormality in his pre-employment chest radiograph in April 2011. An approximately 2x4-cm shadow was present in the left retrocardiac region, with adjacent dilated tubular...
structures suggestive of feeding arteries and draining veins (Figure 1). The respiratory physician suspected that the shadow was a pulmonary AVM. The patient underwent magnetic resonance imaging (MRI) to measure the shunt ratio, as embolisation depended on the degree of arteriovenous (AV) shunting.

However, MRI was not typical of pulmonary AVM. Magnetic resonance angiography (MRA) demonstrated a serpiginous vascular tangle at the left lower lobe, with systemic arterial blood supply and pulmonary venous drainage (Figure 2). Cine phase-contrast MRI for flow volume showed a pulmonary-to-systemic

---

**Figure 1.** Chest radiographs of a patient with anomalous systemic arterial supply to the normal basal segments of the lower lobe of the lung. (a) Posterior-anterior projection demonstrating an approximately 2x4-cm shadow at the left retrocardiac region (arrow), with adjacent dilated tubular structures, suspicious of feeding arteries and draining veins; and (b) left lateral projection confirming the observations in the posterior-anterior projections (circle).

**Figure 2.** Magnetic resonance angiography demonstrating a serpiginous vascular tangle at the left lower lobe, with (a) systemic arterial blood supply (circle), and (b) pulmonary venous drainage (arrow); and (c and d) volume-rendering images demonstrating tangling between the systemic artery and dilated pulmonary vein in the left lower lobe.
flow ratio (Qp:Qs) of 1:1.1, indicative of no significant AV shunting. Pulmonary MRA showed attenuation of segmental branches of the basal segments of the left lower lobe (Figure 3). Bronchopulmonary sequestration was suspected.

Computed tomography (CT) was subsequently performed and demonstrated an anomalous systemic artery, of approximately 1.0 cm in diameter, from the descending aorta at the T8 level to the basal segment of the left lower lobe (Figure 4). Dilated
arteries were paralleling the normal bronchi. No direct communication with the pulmonary veins was evident. Tributaries of the left inferior posterior vein in the left lower lobe were hypertrophied, with variceal formation. However, branches of the left pulmonary artery supplying the basal segments, distal to the origin of the apical segment branch, were attenuated. The underlying lung parenchyma was normal, which made a diagnosis of bronchopulmonary sequestration less likely. The overall findings were supportive of a diagnosis of anomalous systemic arterial supply to normal lung parenchyma in the basal segments of the left lower lobe, with associated pulmonary vein dilatation and attenuated pulmonary artery branches. Since the patient was asymptomatic with no significant shunting, he was treated conservatively.

DISCUSSION
Anomalous systemic arterial supply to the normal basal segments with normal bronchial connection of the lower lobe, which has no normal pulmonary arterial supply and no sequestration, is a rare congenital anomaly. The condition was previously classified as a type of sequestration according to Pryce’s terminology. However, use of the term ‘sequestration’ is controversial because of the normal bronchial connections in this anomaly. Therefore, other terms, including systemic arterial supply to the basal segments of the lung; systemic origin of the sole artery to the basal segments of the lung; or systemic arterialisation of the lung without sequestration, have been used.

The aetiology of a systemic arterial supply to the basal segment of the left lower lobe without a pulmonary artery supply has been contentious. The lungs first appear during the third week of embryonic life as ventral outgrowths on the foregut. By 30 days, a visceral vascular plexus appears around the branching primary lung buds. This plexus is fed by pulmonary channels from the sixth aortic arches and systemic channels from the dorsal aorta. At the sixth week, the systemic channels normally regress once the pulmonary arterial system has become established. Pulmonary oligemia is one of the factors that determines the persistence or re-establishment of pulmonary-systemic collaterals. The most likely theory is that persistence of the early branches arising from the aorta causes an anomalous systemic arterial supply to the lung. Cases reported in the literature with a combined supply from the pulmonary artery and systemic artery to normal lung parenchyma support this theory.

Radiological features of anomalous systemic arterial supply to the normal basal segments are often confused with pulmonary AVM or bronchopulmonary sequestration. In plain radiographs, the anomalous systemic artery can appear as a dilated tubular structure or ill-defined nodular area of increased opacity in the retrocardiac region, as in this patient. The anomalous systemic artery can silhouette the outline of the lower thoracic descending aorta as a focal obscuration. Anomalous systemic arteries are also well demonstrated by CT as vascular structures, with the same attenuation as the thoracic aorta in the left lower lobe. Moreover, CT can also provide information about the morphology of the bronchial tree and the pulmonary parenchyma, which are both normal in this condition. Another CT finding is the absence or attenuation of the interlobar artery distal to the origin of the superior segmental artery. Although MRI can demonstrate an anomalous systemic artery as well as CT, it would not be easy to determine confidently whether the lung tissue supplied

<table>
<thead>
<tr>
<th>Type</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital</td>
<td>Bronchopulmonary sequestration</td>
</tr>
<tr>
<td></td>
<td>Congenital pulmonary venolobar syndrome:</td>
</tr>
<tr>
<td></td>
<td>• involved lung parenchyma is supplied by the aberrant systemic arteries</td>
</tr>
<tr>
<td></td>
<td>• anomalous systemic artery can also supply an area of otherwise normal lung parenchyma</td>
</tr>
<tr>
<td></td>
<td>Systemic arterial supply to the normal basal segments of the lower lobe</td>
</tr>
<tr>
<td>Acquired</td>
<td>Bronchiectasis</td>
</tr>
<tr>
<td></td>
<td>Pulmonary tuberculosis and other pulmonary infections</td>
</tr>
<tr>
<td></td>
<td>Chronic pulmonary artery obstruction:</td>
</tr>
<tr>
<td></td>
<td>• Takayasu's arteritis</td>
</tr>
<tr>
<td></td>
<td>• pulmonary thromboembolism</td>
</tr>
<tr>
<td></td>
<td>Chronic obstructive pulmonary disease:</td>
</tr>
<tr>
<td></td>
<td>• hypertrophied normal systemic arteries supply the lungs</td>
</tr>
</tbody>
</table>

Table. Causes of systemic arterialisation of the lung.
by a systemic artery is normal. Therefore, contrast-enhanced CT is a useful technique for diagnosis of anomalous systemic arterial supply to normal lung tissue.5

There are a number of causes of systemic arterialisation of the lung, including congenital and acquired disease (Table). Bronchopulmonary sequestration has always been a close differential diagnosis of anomalous systemic arterial supply to the normal basal segments. Differentiating the two conditions can be challenging both clinically and radiologically. Debate about their synonyms and relationship with each other remain unresolved. If any pulmonary lesion with a systemic arterial supply is included in the sequestration, it is questionable whether the lung is sequestered in patients with a combined supply from the normal pulmonary artery and anomalous systemic artery to the normal lung, or in patients with a systemic artery-pulmonary vein fistula. Adhering to the definitions of the diseases and anatomical relationships can often provide clues to the correct diagnosis. Radiological findings of the bronchial anatomy and lung parenchymal condition are also determining factors in making a diagnosis. For instance, bronchopulmonary sequestration is defined as a mass of abnormal pulmonary tissue that does not communicate with the tracheobronchial tree through a normally located bronchus, but is supplied by an anomalous systemic artery. According to this definition, the anomaly reported in this patient cannot be considered as a type of true bronchopulmonary sequestration, because the lesion of bronchopulmonary and parenchymal tissue of this disease is never sequestered.

However, despite the differences in bronchopulmonary and parenchymal tissue, a systemic arterial supply to the normal basal segments is not completely unrelated to bronchopulmonary sequestration. The location of an anomalous systemic arterial supply to the normal lung is in the left lower lobe, especially the basal segments, as is the case in most cases of pulmonary sequestration. The characteristics of these aberrant arteries, whose origins are in the pulmonary ligament or below the diaphragm, with an elastic-type arterial wall histologically, are the same as in pulmonary sequestration. Venous drainage is via the pulmonary vein, which is consistent with intralobar sequestration. In some reported cases of bronchopulmonary sequestration, a normal communication with a normal bronchial tree was evident without inflammatory changes.6 Moreover, bronchial growth is dependent on the available coelomic space and pulmonary arteries, which provide the primary blood supply to developing lung tissue, as bronchial blood supply alone is insufficient to supply the nutritional needs of the growing fetal lung. With regard to lung development, it is not possible to separate the relationship between the bronchial bud and adjacent artery.7 These factors suggest the difficulty in considering anomalous systemic arterial supply to the normal basal segments of the left lower lobe as a completely independent disease from bronchopulmonary sequestration.

Other diagnoses have characteristic imaging findings, so differential diagnosis is not difficult (Table). Congenital pulmonary venolobar syndrome consists of a number of components, including hypogenetic lung, partial anomalous pulmonary venous return, absence of pulmonary artery, bronchopulmonary sequestration, systemic arterialisation of the lung without sequestration, absence of the inferior vena cava, and duplication of the diaphragm. Acquired causes of systemic arterialisation of the lungs are often demonstrating features of underlying disease, for example, dilated bronchial tree and mucus retention in bronchiectasis; cavitory lesion or abscesses in tuberculosis and other pulmonary infections; and calcifications, mosaic lung parenchymal attenuation, and pulmonary arterial hypertension in chronic pulmonary embolism. In patients with Takayasu’s arteritis, the diameter of the pulmonary artery is decreased, and a hypertrophied systemic artery, usually the left inferior phrenic artery or bronchial artery, is demonstrated.

Patients with anomalous systemic arterial supply to the normal basal segments of the left lower lobe are often asymptomatic, although haemoptysis occurs occasionally. If clinical symptoms such as haemoptysis and congestive heart failure are not present, treatment is conservative, although surgery is always imperative for correction of a left-to-right shunt. An anomalous systemic artery can be ligated if there is a pulmonary supply to the involved segments of the lung. However, if the aberrant artery represents the sole source of blood flow, lobectomy is required. Either way, contrast-enhanced CT is indispensable for correct diagnosis and proper treatment.

In conclusion, anomalous systemic arterial supply to the normal basal segments of the left lower lobe of the lung without sequestration is a rare congenital anomaly.
Anomalous Systemic Arterial Supply

The condition should be included in the differential diagnosis when a chest radiograph depicts a retrocardiac nodular or tubular shadow with focal obscurity of the descending thoracic aorta. Most of these patients are asymptomatic although haemoptysis is noted occasionally. Both aortic and pulmonary arterial angiographic studies are required to plan the definitive surgical procedure, especially in symptomatic patients. Contrast-enhanced CT and CT angiograms are currently the most appropriate investigations for diagnosing this condition and planning further management.

REFERENCES