Imaging of pulmonary sequestration: what the radiologist needs to know

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Learning objectives

• To provide an overview of the pathophysiology of intralobar and extralobar sequestration.
• To describe imaging findings of pulmonary sequestration in patients with a pulmonary mass or recurrent infection, with particular reference to CT.
• To stress the importance of CT angiography to identify the location of aberrant vessels for treatment planning (either surgical or endovascular).
Background

Bronchopulmonary sequestration (BPS) is a rare congenital malformation of the lower respiratory tract. It consists of a nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree and receives its arterial blood supply from the systemic circulation, typically from an aberrant artery or arteries that may pass through the inferior pulmonary ligament\(^1\) (Fig. 1 on page 6).

It is a rare anomaly representing 0.15% to 6.4% of all pulmonary malformations, reaching 95% of the cases in the left lower lobe\(^2\), and is divided into two types based upon their pleural investment as intralobar or extralobar (Fig. 2 on page 6). Intralobar sequestration (ILS) is more common (75% of cases) and shares the same visceral pleural covering as the native lung (Fig. 3 on page 9) with venous drainage into the pulmonary veins, whereas extralobar sequestration (ELS) (25% of cases) has its own visceral pleural investment outside the normal lung (Fig. 4 on page 8) with venous drainage into a systemic vein, and constitutes an accessory lobe, also called a "Rokitanski lobe". Approximately 10-15% of ELS are found within or below the diaphragm\(^3\).

The age of presentation depends on the type of sequestration and this in turn determines the clinical presentation. ILS presents late in childhood or adolescence with recurrent pulmonary infection while ELS more commonly presents in newborn with respiratory distress, cyanosis, and infection\(^4\).

ORIGIN AND CHARACTERISTICS OF INTRALOBAR SEQUESTRATION

Most cases of ILS appear to have an acquired origin caused by chronic pulmonary infection. It is very rare in infants and is infrequently associated with congenital anomalies (esophagobronchial diverticula, diaphragmatic hernias, skeletal deformities, cardiovascular defects and renal anomalies), but there are numerous examples of ILS discovered on prenatal ultrasound. It is known that lower lobe infection or chronic inflammation, typically with associated bronchiectasis, can result in cystic lung disease with proliferation of systemic arteries (neovascularity) entering the lung through the pulmonary ligament or across the pleura. Small systemic pulmonary ligament arteries arise from the anterior thoracic aorta, contribute to the esophageal arterial plexus, and traverse the pulmonary ligament to ramify in the visceral pleura of the lower lobes of the lung. These vessels are distinct from the pulmonary circulation and from the bronchial, vertebral and intercostal arteries. Some Authors proposed that these normally
occurring arteries may become parasitized to supply then infected portion of a lower lobe if the normal pulmonary arterial supply is compromised. However, in contradistinction to congenital ILS, these cases of acquired lesions have an intact bronchial connection and thus do not have sequestered lung tissue, and the arteries in neovascularity are thin and irregular and distinctly different from the large and regular anomalous artery typical of ILS. Such acquired conditions that can mimic sequestration have been termed pseudosequestration by some authors. ILS is typically located in the medial or posterior basal segments of the left lower lobe.

The anomalous aortic branches that supply the lesion in ILS are characteristically located within the inferior pulmonary ligament. In ILS anomalous systemic arterial supply is via the descending thoracic aorta (72%) (Fig. 5 on page 7), via abdominal aorta, celiac axis, or splenic artery (21%), via intercostal artery (3%), and rarely via the subclavian, internal thoracic, and pericardiacophrenic arteries.

Venous drainage is usually via the pulmonary veins, but it can also occur through the azygos vein/hemiazygos system, portal vein, right atrium, or IVC.

ORIGIN AND CHARACTERISTICS OF EXTRALOBAR SEQUESTRATION

In the ELS more than 60% of patients present in the first 6 months of life with respiratory distress, feeding difficulty and congenital cardiac failure. ELS may manifest in utero, associated with polyhydramnios or fetal hydrops. Patients may also present in childhood or adulthood. Approximately 10% of ELS are found incidentally in asymptomatic individuals. In rare cases, patients may have recurrent pulmonary infections, high output congestive heart failure in infancy, and spontaneous pulmonary or pleural hemorrhage. In contradistinction to ILS, there is a 3:1 to 4:1 male predominance in most reported antenatal series. Sixty-five percent occur on the left side. Only 10-15% of all ELS are subdiaphragmatic. More than 60% have coexistent anomalies, and the most common is congenital diaphragmatic hernia. Twenty-five percent of patients have another congenital abnormality of the lung, such as hypoplasia, cystic adenomatoid malformation, emphysema and bronchogenic cyst. In adulthood, subdiaphragmatic extralobar pulmonary sequestration is found as an incidental retroperitoneal mass on imaging studies, and accompanying symptoms are less common.

The blood supply of ELS is typically from systemic arteries. This arises directly from the thoracic or abdominal aorta in approximately 80% of cases. The feeding vessel is typically single and measures between 0.5cm and 2cm in diameter. In approximately 15% of cases, ELS is supplied by smaller arteries from splenic, gastric, subclavian, and intercostal branches. Approximately 20% of lesions are supplied by multiple arteries. In
5% of cases, the lesion is supplied by branches of the pulmonary artery or by both the pulmonary and systemic circulations. The venous drainage of ELS is usually systemic (80%), through the azygos system, the emiazygos system, or the vena cava to the right atrium. In approximately 25% of ELS, the venous drainage is partly through the pulmonary veins. Less common routes of drainage include the portal, intercostal, suprarenal, and other abdominal veins. An ELS supplied by pulmonary arteries is more likely to have pulmonary venous drainage.

DIFFERENTIAL DIAGNOSIS

Several conditions mimic the imaging appearance of sequestration but do not have systemic arterial supply to the lung. These include primary or metastatic tumor, other congenital lesions, and infection. Other conditions may share the feature of an anomalous systemic artery. Systemic arterial supply to the lung can be found in both acquired and congenital disorders. Acquired systemic arterial supply to the lungs generally develops from hypertrophy of normal systemic arteries in response to chronic inflammation or infection or to chronic pulmonary artery obstruction (eg, chronic pulmonary thromboembolism or Takayasu arteritis). Congenital systemic arterial supply to the lung generally represents anomalous or aberrant arteries and is found with sequestration or hypogenetic lung (scimitar) syndrome and sometimes as an isolated anomaly supplying normal lung tissue.

THERAPEUTIC OPTIONS

Management of the asymptomatic lung sequestration is controversial. However, most authors advocate resection of these lesions because of the likelihood of recurrent infection, the need for larger resection if the sequestration becomes chronically infected, and the possibility of hemorrhage. Surgical resection is the treatment of choice, and ILS often requires ligation of the feeding vessel and lobectomy. Open thoracotomy remains the best approach with safe isolation and division of anomalous systemic feeding arteries. A new promising treatment is the embolization of the aberrant arteries with an endovascular approach, for reducing the complications and risks of surgery. It is mainly attempted by using coils, but also with a self-expandable cylindrical mesh device like the Vascular Plug, when a large and high-flow vessels are present and coil migration is possible or multiple coils may be needed.
**Fig. 1:** The inferior pulmonary ligament is a fused triangular-shaped sheet of parietal and visceral pleura that extends from the hilum to the dome of the hemidiaphragm. It extends from the mediastinum to the medial surface of the lower lobe and is extra-parenchymal to the lung.

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Fig. 2: Intra- and extralobar sequestration. The infradiaphragmatic sequestration is not only extralobar (it has its own pleura), but is also located below the diaphragm.

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**Fig. 5:** Drawing of a lung containing intralobar sequestration. ILS consists of lung tissue that does not have a normal connection to the tracheobronchial tree. A systemic artery (arrow) typically arises from the distal thoracic aorta (A) to supply the lesion.

**Fig. 4:** Graphic illustration of ELS, having its own visceral pleural investment outside the normal lung.

**Fig. 3:** Graphic illustration of ILS, sharing the same visceral pleural covering as the native lung.

Findings and procedure details

In patients with a history of recurrent pneumonia, a persistent left-sided inferior paraspinal mass and the identification of systemic arterial feeding are the most representative radiological findings in pulmonary sequestration. In 80% of cases a venous drainage with left-to-right shunt is present.

IMAGING OF INTRALOBAR SEQUESTRATION

ILS is a great mimicker with diverse imaging features\(^1\). The diagnosis may be suggested by chest radiographic findings alone and should be considered in patients with recurrent pneumonia or localized bronchiectasis, almost always in the posterobasal aspect of a lower lobe.

- **On chest radiography**, there are 3 typical imaging manifestations of ILS: a solitary nodule or mass, a cystic or multicystic lesion, or consolidation (Fig. 6 on page 13). Approximately 26% of cases have an air-fluid level caused by fistulous bronchial communication. The air-fluid level may reflect chronic inflammation and does not necessarily indicate active infection. Occasionally, the only radiographic finding is a tubular or branching opacity representing the anomalous systemic artery. After antibiotic treatment, the radiographic abnormality may diminish or change significantly, and a small irregular, opaque area or a multicystic lesion may remain\(^5\) (Fig. 7 on page 13).

- **Identifying the anomalous arterial blood supply** of a suspected pulmonary mass is the cardinal aspect of imaging diagnosis. CT and MRI (Fig. 8 on page 14) are both reliable imaging modalities for demonstrating the anomalous artery supplying the ILS; conventional angiography is usually not necessary. In most cases, a single systemic artery arises from the thoracic aorta (mean diameter 6.3mm), often passing through the ipsilateral inferior pulmonary ligament, to supply the ILS (Fig. 9 on page 14 and Fig. 10 on page 15). Occasionally there are multiple systemic arteries supplying the ILS (Fig. 11 on page 16 and Fig. 12 on page 17). The supplying artery may also arise from the abdominal aorta, celiac artery (Fig. 13 on page 18 and Fig. 14 on page 19), splenic artery, or even a coronary artery. Occasionally, the systemic artery is small or absent and not visualized at cross-sectional imaging. Calcification is rarely found in ILS, except in the anomalous artery, where its presence reflects premature
atherosclerosis (Fig. 15 on page 20) Most ILSs (95%) drain into pulmonary veins but have demonstrated drainage into the azygos system. Review of multiplanar reformatted and 3D images, in addition to the axial data set, may improve accuracy in identifying anomalous venous drainage.

**IMAGING OF EXTALOBAR SEQUESTRATION**

- The typical appearance of ELS at *prenatal ultrasound* is a solid, homogeneous, well-circumscribed, hyperechoic mass in the fetal thorax near the medial left costophrenic sulcus. Large lesions are associated with a shift of the mediastinal structures. Cystic areas may be detected within the lesion. ELS may manifest with sonographic findings of polyhydramnios, with or without fetal hydrops. Polyhydramnios may result from esophageal compression and diminished swallowing of amniotic fluid by the fetus or from excessive fluid secretion by the sequestration; fetal hydrops may result from venous compression by the mass, with consequent edema, ascites, and pleural effusion. Rarely the lesion manifests as a homogeneous or cystic mass in the fetal abdomen or retroperitoneum.

- **Radiographically** the lesion most commonly manifests as a well-defined pyramidal, oval, or round mass in the pleural space near the posteromedial aspect of the ipsilateral hemidiaphragm. About two-thirds of ELSs occur in the left hemithorax. ELS uncommonly occurs outside the pleural space in the mediastinum, embedded in the diaphragm, or in the upper abdomen or retroperitoneum. Because of its separate pleural investment from aerated lung, ELS almost never contains air.

- **On CT or MRI**, ELS appears as a well-defined mass of uniform soft-tissue attenuation. There is often a single anomalous artery arising from the thoracic or abdominal aorta. The artery may be not be detectable in cases of infarction. In 15% of cases, the lesion is supplied by smaller arterial branches or multiple arteries. In 80% of cases, venous drainage is to the azygos or hemiazygos vein or to the inferior vena cava.

**TREATMENT OF PULMONARY SEQUESTRATION**

Management of symptomatic or complicated pulmonary sequestration can be *surgical* isolation and division of anomalous systemic feeding arteries, or endovascular with the *embolization* of the aberrant arteries with coils or Vascular Plug, for reducing the complications and risks of surgery (Fig. 16 on page 21).
Images for this section:

**Fig. 6:** Chest radiograph showing a right paravertebral lower lobe mass-like consolidation in a woman with recurrent pulmonary infections.

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**Fig. 7:** Pre-surgical chest radiograph showing a complex left lower lobe mass-like consolidation with cystic/cavitary components, in patient with colon cancer.
Fig. 8: Magnetic resonance cholangiography with incidental finding of a left lower lobe mass, with hyper intense signal in T2w and low enhancement in dynamic sequences.
Fig. 9: Axial contrast-enhanced CT showing a well-defined left lower lobe cystic mass, with a systemic artery that arises from the distal descending thoracic aorta to feed the lesion.

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**Fig. 10**: Volume Rendering (VR) and Maximum Intensity Projection (MIP) reconstructions of CT exam showing the arterial feeder of the left lower lobe cystic mass arising from the distal descending thoracic aorta.

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**Fig. 11:** Axial contrast-enhanced CT showing two aberrant vessels arising from descending thoracic aorta and supplying a left paravertebral lower lobe consolidation.

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Fig. 12: Volume Rendering reconstruction showing two aberrant arterial vessels from descending thoracic aorta.

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**Fig. 13:** Axial contrast-enhanced CT showing a large aberrant arterial vessel arising from celiac trunk and supplying a left paravertebral lower lobe consolidation in patient with recurrent pulmonary infections and emophtoe.

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Fig. 14: 3D-VR reconstruction depicting large arterial vessel from celiac trunk feeding the pulmonary lesion.

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**Fig. 15:** Axial contrast-enhanced CT showing an aberrant vessel with parietal calcification arising from aorta and supplying a left paravertebral lower lobe consolidation in the posterior costophrenic recess

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**Fig. 16:** Selective (A) and superselective (B) DSA of celiac artery showing a large aberrant vessel arising from the celiac trunk feeding the pulmonary lesion; complete occlusion of the aberrant vessel after embolisation with Vascular Plug® (C).

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Conclusion

- Pulmonary sequestration is a mass of lung tissue disconnected from the bronchial tree that receives its blood supply from the systemic circulation. It can be classified as intralobar (ILS) or extralobar (ELS) depending on its relationship to the pleura. Arterial supply is via an anomalous systemic artery, usually of the thoracic or abdominal aorta. ILS usually drains via the pulmonary veins and ELS via the azygos or portal venous system.

- Although angiography remains the gold standard for detecting the anomalous vessels, it is now rarely used for diagnostic purposes only. CT and MR are the mainstay for imaging diagnosis of pulmonary sequestration and typically reveal a homogeneous soft tissue mass often having internal cystic areas that show contrast enhancement at the same time as the aorta. Both techniques usually allow to identify the anomalous arterial supply, but are often unable to show the venous drainage. Therefore, the final classification of a pulmonary sequestration as intralobar or extralobar is often made at surgery.

- Surgical resection is the treatment of choice and often requires ligation of the feeding vessel and lobectomy. To reduce the complications and risks of surgery, an endovascular approach with coils or Vascular Plug embolization is possible.
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