Pulmonary sequestration. Imaging features

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Learning objectives

- To review the pathophysiology of the different type of pulmonary sequestration.
- To explain the imaging features of the pulmonary sequestration in the different radiologic techniques.
- To establish a differential diagnosis between different types of pulmonary sequestration.
- To establish a differential diagnosis between pulmonary sequestration and others pulmonary masses.
Background

PULMONARY SEQUESTRATION is defined as nonfunctioning lung tissue that is not normal continuity with the tracheobronchial tree (and is therefore separate from normal lung) and that derives its blood supply from systemic vessels. Is a rare abnormalities that constitute 0.15% - 6.4% of all congenital pulmonary malformations, reaching 95% of the cases in the left lower lobe. The blood supply usually comes from the descending thoracic aorta, but in about 20% of cases it comes from the upper abdominal aorta, celiac artery, or splenic artery. Pulmonary sequestrations are classified as either intralobar or extralobar.

From January 2008 to September 2013 we reviewed 4 pulmonary sequestrations with a histological confirmation in the University Hospital of Salamanca. We found 3 cases of intralobar sequestration and 1 case of extralobar sequestration.

Intralobar sequestration (ILS)

- Consist of lung tissue that lacks normal communication to the tracheobronchial tree, has systemic arterial supply, and shares the pleura of the parent lobe.
- It accounts for approximately 75% - 85% of all pulmonary sequestrations. Male and female are equally affected.
- Occurs almost always within the lower lobe and slightly more often in the left lung than in the right lung.
- ILS is very rare in infants and is infrequently associated with congenital anomalies, including esophagobronchial diverticula, diaphragmatic hernias, skeletal deformities, cardiovascular defects (including tetralogy of Fallot), renal anomalies, scoliosis, rib abnormalities, and vertebral anomalies.
- The pathogenesis of intralobar sequestration is controversial. Although some of these lesions are congenital, there is evidence to support an acquired origin for the majority of intralobar sequestration. Most cases of intralobar sequestration appear to have an acquired origin, as indicated by substantial evidence. ILS is virtually absent in infants, and associated anomalies are relatively uncommon (6% - 12%), compared with their prevalence (26% - 85%) with other congenital lesions of the chest. 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. The systemic arterial supply may be derived from normally occurring aortic branches within the inferior pulmonary
ligament. These facts argue in favor of an acquired origin for most intralobar sequestration.

- ILS is histologically characterized by extensive fibrosis, chronic inflammation, vascular sclerosis, and cystic changes that largely replace the pulmonary parenchyma. The lesion may sharply abut normal lung parenchyma or blend diffusely with it. The alveoli that border the normal lung parenchyma may be hyperinflated or emphysematous, and bronchiectasis may be present. Elastic and muscular arteries within the lesion may demonstrate thrombosis, endarteritis, medial hypertrophy, and atherosclerosis. Atherosclerosis changes within the aortic branches that supply the sequestration are common, even in children.

- The chief pathologic features are chronic inflammation, cystic change, and fibrosis. The anomalous aortic branches that supply the lesion are characteristically located within the inferior pulmonary ligament. The great majority (95%) of intralobar sequestrations are drained by normal pulmonary veins into the left atrium. The vast majority of cases of intralobar sequestration (98%) are located within the lower lobes, and the lesion is slightly more common (55% - 64%) in the left lung than in the right lung. At gross inspection, the visceral pleura overlying the sequestered segment appears thickened by fibrosis and has multiple irregular, cordlike adhesions to adjacent structures, including the mediastinum, diaphragm, and parietal pleura. Infected purulent material may be found within the cysts. The cystic spaces may be arranged in the configuration of ectatic bronchi, and some authors propose that the cysts develop as a result of persistent mucous secretion into obstructed bronchi. The lesion is typically surrounded by nonsequestered, otherwise normal lung tissue. The incomplete, partially fibrous boundary between sequestered and normal lung tissue has long been considered to be the route of collateral air drift into the lesion.

- ILS rarely manifests in neonates, and associated congenital anomalies are rare. Patients usually present before the age 20 years with recurrent infection. Patients with ILS typically present in early adulthood with a history of chronic cough, mucopurulent sputum production, and recurrent pneumonia that progresses in severity and is usually caused by pyogenic bacteria. Half of all patients reach the age of 20 years before the diagnoses is made. Hemoptysis is a common presenting sign. Chest pain, asthma, or pleuritic pain may also be presenting complaints. A small percentage (15%) of patients may be asymptomatic when the lesion is discovered. Physical findings of ILS are nonspecific and sporadic. In unusual cases, patients may have cyanosis, clubbed fingers, chest wall asymmetry or pectus excavatum. A rare manifestation of ILS in the neonatal period is high out-put congestive heart failure, a hemodynamic state that has been attributed to left-to-left shunting caused by the lesion. The combination of congestive heart failure and pulmonary consolidation in an infant should raise the suspicion of sequestration. A continuous or pansystolic murmur, auscultated over the
anomalous feeding artery, may occur in children and young adults. Massive, spontaneous, nontraumatic pleural hemorrhage, which is attributed to fibrinoid necrosis of the systemic artery, is an extremely rare but potentially fatal complication.

- When ILS appears as a mass at the inferior paravertebral aspect of the thorax, the differential diagnosis may include neurogenic tumor, lateral thoracic meningocele, extramedullary hematopoiesis, and pleural tumor. A cavitary ILS can mimic the appearance of a lung abscess, necrotizing pneumonia, fungal or mycobacterial pneumonia, cavitating neoplasm, and empyema. The predominantly cystic form of ILS has broad differential diagnosis that includes pulmonary abscess, empyema, bronchiectasis, emphysema, bronchogenic cyst, foregut cyst, pericardial cyst, eventration of the diaphragm, and congenital cystic adenomatoide malformation.

- Patients are treated with surgical and consist of either segmentectomy or, more commonly, a full lobectomy when chronic infection has obliterated segmental planes. Postoperative complications are uncommon and include hemithorax, empyema, bronchopleural fistula, and bronchopleuralcutaneous fistula. The prognosis following surgical excision of ILS is excellent, and long-term follow-up suggests that these patients do well.

**Extralobar sequestration (ELS)**

- ELS it is an anomaly that consist of pulmonary tissue anatomically separate from normal lung and usually deriving its blood supply from systemic vessels.
- Is a congenital lesion and represents approximately 15% - 25% of all pulmonary sequestrations.
- Occurs more frequently in male than in female patients, with a male - to - female ratio of 4:1.
- Approximately 50% - 65% of patients with extralobar sequestration have associated congenital anomalies. In fact, ELS may be an incidental finding during the surgical correction of another congenital lesion. The most common associated anomaly is congenital diaphragmatic hernia (seen in approximately 20% - 30% of cases). As many as 60% of ELS may be associated with diaphragmatic eventration or paralysis. Pleural effusions, pulmonary hypoplasia. Communication of sequestration with the esophagus or stomach, bronchogenic cystic, pericardial defect, congenital cystic adenomatoide malformation, foregut duplication or diverticulum, ectopic pancreas, vertebral anomalies, and pectus excavatum. Other cardiovascular, genitourinary, and gastrointestinal anomalies also occur.
• ELS accounts for approximately 0.5% - 6% of all congenital lesions of the lung. The normal lung develops from the primitive laryngotracheal groove, which arises on the ventral aspect of the primitive foregut between the 5th and 6th weeks of embryologic development. The lung bud, a ventral outpouching from the laryngotracheal groove, develops and subsequently undergoes multiple divisions to give rise to the tracheobronchial tree. Although there are many theories to explain its origin, ELS is usually thought to result from an anomalous or supernumerary lung bud that derives its blood supply from the primitive splanchnic vessels that surround the foregut. These vessels, which are connected to the primitive dorsal aorta, form the anomalous arterial blood supply to the sequestration. The original connection with the foregut typically involutes but may persist as a fibrous pedicle that accompanies the feeding and draining vessels. In some cases, the original connection to the foregut persists and allows communication with the gastrointestinal tract.

• Microscopically, ELS resembles normal lung except that there is diffuse dilatation of parenchymal structures. The typical findings are dilatation and tortuosity of bronchioles and dilatation alveolar ducts and alveoli. Dilatation of subpleural lymph vessels is seen in greater than 85% of cases and is often associated with dilatation of perivascular and peribronchiolar lymph vessels. A well-formed bronchus is identified in approximately 50% of cases and is typically located at one edge of the lesion. In other cases, there is a rudimentary bronchus with an irregular fibromuscular wall lined by pseudostratified columnar epithelium and poorly formed cartilaginous plates. Congenital cystic adenomatoide malformation type II has been described in 15% - 25% of extralobar sequestrations. ELS is covered by a mesothelial layer overlying thick or thin fibrous connective tissue. Only if the sequestration is intrathoracic can this membrane be called pleura. ELS occurring within the diaphragm lack this mesothelial covering and are surrounded by the diaphragmatic muscle. The arteries supplying ELS demonstrate intimal, medial, and adventitial layers and often show evidence of hypertensive changes including medial hypertrophy. Although normal aortic branches are muscular arteries, the anomalous feeding vessels of extralobar sequestrations have the elastic structure typically found in pulmonary arteries.

• Grossly and microscopically, the lesion resembles lung tissue. It is invested in its own pleura and constitutes an accessory lobe, also called a "Rokitanski lobe". ELS is typically single ovoid, rounded, or pyramidal lesion, either gray, white, or pink, that ranges in size from 3 to 6 cm. However, lesions measuring 0.5 - 15 cm have been describer. By definition, ELS, when in the thorax, has a pleural covering. Its surface is shiny with a smooth or wrinkled texture, and a fine reticular pattern of dilates subpleural lymph vessels is often visible. If infected, the lesion may be adherent to adjacent structures such as the lung, mediastinum, or diaphragm. ELS with foregut
communication are joined to the esophagus or stomach by a thick fibrous stalk. ELS is typically found in the thorax and on the left side (65% - 90% of cases). The typical location is within the pleural space in the posterior costodiaphragmatic sulcus between the diaphragm and the lower lobe. (63& - 77% of cases). The lesions may also found in the mediastinum or within the pericardium. Approximately 10% - 15% of ELS are found within or below the diaphragm. The blood supply of extralobar sequestration is typically from systemic arteries. These arise directly from the thoracic or abdominal aorta in approximately 80% of cases. The feeding vessel is typically single and measures between 0.5 and 2 cm 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 extralobar sequestration is usually systemic (80%), through the azygos system, the hemiazygos system, ir 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.

• The lesion typically manifests in the newborn period or early infancy with symptoms of respiratory distress. Less frequently, patients present in childhood or adulthood. In the majority of the patients (approximately 61%) with extralobar sequestration, the lesion is diagnosed in the first 6 months of life. These infants often have respiratory distress, cyanosis, and feeding difficulties. ELS may manifest in utero, associated with polyhydramnios or fetal hydrops. Patient may also present in childhood or adulthood. Approximately 10% of extralobar sequestrations are found incidentally in asymptomatic individuals. In rare cases, patients may have recurrent pulmonary infection, high output congestive heart failure in infancy, and spontaneous pulmonary or pleural hemorrhage.

• Differential diagnoses: intrathoracic kidney, scimitar syndrome, hepatic herniation through the diaphragm.

• Treatment of extralobar sequestration consists of surgical excision of the sequestered lung. Surgical resection is easily performed because the lesion is distinct and separate from the adjacent normal lung. Delineation of the vascular supply to the lesion may assist the surgeon in the identification and ligation of the anomalous vessels. Prognosis is usually favorable in the absence of associated congenital anomalies, particularly if there is associated pulmonary hypoplasia.
**Fig. 1:** INTRALOBAR SEQUESTRATION (ILS). Graphic shows the morphologic features of intralobar sequestration. Intralobar sequestration is typically a heterogeneous lower lobe lesion with irregular borders that often contains solid and cystic components. The latter may contain air, fluid, and/or air-fluid levels. An anomalous systemic aortic branch courses in the pulmonary ligament to supply the lesion.

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Fig. 2: EXTRALOBAR SEQUESTRATION (ELS). Graphic illustrates the morphologic features of extralobar sequestration characterized by supernumerary lung tissue invested in pleura and located in the left inferior hemithorax. There is no communication with the tracheobronchial tree and the lesion is supplied by the systemic circulation.

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Findings and procedure details

The most representative radiological findings are:

- Persistent left-sided inferior paraspinal mass in patient with history of recurrent pneumonia. (Left lower lobe: 65%, right lower lobe: 35%, mediastinum 14%, intra-abdominal: 10-15%).
- Systemic artery identification from aorta is diagnostic.
- Venous Drainage: 80% systemic venous system (IVC, Azygos, hemiazygos) creating left-to-right shunt.

INTRALOBAR SEQUESTRATION

- Typically appears as a consolidation or mass, with or without cavitation, within a lower lobe. In many cases, cystic change may be present within the affected lobe. Identification of a systemic arterial supply supports the diagnosis.

- **Radiography:** homogeneous consolidation with irregular margins or as uniformly dense mass with smooth or lobulated contours, located in the posterior basal portion of a lower lobe. Focal bronchiectasis, subsegmental atelectasis, decreased lung volume, mediastinal shift, and prominence of the pulmonary hilum have also been described. Pleural effusions rarely occur. ILS is a diagnosis that should always be considered in the setting of recurrent or persistent pneumonia localized to the lower lobe. Although many ILS manifest as parenchymal consolidations as described above, a large number of these lesions contain air. ILS complicated by advanced chronic inflammation may evolve into a predominantly cystic lesion, which typically manifest as closely related "ring" shadows. When cyst contain air-fluid levels or air alone, the cysts have at least partial communication with the tracheobronchial tree. An unusual radiographic manifestation of ILS is localized emphysema without an associated consolidation or mass. In rare cases, punctate or peripheral linear calcifications may be seen within an ILS, but this finding is more readily detected with CT.

- **CT findings:** Homogeneous or heterogeneous soft-tissue mass in the lower lobe or as a heterogeneous consolidation that shares an irregular border with the adjacent lung parenchyma. The lesion is usually located in the posteromedial portion of the lower lobe. Part of the consolidation may be replaced by areas of cavitation containing air or air-fluids levels. The lesion may partially and heterogeneously enhance when contrast material is administered. ILS may be seen on CT as an almost purely cystic mass or as a confluence of multiple thin-walled cyst containing fluid, air-fluid levels, or air alone. The multicystic form can achieve a considerable size.
The bronchovascular bundles of the remaining functional lung may be peripherally displaced by the lesion. Emphysematous changes surrounding the lesion are often seen, a finding interpreted as "air trapping" within the transition zone between the sequestration and normal lung parenchyma. Calcification is an unusual feature, may be diffuse or peripheral, and has been observed within the sequestration and in the anomalous systemic artery. In rare cases, pleural effusion is seen. The anomalous systemic artery that supplies the lesion is visualized in up to 80% of cases after contrast material administration and may be seen in cross section or as an enhancing linear structure adjacent to the aorta in the inferior pulmonary ligament. However, failure to visualize the artery does not exclude the diagnosis of pulmonary sequestration.

EXTRALOBAR SEQUESTRATION

- The typical radiologic finding is a homogeneous soft-tissue mass in the lower hemithorax. However, these lesions can also occur in the mediastinum, within the diaphragm, and, rarely, below the diaphragm. Radiologic diagnosis rests on identification of the systemic vascular supply. Although angiography has been routinely used in the past in evaluating these lesions, other modalities including ultrasound, computed tomography, and magnetic resonance imaging may demonstrate the anomalous feeding and draining vessels.

- **Radiography:** ELS manifest as a single, well-defined, homogeneous, triangular opacity typically located in the lower thorax in close association with the posterior medial hemidiaphragm. ELS has also been described as a small "bump" on the hemidiaphragm or inferior paravertebral region. Because there is no communication with the normal tracheobronchial tree, air bronchograms are absent. The superior border of the lesion typically has well-defined interface with the adjacent lung parenchyma. The lesion may manifest as a mass elsewhere in the thoracic cavity, including the upper thorax, the mediastinum, and the paravertebral region. Rarely, ELS manifest as a subdiaphragmatic soft-tissue mass. Large ELS may produce an opaque hemithorax and may be associated with an ipsilateral pleural effusion. Pleural effusion may result from poor lymphatic drainage through dilated lymph vessels in the lesion. Radiographic manifestations of high output congestive heart failure due to increased blood flow through a sequestration have also been reported. Bronchography demonstrates lack of normal bronchial communication to the sequestered lung. The opacified normal bronchi are typically draped around the lesion.

- **CT findings:** Homogeneous, well-circumscribed mass of solid-tissue attenuation. Emphysematous changes of the adjacent nonsequestered lung
have been described. CT may demonstrate cystic areas within an extralobar sequestration. The anomalous systemic artery is visualized less frequently.

MR in PULMONARY SEQUESTRATION

- Excellent depiction of complex lung lesion (Cystic portions have variable signal manifestations. Hemorrhage)
- May demonstrate abnormal systemic arterial supply.
Fig. 3: INTRALOBAR SEQUESTRATION. Posteroanterior chest radiograph of a 41-year-old man with recurrent pulmonary infection. Shows a complex left lower lobe mass-like consolidation with cystic/cavitary components and intrinsic air-fluid levels.

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**Fig. 4:** Lateral chest radiograph of a 41-year-old man with intralobar sequestration. Shows a consolidation with cystic/cavitary component in the posterior segment of the left lower lobe. The lesion did not resolve with antibiotic treatment.

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**Fig. 5:** Axial contrast-enhanced computed tomography (mediastinal window) shows a well-defined left lower lobe heterogeneous consolidation, with a systemic artery that arises from the distal descending thoracic aorta to supply the lesion.
Fig. 6: Axial contrast-enhanced computed tomography (lung window) shows a left lower lobe heterogeneous lesion with a cystic component. The lesion is supplied by an anomalous systemic artery arising from the descending aorta.

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Fig. 7: Coronal oblique contrast-enhanced computed tomography reconstruction (mediastinal window). Shows a left lower lobe heterogeneous consolidation, with a systemic artery that arises from the distal descending thoracic aorta to supply the lesion.

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Fig. 8: Coronal contrast-enhanced computed tomography (mediastinal window) shows a well-defined right lower lobe heterogeneous consolidation.

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Conclusion

• The pulmonary sequestrations are a rare lung malformation, but it has to be considered in the presence of recurrent pneumonia or persisting consolidation images, especially if involving the lower lobe of the left lung.
• Multidetector CT angiography and magnetic resonance imaging, are very useful methods for demonstrating the presence of the lesion and the location of nutritional blood vessel.
• The pulmonary sequestrations have an excellent postoperative prognosis.
References


