Lung sequestration and Scimitar syndrome

Imaging approaches

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Pulmonary sequestration

- Pulmonary sequestration (PS) is defined as a segment of lung parenchyma separated from the bronchial tree.
- The affected lung parenchyma receives its blood supply from anomalous systemic arteries, usually the abdominal or thoracic aorta.
- The venous drainage of the sequestrated lung is via azygous system, pulmonary veins or inferior vena cava.
- Usually there is no communication between the sequester and tracheobronchial tree.
- PS is divided into two types:
  1. Extralobar
  2. Intralobar
Extralobar sequestration (ELS)

- ELS is a mass of abnormal lung tissue that is surrounded by its own separate pleura.
- It is located in posterior lower chest, 90% on the left side.
- Association with congenital hernia, congenital heart diseases and adenoid cystic malformation are reported.
- Arterial supply from systemic circulation, venous drainage in azygos.
Intralobar sequestration (ILS)

- ILS is contained within the lung and has a visceral pleura covering.
- It is ultimately connected to adjacent lung
- 98% is located within the lower lobe.
- Usually contains air or air-fluid-levels when infection is present.
- The sharing of pleural covering with normal lung and venous draining in pulmonary vein is typical.
Diagnostic approaches

- PS is seen on prenatal sonographam as a hyperechoic mass but can disappear on chestfilm in postnatal period.
- Color Doppler sonography is useful for demonstrating anomalous vessels to trace them to their origin.
- Extralobular PS manifests itself as congenital mass on chestfilm or sonography.
- Intralobar PS on chestfilm can be recognized as a soft tissue mass with or without cystic or fluid levels following recurrent infections.
- The abnormal vascularity of PS is easily demonstrated by CT or MRI, however the CTA is superior to other imaging procedures.
Diagnostic approaches

- In the differential diagnosis of lung sequestration the following other congenital anomalies should be considered.
  1. Congenital cystic adenomatoid malformation
  2. Bronchogenic cysts
  3. Scimitar syndrome

- The vascularity of Scimitar syndrome and PS are nearly similar. Therefore both anomalies are discussed in this presentation.
This presentation includes 14 cases of pulmonary sequestration and 11 cases of Scimitar syndrome, retrospectively analyzed.
Diagnostic procedure in 14 patients with pulmonary sequestration

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Count</th>
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<tbody>
<tr>
<td>Chest X-ray film</td>
<td>14</td>
</tr>
<tr>
<td>Sonography</td>
<td>6</td>
</tr>
<tr>
<td>CT</td>
<td>5 (including 2 cases with CTA)</td>
</tr>
<tr>
<td>MRI</td>
<td>2</td>
</tr>
<tr>
<td>Arterial angiography</td>
<td>2</td>
</tr>
<tr>
<td>Intravenous bolus angiography</td>
<td>5</td>
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Intrathoracic mass in a neonate on chestfilm.

Ultrasound with abnormal vascularity of the mass.

Multidetector CTA shows clearly an extralobar sequestration in the lower part of the left hemithorax.
A neonate with ventricular septum defect and persistent pulmonary infection.

On chest film no pulmonary infiltration is recognized.

CTA demonstrated an intralobular right-sided sequestration.
A 2-months old infant with a left to right shunt.
Chestfilm shows hypervascularity in right paracardial space.
Conventional angiography demonstrated an intralobar sequestration treated by coil occlusion.
11-year old girl with recurrent pulmonary infection in lower lobe. On sonogram pathological vascularisation of lower lobe.

Intralobar sequestration demonstrated by intravenous bolus angiogram.
10-year old girl with recurrent right-sided pulmonary infection. Chestfilm shows consolidation, cystic changes and fluid-levels in different periods. Lung scintigraphy with perfusion defect in paracardial space. Intravenous bolus angiogram shows intralobar sequestration.
A neonate with respiratory insufficiency and right-sided intrathoracic mass on chest film.

Sonogram demonstrated a large mass with prominent central artery.
Umbilical bolus angiogram shows abnormal artery arising from aorta.

MRI shows duplicated trachea belonging to the mass.

The whole finding is interpreted as an extralobular sequestration connected with the duplicated trachea.
2-year-old boy with recurrent infection and vomiting. Chestfilm shows paracardial cystic changes. Esophagram shows duplicated intrathoracic esophagus. Intravenous bolus angiogram demonstrated an extralobar sequestration connected with duplicated esophagus.
Scimitar syndrome

- Partial anomalous pulmonary venous return
- Hypogenetic lung (lobular agenesis, aplasia or hypoplasia); always right-sided
- Absence of pulmonary artery, systemic arterialization of lung
- Pulmonary sequestration
- Absence of inferior vena cava
- Accessory diaphragm
Scimitar syndrome

- The clinical manifestation and prognosis of scimitar syndrome depends on the amount of the resulting left/right shunt.
- 40% of the patients are asymptomatic.
- In unrecognized cases the clinical symptoms usually manifest in the second to third decade of life with fatigue and dyspnea.
Scimitar syndrome

- The diagnosis is based on chest radiograph, the venous anomaly may be visible with a hypoventilated right lung.
- Doppler sonography may document the site of entry of anomalous pulmonary vein.
- CTA and MRA both can detect the vascular anatomy and bronchial abnormalities.
- Cardioangiography is indicated in cases with a large shunt and additional heart anomalies with the possibility of venous occlusion procedure.
Diagnostic procedures in 11 cases with Scimitar syndrome

<table>
<thead>
<tr>
<th>Procedure</th>
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<td>Chestfilms</td>
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<td>Ultrasound</td>
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<td>CT</td>
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<td>MRI</td>
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<td>Conventional angiography</td>
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<tr>
<td>Intravenous bolus angiography</td>
<td>1</td>
</tr>
<tr>
<td>Cardioangiography with coil occlusion</td>
<td>2</td>
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Arterial supply via systemic arteries (aorta) was found in 5 cases.
4-months old boy with dyspnea.

Abnormal pulmonary vein suspected on sonogram.

The scimitar vein is demonstrated on chestfilm as well as on cardioangiogram.
6-month-old girl with Scimitar syndrome and arterial septum defect.

Scimitar vein is visible on chestfilm and CT angiogram.

Cardioangiography is performed with occlusion of scimitar vein.
10-year-old boy with bronchitis.
The scimitar vein is demonstrated on MRI.
Arterial supply from systemic artery was seen during surgery.
Example of adult type of Scimitar syndrome.

14-year-old girl with recurrent pulmonary infection.

Scimitar vein is easily recognized on chestfilm.

Note the small right hemithorax.
Conclusion I

- Any persisting posterior medial basal basal consolidation should raise the suspicion of sequestrated segment.
- An extralobar sequestrated lung manifest itself mostly in neonates as nonaerated intrathoracic mass.
- Nowadays most fetal lung abnormalities, such as sequestration are detected prenatally by sonography.
- Sequestration may incidentally communicate with the gastrointestinal tract.
Conclusion II

- Multidetector CTA is superior to all other modalities.
- Right-sided hypogenetic lung with dextroposition of heart on chestfilm is highly suspected for Scimitar syndrome.
- Cardioangiography is the modality of choice in Scimitar syndrome.