Congenital lung cyst: from antenatal to post natal period

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Aims and objectives

Fetal lung malformations are rare. They are dominated by congenital cystic adenomatoid malformation (CCAM), pulmonary sequestration (PS) congenital lobar emphysema (CLE) and bronchogenic cyst (BC). Currently through imaging development of US and MRI, the diagnosis became easy and precocious from the antenatal period.

Through this work the main objectives are to demonstrate the role of antenatal imaging, based on ultrasound and MRI, in the diagnosis of cystic lesions of lung and to illustrate the different radiologic features of this lesions in prenatal and postnatal period.
This work is elaborated from a retrospective study of 40 children. Five of them were diagnosed in antenatal period. Other were consulting for various clinical symptoms: 45,7% for recurrent bronchopneumopathy, 31,4% for dyspnea, 17,14% for neonatal respiratory distress and 5,7% for hemoptysis

Ultrasonography and MRI were used in antenatal cases. Chest x-ray, ultrasound and MDCT were realized in postnatal period.
Results

There were twelve cases of Congenital pulmonary airway malformation (CPAM), formerly known as congenital cystic adenomatoid malformation (CCAM), eight cases of pulmonary sequestration (PS), six cases of bronchogenic cyst and fourteen case of lobar emphysema. Antenatal diagnosis was made in five cases of CCAM and two cases of PS. Prenatal imaging found echogenic lesion in three cases.

Congenital lung cystic malformations are various. Each one of them is characterized by a typical imaging feature which must be known for the diagnosis. The association is possible.

1/ Congenital cystic adenomatous malformation of the lung (CCAM):

It represents the most common developmental lung malformation diagnosed prenatally which accounts for 50% to 75% of detected fetal lung abnormalities. This increased prenatal diagnosis is a result of improvement of US equipment in recent years.

CCAM is considered as a heterogeneous group of cystic and noncystic lung lesions that largely result from early airway maldevelopment.

Classification schemes for CCAM have evolved, and there are currently five main types, which differ based on the embryologic level of origin and the histologic features:

Type 0: CCAM is the rarest form and arises from the trachea or bronchus. The presentation is severe and usually lethal. Cysts are small.

Type 1: CCAM is the most common form, representing 50% to 70% of cases, and it arises from the distal bronchus or proximal bronchiole. There are usually a small number of large echolucent cysts, measuring 3 to 10 cm. Because these CCAM may be large, they may have significant mass effect, which can lead to hydrops.

Type 2: CCAM account for 15% to 30% of cases and arise from terminal bronchioles. They are composed of smaller cysts, measuring 0.5 to 2 cm, as well as solid areas that may be difficult to distinguish from surrounding tissue.

Type 3: CCAM account for 5% to 10% of cases and are thought to arise from acinar-like tissue. They are composed of cysts that are so small the mass appears to be solid.

Type 4: CCAM account for 5% to 15% of cases. These CCAM contain large cysts that may be as large as 10 cm and have been associated with malignancy, specifically pleuropulmonary blastoma. They are alveolar in origin.
**Imaging features:**

Antenatally, CCAM have been classified as microcystic (< 5 mm) versus macrocystic (> 5 mm). Microcystic lesions are frequently significantly larger than macrocystic lesions and as such have been associated with poorer prognosis.

Types 1, 2, and 4 CCAM are classified as macrocystic or both macrocystic and microcystic. Type 3 CCAM are microcystic.

- **Antenatal:**
  - *Ultrasound:*

    Fetal US of a large cyst CCAM demonstrates numerous variable-sized anechoic spaces intermixed with echogenic soft tissue.

    With microcystic type, classic signs include echogenic unilateral, well-defined, solid lung lesion (fig1). Doppler study shows no feeding arterial vessels arising from the systemic circulation.

    A CCAM is usually unilateral and unilobar with a slight predilection of lower lobes of the lung. The mass is usually detected in the second trimester.

    The study should include fetal echocardiography to screen for potential congenital heart malformations and comprehensive US evaluation to rule out associated anomalies and evaluate signs of fetal hydrops and early manifestations of cardiac failure.

  - *MRI:*

    At fetal MR imaging, large cyst CCAM manifest as hyperintense unilocular or multilocular lesions with discrete walls on T2-weighted images with normal adjacent lung parenchyma (fig2).

- **Postnatal:**
  - *Chest X-Ray:*

    Postnatal radiography shows variable density in the region of the mass depending on the fluid contents of the cysts, and, possibly, mediastinal shift, depending on the size of the CCAM.

    A large cyst CCAM may be seen as a round soft-tissue mass that gradually becomes filled with air, since there is delayed clearance of fetal lung fluid from the cysts through the abnormal airway. It may be seen as a solitary well-defined air-filled cyst with thin walls or as multiple cysts of varying size (Fig.3). Air-fluid levels may be identified.
CT: It is optimal for detailed depiction of the lung parenchyma and airway, and these lesions will be readily detectable as well-defined air-filled spaces (Fig.4)

3/ Pulmonary sequestration (PS):

PS is the second most common lung lesion detected antenatally.

It is characterized by a portion of lung that does not connect to the tracheobronchial tree and has a systemic arterial supply, usually from the thoracic or abdominal aorta.

Two types of sequestration have been described: intralobar and extralobar.

- The extralobar form, which is the most commonly diagnosed in the prenatal, has its own pleural investment and systemic venous drainage
- The intralobar form shares the pleural investment with the normal lung and usually drains into the pulmonary venous system.

**Imaging features:**

- **Antenatal:**
  - **Ultrasound:**
    
    At prenatal US, PS is seen as a homogeneous hyperechoic mass in a paraspinal location, most often the left lower thorax. The feeding artery originating from the descending aorta may be seen at color Doppler US (Fig.5)

    In some cases of extralobar PS can be situated below the diaphragm, making it difficult to distinguish PS from suprarenal masses, such as mesonephric blastoma or neuroblastoma.

    In cases of an intrathoracic mass, the distinction between PS and the microcystic form of (CCAM) is very difficult especially when aberrant vessels may not be identified at Doppler US.

    **MRI:** Prenatal MR imaging shows a solid, well-defined, uniformly hyperintense mass on T2-weighted images and the feeding artery may be identified.

- **Postnatal:**
  
    - Chest X-Ray: On postnatal radiographs, SP is seen as soft-tissue masse with a smooth or lobulated contour, generally in the lung bases (Fig.6).
• CT: CT scan shows a soft tissue lesion and depicts the feeding artery. It may demonstrate an abnormal venous drainage.

CT of intralobar sequestration may show a homogeneous soft-tissue mass, cysts containing air or fluid, focal emphysema, or a hypervascular focus of lung parenchyma.

4/ Congenital lobar emphysema (CLE):

CLE is characterized by progressive lobar overexpansion, usually with compression of the remaining ipsilateral lung.

The underlying cause can be secondary to an intrinsic cartilaginous abnormality with resultant weak or absent bronchial cartilage or extrinsic compression of an airway. Congenital lobar emphysema is usually diagnosed in the neonatal period with respiratory distress.

The left upper lobe is involved in 42.2% of cases, the right middle lobe in 35.3%, the right upper lobe in 20.7%, and either lower lobe in less than 1%.

CLE may be associated with cardiovascular anomalies in 14% of cases and uncommonly, renal, gastrointestinal, musculoskeletal, and cutaneous malformations.

Often CLE, CPAM, and PS are described as a single clinical group representing a spectrum of the same disease which can decrease in size during pregnancy.

Imaging features:

- Antenatal:
  
  • Ultrasound:

  The main fetal sonographic features of CLE include a bright echogenic lung with or without cystic or mixed cystic lesions without abnormal blood flow. A mediastinal shift, polyhydramnios, and fetal hydrops can also be seen and are predictors of severe respiratory distress or mortality (Fig.7).

  • MRI: Fetal MR imaging may demonstrate a mass with homogeneously high signal intensity on T2-weighted images.

- Postnatal:

  • Chest X-Ray::

Radiography performed during the neonatal period may show a radiodense area in the lung, since the involved section is still filled with fluid. Over time, the fluid will resolve via
the lymphatic and capillary systems. Subsequently, the affected lung will progressively overinflate and become hyperlucent on conventional radiographs (Fig.8)

- CT: it shows an overinflated lung which appears on lung window hyperlucent associated often to a mediastinal shift (Fig.9)

5/ Bronchogenic cyst:

Bronchogenic cysts (BC) are part of the spectrum of foregut duplication cysts. They are resulting from abnormal ventral budding of the tracheobronchial tree, probably occurring between the 26th and 40th days of fetal life.

They are mostly situated in the mediastinum near the carina. Less commonly, they may occur within the lung parenchyma, pleura, or diaphragm

The fluid within bronchogenic cysts is usually a mixture of water and proteinaceous mucus. It is ranged from a thin, watery liquid to hemorrhagic fluid to a very viscous, mucoid material

**Imaging features:**

- **Antenatal:**
  - **Ultrasound:**
    
    At prenatal US, BC manifest as unilocular fluid-filled cysts in the middle or posterior mediastinum. The cyst is often anechogenic but may appear echogenic due to mucous content.

    The differential diagnosis includes esophageal duplication cysts and neurenteric cysts

  - **MRI:**

    BC is seen often as a cystic mass of the posterior mediastinum. On T2-weighted MR images, the cysts typically have high signal intensity equal to or greater than that of cerebrospinal fluid; on T1-weighted images, the signal intensity ranges from low to high depending on the cyst contents.

- **Postnatal:**
  - **Chest X-Ray:**
It shows a medio mediastinal or intra parenchymal mass with sharp contours and hydric tonality (Fig.10)

- **CT:**

On CT scan, BC is sharped margined with smooth or lobulated borders. On non-enhanced CT, the attenuation vary depending on the nature of the fluid. In half of cases, it may have a water attenuation and in other half a soft-tissue attenuation. Calcifications may be seen often peripherally in the cyst wall. Sometimes, milk of calcium is seen in the cyst fluid.

On contrast-enhanced CT, there is no enhancement of cyst contents. Air within the cyst, as well as prominent wall enhancement and thickening, are seen when the cysts are infected, although the presence of air could be related to communication with the airway or gastrointestinal tract.
Fig. 1: fetal US shows an echogenic mass of the left lung causing mediastinal shift with displacement of the heart to the right side of the thorax: CCAM type 3.

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Fig. 2: Sagittal T2 MRI showing an hyperintense solid mass of the fetal left lung

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Fig. 3: Postnatal chest radiograph shows multiple large air-filled thin-walled cysts of the upper right lung (arrows)

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Fig. 4: CT scan shows multiple cysts in the right lung (arrow), at least two of which are larger than 2 cm: CCAM type 1

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Fig. 5: echogenic left lung mass vascularized by an aberrant vessel from the abdominal aorta

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**Fig. 6:** Postnatal chest radiograph shows left basi thoracic opacity (arrow)

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**Fig. 7:** axial fetal US showing an echogenic right lung associated to a congenital right diaphragmatic hernia containing the liver (star)

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Fig. 8: Chest radiograph shows a hyperlucent left hemithorax causing rightward mediastinal shift (arrow)

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Fig. 9: Axial CT scan shows an overdistended left upper lobe with right mediastinal shift

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**Fig. 10:** Chest X-ray showing a well-defined mass of the right upper lobe (arrow)

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Conclusion

The natural history of prenatally diagnosed lung masses is variable. Prenatal identification of lung abnormalities has increased with prenatal surveillance. With the advent of improved antenatal imaging over the past ten years, the diagnosis, assessment and management of congenital cystic lung abnormalities have changed.
References


