

## Antenatal Diagnosis of Cystic Adenomatoid Malformation of the Lung

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### Abstract

The Cystic Adenomatoid Malformation (CCAM) of the lung is rare with a frequency estimated between 1/25 000 and 1/35 000 of pregnancies. It consists of a default of alveoli development associated with an abnormal proliferation of terminal bronchioles giving rise to various sizes of cysts. We report a case of cystic adenomatoid malformation type II of the left lower lobe revealed at 22 weeks of gestation by an acute hydramnios. The fetal karyotype was normal. The presence of fetal anasarca and the severity of the adjacent organs compression have justified the pregnancy interruption which has been strongly recommended by the couple. The autopsy confirmed the diagnostic of cystic adenomatoid malformation associated with a complete hypoplastic right lung.

**Keywords:** Pulmonary malformations; Antenatal diagnosis; Foetopatology

### Introduction

The Cystic Adenomatoid Malformation (CCAM) of the lung is rare. They represent 25% of all congenital pulmonary malformations [1]. It consists of a default of alveoli development associated with abnormal proliferation of terminal bronchioles giving rise to various size cysts. Through a case report we will present the diagnosis and evolutionary aspects of this disease.

### Case

Mrs. MM, 32 years old, with no medical history, was referred to our maternity after the discovery of a systematized hyperechoic image of the left lung on the morphological ultrasound at 22 weeks of amenorrhea. Obstetrical examination was normal except an excessive uterine height for the term. On control ultrasound, the lesion extended to the entire lower lobe. The Pulmonary anomaly contained cystic images. This anomaly was associated with acute hydramnios, mediastinal deviation and flattening of the diaphragm (Figures 1 and 2). The fetus also showed an anasarca. The fetal karyotype performed on amniotic fluid was normal: 46 XX. Maternal  $\alpha$  fetoprotein was elevated to 18 times of the normal. Antenatal MRI confirmed the lung asymmetry development in favor of the left side where we found at the lower part a cystic adenomatoid malformation type II made with multiple peripheral T2 high signals. These abnormalities were associated with a significant mediastinal deviation. The left diaphragmatic dome was atrophied (Figure 3).



**Figure 1:** Transverse section of the thorax, the hemichamps pulmonary hyperechoic left, slack, driving back the heart on the right and a lung contralateral completely hypoplastic.

With regard of the signs of severity (adjacent organs compression, anasarca) and the lack of in-utero treatment possibility of these anomalies, a medical interruption of pregnancy has been decided after a formal parents' consent. It was performed by placing 2 tablets Mesoprostol in *intra cervical*. The result was an expulsion of a female newborn with an Apgar score of 1/1/0 and a 700 g of weight. Fetal examination confirmed the diagnosis by showing a cystic adenomatoid malformation of the left lung lower lobe associated with hypoplasia of the left upper lobe and the whole right lung. There were no other associated abnormalities (Figure 4).

### Discussion

The cystic adenomatoid malformations are rare. They were first described by Ch'in and Tang in 1949 as a "maturation arrest in the broncho-pulmonary developing with a deficient parenchyma



**Figure 2:** Axial cut, hyperechoic lung sits many small lower lobar cysts with diaphragmatic eversion.

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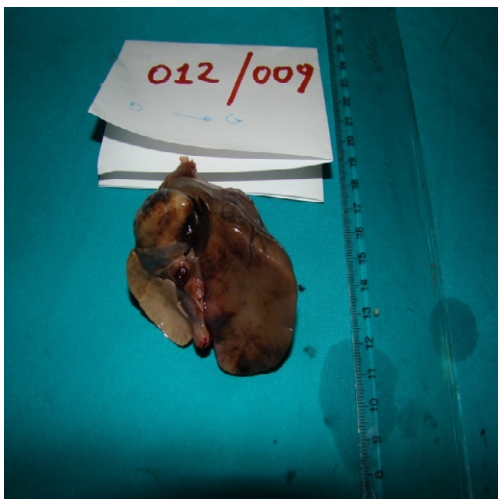
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**Figure 3:** Fetal IRM, cystic adenomatoid malformation type II of the left lower lobe of the lung driven back and horizontalized diaphragm.



**Figure 4:** Autopsy, hypertrophy of the left lung with a right lung completely hypoplastic.

vasculature and a default of proliferation of terminal bronchioles after 15 SA. Their frequency is between 1/25 000 to 1/35 000 of pregnancies [2]. They represent 25% of congenital pulmonary lesions and 71% of lung malformations diagnosed in utero.

This abnormality consists of a cystic distal bronchial and lung expansion communicating with the respiratory tract through abnormal channels. It results from an embryonic development disruption of a lobar or segmental bronchus responsible for a lack of cellular maturation and the formation of an immature broncho-alveolar tissue [3]. This phenomenon can occur at around 15 weeks of gestation; this explains why no diagnosis has been made in the first quarter. In addition, chromosomal abnormalities associated with this malformation have been reported (trisomy 18, trisomy 21). However, there is a fortuitous association.

The pathological diagnosis criteria were defined by Kwitken and Reiner. An anatomical classification was proposed by Stocker. He distinguishes 3 types:

\*Type I: macrocystic: multiple cysts with a diameter greater than 2 cm.

\*Type II: microcystic: multiple cysts of less than 1 cm diameter.

\*Type III: full mass with a voluminous size.

The CCAM is a segmental defect and may affect one or more segments, but it is often located in one lobe (in the left lower lobe classically). It is rarely bilateral [4]. It equally affects both sexes. Our case is female.

The diagnosis of the cystic adenomatoid malformation is made in childhood in 90% of cases on the occasion of a late onset complication which could be fatal. Thanks to advances in ultrasound, antenatal diagnosis of CCAM is made possible from 16-17 weeks of gestation. Since this term, ultrasound may show lung mass composed of multiple anechoic images of various size.

In our patient, an ultrasound done at 16-17 weeks of gestation would be normal. This anatomical classification is not always applicable to ultrasound images of CCAM.

Adzick has proposed an ultrasound classification into two categories [5]:

\*Macrocystic adenomatoid malformation: heterogeneous mass involving a hyperechoic component coexisting with anechoic cystic areas with a diameter greater than or equal to 5 mm.

\*Microcystic adenomatoid malformation: homogeneous limited and systematized mass, hyperechoic with cysts smaller than 5 mm (41% of cases).

Ultrasound remains the cornerstone of antenatal diagnosis of CCAM. These are found mainly at the morphological ultrasound performed after polyhydramnios or anasarca diagnosis [6].

The color-coded Doppler does not find flow within the cysts and do not find either an aberrant vasculature directly from the aorta.

In addition, ultrasound can identify the severity signs such as:

\*A mediastinal compression with deviation or compression of the heart.

\*An eversion of the diaphragm

\*Complications: anasarca, polyhydramnios, and healthy lung hypoplasia.

Finally, ultrasound will be helpful to find other malformations that may be associated [7]: kidney (bilateral renal agenesis, multicystic dysplasia), cardiac (tetralogy of Fallot, VIC), gastrointestinal (duodenal atresia, imperforate anus, omphalocele) or chest (diaphragmatic hernia, pulmonary sequestration). In our case, the ultrasound findings were suggestive of the disease. Moreover, there was a dextrocardia and a pulmonary hypoplasia but no other associated malformation was identified.

The fetal Magnetic Resonance Imaging (MRI) has gradually developed to complement ultrasound in antenatal diagnosis of fetal thoracic malformations in their voluminous and complex forms. Thus, MRI can identify adenomatoid cystic abnormalities and distinguish them from congenital diaphragmatic hernia, but the association of these two lesions is possible. It also allows the assessment of the severity of lesions (content of the malformation and its extension).

In our case, MRI has been helpful to strengthen the diagnosis, to clarify the left upper lobe topography, to identify signs of compression and to eliminate any associated malformation.

The differential diagnosis arises with:

Pulmonary sequestration is the main differential diagnosis. It is characterized by a systemic arterial supply from a branch of the abdominal aorta. The systemic vasculature is visualized with color Doppler ultrasound.

The congenital lobar emphysema: this is an inflation of one or more lung lobes, often in the left upper lobe.

Agenesis pulmonary associated with a complex hypoplasia.

Diaphragmatic hernia, the lack of peristaltic movements orients towards adenomatoid cystic malformation.

The evolution of antenatal CCAM is affected by many factors including:

The anasarca which represents the major complication of this malformation has a frequency that varies from 7 to 33% of cases. In the presence of anasarca, we must examine the fetal brain to search leukomalacia lesions caused by hemodynamic disturbances.

The polyhydramnios associated with the CCAM in 10 to 30% of cases is related to a decrease of fetal swallowing due to esophageal compression with an increase of pulmonary secretions [8].

The healthy pulmonary parenchyma hypoplasia is caused by its compression by adenomatoid cysts. It is suspected when there is a significant mass effect. However, this compression does not necessarily lead to pulmonary hypoplasia [8].

The regression, according to the literature, 6 to 11% of CCAM completely regress in utero [9] and 56% partly regress before birth. It is therefore important to give a period of observation after the diagnosis made.

The fetal and neonatal prognosis depends mainly on the evolution of CCAM in utero. Overall, fetal prognosis at short term is good. In the absence of anasarca and/or extreme prematurity, the survival rate is almost 100% [9]. The Medical interruption of the pregnancy is the leading cause of mortality in fetuses with CCAM [10]. On the other hand, with the presence of anasarca, fetal mortality is very high: nearly 100% of cases in the absence of treatment [10]. The gestational age at diagnosis, the initial size of the lesions and cysts, the CCAM type, the lesions localizations (uni or bilateral) do not influence prognosis.

The antenatal management of cystic adenomatoid malformations includes:

\*A careful morphological assessment to confirm the diagnosis, to search for associated malformations and to assess the impact of the malformation on the fetus.

\*A karyotype is to be done [10].

The antenatal therapeutic indications are not frequent. In the rare cases where complications occur too early during pregnancy to allow the continuation of pregnancy, fetal intervention may be indicated. If cysts are large, they can be emptied by thoracentesis or a thoraco-amniotic shunt; for micro-cystic lesions, an intra-uterine surgery with resection of the mass is the only solution with relative success since that the fetal mortality remains high. A medical interruption of pregnancy can be proposed in presence of associated malformation and/or fetal

anasarca, although observations of spontaneous resolution of anasarca have been observed [7]. Most CCAM do not require intervention before birth.

## Conclusion

The cystic adenomatoid malformations of the lungs are rare. There are three types. Antenatal diagnosis based on prenatal ultrasound at 16 weeks of gestation (30% of cases) has profoundly changed their management. The performance and availability of ultrasound makes the MRI a second-line technique for diagnosis. The prognosis depends basically on evolution in utero. The occurrence of anasarca darkens the prognosis of this disease.

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