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Antenatal Diagnosis of Cystic Adenomatoid Malformation of the Lung

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Abstract:
Introduction: Cystic Adenomatoid Malformation of the Lung (CAML) is a rare developmental anomaly of the respiratory tract accounting for 25% of congenital lung lesions, with a frequency of 1 in 25,000 to 1 in 35,000 pregnancies. Prenatal diagnosis has profoundly transformed the management of these condition allowing for a better proactive monitoring, preventing complications.
Case report: Here, we present a case of antenatal diagnosis of a large CAML in 33-year-old multigravida. Ultrasound imaging as well as a multidisciplinary approach to care, involving maternal–fetal medicine and neonatology, were used from as early as 17 weeks of gestation.
Conclusion: Ultrasound screening has allowed us to have a better understanding of the natural history of especially large cases of CAML as well as a better management of these cases through regular and routine screening.

Keywords: Cystic Adenomatoid Malformation of the Lung, antenatal diagnosis, Ultrasound imaging

Introduction:

Cystic Adenomatoid Malformation of the Lung (CAML), also known as Congenital Pulmonary Airway Malformation (CPAM), is a rare developmental anomaly of the respiratory tract that occurs during an early stage of fetal lung development resulting from an abnormal growth of the terminal bronchial structures with consequent suppression of alveolar growth and formation of intercommunicating cysts[12,13]. CAML is a segmental malformation that can affect one or more pulmonary segments, but most often it is unilobar|6] . It is classically more frequently located on the left side, in the lower lobe. It is rarely bilateral. It communicates with the bronchial tree and is vascularized by the pulmonary circulation. It classically represents 25% of congenital lung lesions[2] and 71% of pulmonary malformations diagnosed in utero and ccording to a Canadian study, the frequency is estimated to be around 1 in 25,000 to 35,000 pregnancies [2].

Prenatal diagnosis has profoundly transformed the management of these condition. Previously, congenital pulmonary airway malformations (CPAM) were only identified when complications arose, which sometimes occurred late. Today, simpler forms are detected early, and affected children are monitored and managed proactively, preventing the onset of such complications.

The objectives of our study, through a clinical case study, are to:

- Describe the ultrasound features of a cystic adenomatoid malformation of the lung.
- Discuss the role of ultrasound in prenatal diagnosis and monitoring.

Case report:

a 33-year-old gravida 3, para 2 woman, with no significant medical history, was referred to our unit by a midwife for obstetric evaluation and ultrasound.

Her first pregnancy in 2012 was uneventful, culminating in a caesarean delivery due to suspicion of fetal macrosomia and breech presentation. The baby was healthy, with a birth weight of 3900 grams.

Her second pregnancy in 2015 was complicated by gestational diabetes, leading to delivery at 38 weeks of gestation for fetal macrosomia in the context of a scarred uterus.

Her current and third pregnancy, was unintended. She did not receive prenatal care during the first trimester. The date of her last menstrual period is uncertain, hence why she was referred by a midwife for an obstetric ultrasound to determine the gestational age.

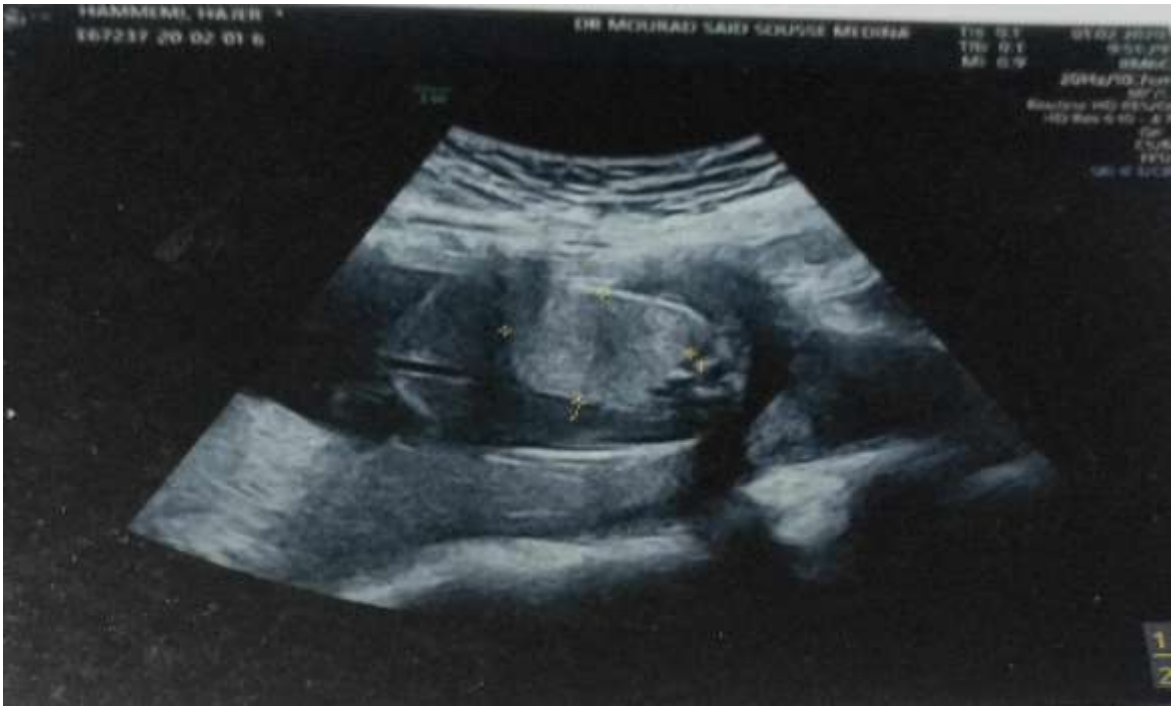
Ultrasound Findings:

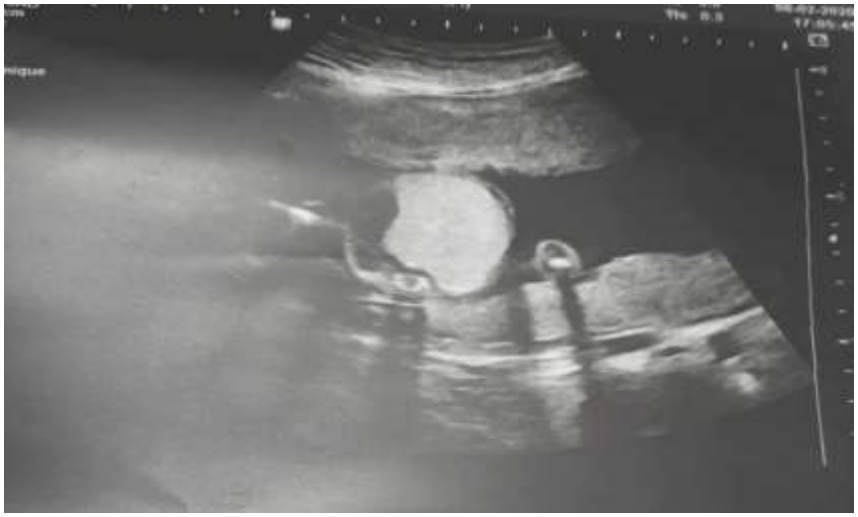
The ultrasound revealed a viable singleton pregnancy at 17 weeks and 1 day. The fetus was male, and the placenta was posterior and fundal. Morphological examination of the brain, abdomen, and all four limbs showed no abnormalities.

However, in the thoracic region, a hyperechoic lesion was shown in the left lung which was significantly enlarged, occupying three-quarters of the thoracic cavity. This mass displaced the heart, mediastinum, and right lung against the thoracic wall. Doppler imaging showed no vascularization within the mass

These findings were consistent with a cystic adenomatoid malformation of the lung (CAML).

The amniotic fluid volume was normal, and there was no evidence of fetal hydrops.





Management:

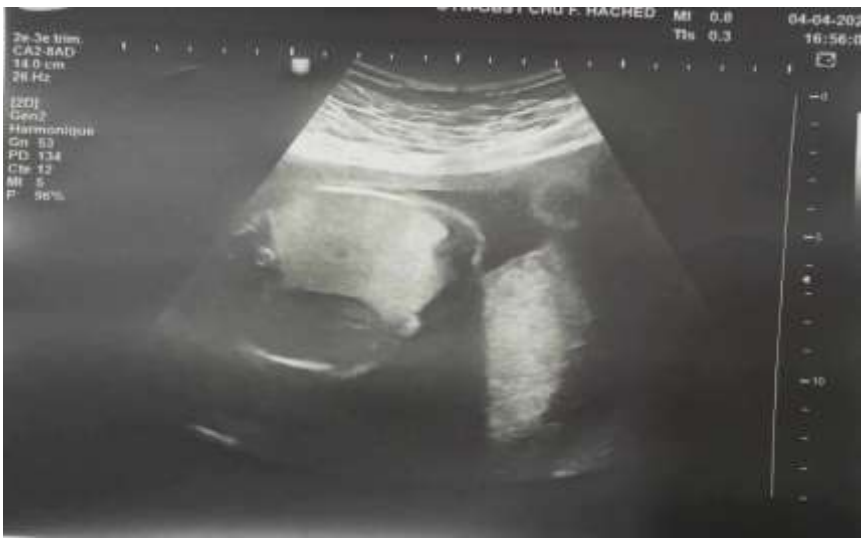
The initial decision was to continue the pregnancy with close monitoring, including biweekly ultrasounds. An amniocentesis combined with a fetal karyotype was performed to rule out chromosomal abnormalities. The fetal karyotype was normal.

At the first follow-up two weeks later, the lung mass remained stable in size and appearance, with no signs of fetal hydrops or polyhydramnios.

Four weeks later, the patient presented with sudden onset dyspnea. On examination, she was hemodynamically stable with an oxygen saturation of 96% on room air. However, the uterine height was significantly increased for the gestational age of 23 weeks and 3 days.

Ultrasound Findings at 23 Weeks:

The ultrasound revealed significant polyhydramnios, with an amniotic fluid index of 21 cm. Fetal findings included skin thickening, ascites, and pleural and pericardial effusions. The left lung mass had increased in size by 1.5 cm compared to the previous scan. A diagnosis of fetal hydrops was confirmed.



Outcome: The patient was admitted to our department, and after consultation with neonatologists, the decision was made to proceed with therapeutic termination of the pregnancy.

Discussion:

The pathogenesis of CAML is still unclear, the formation of this lesion seems to occur at an early stage of lung development as it results from the failure of the pulmonary parenchyma to induce normal bronchoalveolar differentiation. According to Van Leeuwen, this process may occur around 15 weeks of amenorrhea which explains why no diagnosis of CAML is made during the first trimester[3]. Most frequent term of diagnosis appears to be the second trimester and the primary circumstance of discovery is the routine second-trimester ultrasound examination, (although the diagnosis could have been made as early as 16 weeks of gestation). It can also be diagnosed as part of the etiological workup for polyhydramnios (or, more rarely, hydrops fetalis), where the clinical signs would have prompted the performance of an additional ultrasound examination. The ultrasound, according to a classification proposed by Adzick[7], either reveals a homogeneous, hyperechoic intrathoracic mass of variable size (microcystic form) or a heterogeneous mass, combining a hyperechoic component with rounded anechoic areas of varying size and number (macrocystic form). Color Doppler does not detect any flow within these cysts. It also does not reveal any aberrant vascularization directly originating from the aorta[7]. When a diagnosis of CAML is suspected, it is necessary to conduct a thorough morphological assessment, in order to rule out differential diagnoses (particularly temporary atelectasis; Pulmonary sequestration; Diaphragmatic hernia; Bronchogenic cyst..) identify associated malformations (such as renal bilateral renal agenesis, multicystic dysplastic kidney common arterial trunk, ventricular septal defect duodenal/jejunal atresia, jejunal atresia, anal imperforation, omphalocele diaphragmatic hernia, pulmonary sequestration)[8]. —and evaluate the impact of the malformation on the fetus. In case of complications, the ultrasound might find a displacement of the mediastinum with deviation, or even compression of the heart,Eversion of the diaphragm, hydrops fetalis, polyhydramnios, and hypoplasia of the healthy lung. However, fetal growth is usually normal[7]. A prenatal assessment of prognosis can also be partially achieved through ultrasound. Hydrops appears to be the most significant prognostic factor, with reported survival rates , based on large studies , of 0% to 21% for hydropic fetuses compared to 92% for non-hydropic fetuses[15]. Additional prognostic factors have been identified, including mediastinal shift and polyhydramnios, although these seem to be less reliable indicators of outcomes. The size of the mass, particularly when it occupies more than 50% of the thoracic cavity, and pulmonary hypoplasia, also play a role in determining prognosis [11].

A wide spectrum of prenatal evolution is possible for CAML: ranging from spontaneous ``resolution'' leading to asymptomatic newborns at birth, to evolution towards pulmonary hypoplasia and hydrops fetalis. Generally, the favorable prognosis for most cases of CAML diagnosed prenatally is widely recognized. The fetal and long-term prognosis is good in the absence of hydrops[2,6,10]: the survival rate is close to 100% in these fetuses. In the Thorpe-Beeston cases, 4 children (out of 36 whose pregnancy was not terminated) died despite the absence of hydrops: one had associated malformations, and the other three were born prematurely[10]. Medical termination of pregnancy remains the main factor of mortality in fetuses with CAML|16]. However, the clinical course of large CCAMs can be more unpredictable and variable. In the absence of hydrops fetalis, it is always necessary to allow for a period of observation before making any decisions, as some lesions can regress dramatically[9]. Subsequent monitoring should involve regular ultrasounds every 2 to 4 weeks[6,10]. The main challenge lies in managing the risk of

preterm labor . Decompression amniocentesis is indicated in cases of clinically symptomatic severe polyhydramnios. Prenatal therapeutic interventions are relatively uncommon[10]. They are typically considered for fetuses with macrocystic lesions featuring a dominant cyst, those with hydrops fetalis, and cases of severe polyhydramnios. Medical termination of pregnancy may be offered in cases of associated malformations and/or hydrops without the possibility of drainage, although spontaneous resolution of hydrops has been observed in some cases|9]. There is no indication for premature delivery unless signs of decompensation appear and drainage is not possible. Delivery should take place in a maternity hospital equipped with a neonatal intensive care unit.

Postnatally, Rapid surgical intervention is necessary when the newborn presents with respiratory distress (segmentectomy, lobectomy)[16] . In other cases, a postnatal morphological assessment is performed: chest X-ray and thoracic CT scan. If the thoracic CT scan does not reveal any lesions, the child can be monitored clinically. When the CT scan identifies a lesion, the approach is more debated|10]. Some teams advocate for the excision of the lesion, even in the absence of symptoms, before complications such as infection or even malignancy occur. This approach allows us to operate under optimal conditions and on children before the age of 2, during which the healthy lung can still develop to compensate for the tissue loss. even though this approach may also lead to operating on children whose CAML might never have manifested. Other teams only operate on children who have already experienced complications [2].

Conclusion:

Recent progress has been made in understanding CAML. Prenatal ultrasound has enabled a better description of their timing of appearance and progression and has allowed for planning pregnancy monitoring and delivery conditions. As a result, ultrasound screening has allowed us to have a better understanding of the natural history of especially large cases of CAML as well as a better management of these cases by scheduling, in some cases, an early postnatal surgical resection. This strategy, although not universally agreed upon, helps reduce surgical risks and the occurrence of complications.

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