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Antenatal diagnosis and management of congenital cystic adenomatoid malformation

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Summary One of the most enigmatic pulmonary lesions encountered in the prenatal period is the congenital cystic adenomatoid malformation (CCAM). This review presence current thinking on pathogenesis, prenatal assessment, fetal intervention, and management for this pulmonary malformation. Careful delivery planning by utilizing a multidisciplinary approach will optimize neonatal outcomes.

Classification

A congenital cystic adenomatoid malformation (CCAM) is characterized as a bronchopulmonary malformation presenting as an intrapulmonary mass that is usually localized to one lung lobe. A congenital cystic adenomatoid malformations (CCAMs) is characterized by a lack of normal alveoli and an excessive proliferation and cystic dilatation of terminal respiratory bronchioles.[1] Historically, classification schemes for CCAMs reflect the size and histology of the cysts within these lesions. Hence, the initial pathologic classification of CCAMs was based on the size of the cysts and the microscopic appearance of lesions evaluated at autopsy.[1] In this classification scheme, type I lesions are characterized by large cysts of varying sizes (measuring >2 cm in diameter), type II lesions typically contain cysts of a more variable size (≤2 cm in diameter), and type III lesions contain microscopic cysts.

Recently, this classification scheme has been revisited, as several studies suggest that Stocker's classification may not accurately describe the histopathology of CCAMs detected antenatally.[2–6] Analysis of specimens obtained from fetal resections suggests that the antenatal ultrasound features are poorly correlated with the histological findings.[7] Cha et al. identified two histologic patterns of fetal CCAM thought to represent the stage of lung development at which the arrest in pulmonary development occurred.[3] Kreiger et al.[4] proposed a tripartite classification based on the histologic features of the parenchyma between the cysts. The results of these evaluations suggest that there is a histologic difference between CCAMs observed in prenatal versus postnatal life.

Recent studies have also shown that there are hybrid lesions, i.e., masses that not only histologically and

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sonographically appear as CCAMs but also contain a systemic arterial blood supply similar to that of a bronchopulmonary sequestration (BPS).[8,9] The natural history of these intralobar hybrid lesions appears to be dependent on the size of the mass and the resultant physiologic sequelae.[8]

Prenatal assessment

As imaging modalities have become more sophisticated and routine, prenatal identification of these lesions has increased. Once a lung mass is identified on ultrasound, the location, volume, size/appearance (i.e. microcystic versus macrocystic) and blood supply must be evaluated. Additionally, color Doppler should be used to evaluate the origin of the blood supply of the CCAM to exclude the diagnosis of BPS. The initial evaluation should include fetal echocardiography, as there is an increased incidence of structural and functional cardiac anomalies associated with these lesions. It has also been shown that a baseline assessment of cardiac function is useful in monitoring physiologic changes as the pregnancy progresses.[10]

Once the diagnosis of CCAM has been made, careful serial antenatal observation is necessary to monitor the development of hydropic changes in the fetus. Although the large CCAM mass can cause mediastinal shift and esophageal compression, resulting in polyhydramnios, the single best predictor of fetal death is hydrops.[11,12] Depending on the size of the CCAM and associated sequelae, ultrasonographic surveillance should be performed once or twice a week through mid-gestation to monitor the volume changes of the CCAM. Approximately 15% of CCAMs will decrease in size during gestation[2,5,6]; the exact mechanism and reason for the reduction in size is unclear. Peak CCAM growth is expected to occur by 28 weeks, and regression in CCAM volume has been observed in 20% of cases after 29 weeks.[5] In addition to growth, it is also important to monitor the amniotic fluid volume (DVP, AFI), cardiac function, mediastinal shift, measurement of placental thickness (measured at the cord insertion site),[13] and Doppler assessment of the ductus venosus and umbilical vessels for evidence of hemodynamic alterations secondary to the intrathoracic mass effect.

While the majority of fetuses with antenatally detected CCAMs have a good outcome, ongoing surveillance is necessary due to the unpredictable growth patterns for these lesions, and to identify the early occurrence of hydrops. The natural history of CCAMs can be variable, as some lesions increase in size and others will at least partially regress spontaneously. For those lesions that remain small and do not significantly change in size after a 3-week observation period or beyond 28 weeks, expectant obstetrical management and postnatal evaluation and resection are recommended.

To enhance the ability to predict CCAM behavior, we developed a sonographic measurement based on CCAM volume to predict the growth patterns of CCAMs and identify fetuses at risk for progressing to fetal hydrops.[14] The CCAM volume ratio (CVR) is a volumetric ratio based on the elliptical volume of the CCAM [height (cm) $\times$ width (cm) $\times$ depth (cm) $\times$ 0.523 $=$ cm^3] divided by head circumference (cm) for gestational normalization. Specifically, a retrospectively derived CVR $\leq$ 1.6 is correlated with a 94% survival rate and a $<$3% risk of developing hydrops. This study also analyzed gestational changes in CCAM growth and showed that peak CCAM growth occurs at 25 weeks, and that most lesions reach a growth plateau at 28 weeks. An important caveat to the use of CVR is that its predictive utility is most applicable to predominantly solid CCAMs without a dominant cyst. Therefore, the CVR appears to be a useful modality for stratifying patients into categories of high and low risk of hydrops, but is not intended to specifically select fetuses that should have in-utero treatment or surgery prior to developing secondary complications such as hydrops.

Magnetic resonance imaging (MRI; Fig. 1) has been shown to be a useful tool for imaging fetal chest masses, not only for distinguishing CCAMs from other intrathoracic lesions

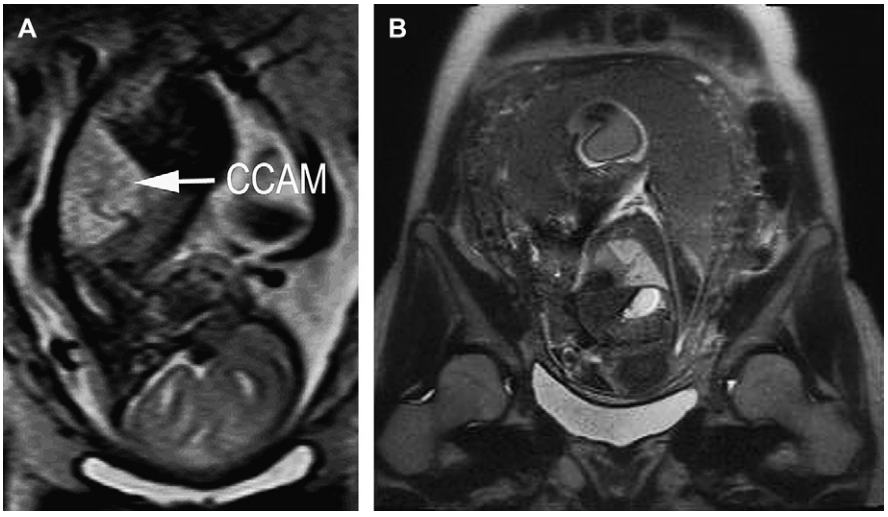

Figure 1 Magnetic resonance imaging (MRI) of (A) a microcystic congenital cystic adenomatoid malformation (CCAM), and (B) a macrocystic CCAM.