Congenital Cystic Adenomatoid Malformation

A Brief Review With Images

LISA MOORE, MD, RDMS*

RACHEL HANDY, RDMS†

Congenital cystic adenomatoid malformation (CCAM) is a rare lung lesion that may be diagnosed antenatally by ultrasonography. It is believed to result from an arrest in lung development. The differential diagnosis of a mass in the fetal thorax includes CCAM, congenital diaphragmatic hernia, and pulmonary sequestration. This brief review discusses classifications of CCAM with images of each type, sonographic findings, factors that affect the prognosis, and important aspects of management once the diagnosis is made.

Key words: CCAM, congenital cystic adenomatoid malformation, thoracic mass

Congenital cystic adenomatoid malformation (CCAM) occurs in approximately 1 in 25,000 pregnancies. The malformation, which most likely occurs between 35 and 55 days of gestation, is caused by excessive proliferation of the terminal bronchioles, which forms cysts in the place of normal alveoli. CCAMs arising from a single lobe of the lung are most common, but up to 15% are either multilobar or bilateral. A genetic cause is not known, although recent research has identified three candidate genes that may contribute to pathogenesis.[1] Slightly more males than females are affected; aneuploidy is rare, but trisomy 13, 18, and 21 have been reported. Between 1% and 15% of CCAMs are associated with additional anomalies.[1,2] The mean gestational age at diagnosis is 22 weeks.[3]

Classification

CCAMs may appear solid or cystic on sonography depending on the size of the cysts in the lesion.

From the *University of New Mexico, Albuquerque, New Mexico, and †New Mexico Sonographics, Albuquerque.

Correspondence: Lisa Moore, MD, University of New Mexico, MSC 10 5580, 1 University of New Mexico, Albuquerque, NM 87131. E-mail: lemoore@salud.unm.edu.

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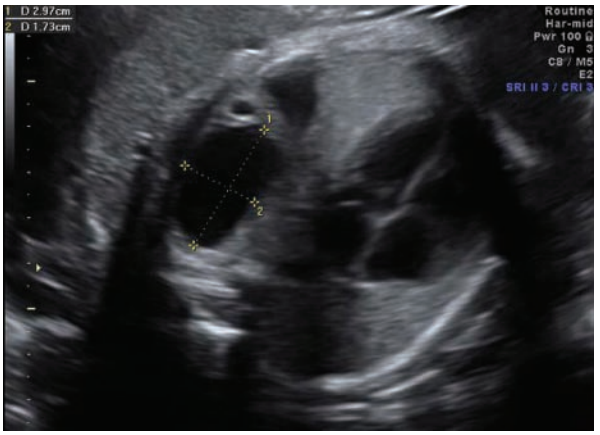

FIGURE 1. Stocker type I congenital cystic adenomatoid malformation. Note the similarity in appearance to a diaphragmatic hernia.

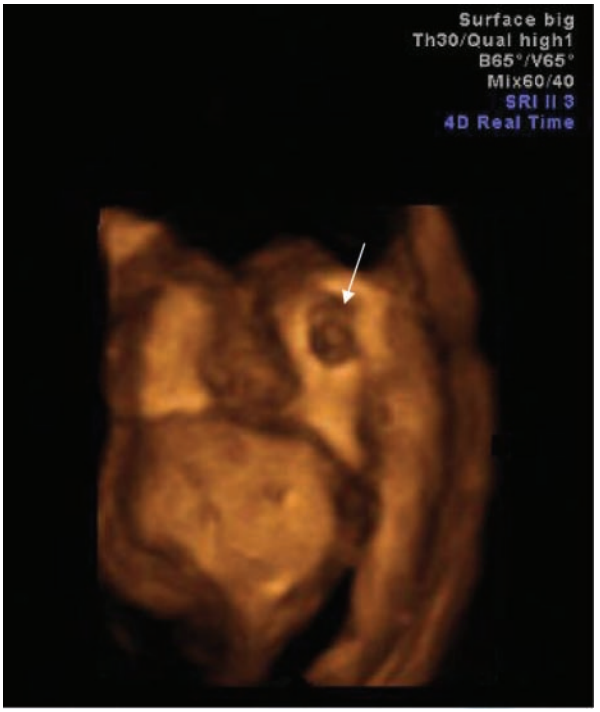

FIGURE 2. A 3D image of Figure 1. The large cyst is noted in the left lung.

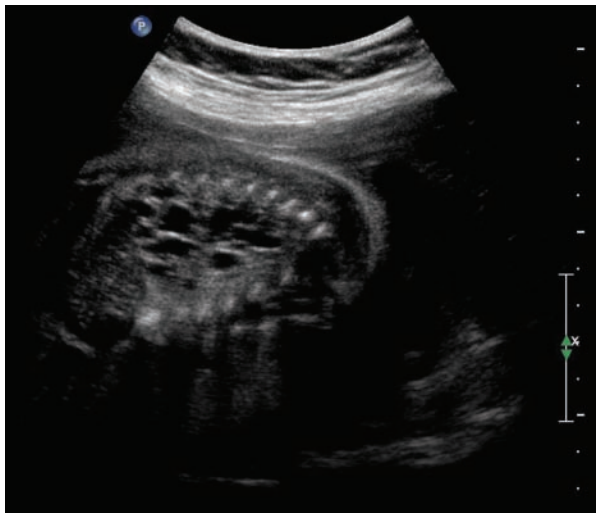

FIGURE 3. Stocker type II congenital cystic adenomatoid malformation with a predominantly cystic appearance.

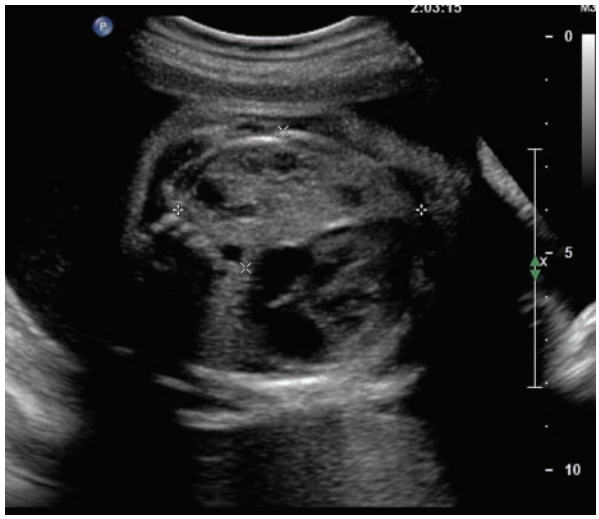

FIGURE 4. Stocker type II congenital cystic adenomatoid malformation with mixed solid and cystic components. Note the echogenic parenchyma between the cysts.

The Stocker classification is the most commonly used method of categorization and is based on cyst size.[4] In Stocker type I CCAM, the most common type, there are multiple large cysts (>2 cm in diameter) or a single large cyst surrounded by several smaller cysts (Figure 1). Figure 2 shows a 3D image of the large cyst shown in Figure 1. In this case, 3D imaging greatly facilitated the diagnosis of CCAM versus congenital diaphragmatic hernia. Type II contains multiple small cysts (<1 cm in diameter) and is found in 40% of cases. Figures 3, 4, and 5 are images of type II CCAMs. Stocker type III occurs in 5% of cases and consists of multiple microcysts, which give a solid appearance on sonograms, as shown in Figure 6.

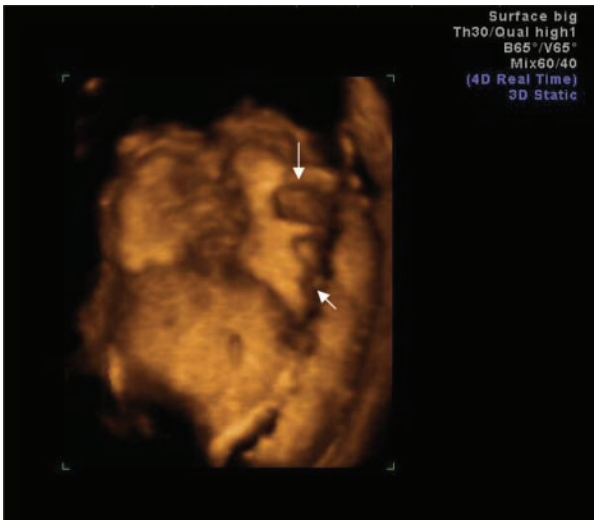

FIGURE 5. Three-dimensional image of type II congenital cystic adenomatoid malformation.

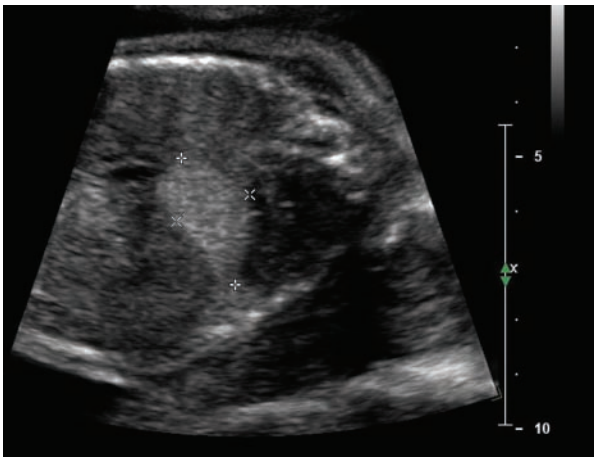

FIGURE 6. Stocker type III congenital cystic adenomatoid malformation.

An alternate simpler classification based on sonographic appearance has been proposed by Adzick and colleagues.[5,6] In the Adzick classification, a CCAM is either macrocystic with a visible cyst of any size or microcystic with a solid echogenic appearance or mixed.

Sonographic Findings

On sonogram, an echogenic mass is seen in the fetal thorax. The mass may or may not contain cysts. The differential diagnosis of a fetal thoracic mass includes pulmonary sequestration and congenital diaphragmatic hernia (CDH). A left-sided CDH usually has herniation of the stomach into the thorax. Figure 1 demonstrates how the two entities may be confused. Locating the stomach in its appropriate location below the heart and the diaphragm helps to make the diagnosis. In a right-sided CDH, the liver is most often herniated into the thorax and can be mistaken for a solid, type III CCAM. Careful attention to anatomic detail by outlining the continuity of the diaphragmatic muscle on the right and the liver located in the appropriate place below the diaphragm may help distinguish between the two types of lesions. Stomach and small bowel may demonstrate peristalsis, which, when noted above the diaphragm, confirms the diagnosis of CDH. A pulmonary sequestration will have systemic arterial blood supply and a wedge-shaped appearance, whereas a CCAM should not. To rule out pulmonary sequestration, look for an arterial blood supply from the aorta to the mass.

In utero, the finding of CCAM may be associated with deviation of the cardiac axis or mediastinal shift if the lesion is large. Polyhydramnios occurs when fetal swallowing is impaired by the lesion. If venous return to the heart is restricted, hydrops may develop.

Prognosis

The prognosis depends most on the size of the lesion rather than the type. A large CCAM may cause pulmonary hypoplasia due to lung compression. Between 43% and 86% of CCAMs will regress or disappear during the course of the pregnancy.[7,8]

The incidence of hydrops associated with CCAM is reported between 12% and 43%.[9] Because hydrops is a harbinger of poor neonatal outcome, several studies have attempted to predict which fetuses will become hydropic. Vu and colleagues[9] identified three characteristics associated with the development of hydrops: a large lesion with a mass-to-thorax ratio of .56 (i.e., the lesion occupied more than 50% of the thorax), mostly cystic, and sonographic evidence of mass effect of the lesion on the fetal diaphragm. Crombleholme et al.[10] developed the congenital

cystic adenomatoid malformation volume ratio (CVR) as a predictor of intrauterine outcomes. Using the maximum measurements in three planes, the CVR is calculated as length × width × height × 0.52/head circumference, where 0.52 is a constant, and dividing by the value of the head circumference compensates for differences in gestational age. A CVR >1.6 predicts an increased risk of developing hydrops. If the CVR is ≤1.6, the risk of developing hydrops in utero is less than 3%. Fifty percent of fetuses who develop hydrops in utero survive.[11] Approximately 95% of fetuses with antenatally diagnosed CCAM are born alive.[5,12,13]

Management

After the diagnosis is made, the patient should be referred to a high-risk specialist for counseling and additional evaluation, which may include an amniocentesis. A fetal echo should be performed to rule out structural cardiac anomalies and to evaluate cardiac function. At the time of diagnosis and at each subsequent visit, the size of the lesion and any cysts should be measured. This will allow an accurate determination of changes to the lesion over time. CCAM may show rapid growth between 20 and 26 weeks. Maximum size is usually reached between 25 and 28 weeks.[14] A general anatomic survey should rule out hydrops at each visit. Stable lesions without mediastinal shift or other evidence of fetal compromise and regressing lesions may be evaluated at 3- to 4-week intervals. Large lesions at risk for the development of hydrops or other complications should be evaluated weekly once a viable gestational age is reached. Magnetic resonance imaging (MRI) with ultrafast imaging sequences is useful in determining lobe localization and the presence of normal compressed lung as well as to differentiate between CDH, CCAM, and pulmonary sequestration.[15] Delivery should be at a tertiary care center. Postnatal evaluation by computed tomography (CT) is recommended even if the lesion resolves in utero.[13]

Conclusion

Congenital cystic adenomatoid malformation is rare, and careful sonographic evaluation allows differentiation of CCAM from other thoracic lesions such as congenital diaphragmatic hernia or pulmonary sequestration. Once diagnosed, a fetal echo and serial sonograms should be performed. Large lesions are more likely to develop complications. Most CCAMs will regress in utero and have an excellent prognosis.

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