Journal of Pediatric Surgery (2007) 42, 404–410

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Journal of Pediatric Surgery

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Large fetal congenital cystic adenomatoid malformations: growth trends and patient survival

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Index words:
Congenital cystic adenomatoid malformation;
Magnetic resonance imaging;
Fetus;
Prenatal diagnosis;
Hydrops;
Ascites;
prognosis

Abstract

Purpose: The prognosis for fetuses with large congenital cystic adenomatoid malformations (CCAMs) remains uncertain. This study examined the natural history of large fetal CCAMs managed expectantly at a major referral center.

Methods: A 5-year retrospective review was conducted on fetuses diagnosed with a thoracic lesion (n = 59). Large CCAMs were identified on prenatal imaging and followed longitudinally. Perinatal outcomes were assessed.

Results: Twelve (20.3%) fetuses had large CCAMs in the absence of other congenital anomalies. Peak CCAM size occurred at 25.3 ± 3.6 weeks' gestation. Serial magnetic resonance volumetry demonstrated a trend toward decreasing CCAM mass volume relative to thoracic cavity volume over time. Overall, 6 patients, including 3 with signs of early hydrops, showed a marked regression of their lesions relative to estimated fetal weight. Five fetuses required an emergent intervention postnatally, including extracorporeal membrane oxygenation support (n = 1), cyst aspiration (n = 1), and lung resection (n = 5). Overall survival was 75%, with severe hydrops before 30 weeks seen in all 3 deaths.

Conclusion: Large fetal CCAMs tend to peak in size at 25 weeks' gestation and are characterized by in utero diminution relative to overall fetal growth. The prognosis for most fetuses with large CCAMs remains quite favorable under careful perinatal management.

Since the widespread use of screening prenatal ultrasonography more than 2 decades ago, the natural history of most fetal congenital cystic adenomatoid malformations (CCAMs) has been elucidated [1-5]. Spontaneous in utero regression of CCAMs is now recognized as a common event, occurring in up to 43% to 86% of lesions based on some of the more recent published series [5-7]. As a result, the majority of prenatally diagnosed CCAMs have good outcomes under a management strategy of routine prenatal observation with early postnatal resection [4,5,8].

Nevertheless, much less is known about the outcomes of fetuses who are initially diagnosed with large CCAMs,

Presented at the British Association of Paediatric Surgeons 53rd Annual International Congress, Stockholm, Sweden, July 18-22, 2006.

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doi:10.1016/j.jpedsurg.2006.10.014

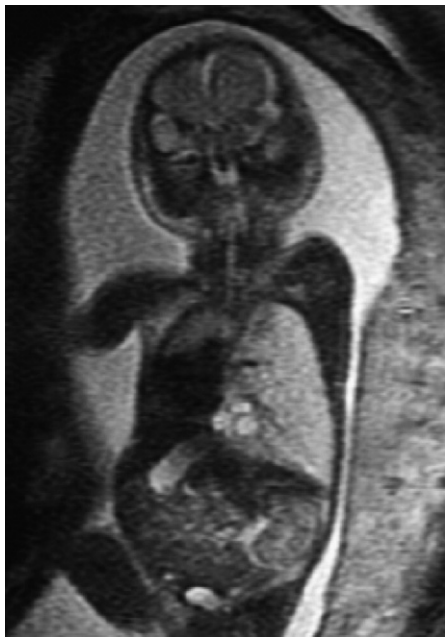

Fig. 1 Coronal fast spin-echo T2-weighted MR image of a 22-week gestation fetus with a large, hyperintense mass consistent with a congenital cystic adenomatoid malformation. Severe mediastinal shift can be appreciated.

defined as lesions that are sizeable enough to cause significant mass effect on adjacent thoracic structures [4,9]. Many fetuses with large CCAMs are believed to be at considerable risk for hydrops and in utero demise [3,10]. To date, however, there remain no widely accepted predictors of outcome for fetal CCAMs except for the presence of hydrops. In the present study, we reviewed our own experience with large fetal CCAMs in an effort to better counsel expectant parents. Specifically, we sought to better define the growth patterns of large CCAMs and to determine overall survival and postnatal outcomes in these patients when managed expectantly at a major fetal care referral center.

1. Materials and methods

A retrospective review of records from all prenatally diagnosed fetal thoracic lesions (n = 59) referred to Children's Hospital Boston between October 1, 1999, and May 31, 2005, was conducted. The maternal-fetal data obtained for this study were collected from a prospectively maintained database under an approved institutional review board protocol.

After a thorough evaluation of all imaging performed by the referring institution, a repeat fetal ultrasound with color Doppler interrogation (ATL 5000, Philips, Bothell, Wash, or Sequoia, Siemens, Mountain View, Calif) was performed in our Advanced Fetal Care Center clinic. In each study, mass location, homogeneity, and overall dimensions were assessed based on methods as previously described [11]. The degree of mediastinal shift, distortion of the hemidiaphragm, and hydrops were also noted. Early hydrops was defined as the presence of fluid in the abdominal and/or pleural cavity. Severe hydrops was defined as the abnormal

Table 1 Baseline characteristics and outcomes of large fetal CCAMs

Case	Location	Stocker	Severe midline shift	Diaphragm distortion	CVR	Hydrops	Follow-up scans	Pathology	Outcome
1	Right	III	No	Yes	1.7	Ascites	Severe hydrops by 24 wk	N/A	Fetal demise
2	Left lower	II	Yes	Yes	1.0	None	Massive growth then plateau phase at 24 wk	Mixed	ECMO, alive
3	Right lower	I	Yes	No	2.3	None	Stable lesion	CCAM	Alive
4	Left lower	II	No	No	1.6	Effusion	Effusion resolved by 33 wk	Mixed	Alive
5	Right lower	II	Yes	Yes	3.2	Ascites	Ascites resolved by 34 wk	Mixed	Alive
6	Right upper	II	Yes	No	2.4	None	Regression by 34 wk	CCAM	Alive
7	Left upper	I	Yes	Yes	2.2	None	Stable at 29 wk	CCAM	Preterm labor, alive
8	Right lower	III	No	Yes	0.8	None	Regression by 29 wk	Mixed	Alive
9	Right	II	No	Yes	2.8	Ascites	Severe hydrops	N/A	Fetal demise
10	Right lower	II	No	Yes	1.0	None	Regression by 35 wk	Mixed	Alive
11	Right upper	II	Yes	No	3.1	Ascites	Stable lesion with severe ascites	CCAM	Preterm labor, neonatal demise
12	Left upper	III	No	No	1.0	Ascites	Massive growth by 24 wk, regression by 30 wk	CCAM	Alive

N/A indicates not available; mixed, combined CCAM and bronchopulmonary sequestration; ECMO, extracorporeal membrane oxygenation; CVR, cystic adenomatoid volume ratio.