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Antenatal Ultrasound Findings in Cystic Adenomatoid Malformation

S. M. Donn[1], J. N. Martin, Jr.[2], and Susan J. White[3]

Departments of [1]Pediatrics and Communicable Diseases, Division of Newborn Services, [2]Obstetrics and Gynecology, Division of Maternal-Fetal Medicine, and [3]Radiology, University of Michigan Medical Center, Ann Arbor, Michigan, USA

Abstract. A case of congenital cystic adenomatoid malformation of the lung is presented with prenatal ultrasound findings. The present case appears to be the third instance reported in which the diagnosis was made antenatally by ultrasound, and the first which resulted in a live birth, although the patient succumbed to pulmonary hypoplasia.

Key words: Cystic adenomatoid malformation (CAM) – Ultrasound – Hydramnios

Cystic adenomatoid malformation of the lung (CAM) is a rare congenital anomaly which often results in severe respiratory distress in the newborn. Acute maternal hydramnios is the most common initial clinical event which can alert the obstetrician to the possible occurrence of this potentially treatable condition. We wish to report a case in which the diagnosis was made antenatally, based on the maternal clinical course and ultrasonographic findings.

Case Report

A 20 year-old white primigravida was admitted to Women's Hospital on November 26, 1979 with acute hydramnios. Her initial ultrasound examination on November 20 had revealed a single fetus, hydramnios, and possible fetal hydrothorax and ascites. Gestational age was estimated at 27 weeks, consistent with the previously estimated date of confinement. Significant laboratory findings included maternal blood type AB+ with negative antibody screen, nonreactive serology for syphilis, and negative liver function and coagulation studies. A repeat sonogram (Fig. 1) disclosed a single fetus in the vertex position, biparietal diameter of 72 mm (consistent with 29 weeks gestation), marked hydramnios, fetal ascites, and what appeared to be a cystic lesion in the right fetal hemithorax. A realtime ultrasound sector scan confirmed the mass

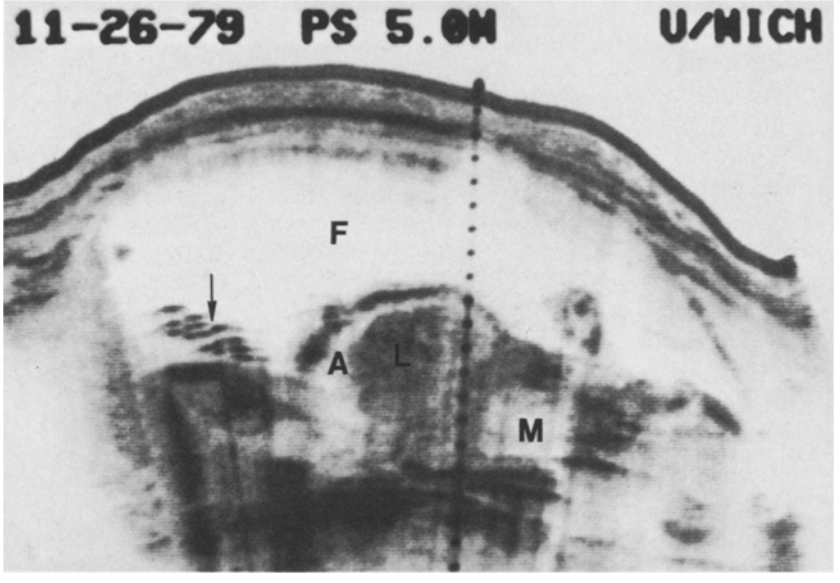

Fig. 1. Longitudinal sonographic scan of gravid uterus at 29 weeks gestation. Note fetal ascites (A) and cystic mass (M) in the fetal chest. Marked hydramnios (F) is present. (L) indicates fetal liver. Arrow indicates umbilical cord

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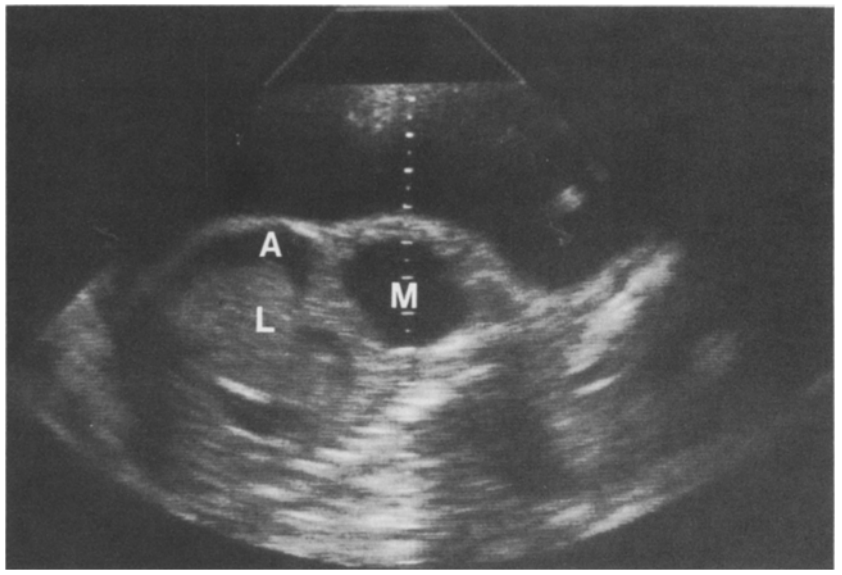

Fig. 2. Real-time sector scan shows fetal liver (L), fetal ascites (A), and cystic mass (M) in the fetal chest

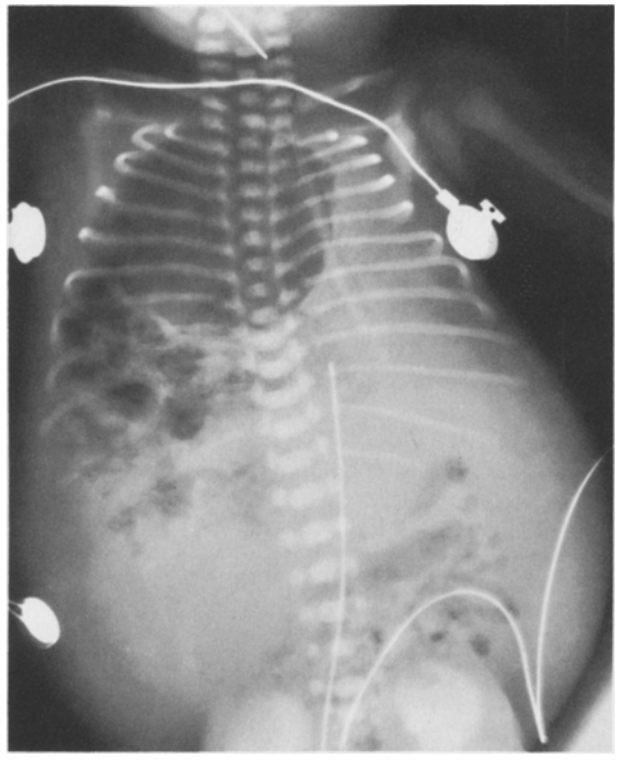

Fig. 3. Antero-posterior radiograph of the infant's chest and abdomen at one hour of age demonstrates large lucent mass in the right hemithorax with shift of mediastinal structures to the left. Ascites is evident

to be distinct from the fetal heart and non-pulsatile (Fig. 2). Initial diagnostic considerations had included non-isoimmune hydrops fetalis, diaphragmatic hernia, and cystic adenomatoid malformation of the lung (CAM). The latter was favored because of the association of hydramnios, fetal ascites, and the presence of the lesion in the right hemithorax. Spontaneous rupture of the membranes occurred on December 5, and a follow-up sonogram suggested that the mass was enlarging, now occupying nearly one-half of the right thoracic cavity, displacing the heart and mediastinum to the left. This again favored the diagnosis of CAM, where the presence of "mucigenic" cells can give rise to an expansile lesion [5]. Because of the risk of possible body dystocia and fetal trauma, a primary low transverse cesarian section was performed under epidural anesthesia.

An 1180 gram male infant was delivered. The Apgar scores were 1 and 3 at 1 and 5 minutes necessitating endotracheal intubation and ventilation. The physical examination revealed markedly diminished breath sounds over the right hemithorax, with asymmetrical movement of the chest wall. There was pronounced ascites, though the infant was not hydropic.

The initial radiographs of the chest and abdomen (Figs. 3 and 4) showed a large, 4 cm spherical cyst-like structure in the upper right hemithorax with extension across the midline. An air-fluid level was seen within the mass in the cross-table lateral view. Multiple cyst-like lesions resembling air-filled loops of bowel were seen in the lower right chest, and hypoaeration of the left lung was present. Ascites was evident.

The infant underwent immediate surgical exploration. The primary operative finding was a large cystic mass in the right hemithorax, grossly consistent with CAM. A right lower lobectomy with removal of the cystic mass was performed. The infant remained in marked respiratory distress with hypotension, hypothermia, and acidosis and expired on the first post-operative day.

Histologic examination of the operative specimen confirmed the diagnosis of CAM. The surgical specimen of the lung was enlarged to two to three times the normal size. On sectioned surface the normal parenchyma was completely replaced by cysts of varying sizes. Microscopically there were sheets of glandular structures lined by bronchiolar rather than alveolar cells. Additional findings at postmortem examination included marked pulmonary hypoplasia, a 20 ml intraperitoneal hemorrhage, and extensive hemorrhage within the fourth ventricle. Ascites was present. There were no other congenital anomalies.

Discussion

Cystic adenomatoid malformation of the lung is a rare congenital anomaly. However, the association of CAM with maternal hydramnios and fetal anasarca

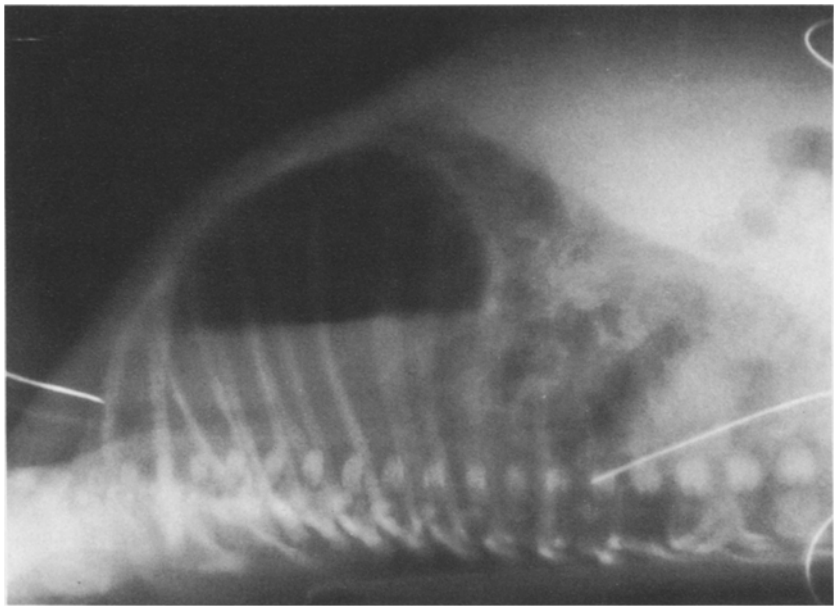

Fig. 4. Cross-table lateral radiograph of the infant's chest and abdomen at one hour of age demonstrates air-fluid level within the mass

has been reported frequently [1, 3, 5]. Östör [5] reported that 80% of affected pregnancies demonstrated acute hydramnios, with the onset usually one to three weeks prior to delivery. Two of ten fetuses were stillborn and the remaining eight infants expired by three hours of age. Though the mechanism responsible for production of hydramnios is unknown, excess amniotic fluid may result from decreased fetal swallowing secondary to esophageal compression; additionally, excessive lung fluid may be produced directly by the abnormal lung tissue [5]. Kohler [3] also points to possible decreased absorption of lung fluid by the hypoplastic, malformed lungs.

Similarly, the mechanism of edema formation in the fetus may be secondary to vascular compression. The expanding pulmonary mass may obstruct venous return and compress the fetal heart leading to decreased myocardial contractility, heart failure, and subsequent edema formation.

Almost all of the infants with CAM will have respiratory distress. According to Madewell et al. [4] about two-thirds of the affected infants have onset of distress on the first day of life, but over one-quarter may remain asymptomatic for a week or longer. Respiratory function is compromised by pulmonary hypoplasia, compression of thoracic viscera and concomitant heart failure (if present). There is a tendency for the cystic mass to expand progressively after birth, leading to air trapping [2]. Immediate surgical decompression is the treatment of choice, and though this has been shown to be well-tolerated [1, 2] ultimate survival depends upon both the extent of pulmonary hypoplasia and the state of maturation of the normal lung tissue. Analogous to the emergency presented by diaphragmatic hernia, immediate and proper surgical intervention might salvage many of the affected infants whose lungs would otherwise support life.

This pulmonary anomaly should be considered by obstetricians and sonographers in the differential diagnosis of acute hydramnios, with ultrasound examination allowing early recognition and adequate preparation for management of affected infants.

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Steven M. Donn, M. D.
Department of Pediatrics and Communicable Diseases
Division of Newborn Services
Box 007, L3018 Women's Hospital
University of Michigan Medical Center
Ann Arbor, MI 48109
USA