J Clin Ultrasound 17:675–677, November/December 1989

Case Report

Prenatal Treatment of Type I Congenital Cystic Adenomatoid Malformation by Intrauterine Fetal Thoracentesis

Clark E. Nugent, MD,* Robert H. Hayashi, MD,* and Jonathan Rubin, MD, PhD†

Congenital cystic adenomatoid malformation of the lung is a developmental abnormality characterized by abnormal proliferation of terminal bronchioles forming cysts of varying sizes. Extensive lesions are associated with a poor prognosis due to the development of nonimmune hydrops and/or pulmonary hypoplasia. The advent of high-resolution ultrasonography has made it possible to identify these lesions during the antenatal period. With the prenatal identification of cystic adenomatoid malformations comes the potential for in utero intervention to modify the natural history of this process and prevent the development of hydrops and pulmonary hypoplasia. An instance of successful intrauterine treatment of congenital cystic adenomatoid malformation is presented.

CASE REPORT

A 32-year-old primagravida was referred at 28 weeks, menstrual age, because of a cystic chest mass noted on ultrasound examination. Ultrasound evaluation revealed a 28-week fetus with a 4 cm × 3.5 cm × 2.5 cm unilocular cystic mass filling the left side of the fetal thorax, displacing the heart into the right hemithorax (Figure 1). The fetal stomach was present in the appropriate location and the diaphragm appeared intact. No evidence of fetal hydrops was identified. Based on the ultrasound findings, congenital cystic adenomatoid malformation was felt to be the most likely diagnosis followed by pulmonary sequestration or bronchogenic cyst.

Given the relative size of the cyst, the fetus was felt to be at significant risk for pulmonary hypoplasia. Likewise, with the degree of mediastinal shift present, there was concern that cardiac function could ultimately be compromised leading to nonimmune hydrops. These concerns were discussed with the parents, who strongly desired that an attempt at intrauterine therapy be made. It was therefore planned to proceed with fetal thoracentesis followed by interval placement of an indwelling shunt only if there was significant fluid reaccumulation.

Cessation of fetal movement was achieved by injecting 0.3 mg of pancuronium bromide into the fetal thigh under ultrasound guidance (Acuson 128). An 18-gauge Tuohy needle was then guided into the center of the cyst. Immediately on removing the stylet, there was brisk flow of clear viscous fluid. After removal of 22 mL of fluid, the cyst had completely disappeared and heart shifted back to the left. An umbilical vein blood sample for cytogenetic analysis was then obtained from the placental insertion of the cord. Fetal heart activity remained stable throughout the procedures.

Analysis of the fluid included a cell count and differential with 545 red blood cells and 355 white blood cells/cmm with the differential showing 13% segs, 36% lymphs, and 51% histiocytes. Protein was less than 1.0 g/dL. Cytologic examination of the fluid revealed numerous histiocytes, karyorrhectic debris, and ciliated columnar epithelial cells. Cytogenetic analysis of the fetal blood sample yielded a normal 46,XY karyotype.

Serial follow-up ultrasound scans were obtained. On the first day following the procedure there was only minimal fluid reaccumulation. The cyst had increased in size to approximately 2 cm four days following the thoracentesis. The cyst exhibited minimal expansion during the re-

From the Departments of *Obstetrics and Gynecology, and †Radiology, University of Michigan Medical School. For reprints contact Clark E. Nugent, MD, Department of Obstetrics and Gynecology, University of Michigan Medical School, L3100 Women's Hospital, Box 0264, Ann Arbor, Michigan 48109-0264.

0091-2751/89/090675-03 $04.00

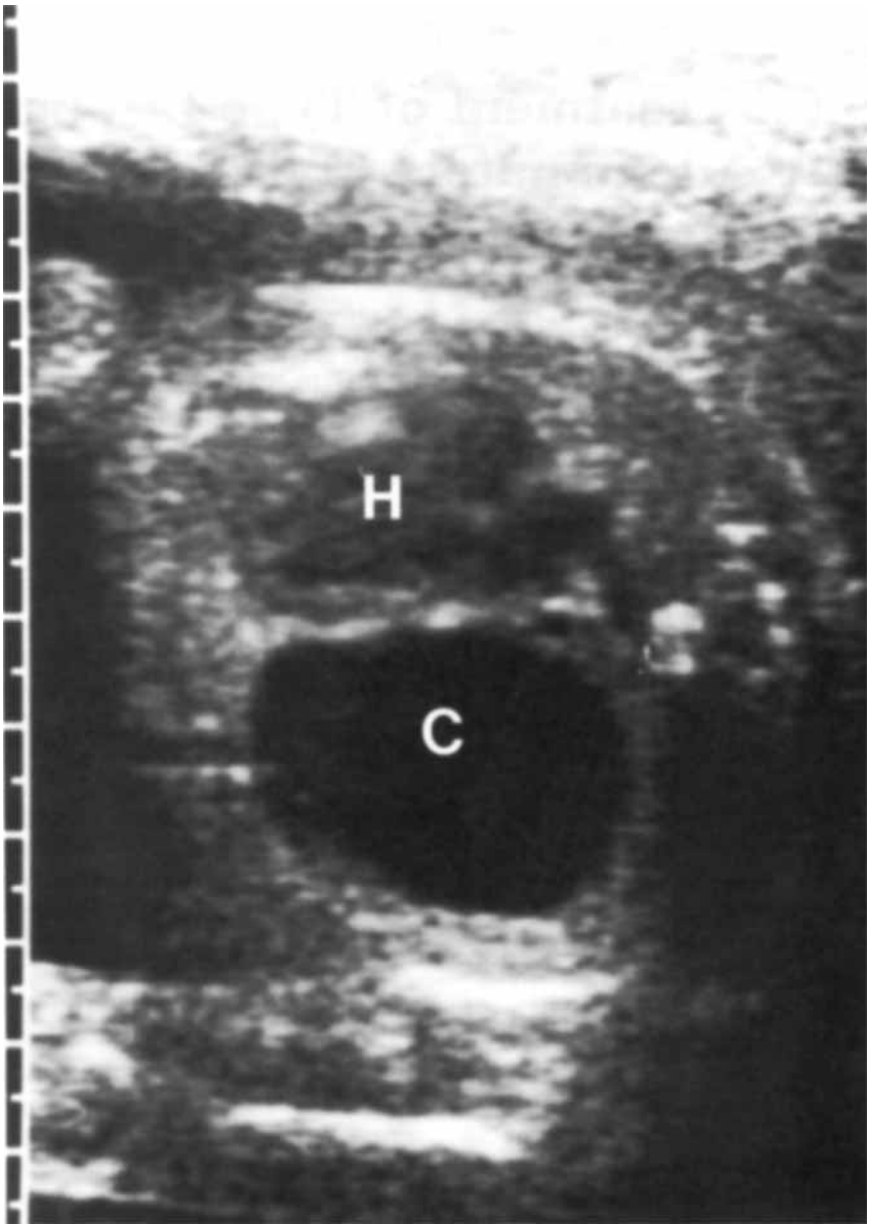

FIGURE 1. Transverse view of the fetal chest. The heart (H) is displaced into the right hemithorax by the cystic adenomatoid malformation (C).

mainder of the pregnancy and actually occupied a relatively smaller proportion of the thorax.

A 3440 g male infant with Apgar scores of 7 at one minute and 9 at five minutes was delivered at 40 weeks. Careful examination revealed no evidence of trauma from the thoracentesis. The infant had no evidence of respiratory compromise in the nursery. Chest radiographs and computed tomography scans verified the presence of a cystic mass in the region of the left upper lobe. A left upper lobectomy was performed on the fourth day of life. The specimen weighed 30 g and contained a 4 cm × 3 cm × 2.5 cm smooth-walled unilocular cyst. Microscopic examination verified the diagnosis of Type I cystic adenomatoid malformation. The infant was discharged home on the seventh postoperative day.

DISCUSSION

Congenital cystic adenomoid malformations have typically been detected in infancy at the time of evaluation of respiratory distress or recurrent respiratory infections. The advent of high-resolution ultrasonography has shifted the ability to diagnose these lesions into the antena-

tal period with the potential for intrauterine intervention. Although fetuses with macrocystic lesions have a more favorable prognosis than those with microcystic lesions,[1] there is currently no reliable marker to indicate which fetuses will be benefited by in utero decompression (short of those instances in which hydropic change is present).

A review of the literature yielded 16 other reported instances of antenatally detected macrocystic congenital cystic adenomatoid malformations with 12 surviving infants. In three instances, intrauterine drainage procedures were performed with favorable outcomes. Adzick et al.[1] performed thoracentesis at 34 and 36 weeks, menstrual age, but noted reaccumulation of the fluid in less than 16 hours. They concluded that thoracentesis of macrocystic lesions in utero did not appear to provide lasting decompression of normal lung tissue. Nicolaides et al.[2] accomplished chronic drainage of a cyst due to Type I cystic adenomatoid malformation by placing a double pigtain catheter between the cyst and amniotic fluid cavities at 24 weeks. Clark et al.[3] reported resolution of fetal hydrops following drainage of a cystic adenomatoid malformation (CAM). The cyst was initially drained by thoracentesis, but reexpanded within 48 hours. The cyst was then chronically drained with an indwelling catheter with complete resolution of the fetal hydrops documented within three weeks.

The major dilemma confronted in managing these lesions is knowing whether intervention prior to the onset of hydrops is truly altering the ultimate outcome. Although all reported instances of intrauterine drainage to date have been associated with survival of the infant, there are reports of equally dramatic lesions that have not been treated with intrauterine drainage that have ultimately had a favorable outcome.[4] This is surprising in view of the severity of pulmonary hypoplasia seen with other space-occupying lesions of the fetal chest such as diaphragmatic hernia.

Despite the initial complete drainage of the cyst in this instance, the dimensions of the cyst in the surgical specimen were essentially identical to the ultrasound measurements prior to thoracentesis. Based on the outcome of this patient, it does not appear that in utero decompression of Type I cystic adenomatoid malformation is necessary in the absence of hydrops.

REFERENCES

1. Adzick NS, Harrison MR, Glick PL, et al: Fetal cystic adenomatoid malformation: Prenatal diagnosis and natural history. *J Pediatr Surg* 20:483, 1985.
2. Nicolaides KH, Blott M, Greenough A: Chronic drainage of fetal pulmonary cyst. *Lancet* 1:618, 1987.
3. Clark SL, Vitale DJ, Minton SD, et al: Successful fetal therapy for cystic adenomatoid malformation associated with second-trimester hydrops. *Am J Obstet Gynecol* 157:294, 1987.
4. Asher JB, Sabbagha RE, Tamura RK, et al: Fetal pulmonary cyst: Intrauterine diagnosis and management. *Am J Obstet Gynecol* 151:97, 1985.