A Case of Congenital Cystic Adenomatoid Malformation of the Lung (CCAM)

Yoshiyuki Ihno, Masahiro Saitoh,
Shin-ichi Takada, Satoru Takeda,
Katsuyuki Kinoshita, Shouichi Sakamoto.

Department of Obstetrics and Gynaecology,
Saitama Medical Center,
Saitama Medical School,
Kawagoe, Japan

Abstract : *A case of the fetal intrathoracic abnormality with ascites detected by ultrasonography at 27 weeks of gestation was demonstrated to be congenital cystic adenomatoid malformation of the lung after delivery. The prognosis of the infant after the resection of the tumour was favourable. This case was reported and the problems of diagnosis and management were discussed.(Thai J Obstet Gynaecol 1991;3:121-125.)*

Key words : cystic adenomatoid malformation, congenital anomaly, fetal ascites, amniocentesis

Congenital cystic adenomatoid malformation of the lung (CCAM) has been reported with increasing frequency with regard to prenatal diagnosis by ultrasonography in the past few years[1]. This malformation was classified histologically into three types by Stocker et al[2] and its spectrum of clinical presentations is broad by the extent of the affected pulmonary segment or lobe[3]. Our report describes the prenatal detection of fetal intrathoracic abnormality with ascites at 27 weeks of gestation, followed by the surgical excision, revealing a type II lesion of CCAM.

Case Report

A 31 years old woman (gravida 2, para1) was referred to our hospital at 27 weeks gestation because of fetal ascites detected by ultrasonography (Figure 1). A more detailed examination demonstrated maternal polyhydramnios, and a large, homogenous, solid mass occupying the fetal left hemithorax with mediastinal shift to the right (Figure 2). The fetal growth was appropriate for dates of gestation and the NST was reactive. Amniocentesis was performed. Alpha - fetoprotein (AFP) levels in the amniotic fluid

were not elevated at 2060 ng/ml and cytogenetic analysis showed a normal 46 XX karyotype. Fetal ascites were aspirated and the fluid was serous and no germs were detected by culture. Amniographic examination revealed no surface malformation of the fetus. These findings suggested that the diagnosis was likely to be CCAM in the left hemithorax, but herniation of the fetal diaphragm was not completely ruled out.

Preterm premature rupture of membranes occured at 30 weeks and 4 days of pregnancy and the volume of amniotic fluid was diminished, followed by variable deceleration of the fetal heart rate. Cesarean section was performed because of fetal distress. A 1552g female infant was delivered. The pH value in the umbilical artery was 7.29, and Apgar scores were 5 at 1 min and 9 at 5 min.

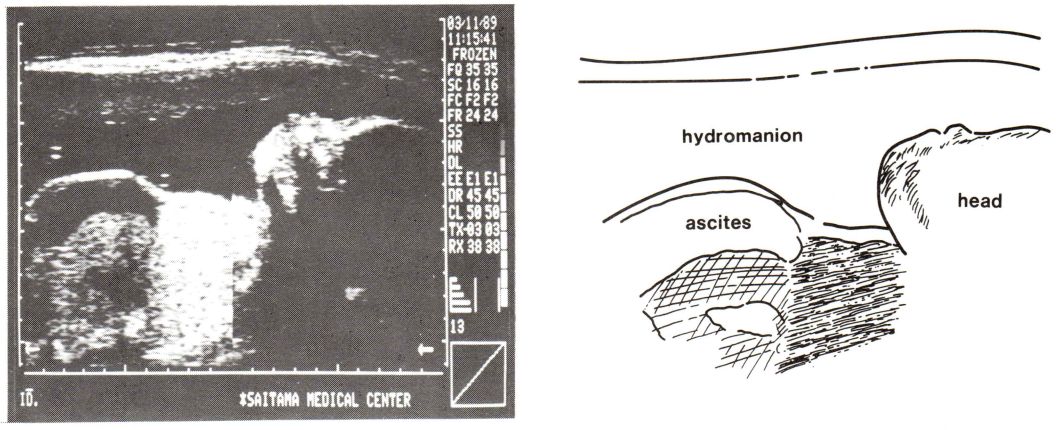

Fig. 1 Polyhydramnios and fetal ascites detected by ultrasonography.

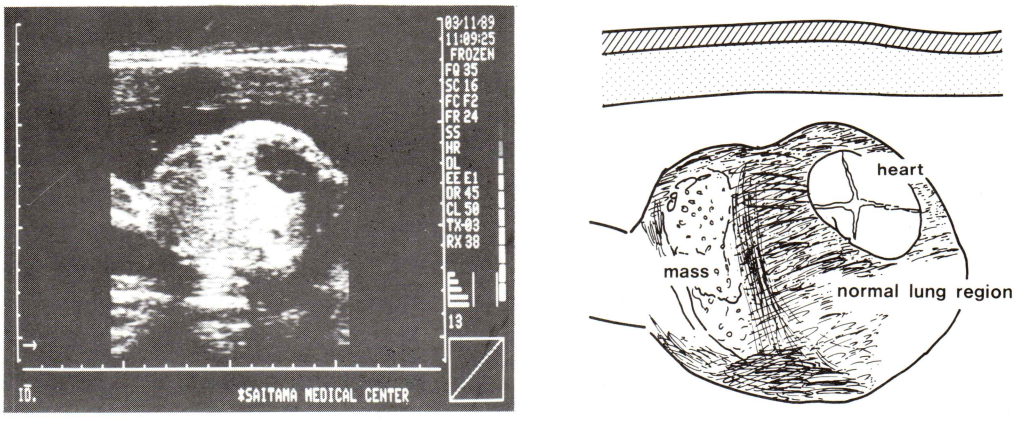

Fig. 2 The left thoracic mass, shifting the heart to the right side, detected by ultrasonography.

Immediate intubation and respiratory management were effective to overcome the respiratory distress for the first few days. Chest X-rays, ultrasonography, and CT scan revealed a mass in the left lower lobe, entirely consistent with a diagnosis of CCAM (Figure 3). A left lower lobe resection was performed on day 39 because of the development of respiratory distress caused by the compression of the lung by the mass. Postoperative course of the infant was good and on follow up visit, the infant was judged to be growing normally. Upon gross examination, the cut surface of the resected lobe showed numerous, small cysts filled with mucinous fluid (Figure 4). Histological examination of the mass revealed a lot of small cysts of varying size that were lined by ciliated columnar cells (Figure 5). Cartilage and inflammatory lesion were not present, whereas, some bands of smooth muscle were found. The pathologic diagnosis was CCAM, Stocker type II.

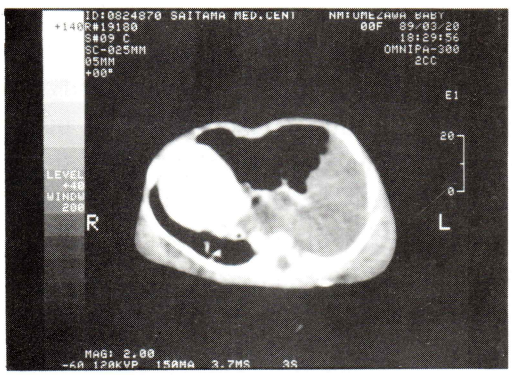

Fig. 3 Computed tomography taken on day 2 after birth demonstrates a marginal clear-cut mass in the left posterior part of the lower lobe consistent with a CCAM.

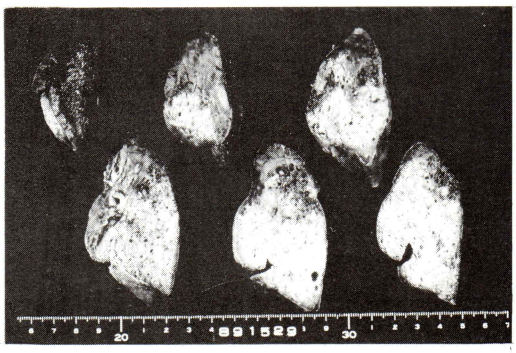

Fig. 4 Macroscopic findings of the resected lung mass show many microcysts with mucus, revealing type II CCAM.

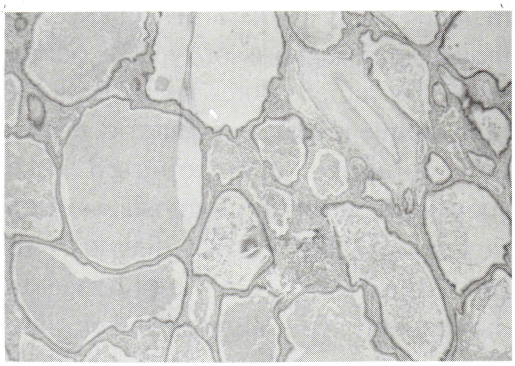

Fig. 5 Histopathological findings of the mass show small cysts lined by ciliated columnar cells looking like bronchal cell and scattered of residual lying tissue in the cyst (x100).

Discussion

Congenital adenomatoid malformation of the lung was first described by Ch'in and Tang in 1949[4]. Several reports on the rare congenital cystic disease of the lung discussed the nature of the malformation and the type of pathologic lesion[5,6]. In 1977, Stocker et al[2] categorized the lesion by clinical, gross, and microscopic

Table 1 Stocker's classification

Types		Prognosis	Frequency
I	single or multiple large cysts (>2cm in diameter)	: with no associated anomalies better prognosis than others	75%
II	multiple cysts (<1cm in diameter)	: frequently associated with other congenital anomalies	10-15%
III	bulky non-cystic lesion	: invariably poor with stillbirth or neonatal death	10%

findings into three types : cystic, intermediate and solid (Table 1). This case was classified as type II, of which the incidence was 10-15% of CCAM. Although the prognosis of type II, on the whole, is rather poorer because of the high frequency of associated congenital anomalies, this reported case showed as good a prognosis as seen in type I[3]. The correct diagnosis of CCAM in utero is of importance to the patient. The diseases taken into consideration as differential diagnosis are listed in Table 2[7,8]. Diaphragmatic hernia has to be excluded since it shows a similar clinical picture. Ultrasonography is a very useful tool in the diagnosis of CCAM, and MRI has recently been advocated as a very powerful and precise tool to detect CCAM. Furthermore, AFP levels in the amniotic fluid or in umbilical cord blood have been reported to be high in patients with CCAM[9,10] because of tumour-like lung masses of embryonic origin, though no elevation of AFP was shown in this case. In order to permit therapeutic planning, the early diagnosis by ultrasonography is of utmost importance. The presence of maternal hydramnios, in general, is a strong indication of poor prognosis as well. Therefore, its intensive prenatal care of the patient by the fetal heart monitoring, prevention of premature labour and the frequent of the polyhydramnios and fetal ascites, is mandatory to decide the mode of delivery and its timing of termination of pregnancy. After delivery, the prognosis of the newborn depends on the type and extent of its malformation as well as of maturity and hypoplasia of the unaffected lung. Therefore, avoidance of fetal distress has to be considered during vaginal delivery, and cesarean section is the choice, if fetal distress is expected. Surgical resection is the treatment of choice in the newborn

Table 2 Differential diagnosis

1. Mediastinal origin
   teratoma, enteric cyst, lymphangioma, meningocele
2. Congenital diaphragmatic hernia
3. Pulmonary origin (rare)
   CCAM, giant lobar emphysema, bronchial atresia
4. Cardiac origin (rare)
   rhabdomyomata

infant, when it has emergency indication[11]. In this case, the newborn baby was under observation in the NICU, in which the complication of sepsis and DIC took place after the delivery and the dyspnea progressing indicated lobectomy at 39 days of age. Complete recovery was accomplished by the procedure.

In conclusion, the proper diagnosis of CCAM in utero by ultrasound is of importance to permit the therapeutic planning after delivery with good survival rate.

References

1. Fine C, Adzick S, Doubilet PM. Decreasing size of a congenital cystic adenomatoid malformation in utero. J Ultrasound Med 1988;7:405-8.
2. Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung: classification and morphologic spectrum. Human Pathol 1977;8:155-71.
3. Beluffi BG, Brokensha C, Kozlowski K, et al. Congenital cystic adenomatoid malformation of the lung. Presentation of 16 cases. Fortschr Rontgensty 1989;5: 523-30.
4. Ch'in KY, Tang MY. Congenital adenomatoid malformation of one lobe of a lung with general anasarca. Arch Pathol 1949;48:221-9.
5. Adzick S, Harrison MR, Glick PL, et al. Fetal cystic adenomatoid malformation: Prenatal diagnosis and natural history. J Pediatr Surg 1985;5:483-8.
6. Mendoza A, Wolf P, Edwavds DK, Leopold GR, Voland JR, Benirschke K. Prenatal ultrasonographic diagnosis of congenital adenomatoid malformation of the lung. Arch Pathol Lab Med 1986; 110:402-4.
7. Alhright EB, Crane JP, Shachelford GD. Prenatal diagnosis of a bronchogenic cyst. J Ultrasound Med 1988;7:91-5.
8. Fitazgerald EJ, Toi MA. Antenatal ultrasound diagnosis of cystic adenomatoid malformation of the lung. J Canadian Ass Radiol 1986;37:48-9.
9. Albright SG, Katz VL. Alpha-fetoprotein findings in a case of cystic adenomatoid malformation of the lung. Clin Genet 1989;35:75-6.
10. Petit P, Bossens M, Thomas D, Moerman P, Fryns JP, Berghe HV. Type II congenital cystic adenomatoid malformation of the lung; another cause of elevated alpha fetoprotein? Clin Genet 1987;32:172-4.
11. Rempen A, Feige A, Wunsch P. Prenatal diagnosis of bilateral cystic adenomatoid malformation of the lung. J Clin Ultrasound 1987;15:3-8.

7th Scientific Annual Meeting

The College of Obstetricians and Gynaecologists of Thailand
October 19-21, 1992
Chiang Mai Plaza Hotel
Chiang Mai

Informations : Kittipong Vairojanavong, MD
C/O Department of Obstetrics and Gynaecology
Rajavithi Hospital
Bangkok 10400
Tel. 245-6072-3
Fax. 246-8270