Prenatal Diagnosis of Congenital Cystic Adenomatoid Malformation

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Congenital cystic adenomatoid malformation (CCAM) of the lung is a rare pulmonary lesion. The prognosis is variable ranging from perinatal death to spontaneous in utero regression, with no neonatal morbidity. We present a case of CCAM diagnosed at 23 weeks' gestation using results of prenatal sonograms. Both prenatal Two-Dimensional and Three-Dimensional sonograms revealed multiple cystic lesions at the right lower lung field. Regular serial antenatal sonograms revealed the fetus had persistent right lower lung multicyst lesions but had no hydrops fetalis or associated congenital anomalies. The results of the antenatal 50 g glucose diabetic screen at 24 weeks' gestation were normal. A live male baby was vaginally delivered smoothly at 40 weeks' gestation. Body weight was 4170 g. Apgar scores were 7 at 1 minute and 8 at 5 minutes. The newborn had no signs of respiratory distress at birth. Postnatal chest computed tomography (CT) also revealed multiple fluid-filled cysts in the right lower lobe compatible with congenital cystic adenomatoid malformation. Because of the potential risk of perinatal death, repeated lung infection and malignant change of CCAM, the newborn received right lower lung lobectomy 1 week after delivery. Pathology confirmed the diagnosis of CCAM type II. The recovery was smooth postoperatively and the baby still receives regular follow-up. We emphasize the importance of prenatal diagnosis of CCAM and early removal of the congenital cystic lesions of the lung to achieve a good outcome. (Chang Gung Med J 2004;27:61-5)

Key words: congenital cystic adenomatoid malformation, prenatal diagnosis.

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ry distress such as dyspnea, tachypnea, or cyanosis at birth. Postnatal chest radiography and chest computed tomography (CT) of the newborn were performed. Chest X-ray showed consolidation of the right lower lung field (Fig. 3) and chest CT revealed multiple fluid-filled cysts in the right lower lobe compatible with congenital cystic adenomatoid malformation (Fig. 4). Because of the potential risk of perinatal death, repeated lung infection and malignant change of CCAM in the future, the newborn received a right lower lung lobectomy 1 week after delivery. Results of pathology examinations confirmed the diagnosis of CCAM type II. The recovery was smooth postoperatively and the baby still receives regular follow-up.
CCAM of the lung is a rare pulmonary lesion characterized by an excessive overgrowth of the terminal respiratory bronchioles. The lesion is almost always unilateral and may occur in any lobe. There are three distinct types of CCAM based on the size of the cysts and the microscopic appearance. Type I cyst is notable for its large size (measuring more than 2 cm in diameter). Type II cyst has smaller cysts, usually less than 2 cm and more than 5 mm in greatest diameter. Type III cysts are noted to be firm, bulky masses of lung tissue on gross inspection. Histologically, these lesions are described as having cysts that were 2 to 5 mm in the greatest diameter. Some authors prefer to classify the CCAM as microcystic (cysts less than 5 mm in diameter) and macrocystic (cysts equal to or greater than 5 mm in diameter) by gross anatomy, ultrasonographic findings, and prognosis. Microcystic lesions are often associated with hydrops fetalis and the patients have poor prognoses. Macrocystic lesions (single or multiple cysts > 5 mm) are not usually associated with hydrops fetalis and the patients have more favorable prognoses.

According to the classification of CCAM using ultrasonographic findings, our case was CCAM type II and has a favorable prognosis.

The characteristic appearance of CCAM by prenatal sonogram is multiple cystic lesions at the lung field as our case showed. The differential diagnosis of CCAM includes extralobar and intralobar bronchopulmonary sequestration, bronchogenic cyst, diaphragmatic hernia and mediastinal lesions, such as neurenteric cyst, or cystic teratoma. Bronchopulmonary sequestration typically produces a well-defined, homogenous, highly echogenic mass in the inferior aspect of the hemithorax or upper abdomen. The extralobar sequestration is spherical in shape, whereas the fetus with extralobar sequestration has a conical or triangular shaped mass. A bronchogenic cyst is a simple cyst near the mediastinum or centrally located in the lung. Patients with diaphragmatic hernias typically have the peristaltic bowels in the thorax and absence of normally positioned stomachs. A neurenteric cyst is often a simple cyst near the mediastinum or centrally located in the lower lobe. A mediastinal teratoma is often echogenic and occasionally has cystic components.

Some cases of CCAM may become inconspicuous, or disappear on serial prenatal sonograms, thus, emphasizing the importance of serial prenatal sonograms and postnatal imaging studies, including radiography and computed tomography.

Although the etiologic factors predisposing infants and children to pulmonary neoplasms are unknown, pulmonary developmental abnormalities may play a pathogenetic role. As CCAM can host metaplastic mucous cells, primitive mesenchymal cells and differentiated but poorly organized striated muscle fibers, it has been proposed that CCAM may act as a predisposing condition for oncogenesis. Because of the risk of malignant change of rhabdomyosarcoma and bronchioloalveolar carcinoma and the possibility of repeated respiratory tract infection, early removal of the congenital cystic lesions of the lung is advisable.

In the past, patients whose fetuses presented with presumed cystic adenomatoid malformation were advised of the poor prognoses. However, recent studies have demonstrated that aggressive obstetrical management and termination of pregnancy are not appropriate for patients with affected fetuses, as with conservative management (in the absence of hydrops) patients tend to have good prognoses. Based on this concept, we adopted conservative management in this case. Serial sonographic evaluations were performed to monitor the growth of the congenital cystic lesions, fetal growth and fetal well-being.

Fetuses with CCAM but without hydrops have a good chance for survival with support of maternal transport, planned delivery, and early surgery. Fetal lung masses associated with hydrops are nearly 100% fatal. Attempts have been made to treat fetuses with lung masses and hydrops using in utero surgical intervention. After more than two decades of experimental and clinical work, fetal surgery has become an accepted treatment modality for selected fetuses with life-threatening anomalies. Because some cases of CCAM may become inconspicuous, or disappear on serial prenatal sonograms, the role of fetal surgery in lesions such as CCAM is currently reserved for those fetuses with hydrops remote from term.
REFERENCES

產前診斷先天肺部腺體囊腫性構造異常

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先天肺部腺體囊腫性構造異常乃一種罕見的肺部疾病。此一先天肺部構造異常，嚴重時可導致胎兒於週產期死亡，亦可能自產於宮內回復正常而不造成新生兒的罹病。我們報告一例於妊娠23週時倉由產前二維及三維超音波診斷胎兒先天肺部腺體囊腫性構造異常。順利自然產後，安排肺部電腦斷層檢查亦發現新生兒右下肺部有多囊腫性構造異常。此新生兒於產後一週後接受右下肺葉切除，術後病理診斷為第二型先天肺部腺體囊腫性構造異常。由於先天肺部腺體囊腫性構造異常可能導致胎兒於週產期死亡、反覆性肺部感染及轉變為惡性的可能性，故於產前診斷此一先天肺部構造異常並於產後儘快切除肺部病灶，可以得到良好的預後。(長庚醫誌 2004;27:61-5)

關鍵字：先天肺部腺體囊腫性構造異常，產前診斷。