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Case report

CONGENITAL CYSTIC ADENOMATOID MALFORMATION OF THE LUNGS: A CASE REPORT

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Abstract

Congenital cystic adenomatoid malformations (CCAM) of the lung are a rare congenital disorder. The prevalence of this condition is 9 per 100,000 births, with a slight predominance in males. Most cases present clinically in the neonatal period, but may remain asymptomatic until adulthood. The etiology of CCAM is not fully understood. Fetal cystic changes of the lungs are detected prenatally by ultrasound, and postnatally, the definitive method is chest computed tomography (CT) or magnetic resonance imaging (MRI). Resection of CCAM is recommended due to the risk of malignancy. We present the case of a term-male hypotrophic newborn, in whom cystic changes of the right lung were detected prenatally. The chest X-ray and CT scan postnatally confirmed right-sided congenital adenomatoid cysts. At the age of 2 months, the infant underwent surgical resection of cystic changes, and CCAM type 1 was histologically proven.

Keywords: newborn, congenital cystic adenomatoid malformations

Introduction

Congenital cystic adenomatoid malformation (CCAM) of the lung are developmental hereditary congenital change in the lung parenchyma. These hamartomatous lesions suppress alveolar growth and lead to the replacement of normal lung structure with a multicystic mass[1]. This is a relatively rare condition. The incidence of congenital cystic adenomatoid malformation of the lung is 1 in 25,000 to 35,000 pregnancies. The prevalence is approximately 9 per 100,000 births[2]. This congenital anomaly accounts for 25% of congenital lung malformations[3]. There is a slight predominance of males over females with a ratio of 1.8:1[4]. Most CCAM cases present in neonatal period (85%), followed by infancy, childhood, and very rarely in adulthood[4]. In the neonatal period, it presents clinically as acute respiratory distress, while in the infancy and childhood, recurrent lung infections are the most common clinical manifestations.

CCAMs can affect any part of the fetal respiratory system, starting from the trachea to the alveoli[6]. Normal embryonic development of the respiratory system begins in the 3rd week of pregnancy. Lung development is divided into embryonic (3-7 weeks), pseudo-glands (7-17 weeks), canalicular (17-29 weeks), saccular (24-36 weeks) and alveolar stage (36 weeks to maturity)[5]. Changes in the embryonic and pseudoglandular stages may lead to a group of structural abnormalities termed bronchopulmonary malformations, including congenital cystic

abnormalities of the lung. There are four types of congenital cystic lung abnormalities: pulmonary sequestration, congenital cystic adenomatoid malformation, congenital lobar emphysema, and bronchogenic cysts[7]. The etiology is unknown. However, various growth factors and signaling mechanisms involved in the morphogenesis of the lung stem branching could be implicated[8].

CCAMs are probably a result of impaired embryonic differentiation before the 35th day of pregnancy, leading to local aberration of parenchymal tissue caused by excessive adenomatoid growth of the terminal bronchioles and suppression of alveolar growth[9].

Advances in fetal ultrasonography allow early prenatal detection of congenital cystic adenomatoid malformation of the lung, enabling timely planning of appropriate postnatal intervention and treatment.

Case report

We present the case of a term-male newborn. The 30-year-old mother had uneventful second pregnancy. During regular follow-up ultrasound examination in the 28th week of gestation, a large cystic formation was detected in the right lung of the fetus. There was no polyhydramnios. Due to the occurrence of fetal distress, the baby was born with emergency caesarean section at 40 weeks of gestation. A male hypotrophic newborn was born with birth weight of 2750 g, birth length of 47 cm and Apgar score of 7/8. The initial examination showed noticeable grunting crying and auscultatory weakened vesicular breathing in the right hemithorax. The first chest X-ray showed multiple cystic changes in the right lung, predominantly in the upper and middle lobe, with widened intercostal spaces in the right thoracic wall and mediastinal structures pushed to the left (Figure 1).

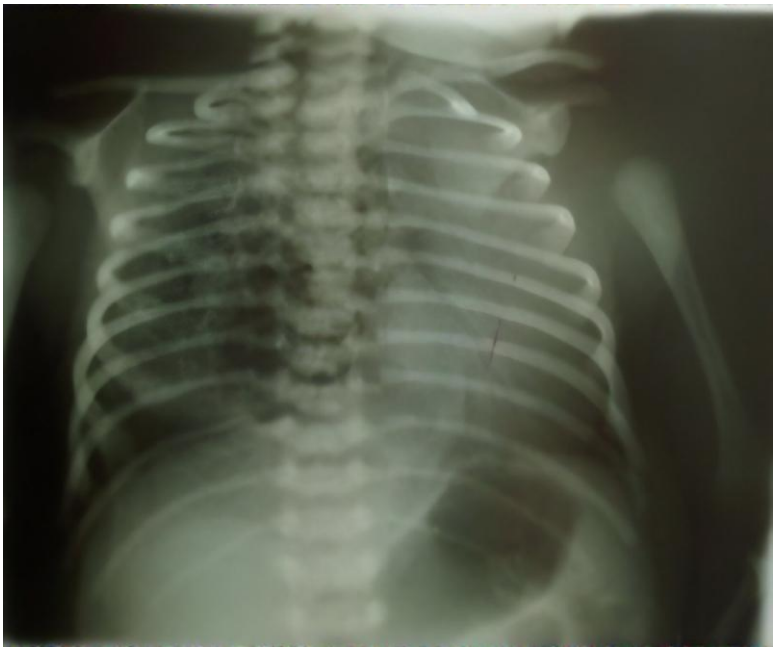

Fig. 1. Chest X-ray of the newborn with right cystic lung formations

These changes were confirmed by chest computer tomography (CT), where several cystic changes were seen, measuring over 20 mm, some of them filled with fluid (Figure 2). CCAM was suspected by the radiologist.

Initially, the newborn was treated with diffuse oxygen support. In the first three weeks, broad spectrum antibiotic treatment was administered due to elevated inflammatory markers (CRP 139 mg/L) and suspected bacterial infection; blood culture was negative.

Echocardiography, cranial, abdominal and urogenital ultrasound examinations did not show any other abnormalities.

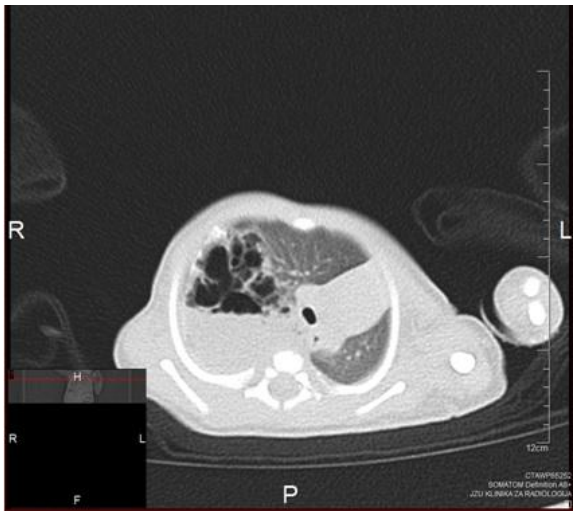

Fig. 2. Chest CT scan - right congenital cystic adenomatous malformation

On the 24th postnatal day, the newborn developed dyspnea and cyanosis. Spontaneous pneumothorax was detected, probably due to rupture of the pulmonary cysts. The newborn was intubated and put on conventional mechanical ventilation for a period of 5 days. After that, the newborn was extubated and treated with diffuse oxygen support, maintaining stable vital parameters.

At the age of two months, the cystic lung changes were resected by a pediatric surgeon (Figure 3).

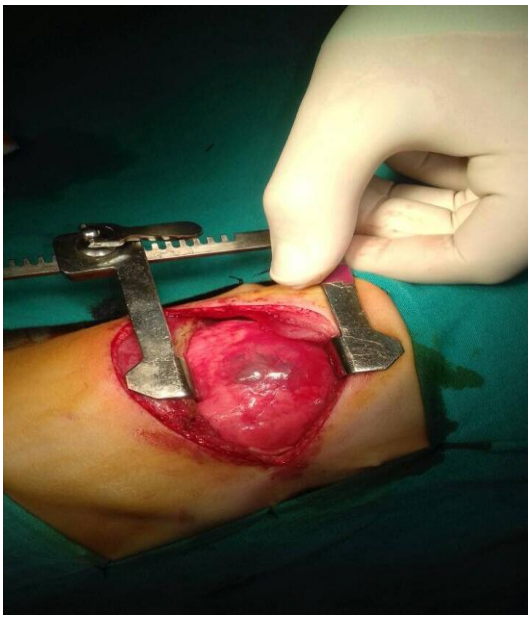

Fig. 3. CCAM surgical resection

Histopathologically, CCAM type 1 was diagnosed. Twenty-seven days following surgery, the infant was discharged from the hospital. The postoperative period was uneventful.

Discussion

Congenital cystic adenomatoid malformation (CCAM), currently referred to as congenital pulmonary airway malformation (CPAM), is a rare developmental anomaly of the

lung characterized by cystic proliferation of abnormal bronchiolar structures and distortion of normal lung parenchyma. This relatively rare sporadic congenital lung anomaly was first described in 1949 by Ch'in and Tang[10].

Congenital cystic adenomatoid malformations of the lung are histologically divided into five types according to Stocker[11].

Type 0 (acinar dysplasia): The origin of this type of cysts is trachea or bronchi. Microscopically there is cartilage, smooth muscle and glands with abundant mesenchymal tissue. The ultrasound examination shows a solid homogeneous mass in the lung tissue. This type is found in 1-3% of cases and has a lethal outcome immediately after birth.

Type 1 (bronchial type): The site of origin of this type of cysts is the distal bronchus or proximal bronchioles lined with ciliated pseudostratified epithelium. These macrocytic changes can grow up to 2 cm in diameter and are usually localized in a single lobe. On ultrasound, they are identified as cystic changes in the lung parenchyma. This group includes 50-70% of cases and has a good prognosis and survival.

Type 2 (bronchiolar type): CCAM type 2 originates from the terminal bronchioles and is characterized by multiple cysts <2 cm in diameter, lined with cuboidal epithelium. They occur in about 25% of cases and are associated with other congenital anomalies (renal agenesis, hydrocephalus, diaphragmatic hernia, jejunal atresia, tracheoesophageal fistula, ventricular septal defect (VSD), Tetralogy Fallot ...).

Type 3 (bronchoalveolar type): This type is a rare form that occurs in 5-10% of cases and has a poor prognosis. It is a mixture of microcystic changes coated with ciliated cuboidal epithelium and solid tissue. On ultrasound, it is visualized as an echogenic solid mass.

Type 4: They are macrocystic changes (>10 cm) lined by flattened epithelium and resting on loose mesenchymal tissue. They occur in 5-15% of cases and are associated with malignancy (pleuropulmonary blastoma).

The presented case was a type 1 CCAM with a favorable outcome. In our patient, the lesion involved both the upper and middle lobes, whereas most cases in the literature are reported as involving a unilateral single lobe[6].

In utero this congenital anomaly may present as lung hypoplasia with fetal death, severe fetal hydrops, or complete regression by an unknown mechanism[7]. From a pathophysiological point of view, large CCAM can compress the esophagus, leading to difficulty swallowing in the fetus and resulting in polyhydramnios. This compression on the ipsilateral side of the lung leads to pulmonary hypoplasia and displacement of the mediastinal structures and impaired cardiac venous return[12].

Polyhydramnios was not detected. However, ipsilateral lung compression with displacement of the mediastinal structures was observed in our case.

Postnatally, CCAM of the lung may be asymptomatic or symptomatic. In the neonatal period, dominant signs are respiratory distress, tachypnea, grunting, retraction, and cyanosis. Compression of mediastinal structures is also possible, leading to compromise of the cardiovascular system. Spontaneous pneumothorax is a rare initial manifestation in newborns.

Congenital cystic adenomatoid malformation of the lung should be distinguished from diaphragmatic hernia, pulmonary sequestration, bronchogenic cysts, congenital lobar emphysema, and cystic fibrosis[5].

Congenital cystic adenomatoid malformation of the lung can be detected *in utero* by ultrasonography, usually around 21 weeks of gestation. The intrauterine prognosis of the fetus is determined by the size of the fetal lung mass or by the CCAM volume ratio (CVR). CCAM volume is calculated by using a formula for prolate ellipse, with measurements of the lung mass in three perpendicular planes[13]. Also, CCAM type 3 is associated with higher levels of alpha-fetoprotein in amniotic fluid[3].

In our case, a cystic lung lesion was detected at 28 weeks of gestation and no further investigation was done. The mother was counseled regarding the fetal condition and the potential neonatal outcome. After delivery, the newborn was admitted to the neonatal intensive care for observation, further evaluation, and management.

A chest X-ray is not always sufficient to diagnose CCAM postnatally. The preferred diagnostic method is chest CT scan or chest magnetic resonance imaging (MRI). CT scan can detect multilocular thin-walled cystic masses. If there is an infection, the changes are a mixture of a solid and cystic lesion with thicker walls. On MRI, the changes are determined by the size of the lesion, the number of the cysts and appear as non-homogeneous masses in the lung parenchyma[3].

In highly developed centers, antenatal fetal intervention is possible that includes excision of the lobe with microcystic changes, aspiration of macrocystic changes, or open fetal surgery[8].

Postnatal symptomatic cases are resolved surgically by lobectomy or excision of the cyst. The attitude towards treatment in asymptomatic patients is controversial. Approximately 3.2% of non-operated patients may develop complications, such as hemorrhage, pneumothorax, nutritional difficulties, sudden respiratory compromise, and malignant changes[14].

In our case, there was spontaneous pneumothorax, indicating a necessity for surgical treatment.

Due to the high potential for possible malignant alteration and occurrence of rhabdomyosarcomas (35%) and lung blastomas (20%), surgical removal of the cystic change is recommended. The optimal age for surgical treatment is 6-12 months[7].

Mortality rate in prenatally diagnosed cases ranges from 9 to 49%[15].

Conclusion

The prognosis of CCAM in the lung depends on the type, size, and location of the cysts. The treatment of choice should be minimally invasive surgery where possible. Associated congenital anomalies in other organs can lead to a worse prognosis. Regular antenatal examinations, good obstetric care, and multidisciplinary postnatal care are essential to ensure better outcomes in these patients.

Conflict of interest statement. None declared.

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