Successful Surgical Management of Symptomatic Antenatally Diagnosed Congenital Pulmonary Airway Malformation: A Case Report

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Key Clinical Message

Routine obstetric ultrasound is important in detecting CPAM and it should be followed up routinely throughout the pregnancy. Management of CPAM is debatable and varies from conservative to invasive approaches.

ABSTRACT

Congenital pulmonary airway malformations (CPAM) are rare developmental anomalies often diagnosed on prenatal ultrasound. This case presents a 30-year-old female who had a routine ultrasound in the 13th gestational week which revealed cystic changes in the fetal left lung with polyhydramnios. The delivered baby underwent an uneventful open left lower lobectomy.

Keywords: CPAM; Congenital lung lesions; Congenital pulmonary airway malformations; Cystic adenomatoid malformation ; Case Report.

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Introduction

Congenital pulmonary airway malformations (CPAM), characterized by an abnormal airway pattern, are developmental malformations of the lower respiratory tract that occur during lung branching morphogenesis. CPAM has the potential to cause cystic and/or adenomatous pulmonary areas. Despite being a rare condition with a prevalence of only 0.81/10,000 fetuses, CPAM is considered to be the most common congenital lung abnormality, being responsible for 30–40% of all malformations (1). The pathogenesis and exact cellular mechanisms behind CPAM are not fully understood yet; however, many potential genes were suspected to play a part in the formation of lung cysts (2). Nowadays, with the advance of prenatal ultrasound (90% sensitivity and 77% specificity for CPAM), CPAM is often diagnosed antenatally, by detecting an echogenic mass within the fetal lungs usually in the second trimester of gestation (21–24 weeks), then followed up routinely during the course of pregnancy (1). Magnetic resonance imaging (MRI) also plays an important diagnostic role complementary to ultrasound since it is able to define the type and extent of these lung

lesions, thus guiding prenatal intervention and postnatal care (2). CPAM can be classified into five different types, each one of them vary in appearance and originate from a different area in the lung (3). Prognosis, and therefore, management usually depends on the classification and presence of symptoms and any other fetal anomalies (3). We present a symptomatic case of CPAM with the associated clinical, radiological and pathological details. The work has been reported in line with the SCARE criteria (4).

Case Report

A 30-year-old pregnant female with an unremarkable surgical, medical, drug or family history presented in the 13th week of gestation for a routine Ultrasound (US) examination which revealed a cystic formation in the left lung of the fetus associated with mild polyhydramnios. The patient was asymptomatic and physical examination was unsignificant, By monitoring, and during the 23rd and 26th weeks of gestation, the lesion was stable and there was a slight increase in the amniotic liquid (Fig.1). In the 39th week of gestation, the woman underwent caesarean delivery and had a 3400 g weighing male who was admitted to the neonatal intensive care unit by their parents and referred to the Department of Pediatric Surgery complaining from severe respiratory distress worsening during lactation, and it was decided to stay in the incubator for 24 hours. The chest radiography indicated the deviation of the mediastinal structures toward the right-side Fig.2 (A). Chest radiography with oral contrast excluded the presence of diaphragmatic hernia Fig.2 (B). At the age of 14 days, a (computed tomography) CT scan of the chest with contrast was performed and the diagnosis of CPAM was confirmed at the expense of the lower left lobe (Fig.3). Except for the right axis deviation, echocardiography did not indicate any cardiac abnormalities. Upon completion of preoperative procedures, the 14-day-old patient underwent an open left lower lobectomy via a left posterior thoracic approach (Fig.4), and pathology studies of the resected part confirmed the diagnosis of CPAM. The infant received post-operative care with incubation for 3 days. On the third day after the operation, chest radiography demonstrated the return of mediastinum to the midline, and a compensatory hyperinflation of the residual lobes of the left lung. Finally, the infant weaned off the respiratory support and was discharged home at the age of 17 days without any complications. During follow-up, the infant's condition was found to be satisfactory.

Discussion

CPAM, once referred to as congenital cystic adenomatoid malformation (CCAM), is an uncommon, developmental lung anomaly of unknown cause, presenting as cystic changes to the terminal bronchioles (5). There are a few differential diagnoses to CPAM that include bronchogenic cysts (BC), congenital lobar emphysema (CLE), and bronchopulmonary sequestration (BPS) (1). Antenatally, CPAM can cause some complications in the fetus, the most important of which is Hydrops Fetalis due to the compressing of the mass on other structures, which usually requires an emergency cesarean section (3). At birth, 70% of CPAM are asymptomatic. Among antenatally diagnosed patients, 70% of antenatally diagnosed CPAM are asymptomatic; while the other 30% cause neonatal respiratory distress, with 10% have severe respiratory distress requiring assisted ventilation (1). In some cases, CPAM remains unrecognized until adolescence or later life when complications, such as recurrent pulmonary infections, pneumothorax and lung abscess, start to happen (5). In our case, there was symptomatic, antenatally diagnosed CPAM, associated with Hydrops Fetalis antenatally (requiring caesarean delivery), and severe respiratory distress postnatally (requiring neonatal intensive care unit). Therefore, left lower lobectomy was decided, and open surgery was chosen over thoracoscopy due limited resources (lack of equipment and expert hands). The management of CPAM is highly debatable, and varies from administering maternal betamethasone prenatally (to decrease the size of large CPAMs and reverse hydrops) to minimally invasive or even open surgical resection postnatally, with the most controversial area remains the postnatal management of asymptomatic cases. Prophylactic lung resection in asymptomatic patients is sometimes done, and has advantages that include reduced risk of infection, malignancy and decreased complication rate, while disadvantages include mortality and morbidity associated with operation and cancer risk despite excision. Due to the low incidence and wide spectrum of manifestation of CPAM, it is not surprising that the quality of the evidence on management styles is generally poor (6). David et al (2016) proposed prenatal Fig.5 (A) and postnatal Fig.5 (B) management algorithms (2). A meta-analysis

by Naser and Bass (2012), suggested that thoracoscopic resection is a safe and feasible alternative to open resection of congenital lung lesions in experienced hands, and no differences between thoracotomy versus thoracoscopy for management of CPAMs were observed in terms of overall complications and the duration of surgery. However, days with chest tube in place and length of hospitalization period were longer after open surgery (7).

Conclusion

CPAM, previously known as CCAM, is the most common congenital lung lesion. CPAM is classified into five different types, varying in origin, and appearance. Routine obstetric US is particularly important in detecting CPAM at around the second trimester of gestation and is followed up routinely through the term of the pregnancy. Management of CPAM is debatable and varies from conservative to invasive approaches.

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Figure Legends

Fig.1: Antenatal sonography monitoring (a) in the 23rdweek, there were well-defined cystic structures in the left lung accompanied by mild polyhydramnios (b) in the 26thweek, a 20×40 cm anechoic multicystic lesion was occupying the left side of the thoracic cavity with moderate polyhydramnios.

Fig.2: (A) postnatal plain trunk X-ray with mild diffuse opacity on the left side that suggests a large cystic lesion and mediastinal shifting to the right. (B) plain trunk X-ray with oral contrast assures the absence of the diaphragmatic hernia.

Fig.3: CT scan of the chest with contrast suspects of 20×40 cm CPAM in the left lower lobe consisting of enhanced solid mass in the posterior part associated with a multicystic structure located anteriorly with compensatory hyperinflation of the left upper lobe, which causes the lung herniation to the right side.

Fig.4: Open left lower lobectomy via a left posterior thoracic approach.

Fig5: (A) Prenatal management algorithms of CPAM by David et al (2016).

ECHO = Echocardiogram. * Multiple courses of betamethasone may be an option. (B) Postnatal management algorithms of CPAM by David et al (2016).

ECHO = Echocardiogram; PPHN = persistent pulmonary hypertension of the newborn; ECMO = extracorporeal membrane oxygenation.