Ultrasound as a screening tool in the follow-up of asymptomatic congenital cystic adenomatoid malformation

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Abstract

Congenital cystic adenomatoid malformation accounts for the vast majority of congenital cystic lung pathologies. The clinical spectrum of congenital cystic adenomatoid malformation is wide and ranges from silent incidental lesions to severe congenital respiratory distress or stillbirths. Management of clinically silent lesions is still a matter of debate, with conservative follow-up being a popular approach during the last decade. However, an optimal methodology of follow-up is yet to be determined. In this study, ultrasound is proposed as a follow-up tool for clinically silent lesions based on the findings in a case with known congenital cystic adenomatoid malformation from the antenatal period, which has been followed up for four years.

Keywords

Congenital cystic adenomatoid malformation, ultrasound, fetal lung, cystic adenomatoid malformations

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Introduction

Congenital cystic adenomatoid malformation (CCAM) is a rare multicystic mass arising in the lung parenchyma from abnormal proliferation of the immature terminal respiratory bronchioles and suppression of alveolar development in the affected site during the first trimester of fetal life.¹ Despite its rarity, CCAM encompasses the vast majority of congenital cystic lung pathologies with a reported incidence as high as 95%.² Stocker et al.³ have classified CCAM into three subtypes from I to III. Type I is the most commonly seen and favorable type of all. Type II has smaller sized cysts, measuring less than 2 cm in diameter. Type III is the most infrequent and lethal form of all and unlike the multicystic appearances of types I and II, has a hyperechogenic solid mass-like appearance on ultrasound. Ranging from silent underlying lesions to severe congenital respiratory distress or stillbirths, congenital cystic lung lesions possess a wide clinical spectrum. The most common complication in those who are being conservatively followed up is recurrent infection related

to the underlying lung lesion, mainly in the form of pneumonia or abscess.² During the last decade, conservative management in asymptomatic individuals with known CCAM gained popularity among authors.^{4,5} Nonetheless, the optimum methodology in follow-up is not clear yet. In this study, ultrasound is proposed as a follow-up tool based on findings in a case with known CCAM from the antenatal period which has been followed up for four years.

Case report

A 30-year-old woman (gravida 3, para 2) with an unremarkable pregnancy and medical history was referred at the 21st week of pregnancy for routine fetal anomaly

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Figure 1. Fetal axial sonogram obtained at 21st week of gestation showing multiple cystic structures separated with irregular septa formation filling almost the entire left hemithorax. Note mild rightward displacement of the normal-sized fetal heart and slight eversion of the left hemidiaphragm (not shown here) secondary to mass effect. No sign of fetal hydrops was evident.

screening. High-resolution trans-abdominal ultrasound and color Doppler interrogation were performed using an Aplio XG scanner equipped with a 5-MHz convex transducer (Toshiba Medical Systems, Tokyo, Japan). Sonographic fetal gestational parameters were compatible with pregnancy age based on the beginning of mother's last menstrual period. Ultrasound revealed a 4×3 cm cystic mass located in left hemithorax (Figure 1). The cysts measured up to 2 cm in diameter. Being separated with an irregular septa formation, the mass filled almost the entire left hemithorax and caused mild rightward displacement of the normal sized fetal heart with slight eversion of the left hemidiaphragm. A small portion of ipsilateral lung parenchyma was visible above the multicystic structure and was significantly hyperechoic compared with normal appearing right lung parenchyma. There was no significant change in appearance and size of the mass on follow-up ultrasonography at the 35th week of pregnancy. However, the proportion of mass to the left hemithorax was notably decreased due to development of the ipsilateral lung parenchyma. Two weeks later (at the 37th gestational week), the fetus was delivered by cesarean section because of the breech presentation. The male newborn did not suffer clinically apparent respiratory symptoms, and the family was discharged in good condition after two days of hospital stay. After one uneventful year, the infant was admitted to hospital for a slight fever and cough. A computed tomography (CT) scan revealed bilateral normal lung parenchyma and a well-defined, slightly hypodense region of soft tissue located just above the diaphragm at the posterior base of the left lung (Figure 2). The infant was

Figure 2. Sagittal reformatted unenhanced CT image obtained at age 1 showing normal aeration of lung parenchyma and a sharply defined, slightly hypodense soft tissue mass (arrow) sitting on the left hemidiaphragm at the posterior base of the left lung.

discharged from hospital in good condition after inpatient management for one day, and has been doing well since then. Because the mass was located at a relatively blind spot in the lung, it was overlooked on CT scan at age 1, after which the patient inadvertently quitted the follow-up. No additional CT scan was performed. The patient has been asymptomatic since then, and was seen at age 3 after a follow-up phone call with the family. A thoracic ultrasonography revealed a 3×1 cm multicystic ovoid mass containing slightly irregular septa formation that showed weak vascular encoding on power Doppler interrogation (Figure 3). The lesion did not show significant morphological changes on repeat ultrasound scan at age 4. The child is now aged four years and remains asymptomatic.

Discussion

The case presented here shows fairly long-term findings of a child who was born with an antenatally diagnosed large unilateral CCAM and followed up for four years. Despite showing no significant decrease in size compared with a single CT scan that was performed at age 1, the proportional size of the mass relative to the ipsilateral normal lung was significantly decreased over the four-year follow-up period. Analogous to the conditions in congenital high airway obstruction syndrome (CHAOS), the hyperechogenic lung above the cystic mass found in the fetus presented here must result

from blockage of the fluid passage through the bronchial tree of normal lung parenchyma that was compressed by the cystic mass (Figure 4). A similar appearance with a distinct pathophysiology is seen in

Figure 3. High-frequency linear sonogram (obtained at age four) showing multicystic (asterisks show individual cyst formations) ovoid mass containing slightly irregular septa formation that showed weak vascular encoding on power Doppler interrogation. Note posterior acoustic enhancement typical of cystic lesions on ultrasound. CW: chest wall; LP: lung parenchyma.

CCAM type III, where the lungs are filled with myriad small cysts (Figure 4). Unlike in CCAM type III, all sonographic findings, including lung hyperechogenicity, have been shown to revert to normal appearance once the tracheal fluid passage is reestablished in fetuses with CHAOS.⁶ The hyperechogenic appearance of compressed lung tissue above the cystic mass found in the present fetus was also reversible and the ipsilateral lung proved to be completely normal on follow-up studies. Reversal of the hyperechogenic appearance on ultrasound most likely results from elimination of accumulated fluid within the bronchial tree, once the airway was reestablished and lung expanded after the first inspiration of the newborn.

Significant technological developments made early diagnosis of congenital lesions like CCAM possible.⁷ Today, in developed countries, the majority of congenital cystic lung lesions are being diagnosed antenatally by virtue of widespread use of antenatal ultrasound. The more we detect the lesions antenatally, the more we have asymptomatic 'sick' newborns. Moreover, certain cystic lung lesions that were detected by antenatal ultrasound screening were shown to disappear in the late gestational stages or during infancy.⁸ Detection of small incidental lesions that would not have been noticed otherwise in a lifetime, also increases the number of such instances. This situation has raised questions about the correct management of such cases. Up until recent decades, surgical excision of the abnormal mass was one of the main considerations in management of congenital cystic lung lesions. Nonetheless, over the last decade, there has been an increase in the popularity of conservative management

Figure 4. (a) Oblique-axial sonogram obtained from a male fetus at 15th week of gestation shows typical findings in CHAOS with bilateral hyperechogenic swollen lungs (arrow heads) that centrally relocated and severely compressed the fetal heart (arrow). This fetus suffered severe hydrops and was eventually demised at 28th week of gestation. (b) Axial sonogram of a male fetus at 31st week of gestation with type III CCAM. Unlike in CHAOS, the lung is unilaterally affected and caused the leftward displacement of the normal appearing fetal heart. The volume and echogenicity of the left lung are notably lower than that of the right lung.

of asymptomatic individuals.^{2,4} That being said, there is still no consensus between authors on whether these children should be followed up conservatively or undergo surgical removal, due mainly to the lack of long-term scientific evidence supporting the conservative approach.⁹ A recent meta-analysis, including one non-randomized prospective and eight retrospective studies, suggests elective resection of congenital pulmonary airway malformations to prevent further surgical complications during expectant management.¹⁰ The timing and indications of surgery in conservative management are still a matter of debate. There has been an assumption of increase in potential complications and morbidity rates of surgical performance once the patient becomes symptomatic, given infection of the underlying lesion.¹⁰ Lung function tests were also proposed in determination of necessity for surgical excision by some authors.¹¹ Those who advocate surgical treatment suggest an increase in potential side-effects such as pneumonia, long hospital stay, pneumothorax and bleeding after a procedure that has been delayed until the patient becomes symptomatic.^{1,8,10,12} Apart from that, perhaps the most important concern in conservative management is that a malignant lesion may arise within the cystic lung lesion. Little is known about the exact pathophysiology and etiology in such cases due to the extreme rarity of reported cases of malignancy.¹³

On the other hand, appropriate management of those who opted for follow-up is yet to be determined. Different groups have practiced various approaches.^{5,8,9} If the patient is being followed up conservatively, the method of follow-up should also be conservative as the cumulative ionizing radiation dose of frequent CT scans has potentially undesirable long-term outcomes. Direct posterior–anterior chest X-ray films are the initial work up in the evaluation of symptomatic presentations during follow-up period of children with lung lesions. Nevertheless, because of their inefficiency in showing small-sized lesions, X-ray studies are today being replaced by CT scans in most clinical settings. Ultrasound scans may provide excellent details of the lesion, particularly when they are adjacent to the thoracic wall. Owing to the relatively superficial location of the lesion, it was almost excellently depicted on ultrasound in the case presented here. With its low cost, real-time examination chance, and lack of ionizing radiation, ultrasound may emerge as an excellent modality of choice in such cases, especially in countryside hospitals where access to sophisticated tools is limited.

The relatively short duration of follow-up in the present case is the main limitation of this study. Well-designed prospective studies with large cohorts are needed to verify the efficacy and safety of this practice. In conclusion, ultrasound is proposed as a practical and

safe follow-up tool in amenable CCAM cases that have been conservatively followed up for a long period of time.

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Ethical approval

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Guarantor

MA.

Contributorship

MA researched literature, conceived and designed the study, wrote the manuscript and revised the final version of the manuscript. MA approved the final version of the manuscript.

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