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CASE REPORT

Early sonographic manifestation of fetal congenital lobar emphysema

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Abstract

Advanced fetal sonographic equipment has contributed to the increase in prenatal diagnosis of congenital thoracic malformations. Among these anomalies is congenital lobar emphysema (CLE), a rare congenital anomaly characterized by over distention and overexpansion of the involved fetal pulmonary lobe. Several studies addressed the prenatal diagnosis of CLE in mid second or early third trimester. The early prenatal diagnosis and the outcome of a case of CLE are reported in this study.

KEYWORDS

congenital, lobar emphysema, lung, prenatal diagnosis, ultrasound

1 | INTRODUCTION

Congenital lobar emphysema (CLE) is a rare lung malformation, characterized by overinflation of lung tissue, and is generally diagnosed postnatally.[1] The most common cause is the absence or hypoplasia of bronchial cartilage, which causes bronchial collapse during expiration; extrinsic bronchial compression is another possible cause.[2]

Most cases are diagnosed postnatally, with moderate respiratory distress after a few hours of life. A small percentage of the cases remain asymptomatic and are diagnosed incidentally.[3] Few studies have been published in the medical literature addressing the antenatal diagnosis of CLE. In most cases, the final diagnosis was made postnatally.[4,5] Oluyinka et al. reported two cases of CLE diagnosed at mid-gestation by ultrasound (US) and magnetic resonance imaging (MRI).[6] Here, we present the early prenatal US manifestations of CLE.

2 | CASE REPORT

A 33-year-old woman, gravida 2, para 0 was referred to our department for early US anatomical screening at 16 weeks. Both pregnancies were achieved after fertility treatments. The first pregnancy was terminated due to central nervous system fetal anomalies. The US examination showed an echogenic homogenous lesion in the left lung without cysts, with displacement of the heart to the right (Figure 1). The rest of the lung showed normal echogenicity, and no other fetal abnormalities were demonstrated. The US examination was repeated at 22 and 30 weeks, and confirmed the previous findings. Fetal echocardiography revealed displacement of the heart to the right without additional abnormal findings. The woman underwent amniocentesis, and a normal karyotype was detected. The differential diagnosis included CLE, congenital cystic adenomatoid malformation (CPAM), and pulmonary sequestration (PS). MRI performed at 33 weeks supported the diagnosis of CLE (Figure 2). At term, a female fetus weighing 3300 g was delivered vaginally. Postnatal chest radiograph confirmed the diagnosis of CLE (Figure 3), and lobectomy was performed at the age of 3 months. Seven years later, the girl is developing well; no pulmonary complications have been reported.

3 | DISCUSSION

With the advance of prenatal fetal US screening, most congenital thoracic malformations are currently detected on routine screening at the age of 18-20 weeks. The most common finding is a solid-appearing echogenic tumor or cysts, which may be accompanied by mediastinal

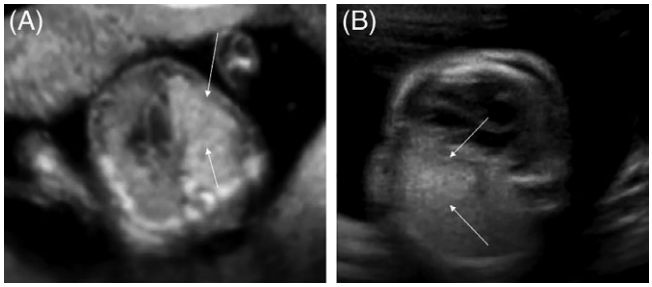

FIGURE 1 A and B, Sonograms of the chest showing an echogenic lung mass (arrows) and displacement of the heart to the right

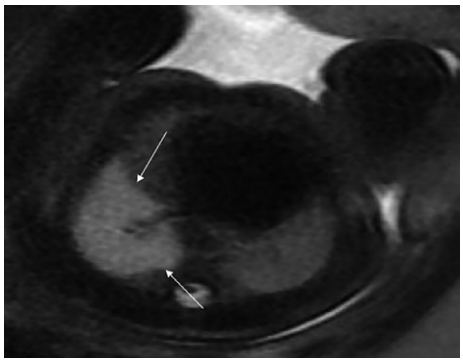

FIGURE 2 MR image at 33 gestational weeks revealed overexpanded lung parenchyma (arrows) and confirmed the diagnosis of CLE

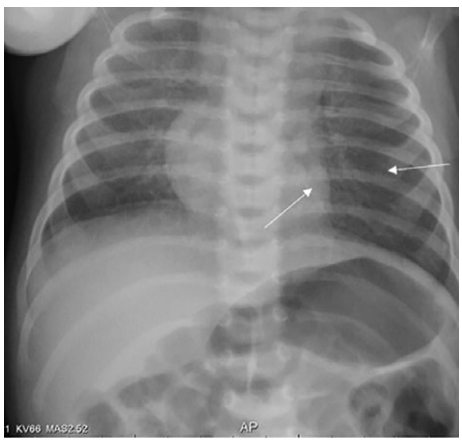

FIGURE 3 Postnatal chest X-ray showing overexpanded left lung

shift, and/or pleural effusion. The differential diagnosis includes CLE, CPAM, PS, bronchogenic cysts, and congenital diaphragmatic hernia. The combination of high-resolution US with MRI can differentiate the majority of lesions, and evaluate the lung, heart, and major vessels.[7]

The diagnosis of CLE remains a challenge and is not always included in the differential diagnosis of echogenic lung, presumably due to the rarity of its prenatal diagnosis. The exact cause of CLE is difficult to determine, and no apparent cause is found in 50% of cases. The most common identified cause, which accounts for 25% of cases, is a congenital bronchial cartilage defect, ranging from hypoplastic and flaccid tissue to its complete absence, with collapse of the bronchi in expiration and obstruction in deflation. Some of these cases have extra-pulmonary congenital cartilage defects, such as chondrodystrophy and chondroectodermal dysplasia. The remaining 25% is constituted by other causes of bronchial obstruction including—intramural, intraluminal, or external compression. Redundant mucosal folds or septum, mucous plugging, anomalous vessels, and, rarely, intrathoracic masses, may be involved.[3] Carrol et al. reported a case of CLE associated with cytomegalovirus.[4]

The postnatal course of CLE is variable. Most patients present with moderate respiratory distress, immediately or soon after birth. The symptoms worsen as the emphysematous lobe gradually enlarges. Cyanosis is the second most common finding. The next common mode of presentation is mild respiratory distress after the neonatal period. These patients suffer from recurrent respiratory infections and cough.

A small percentage of cases are asymptomatic and are picked up incidentally on a chest radiograph obtained for other reasons. Severe life-threatening respiratory distress is the least common form of presentation, but requires immediate surgical intervention.[3]

The symptoms developing after birth are attributed to the fetal lung physiology. During the fetal intrauterine life, the lungs have positive pressure due to liquid production which keeps the airways patent. Postnatally, airway collapse and air trapping occur as result of the negative intrathoracic pressure, leading to the clinical manifestations of CLE. In severe cases, the respiratory distress may be life-threatening, unless prompt treatment is applied. This scenario underscores the necessity of a correct diagnosis and immediate treatment.

Babu et al.[8] attempted to summarize the fetal sonographic findings in the medical literature, aiming to find any similarities that may help in prenatal prediction of CLE. No such similarities were found, and he concluded that the main advantage of prenatal US is to alert for the need of early postnatal investigations and treatment. In the present case, the prenatal US revealed a homogenous echogenic lung mass without a cystic component. Absence of aberrant blood vessels eliminated the diagnosis of PS, leaving the physician with the diagnoses of CLE or CPAM. Although the echogenic lung was demonstrated early in pregnancy, the diagnosis of CLE was confirmed at 33 weeks by MRI. Obviously, MRI could have been done earlier, which would have allowed a definite diagnosis in the first half of pregnancy. To the best of our knowledge, this is the first case of early prenatal sonographic diagnosis of CLE.

CONFLICT OF INTEREST

The authors declared no conflict of interest.

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REFERENCES

1. Kravitz RM. Congenital malformations of the lung. *Pediatr Clin North Am*. 1994;41:453-472.
2. Andrade CF, Ferreira HP d C, Fischer GB. Congenital lung malformations. *J Bras Pneumol*. 2011;37:259-271.
3. Karnak I, Şenocak ME, Ciftci AO, Büyükpamukçu N. Congenital lobar emphysema: diagnostic and therapeutic considerations. *J Pediatr Surg*. 1999;34:1347-1351.
4. Carrol ED, Campbell ME, Shaw BN, et al. Congenital lobar emphysema in congenital cytomegalovirus infection. *Pediatr Radiol*. 1996;26(12):900-902.
5. Lacy DE, Shaw NJ, Pilling DW, Walkinshaw S. Outcome of congenital lung abnormalities detected antenatally. *Acta Paediatr*. 1999;88:454-458.

6. Olutoye BOO, Coleman BG, Hubbard AM, et al. Prenatal diagnosis and management of congenital lobar emphysema. *J Pediatr Surg*. 2000;35:792-795.
7. Azizkhan RG, Crombleholme TM. Congenital cystic lung disease: contemporary antenatal and postnatal management. *Pediatr Surg Int*. 2008;24:643-657.
8. Babu R, Kyle P, Spicer RD. Prenatal sonographic features of congenital lobar emphysema. *Fetal Diagn Ther*. 2001;16:200-202.

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