FETAL CONGENITAL LOBAR EMPHYSEMA

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SUMMARY

Objective: To report a rare fetal congenital lung anomaly characterized by overinflation of a pulmonary lobe.

Case Report: A 28-year-old systemic lupus erythematous mother, gravida 1 para 0, who had normal prenatal care in our department, was admitted for labor pain and an abnormal fetal heart location was noted incidentally during labor. The baby showed rib retraction in room air but no obvious cyanotic change after delivery. Both the fetus chest X-ray and ultrasound showed a hyperechogenic tumor in the left thoracic cavity with a right-side-shifted heart and trachea. Computed tomography showed a hypodense and multiseptal tumor in the left thoracic cavity with right-sided shift of the heart and trachea. It was a soft, solid tumor in the parenchyma of the left lung and the histopathology confirmed it to be benign congenital lobar emphysema.

Conclusion: The favorable outcome in both asymptomatic and mildly symptomatic children suggests that a nonsurgical approach should be considered for these patients. [*Taiwanese J Obstet Gynecol* 2007;46(1):73–76]

Key Words: congenital lung tumor, lobar emphysema

Introduction

Congenital lobar emphysema (CLE) is a rare congenital abnormality characterized by overinflation of a pulmonary lobe, and it often presents a diagnostic and therapeutic dilemma. Traditionally, lobectomy has been advocated for all cases of CLE. However, with the use of ventilation/perfusion scintigraphy, cases may be managed nonsurgically while maintaining observation of regional anatomy and function.

Prenatal diagnosis of CLE has been rarely reported in the literature; here we report a fetus with an echogenic lung diagnosed during labor. The final diagnosis was made in the postnatal period as a result of follow-up of the prenatal findings, and this reinforces the importance of continuing postnatal investigation of prenatal sonographic hyperechogenic lung abnormalities [1].

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Case Report

A 28-year-old systemic lupus erythematous mother, gravida 1 para 0, was admitted to our ward for labor pain. She had received regular prenatal care in our obstetrics outpatient department since earlier in the first trimester of her pregnancy. No abnormalities, including thalassemia, Down's, gestational diabetes mellitus, and hepatitis B were found on screening. The prenatal ultrasounds (Level II), performed at 20 weeks of gestation, showed an appropriate gestational age fetus without obvious structural anomalies except a low-lying placenta. The last prenatal fetal ultrasound survey was 2 weeks before the onset of labor pain and it showed negative findings except for a low-lying placenta.

During admission, the nonstress test was reactive until the active phase. Variable deceleration was noted and profuse vaginal bleeding occurred when the cervical os was 6 cm. The ultrasound showed an abnormal presentation of the fetal head, which was in the occiput posterior position, and a low-lying placenta. An abnormal fetal heart location was noted incidentally. The fetal heart had been pushed to the right side of the thoracic cavity and a left thoracic tumor was suspected

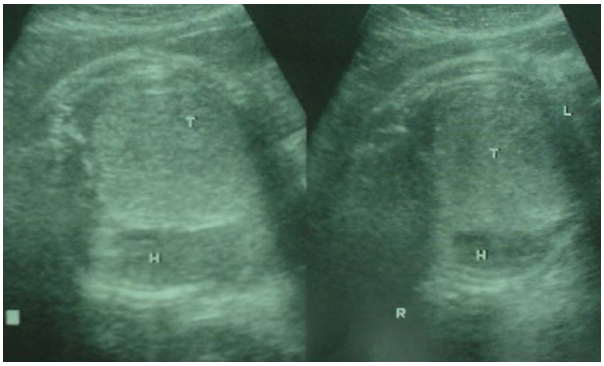

Figure 1. Prenatal ultrasound shows that the fetal heart (H) has been pushed to the right side of the thoracic cavity and a left thoracic tumor (T) was suspected.

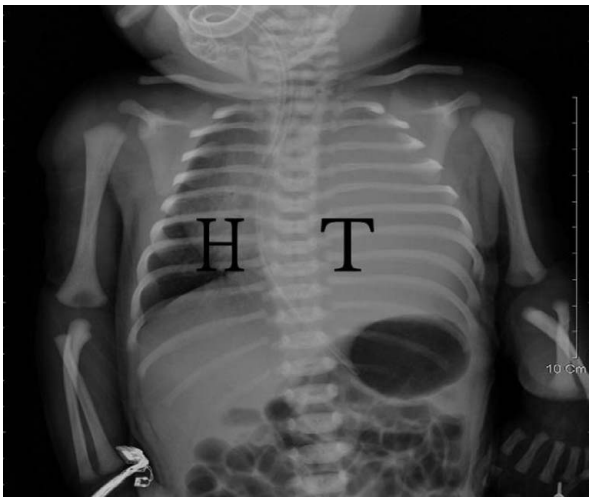

Figure 2. Fetal chest X-ray shows a hyperdense tumor (T) in the left thoracic cavity with a right-side-shifted heart (H) and trachea.

(Figure 1). Under the impression of acute fetal distress, with low-lying placenta and a congenital lung tumor, emergency cesarean section was performed.

During surgery, massive bleeding occurred during low-segment uterine incision, and a live baby was delivered occiput posteriorly by vacuum and smooth fundus pressure. The baby showed rib retraction in room air without obvious cyanotic change and was sent to the pediatric intensive care unit for further evaluation.

Both the chest X-ray and ultrasound of the fetus showed a hyperdense tumor in the left thoracic cavity with a right-side-shifted heart and trachea (Figure 2). The baby's arterial blood gas oxygen saturation was around 95% in room air without intubation and rose to 95~98% after oxygen supplementation. The chest and abdominal computed tomography (CT) scan showed a hypodense, multiseptal tumor in the left thoracic cavity with right-sided deviation of the heart and trachea (Figure 3). CT-guided aspiration biopsy was done but failed to obtain tissue proof before surgery. Surgery was

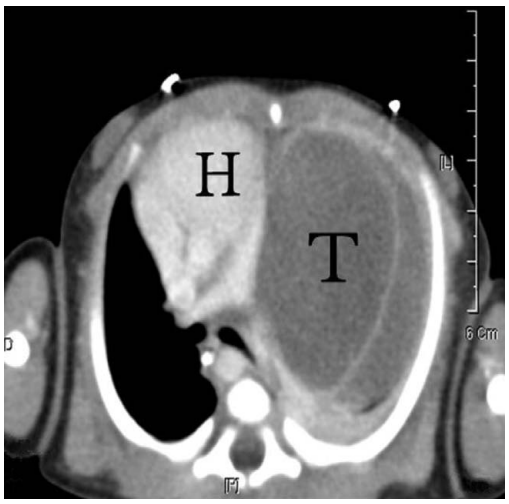

Figure 3. Chest and abdominal computed tomography shows a hypodense multiseptal tumor (T) in the left thoracic cavity with right-side-shifted heart (H) and trachea.

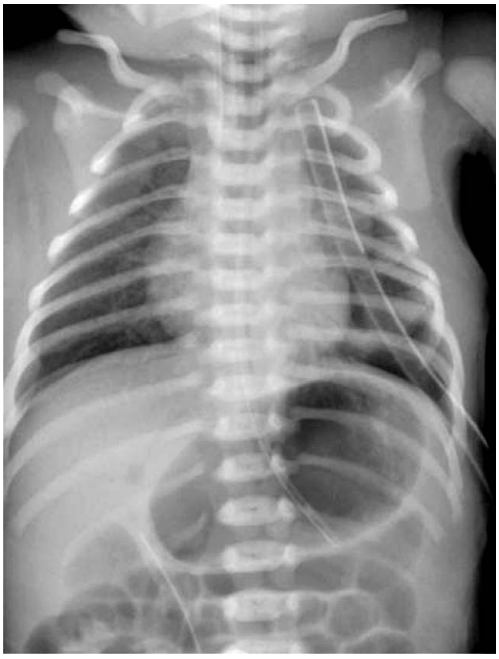

Figure 4. Chest X-ray on the second day after surgery shows a well-expanded left lung.

performed on the baby on the 2nd day after birth; a soft, solid tumor was found in the left lung parenchyma, and was totally resected. The postoperative chest X-ray showed a well-expanded left lung (Figure 4).

The final histopathology report confirmed that the tumor was a fluid-filled benign CLE (Figure 5). The specimen was a well-circumscribed tumor measuring 7.5 × 6 × 4.5 cm and weighing 65 g. The outer surface was smooth, glistening, and of a brownish color. On dissection, it revealed a yellow-tan cut surface with cystic areas containing clear, yellowish fluid and areas of hemorrhage. The postoperative period was uneventful

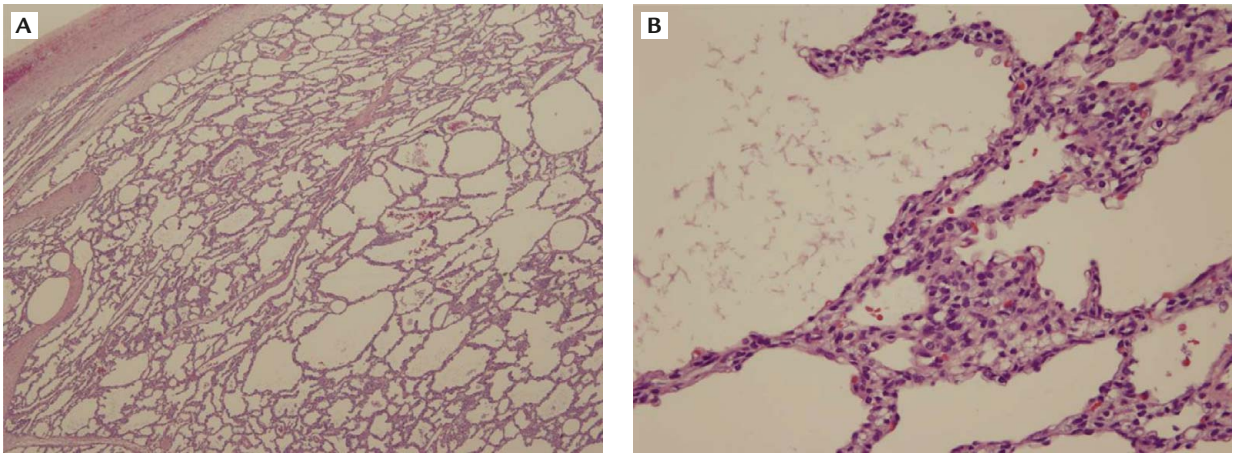

Figure 5. Histopathology shows a fluid-filled benign congenital lobar emphysema: (A) 40×; (B) 400×.

and the baby was discharged on the 11th day after surgery. Neither infant nor maternal chromosome studies were available.

Discussion

Primary tumors of the fetal lung are rare and most diagnoses are made during the neonatal period [2]. Primary pulmonary tumors reported in the perinatal period include pulmonary blastoma, fibrosarcoma, myofibromatosis, and hemangiomas [3]. Other mass lesions detected within the lung fields include vascular malformations, CLE, and bronchogenic cysts [4]. Mediastinal and thoracic wall masses are also reported, such as bronchogenic cyst, neurenteric cyst, esophageal duplication cyst, diaphragmatic hernia, pericardial cyst, lymphangioma, teratoma, and neuroblastoma [5]. Despite the feasibility of detecting lung lesions by antenatal ultrasound, there are problems in correlating the prenatal diagnosis with the final histology and in predicting the outcome. However, prenatally diagnosed echogenic lung has a good prognosis in the absence of hydrops. The ability to correctly assess echogenic lung lesions and the need for surgery by prenatal ultrasound is limited. [6]. Quinton et al [7] presented a fetus with an echogenic lung diagnosed at 18 weeks of gestation that resolved completely during the pregnancy. This case is unique in that sonographic-increased echogenicity was first noted at 18 weeks' gestation and disappeared by the 29th week. The diagnosis of CLE was made in the postnatal period as a result of follow-up of the prenatal findings. This reinforces the difficulty of correct prenatal diagnosis of CLE and the importance of continuing postnatal investigations of prenatal sonographic abnormalities even when they appear to have disappeared.

The management of CLE has traditionally been through surgical intervention but this is controversial for the asymptomatic patient, and should be considered in the light of the risk of infection and malignancy. Truitt et al [8] reported all CLE could be safely removed using endosurgical techniques. Due to the increased use of images, this lesion is frequently found in asymptomatic and mildly symptomatic children prompting some authors to adopt a more conservative approach in these children. CLE is thought to primarily occur by a check-valve mechanism and pulmonary resection is indicated for the majority of cases. Ayed et al [9] reported 47 infants with congenital lung diseases and the indications for surgery were respiratory distress in 32 patients (68%), respiratory tract infections in 12 patients (26%), and the presence of asymptomatic chest radiographic findings in three patients (6%). A lobectomy was performed in 42 patients (89%), bilobectomy in two patients (4%), left pneumonectomy in one patient (2%), and excision of a mass in two patients with extralobar sequestration (4%). Truitt et al reported that the management of asymptomatic congenital cystic lung lesions should be individualized according to the nature of the tumor: congenital cystic adenomatoid malformation, intralobar sequestration, and bronchogenic cyst needed surgical intervention at 3–6 months of age at the latest, while asymptomatic CLE may regress spontaneously and observation is warranted [10].

The natural history of prenatally diagnosed lung masses is variable and associated anomalies are rare. Planned term delivery and postnatal resection is advisable. Many extralobar pulmonary sequestrations dramatically decrease in size before birth and may not need treatment after birth. Fetal therapy is now an option for lung lesions associated with nonimmune hydrops [11].

In conclusion, the prenatal diagnosis of an echogenic lung has a good prognosis in the absence of hydrops,

and the ability to correctly assess echogenic lung lesions and the need for surgery prenatally is limited [12]. The favorable outcome in both asymptomatic and mildly symptomatic children suggests that a nonsurgical approach should be considered for these patients.

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