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CLINICAL CASE REPORT

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A Congenital Peribronchial Myofibroblastic Tumor Detected in a Premature Infant at 28 Weeks but That Resolved in the Late Stage of Pregnancy

A Case Report

Bo Xia, MD, Gang Yu, MD, Chun Hong, MD, Lei Zhang, MD, Jing Tang, MD, and Cuifen Liu, BA

Abstract: A congenital peribronchial myofibroblastic tumor (CPMT) is a rare benign tumor arising from the lungs. Although CPMT is a benign tumor, it is characterized by rapid growth, and is easily misdiagnosed during the prenatal period when the symptoms are nonspecific. The authors present a rare case of CPMT in a premature infant, which was detected at 28 weeks on ultrasonography (US) but resolved at a later stage of pregnancy. The knowledge concerning the diagnosis and management of CPMT is reviewed. Herein, the authors report of a 30-minute-old premature newborn infant in whom a pulmonary mass was discovered 1 month before delivery. Maternal prenatal US demonstrated a 0.8 × 1 cm well-defined oval-shaped mass in the left hemithorax in the 28th week of gestational age. The pulmonary mass, however, was not apparent on repeat US examination at 32 weeks. The child was delivered by cesarean section at 34 weeks estimated gestational age. Chest radiography and computed tomography revealed a mass-like lesion in the left lower pulmonary lobe. The chest computed tomography characteristics of the tumor included large size (4 cm), an irregular margin, and surrounding ground-glass opacity, which led to misdiagnosis as a malignant tumor. The patient underwent a left inferior lung lobectomy and was pathologically diagnosed with CPMT. He is currently alive 12-month postresection with no evidence of disease recurrence.

The authors report this rare case of CPMT, which was detected at 28 weeks and resolved at a later stage of pregnancy. Congenital peribronchial myofibroblastic tumor is an uncommon benign tumor. Lobectomy or pneumonectomy is often required. The prognosis after surgery is good.

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Abbreviations: CPMT = congenital peribronchial myofibroblastic tumor, CT = computed tomography, US = ultrasonography.

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From the Department of Maternal-Fetal Medicine, Guangdong Women and Children Hospital, Guangzhou, China.
Correspondence: Gang Yu, MD, Department of Maternal-Fetal Medicine, Guangdong Women and Children Hospital, Guangzhou 511400, China (e mail: yugang1959@126.com).
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INTRODUCTION

A congenital peribronchial myofibroblastic tumor (CPMT) is a rare benign lung tumor, and was originally described as an unusual hamartoma by Jones in 1949.[1] In 1993, McGinnis et al[1] coined the term CPMT for morphologically similar tumors. Congenital peribronchial myofibroblastic tumor is usually detected by prenatal imaging or in the immediate postnatal period because of associated symptoms, including polyhydramnios, hydrops fetalis, heart failure, intrauterine fetal death, and respiratory distress, among others.[2,3] The ultrasonography (US) features of CPMT are nonspecific, and it is usually diagnosed by biopsy or pathology. To our knowledge, less than 30 cases of CPMT have been reported in the English language literature. Herein, we report of a premature infant with CPMT, which was detected during pregnancy, but disappeared at a later prenatal stage. The literature was reviewed and analyzed to further understand this disease.

Case Report

A 30-minute-old premature infant initially presented to our hospital after detection of a pulmonary mass 1 month before delivery. The infant weighed 2570 g at 34 week estimated gestational age; cesarean section was performed for premature rupture of membranes. The Apgar scores were 9, 10, and 10 at 1, 5, and 10 minutes, respectively. He had normal vital signs, color, activity, and feeding, no respiratory difficulty, and normal stool and urine output.

The mother was 32 years old, and had an uncomplicated pregnancy; regularly scheduled prenatal US did not reveal polyhydramnios. A first-trimester US examination showed no significant abnormalities, but US at 28 weeks of gestational age showed a 0.8 × 1 cm relatively well-defined oval-shaped mass in the left hemithorax, without mediastinal shift (Fig. 1). When rechecked by using US at 32 weeks, the pulmonary mass, however, was not seen. Amniocentesis was performed, but karyotyping showed no distinct abnormality. Maternal blood test results were all negative for active infection by adenovirus, cytomegalovirus, or toxoplasma.

Chest radiography after admission demonstrated a mass-like opacity (3.3 × 2.8 cm) in the left lower lung. There were diffuse interstitial infiltrates in both the lungs, which suggested pneumonia (Fig. 2). Subsequent computed tomography revealed an iso- or hypo-dense mass located in the left lower lung. The tumor had an irregular margin and surrounding ground-glass opacity. After contrast administration, the tumor showed heterogeneous enhancement (Fig. 3). The results of a complete blood count, renal and liver biochemistry analysis, and resting electrocardiography were all normal.

Considering the findings and the history, malignancies, such as pleuropulmonary blastoma, and congenital pulmonary airway malformation were thought to be the most likely diagnoses.

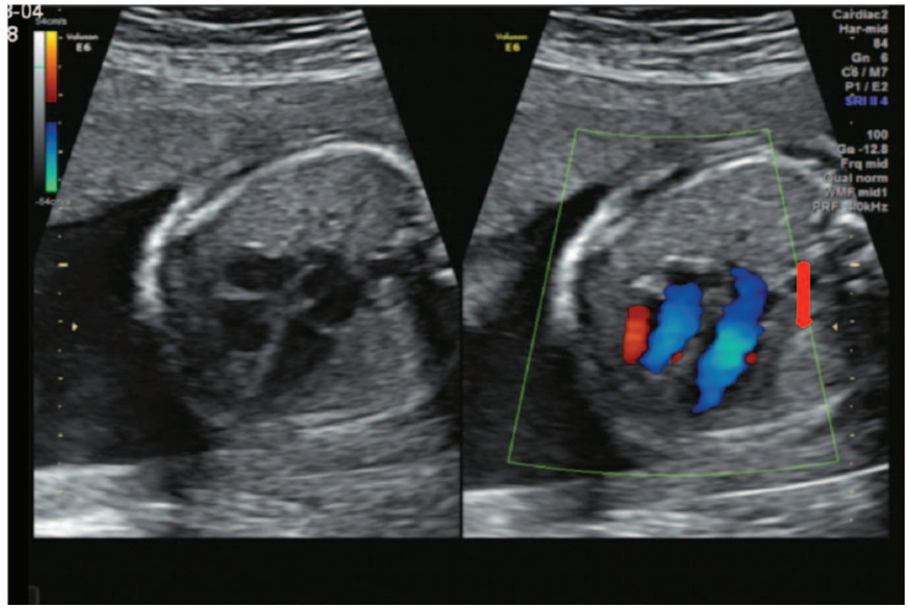

FIGURE 1. Ultrasonography at 28 weeks of gestational age showed a 0.8 × 1 cm relatively well-defined oval-shaped mass in the left hemithorax.

A left lower lobectomy revealed a 4.4 × 3.2 cm, large, grayish-white, rubbery mass replacing most of the left lower lobe. The mass did not have an anomalous blood supply and could not be separated from the pulmonary vasculature. Pathologically, the characteristics were those of immature cartilaginous tissue hyperplasia with muscle spindle-cell proliferation, with marked atypia, and increased karyokinesis (Fig. 4). Immunohistochemical staining of the tumor cells showed positivity for vimentin, smooth muscle actin, S-100, CD99, and p53, and negativity for creatine kinase, caldesmon, desmin, and CD34. Thus, the diagnosis was revised to CPMT, and no further adjuvant treatment, such as chemotherapy and/or radiotherapy was administered. The patient is currently alive 12 months postresection with no evidence of disease recurrence.

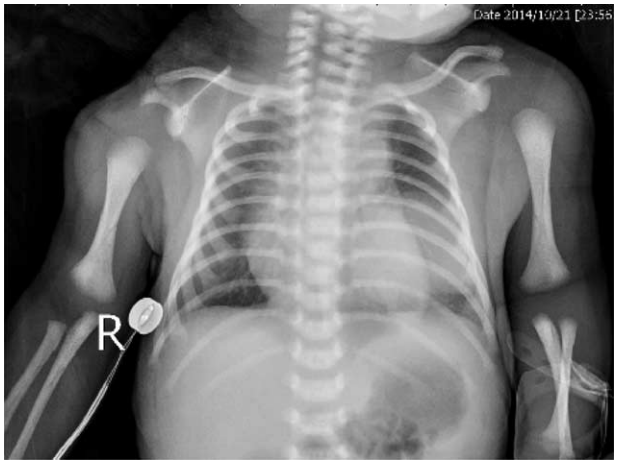

FIGURE 2. Chest x-ray demonstrated a mass-like opacity (3.3 × 2.8 cm) in the left lower lung. There were diffuse interstitial infiltrates in both the lungs, which suggested pneumonia.

DISCUSSION

Congenital peribronchial myofibroblastic tumor is a rare benign congenital primary lung mesenchymal tumor, accompanied by various degrees of cytologic atypia. There is no predilection for sex, lobe, or laterality. It is associated with respiratory distress, heart failure, polyhydramnios, fetal hydrops, and intrauterine fetal demise, because of the large size. In our case, there were no prenatal or postnatal symptoms, except for premature delivery. This may partly be because of the small size of the mass prenatally and because timely surgery was performed after delivery. Premature delivery was also reported in other literature, suggesting that CPMT may to some extent be related to premature delivery.[4,5] Because of the rarity of the case, this inference needs to be investigated in future studies.

Congenital peribronchial myofibroblastic tumor is generally detected on imaging in the prenatal or the immediate postnatal period. It typically presents as a large solid chest mass with mediastinal shift, potentially leading to intrauterine fetal demise, or respiratory distress at birth.[3] In this case, the tumor was detected at a diameter of 1 cm at 28 weeks, and grew to 4 cm by postnatal day 1. This shows that this tumor grew rapidly, which is consistent with previous reports.[2,6] This is a noteworthy characteristic of CPMT, because a mass with a rapid growth rate and the chest CT characteristics of a tumor usually leads to a diagnosis of malignancy. Therefore, CPMT should be considered in the differential diagnosis when a mass with a rapid growth rate is observed in the fetus. In addition, it is also worth noting that the mass could not be found on US in a later stage of pregnancy in our case. We speculate that the mass was present prenatally, but that it was difficult to distinguish between the tumor and surrounding lung tissue in late pregnancy US. This phenomenon was also reported in patients with congenital cyst adenomatoid malformations.[7] Thus, it may be difficult to diagnosis a CPMT without symptoms late in pregnancy.

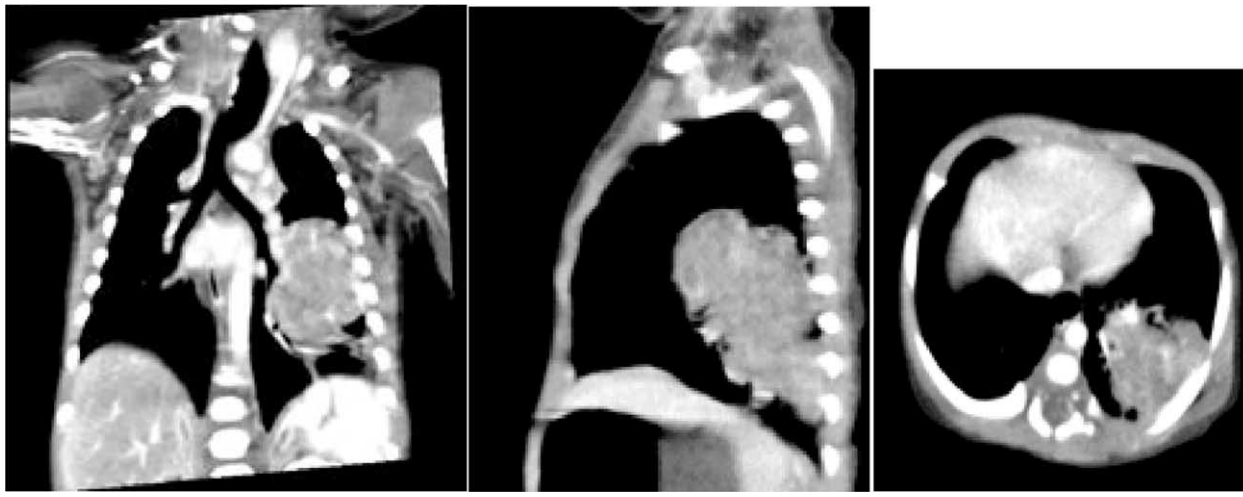

FIGURE 3. Computed tomography scanning revealed an iso- or hypo-dense mass located in left lower lung. The tumor had irregular margin and ground-glass opacification surrounded. After contrast administration, the tumor had heterogeneous enhancement.

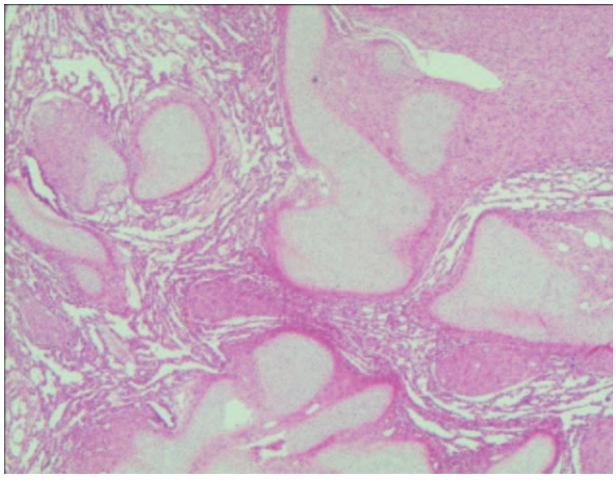

FIGURE 4. Pathology of the tumor, the characteristics were immature cartilage tissue hyperplasia, and muscle spindle-cell proliferation, the spindle cells proliferation were markedly atypical, and karyokinesis is increased.

Although CPMT is a benign tumor, the survival rate is only 36%, after excluding cases of elective termination. Because associated complications, such as mediastinal shift, hydrops, and polyhydramnios, can be caused by large lesions,[5] early surgical excision is recommended. The prognosis after surgery, however, is good; there are no reports of recurrence or metastasis after surgery.[6] Lobectomy or pneumonectomy is often required, as the tumor is associated with the bronchial tree. Chemotherapy and/or radiotherapy are not recommended for this benign tumor.[8] Our patient is currently alive 12 months postresection, with no evidence of disease recurrence.

CONCLUSIONS

In summary, we report this rare case of CPMT, which was detected at 28 weeks and resolved at a later stage of pregnancy. Congenital peribronchial myofibroblastic tumor is an uncommon benign tumor characterized by rapid growth. Early surgical excision is recommended. The prognosis after surgery is good.

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