Radiology Case Reports 19 (2024) 1463–1467

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Prenatal features of congenital peribronchial myofibroblastic tumor ☆,☆☆

Hitoshi Isohata[a], Tsutomu Yoshida[b], Itaru Sanoyama[c], Yu Yamazaki[a], Hiroyuki Goto[a], Yoshihiro Yoshimura[a], Kyoko Hattori[a], Takao Shimaoka[a], Kazuki Sekiguchi[a], Yoko Onishi[a], Kiyoshi Tanaka[d], Daigo Ochiai[a,]*

[a] *Department of Obstetrics and Gynecology, Kitasato University School of Medicine, Sagamihara, Japan*
[b] *Division of Molecular Pathology, Department of Comprehensive Medicine, Research and Development Center for New Medical Frontiers, Kitasato University School of Medicine, Sagamihara, Japan*
[c] *Department of Pathology, Kitasato University School of Medicine, Sagamihara, Japan*
[d] *Department of Pediatric Surgery, Kitasato University School of Medicine, Sagamihara, Japan*

ARTICLE INFO

Article history:
Received 30 October 2023
Accepted 1 January 2024

Keywords:
Congenital peribronchial myofibroblastic tumor
Prenatal diagnosis
Fetal MRI
Ultrasound

ABSTRACT

Here, we report a case of a congenital peribronchial myofibroblastic tumor (CPMT). A 34-year-old primigravida was referred to our hospital at 31 gestation weeks because of suspected congenital pulmonary airway malformation (CPAM). Fetal ultrasonography showed a mass measuring 4.6 × 4.0 × 3.9 cm with mixed high and low echogenicity in the left lung, which was associated with microvascular blood flow in the tumor. Fetal magnetic resonance imaging (MRI) revealed a low-intensity left lobe lung lesion on a T2-weighted image. These findings suggested that the mass was a CPAM with atypical hypointense findings on MRI T2-weighted images or a rare primary pulmonary tumor, such as a CPMT. Unfortunately, the fetus died in utero at 34 gestation weeks due to cardiovascular failure, which could have resulted from direct encasement of the great vessels or cardiac compression due to rapid tumor growth. The autopsy findings confirmed the diagnosis of CPMT. Primary pulmonary tumors, such as CPMT, are extremely rare lung diseases that develop in utero. These tumors often rapidly grow during pregnancy, resulting in intrauterine fetal death. However, if the patient survives surgical mass resection, the prognosis is good. Given the adverse outcomes observed in our case, careful fetal monitoring is required in case of suspected CPMT during the third trimester of pregnancy. Moreover, in case the well-being of the fetus cannot be assured, immediate delivery should be considered, even in the preterm period, followed by surgery.

☆ Acknowledgments: This work was supported by JSPS KAKENHI (grant numbers: JP21H03080)
☆☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
* Corresponding author.
E-mail address: ochiaidaigo@gmail.com (D. Ochiai).
https://doi.org/10.1016/j.radcr.2024.01.002

Introduction

Congenital lung lesions occur in 1 in 15,000 live births [1] and include abnormalities related to airway obstruction and malformations, such as congenital pulmonary airway malformation (CPAM) and primary pulmonary tumors, including congenital peribronchial myofibroblastic tumor (CPMT). CPMT is an extremely rare lung disease that develops in utero or in infancy [2,3]. CPMT can cause polyhydramnios, fetal hydrops, and intrauterine fetal death [2,4]. However, the characteristics of CPMT on fetal ultrasonography and magnetic resonance imaging (MRI) remain unclear [5,6]. This article describes a case of CPMT in which the tumor rapidly grew in the third trimester, which resulted in intrauterine fetal death.

Case report

A 34-year-old primigravida was referred to our hospital at $31^{2/7}$ gestation weeks due to a left lung lesion suggestive of CPAM. Fetal ultrasonography revealed a mass measuring 4.6 × 4.0 × 3.9 cm with a CPAM with a volume ratio of 0.41 as well as mixed high and low echogenicity in the left lung. Color Doppler ultrasonography revealed microvascular blood flow within the tumor, which was clearly visualized in the SlowflowHD mode (Fig. 1). The fetus was appropriate for date, and ultrasonography did not reveal structural defects; however, polyhydramnios (maximal vertical pocket, 9.2

Table 1 – Changes in the fetal ultrasound parameters. Note the increased CVR; however, the amniotic fluid volume, which reflects mediastinal compression, decreased to the normal range with the progress of pregnancy. CVR, CPAM volume ratio; MVP, maximum vertical pocket.

GW	CVR	MVP (cm)
$31^{2/7}$ wk	0.41	9.2
$32^{2/7}$ wk	0.30	6.9
$33^{1/7}$ wk	0.64	7.1
$34^{0/7}$ wk	1.02	5.5

GW, Gestational weeks.

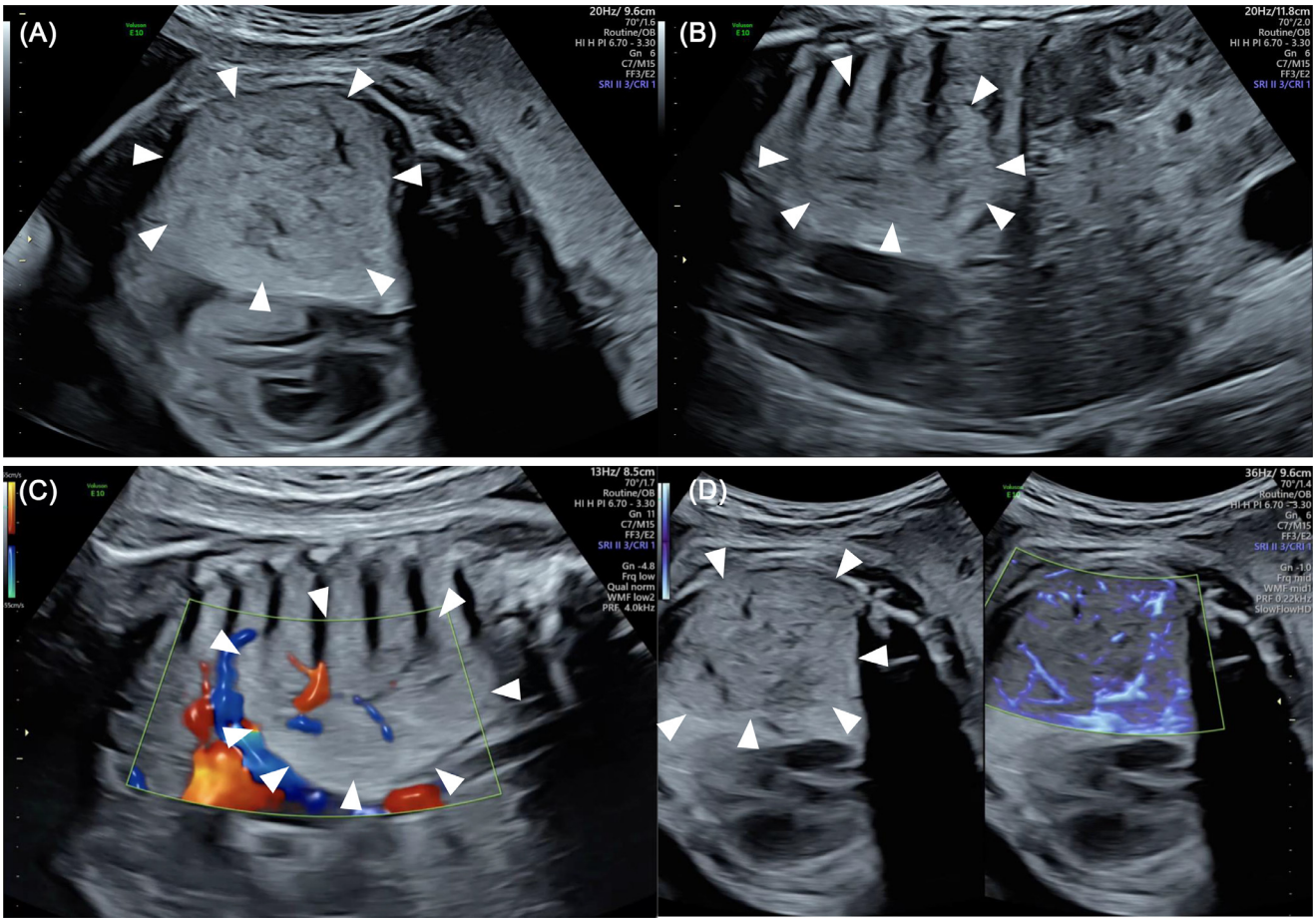

Fig. 1 – Fetal ultrasonography at 33 gestation weeks. Transverse (A) and sagittal (B) ultrasound sections demonstrate a heterogeneous lung lesion (arrowhead). Color Doppler (C) and SlowflowHD mode (D) showed blood flow within the tumor (arrowhead). The SlowflowHD mode allowed clear visualization of the microvascular flow in the tumor.

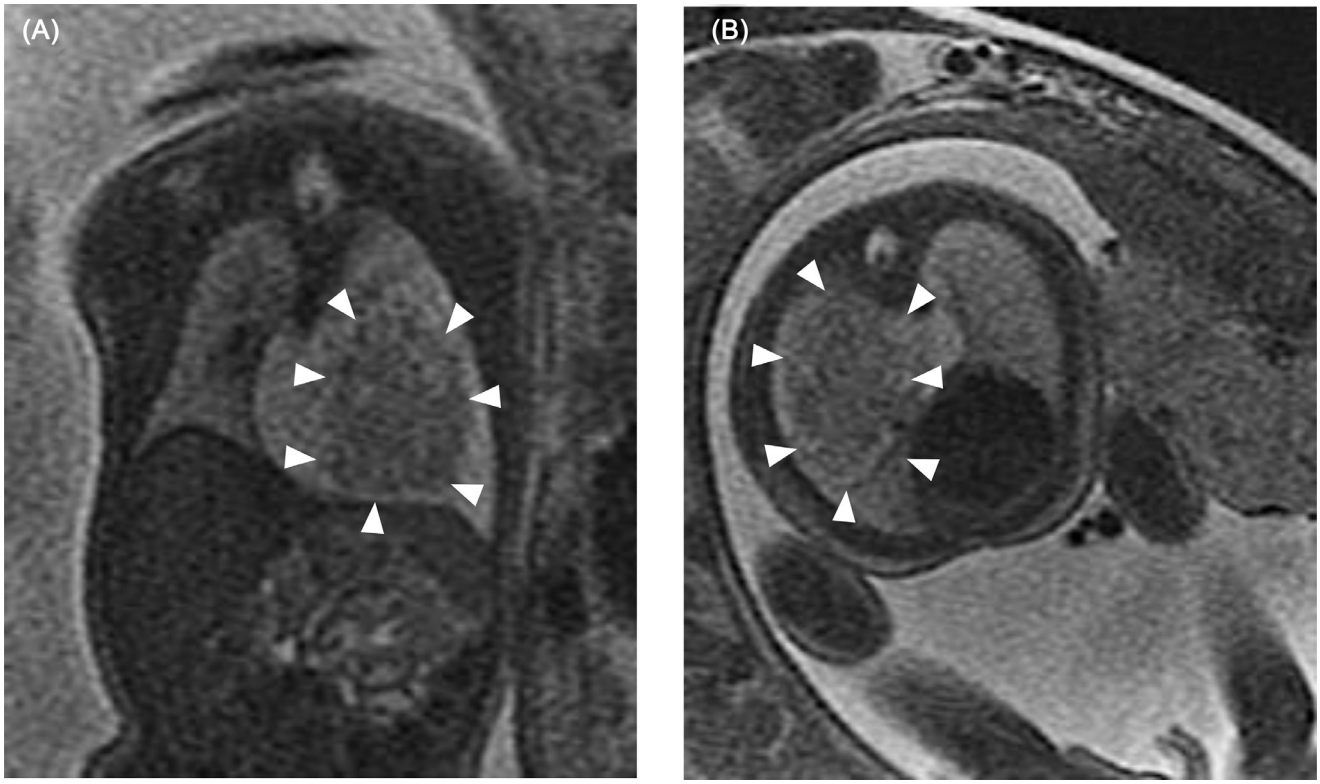

Fig. 2 – Coronal (A) and transverse (B) sections of T2-weighted MRI image of fetal CPMT. Note the T2- hypointense lung mass within the left lung (arrowhead).

cm) was observed, which was suggestive of mediastinal compression due to a mass (Table 1). Further evaluation using fetal MRI revealed a low-intensity left lobe lung lesion on T2-weighted images (Fig. 2). Based on these findings, we assumed that the mass was a CPAM with atypical hypointensity on T2-weighted images or a CPMT, which is an extremely rare condition.

The patient was followed up once per week. Although the mass gradually increased in size, the polyhydramnios improved (Table 1). Moreover, the biophysical and cardiovascular profile scores were within the normal ranges. The patient was followed-up as an outpatient. Unfortunately, the fetus died in utero at 34 $^{5/7}$ gestation weeks. After intravenous administration of oxytocin, the patient delivered the dead fetus, which weighed 2330 g.

Autopsy findings showed that the lower left lung lobe was occupied by a grayish-white solid mass (Fig. 3). Moreover, the findings were suggesting of vascular compression and circulatory failure due to the mass were also presumed. Histopathological examination revealed proliferation of uniform spindle-shaped cells along the bronchioles, with some of them showing chondrogenesis. Immunohistochemical staining was positive for vimentin and weakly positive for alpha-smooth muscle actin (Fig. 4); contrastingly, it was negative for desmin, CD34, AE1/AE3, TTF-1, and HHF-35. S-100 was focally positive in tumor cells near the cartilage. These pathological findings confirmed the diagnosis of CPMT.

Discussion

This article describes the prenatal features of CPMT, which is a rare congenital primary lung tumor. Differential diagnoses for primary lung masses in fetuses include various developmental abnormalities, such as CPAM, as well as rare congenital primary lung tumors, including CPMT, cystic pleuropulmonary blastoma, fetal lung interstitial tumor, and congenital fibrosarcoma [3,5–8]. CPMT is a rare solid fibroblastic/myofibroblastic tumor that develops in utero or during infancy and originates from pluripotent mesenchymal cells around the proximal bronchial branches [2].

Differentiation between CPAM and primary lung tumors such as CPMT based on fetal imaging findings is often challenging [5,6]. In our case, the sonographic findings of the CPMT included a solid pattern mixed with hyperechoic and hypoechoic components (Fig. 1), which differed in appearance from typical findings of CPAMs. There remains no consensus regarding the presence or absence of internal blood flow within CPMTs [7–10]. Advances in ultrasound devices have improved the accuracy of detecting blood flow inside tumors. For example, in our case, the SlowflowHD mode, which is a Doppler Ultrasound technology used to assess microvasculature characterized by small-sized vessels and low-velocity blood flow [11,12], allowed clarification of the blood flow within the tumor (Fig. 1). To our knowledge, this is the first report of the appli-

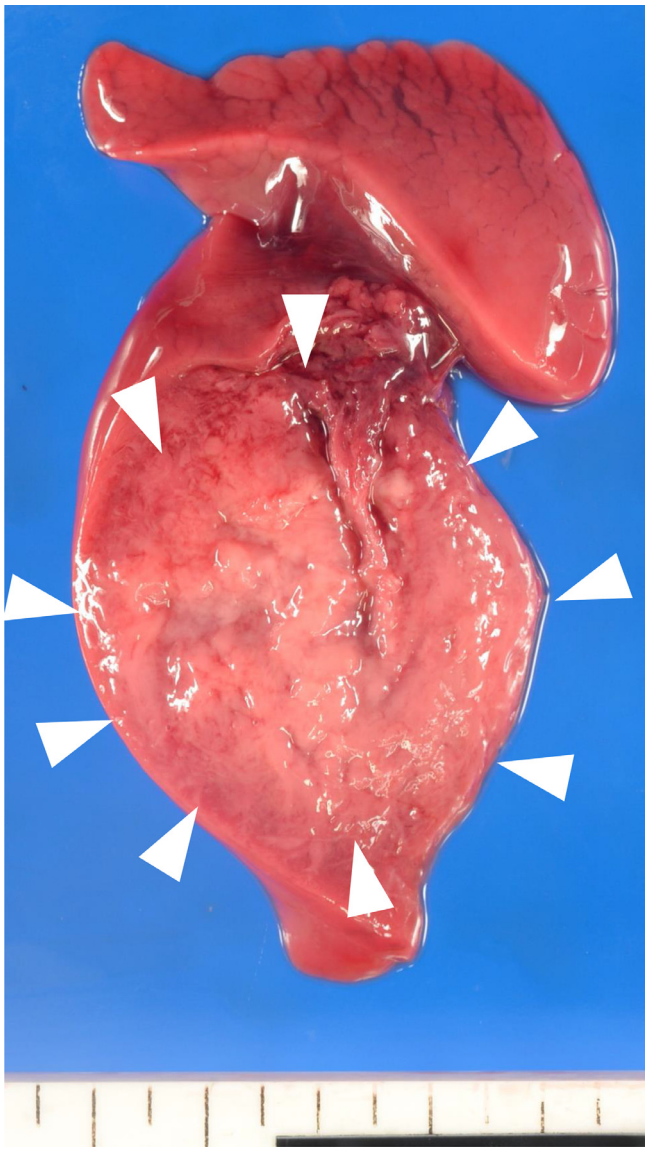

Fig. 3 – Gross pathological image shows that a tumor involves the entire lobe (arrowhead).

cation of this technology to fetal primary lung tumors, especially CPMT. Pathological examination suggested that the degree of mitosis might contribute to the speed of tumor growth. Therefore, quantitative assessment of microblood flow within a tumor may facilitate prediction of its subsequent growth rate.

In our case, the fetal MRI findings of CPMT demonstrated T2-hypointense elements (Fig. 2). CPAM lesions are often predominantly T2 hyperintense on fetal MRI. Consistent with our findings, a previous report characterized lung CPMT lesions as predominantly T2 hypointense and indicated that the degree of T2 hypointensity was correlated with the degree of immaturity [5]. Accordingly, we considered our case as atypical for CPAM and considered the possibility of CPMT.

Since the first report of CPMT in 1948, 25 cases have been reported [2]. Among them, 18 case reports described information regarding the prenatal period. These included 10 cases (10/18, 5 %) of fetal hydrops, 5 cases (5/18: 28%) of pleural effusion, and 7 cases (10/18, 34%) of polyhydramnios. The prognosis was poor in 9 cases (9/18, 50%), including fetal death, early neonatal death, or elective termination, while the remaining cases had a good prognosis after tumor resection during the neonatal period. Therefore, early surgical excision is recommended in case of fetal complications such as mediastinal shift, hydrops, and polyhydramnios, which can be attributable to large lesions. Although we observed improvement of polyhydramnios caused by mediastinal tumor compression, it caused fetal death due to cardiovascular failure, which could have resulted from direct encasement of the great vessels or cardiac compression from rapid tumor growth.

Surgical resection of the involved lung lobe remains the sole treatment option for CPMT. The prognosis is generally good if the infant can survive long enough for mass resection [2]. Given the adverse outcomes observed in our case, careful fetal monitoring is required in case of suspected CPMT during the third trimester of pregnancy. Moreover, in case the well-being of the fetus cannot be assured, immediate delivery should be considered, even in the preterm period, followed by surgery.

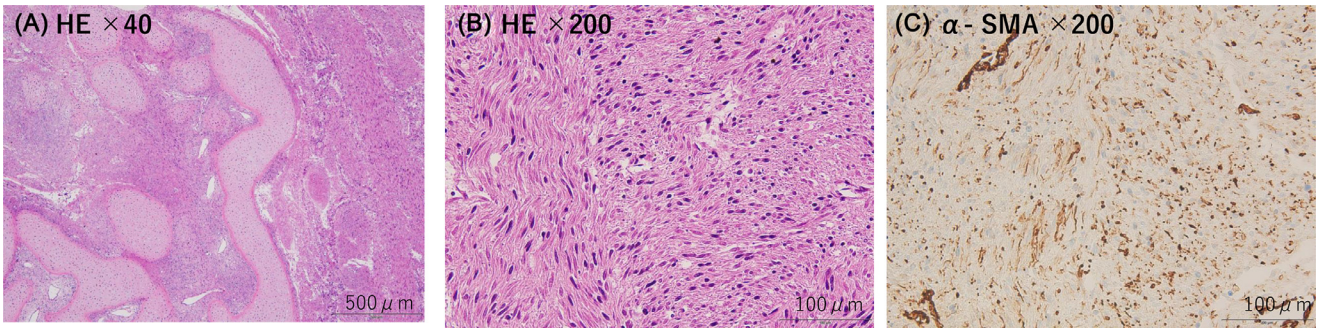

Fig. 4 – Microscopical images and immunohistochemical staining of the CPMT (A and B). Tumor cells surrounded the bronchi with enlarged immature cartilage plates (A: HE × 40). Spindle-shaped cells showing proliferation (B: HE × 200) and positivity for alpha-smooth muscle actin. (C: α-SMA × 200).

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

REFERENCES

[1] Cass DL, Olutoye OO, Cassady CI, Moise KJ, Johnson A, Papanna R, et al. Prenatal diagnosis and outcome of fetal lung masses. J Pediatr Surg 2011;46(2):292–8.

[2] Zhou P, Li S, Wang W, Tang Y, Jiang L. Congenital peribronchial myofibroblastic tumor (CPMT): a case report with long term follow-up and next-generation sequencing (NGS). BMC Pediatr 2023;23(1):184.

[3] Hotokebuchi Y, Kohashi K, Toyoshima S, Matsumoto N, Nakashima T, Oda Y. Congenital peribronchial myofibroblastic tumor. Pathol Int 2014;64(4):189–91.

[4] Brock KE, Wall J, Esquivel M, Newman B, Marina N, Albanese C, et al. Congenital peribronchial myofibroblastic tumor: case report of an asymptomatic infant with a rapidly enlarging pulmonary mass and review of the literature. Ann Clin Lab Sci 2015;45(1):83–9.

[5] Victoria T, Srinivasan AS, Pogoriler J, Kreiger PA, Laje P, Oliver ER, et al. The rare solid fetal lung lesion with T2-hypointense components: prenatal imaging findings with postnatal pathological correlation. Pediatr Radiol 2018;48(11):1556–66.

[6] Adams NC, Victoria T, Oliver ER, Moldenhauer JS, Adzick NS, Colleran GC. Fetal ultrasound and magnetic resonance imaging: a primer on how to interpret prenatal lung lesions. Pediatr Radiol 2020;50(13):1839–54.

[7] Tu YA, Lin WC, Chen HJ, Shih JC. Prenatal imaging and immunohistochemical analysis of congenital peribronchial myofibroblastic tumor. Ultrasound Obstet Gynecol 2015;46(2):247–9.

[8] Calvo-Garcia MA, Lim FY, Stanek J, Bitters C, Kline-Fath BM. Congenital peribronchial myofibroblastic tumor: prenatal imaging clues to differentiate from other fetal chest lesions. Pediatr Radiol 2014;44(4):479–83.

[9] Horikoshi T, Kikuchi A, Matsumoto Y, Tatematsu M, Takae K, Ogiso Y, et al. Fetal hydrops associated with congenital pulmonary myofibroblastic tumor. J Obstet Gynaecol Res 2005;31(6):552–5.

[10] Reiss A, Goldberg Y, Monichor M, Drugan A. Congenital pulmonary myofibroblastic tumor–pathology and prenatal sonographic appearance. Prenat Diagn 2005;25(11):1064–6.

[11] Abi Habib P, Seger L, Cagliyan E, Turan S. Topography of the heart: mapping the fetal heart through SlowflowHD. J Ultrasound Med 2023;42(8):1893–8.

[12] Hata T, Koyanagi A, Yamanishi T, Bouno S, Takayoshi R, Miyake T. Natural course of fetal hyaloid artery: SlowflowHD longitudinal study. J Ultrasound Med 2022;41(9):2259–67.