J Ped Surg Case Reports 3 (2015) 154–157

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Congenital peribronchial myofibroblastic tumor: Case report and review of literature

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ARTICLE INFO

Article history:
Received 30 December 2014
Received in revised form
10 February 2015
Accepted 19 February 2015

Key words:
Congenital peribronchial myofibroblastic tumor
Pulmonary neoplasm
Polyhydramnios

ABSTRACT

Congenital peribronchial myofibroblastic tumor (CPMT) is a rare entity recognized in the WHO classification of pulmonary neoplasms. According to available literature, it is a benign tumor with a high mortality rate exceeding 50%. It is partially attributed to polyhydramnios, hydrops, prematurity, respiratory distress or adverse surgical outcomes due to intraoperative bleeding. Herein we present a case of congenital peribronchial myofibroblastic tumor in a premature male infant who was born at 31 weeks gestation due to polyhydramnios and premature rupture of membranes. Soon after birth, he required intubation due to worsening respiratory distress. Imaging demonstrated a large right chest mass causing mediastinal shift. Surgical intervention was attempted, which was challenging due to intraoperative bleeding and tumor retraction. The patient expired soon after the surgery. Hence, in this report we would like to share our experience with this difficult diagnosis and treatment of this rare tumor.

1. Case report

A 27-year-old primigravida female at 31 weeks gestational age and previously uncomplicated pregnancy presented with premature rupture of membranes. Ultrasonogram demonstrated an intrauterine gestation with polyhydramnios and a fetus with a large solid and cystic mass in the right chest. After transfer to a tertiary care facility, a male infant weighting 2060 g was delivered vaginally with Apgar scores of 2 at 1 min, 3 at 5 min and 7 at 10 min. He required intubation with high frequency oscillator soon after the delivery. He was given surfactant to help with respirations. A chest CT scan demonstrated a homogenous round soft tissue density in the right hemithorax with a few small cystic spaces (Fig. 1A), compatible with a diagnosis of congenital pulmonary airway malformation (CPAM).

Due to mass effect leading to progressive collapse of the left lung, urgent resection of the mass was pursued on the 2nd day of life. Right posterior lateral thoracotomy was performed with intent to perform a left lung lobectomy. Surgical intervention proved to be challenging. The tumor invaded the right mainstem bronchus and gave the bronchus the character of thin glass. The surgeons retracted the lung to provide exposure of the hilum and the mainstem bronchus completely fractured off the trachea. The tumor itself appeared to be growing into the hilum and after fracturing the bronchus through what appeared to be the tumor bed, the surgeon was forced to place a Satinsky clamp across the hilum. Moreover, a tear of the superior pulmonary vein back into the atrium was noted. To control the bleeding and the airways, an emergency completion pneumonectomy was performed. A pediatric cardiac surgeon was also involved to prevent injury to the pericardium. Unfortunately, the contralateral lung, which was suspected to be hypoplastic on antenatal ultrasound, proved to be insufficient to support the child. After completion of the procedure the child's vital signs were heart rate of 130 bpm, blood pressure of 55/25 mmHg and oxygen saturation of 53%. The child was moved to the intensive care unit and required respiratory and vascular support. The patient passed away approximately 36 h after the operation due to inadequate pulmonary reserve.

Pathologic examination revealed a grossly trabeculated solid mass without a cystic component measuring 5.3 × 4.5 × 3.4 cm (Fig. 1B). On microscopy, the mass partially involved all three lung lobes, particularly at the bronchovascular margins. No hemorrhage or necrosis was identified. The tumor cells had uniform spindle

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http://dx.doi.org/10.1016/j.epsc.2015.02.012

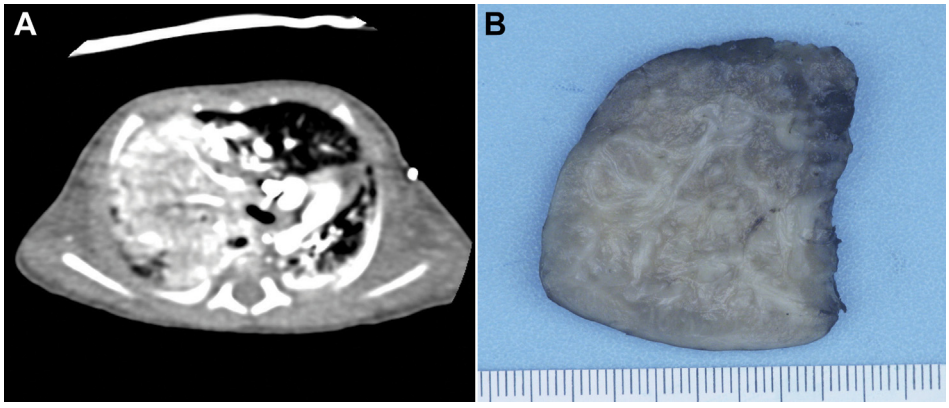

Fig. 1. A) Contrast CT of the chest, showing a large mostly solid and partially cystic right pulmonary mass; B) Gross photograph of the tumor showing trabecular solid cut surfaces.

morphology and showed bland nuclei with inconspicuous nucleoli and pink cytoplasm. They grew predominantly in broad fascicles, replacing normal lung parenchyma and abutting normal structures including bronchi and vessels (Fig. 2A–D). Brisk mitotic activity was present. The tumor cells were positive for smooth muscle actin, CD34, calponin and bcl2 immunohistochemical stains, supporting myofibroblastic differentiation (Fig. 2E and F). The AE1.3 cytokeratin cocktail (an epithelial marker) immunohistochemical stain is usually positive in the epithelial component of pleuropulmonary blastoma and CPAM but was negative in this case. The desmin (a muscle marker) immunohistochemical stain was also negative, ruling out leiomyosarcoma. The Ki-67 proliferation index was approximately 10%, which was consistent with a mitotically active tumor. Fluorescent in situ hybridization for the ETV6 rearrangement using a break apart probe was negative excluding congenital fibrosarcoma (congenital fibrosarcoma typically harbors an ETV6-NTRK3 gene fusion). Additionally, a Formalin-Fixed Paraffin-Embedded (FFPE) Molecular Inversion Probe Array was performed using the Affymetrix OncoScan FFPE Assay. A 777 Kb deletion at 2q21.1, Chr2: 130,701,189–131,478,017 (hg19) was identified in the tumor tissue as well as in normal lung tissue. This deletion overlaps with a database of the Genomic Variant Region and therefore, this copy number change was predicted to be most likely constitutional or benign. Overall, the morphologic assessment and immunohistochemical stains supported a diagnosis of congenital peribronchial myofibroblastic tumor.

2. Discussion

CPMT is a rare entity recognized in the WHO classification of pulmonary neoplasms of fetuses and infants. Only 23 reported cases are found in the literature (Fig. 3) [1]. Usually, this tumor presents with non-immune hydrops, polyhydramnios and premature rupture of membranes in the 3rd trimester. On imaging, a large solid mass in either left or right lung is identified that may exceed 5 cm [1]. There are no reports describing bilateral lung involvement. The differential diagnosis includes CPAM, bronchopulmonary blastoma, congenital fibrosarcoma and leiomyosarcoma. Prenatal

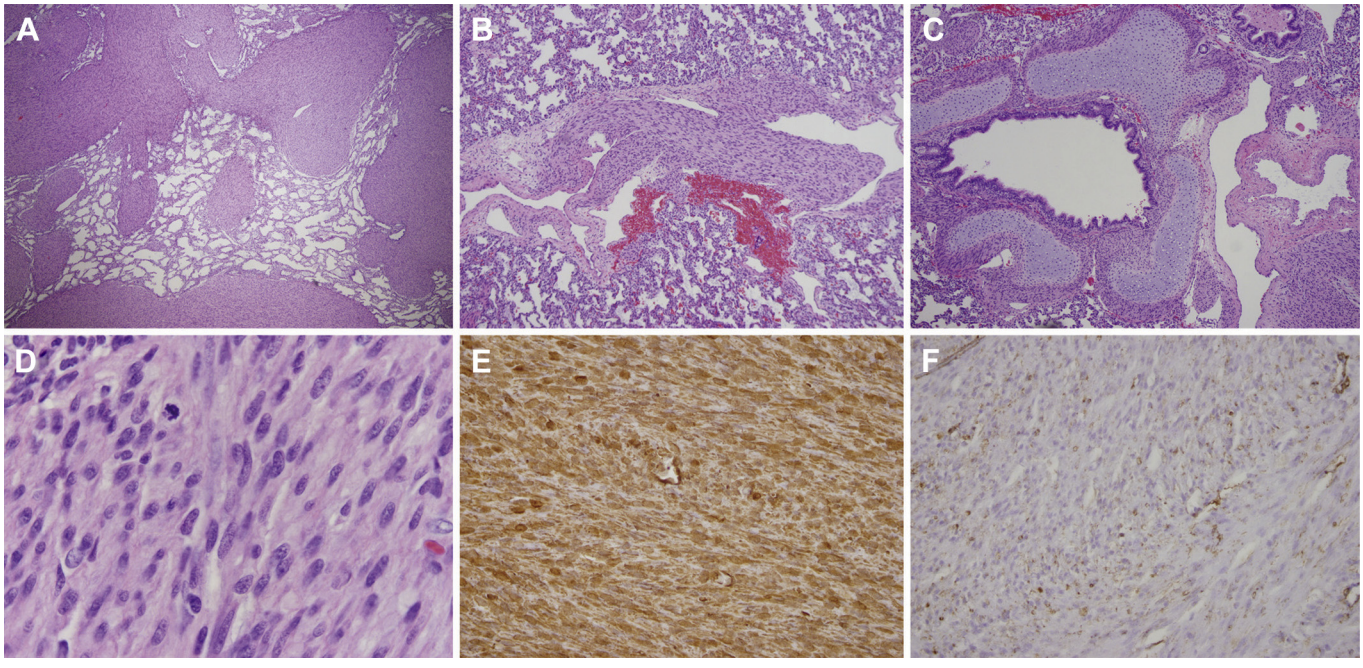

Fig. 2. A) Congenital peribronchial myofibroblastic tumor at low power, H and E stain 2×; B) Tumor tracking along the pulmonary septa, H and E stain 10×; C) Tumor abutting a larger bronchus, H and E stain 10×; D) Neoplastic cells on high power, H and E 100×; E) Positive vimentin immunohistochemical stain, 40×; F) Positive smooth muscle actin immunohistochemical stain, 40×.