Pleuropulmonary Blastoma (PPB) in an infant: Is the timing of an elective resection of neonatal lung lesions challenged?

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Abstract

Congenital Pulmonary Airway Malformations (CPAMs) are abnormalities of lung parenchyma that are often diagnosed upon prenatal imaging as opposed to postnatal symptoms. With a clinical presentation identical to CPAMs, Pleuropulmonary Blastoma (PPB) is a rare pulmonary neoplasm of highly malignant potential. We present a rare case of a female infant with a vague medical history of respiratory distress syndrome (RDS) at birth, presenting with a tension pneumothorax at three months of age, thought initially to be secondary to CPAM, but found to be PPB upon surgical resection and histological analysis. PPB is a rare pulmonary neoplasm of childhood that originates from the primitive interstitium of the lung, resulting in lesions that can be highly malignant. It is classified as type I (cystic), type II (cystic/solid) or type III (solid), with a progression of disease and worsening prognosis from type I to type III. Due to the cystic nature of CPAM and PPB it is difficult to differentiate on imaging alone; diagnosis must be made based on histological analysis. The highly malignant nature and potential for morbidity and mortality of PPB should make clinicians consider early resection of cystic lung lesions preferentially on an elective basis.

1. Case report

CD is a 3.17 kg female who was born at 39 weeks gestation with a suspicion of chorioamnionitis given malodorous amniotic fluid. Born apneic with decreased tone and bradycardia, she was transferred to the Neonatal Intensive Care Unit (NICU) for antibiotics, intravenous fluids and work up for sepsis. She had no further respiratory issues and therefore did not require any postnatal imaging. She was discharged from the hospital on day of life three.

She was seen by pediatric gastroenterology at approximately 1.5 months of life due to “fussiness.” Conservative dietary modifications were attempted for presumed milk protein allergy and gastro-esophageal reflux disease without improvement in her symptoms.

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Fig. 1. A) Chest X-ray on presentation to the Emergency Department, showing left sided pneumothorax with mediastinal shift to the contralateral side (left). Post chest tube insertion chest X-ray, showing resolution of pneumothorax and return of the mediastinum to the midline (right). B) Computed tomographic image of the chest with lesion in left upper lobe that is combination of solid and cystic components. C) Intraoperative photograph of the patient’s left upper lobe prior to its blood supply being ligated (left). The patient’s left upper lobe after being successfully removed from the patient (right). The gross specimen appeared cystic without much lung parenchyma. It was approximately 6 cm in diameter. D) The cyst walls are lined by cuboidal to columnar epithelial cells that overlie cellular fibrous stroma, with focal cartilage (left) (Hematoxylin and eosin stain, original magnification x100). The fibrous stroma of the cyst wall is focally composed of primitive small cells condensed in a cambium-like arrangement beneath the epithelium (right) (Hematoxylin and eosin stain, original magnification x400).
At 3 months of life she presented to the emergency department for new onset respiratory distress. A chest X-ray at this time showed a left sided tension pneumothorax with mediastinal shift. Angiography catheter decompression was performed followed by a chest tube placement (Fig. 1a).

A computed tomography of the chest was eventually obtained after re-expansion of her lung (Fig. 1b). An area in the left upper lobe of her lung appeared to have a combination of solid and cystic lesions, suspicious for a CPAM. On hospital day 3, she underwent a left upper lobectomy to ensure clear margins via a muscle sparing thoracotomy without complication (Fig. 1c) and was discharged home on hospital day 5. The final pathology demonstrated a Pleuropulmonary Blastoma Type I with subtile findings of rare cells that are small, rounded or spindle shaped and hyperchromatic located within the walls of the cyst (Fig. 1d). The cyst was completely excised with at least 0.4 cm margins.

A thorough family history was obtained identifying multiple other family members with cystic lung lesions suspicious for other CPAMs. On post-operative follow up the patient continues to do well 11 months after surgery.

2. Discussion

PPB was first described in 1988 by Manivel et al. with an overall prevalence of approximately 1 in 250,000 live births [1]. There are three morphologic types of PPB: Type 1 lesions are purely cystic, Type 2 have a combination of cystic and solid components, and Type 3 lesions are purely solid [2–5]. The type of lesions also reflects a continuum of disease progression. The age of presentation of each type represents a continuum of more aggressive lesions presenting later in time: for instance Type 1 PPB presents at 9 months (n = 25), Type 2 presents at 31 months (n = 67) and Type 3 PPB presents last at 42 months (n = 64) [5]. The prognosis is inversely proportional to the type of PPB, with Type 1 having a >90% cure rate, compared to approximately 50% survival for Types 2 and 3 [4,6,7]. PPB is also known to metastasize, most commonly to the brain, followed by bone and liver. The risk of metastasis also increases with advanced type. There are no reports of metastases for Type 1 PPB, whereas the 5 year risk of brain metastasis is 11% for Type 2 compared to 54% in Type 3. Most metastases occur within 24 months, with the median time to metastasis being 11.5 months [8].

Due to the rare nature of PPB, this case report demonstrates that although most congenital cystic lung lesions are benign, there is a rare subset that upon resection and pathologic analysis are found to be PPB, presenting as a congenital CPAM, but with dramatically different consequences. At 3 months of age, our patient is among the youngest patients to be diagnosed with PPB documented in the literature, including the PPB registry database [9–14]. The current treatment standards for PPB are surgical resection with clear margins, usually requiring at least a lobectomy. Adjuvant chemotherapy is needed only if the lesion is not completely resected or given an advanced type. It is important to note that surgery for CPAMs can be completed successfully in this age group, but is often delayed because of the conservative approach to CPAMs due to the fact that some cystic lung lesions regress spontaneously and a larger baby more easily allows for a minimally invasive resection. This case of PPB lesions in those 3 months of age highlights the importance of optimal timing for elective resection of cystic lung lesions, since delaying surgery should be weighed against a potential diagnosis of PPB.

3. Conclusion

PPB may be a rare entity, but presuming all CPAMs are benign, may overlook a potentially aggressive disease process, that if caught early, can be easily curable [4,6].

References