

Pleuro-pulmonary blastoma presenting as a chest wall deformity: a case report

Deformación de la pared torácica como presentación de un blastoma pleuropulmonar, caso clínico

Josefina Sáez B.^a, Juan Carlos Pattillo S.^b, José Fernando Vuletin S.^b

^aGeneral Surgery Resident, School of Medicine, Pontificia Universidad Católica de Chile

^bPediatric Surgery, Division of Surgery, School of Medicine, Pontificia Universidad Católica de Chile

Received: 25-07-2017; Accepted: 18-11-2017

Abstract

Introduction: Pleuropulmonary blastoma (PPB) is the most common primary malignancy of the lungs in childhood. It occurs more frequently in children between one and four years of age, and respiratory symptoms are a common manifestation. Three types have been defined (type I, II and III), which are related to survival and prognosis. **Objective:** To report the first case of a patient with a PPB who presented with a chest wall deformity. **Case report:** One year old male patient who had a chest wall deformity at ten months of age. Imaging revealed a giant cyst in the right hemithorax. He did not develop respiratory symptoms until hospital admission. A right upper lobectomy was performed and the biopsy confirmed a type I pleuropulmonary blastoma. He was considered successfully treated with complete surgical excision and routine follow-up with thoracic imaging is conducted. **Conclusions:** PPB is a very rare cancer that needs to be considered in the differential diagnosis of cystic lung diseases in children. The recognition of this lesion as a malignant tumour rather than a developmental cystic malformation is vital so the child can receive complete excision and appropriate follow-up care.

Keywords:

Lung neoplasm;
pleuropulmonary
blastoma;
chest wall deformity

Introduction

Pleuropulmonary blastoma (PPB) is the most common malignant pulmonary neoplasia in childhood¹. It is a rare and potentially aggressive tumor, most often reported in children under 5 years of age². Between 25 and 50 patients are diagnosed annually in the United States³. They usually develop respiratory symptoms: dyspnea, chest pain, cough, respiratory distress and/or pneumothorax⁴.

Manivel was the first to report it in 1988⁵. Since then, three types have been defined by histopathology: I or cystic, II or cystic/solid and III or solid¹. Type I PPB has the best prognosis, with a 5-year survival rate of 91%, types II and III reach 62% and may be associated with metastasis, most frequently in the central nervous system¹. PPB can evolve from cystic to solid states over time, cases that do not progress have been called I-r¹. The biopsy is the study that allows to classify lesions and, therefore, to define prognosis, guide future adjuvant therapies and appropriate follow-up⁴.

The objective of this study is to report the first case of a patient presenting a PPB as a deformation of the chest wall.

Clinical case

Healthy patient, male, 10 months old, with no family history of lung tumors or other neoplasms and no history of respiratory symptoms. Thoracic asymmetry was detected in routine pediatric outpatient care. In the physical examination, the right hemithorax was

bulging, without alterations in pulmonary auscultation. A chest x-ray revealed a lesion with multiple cystic images in the right hemithorax (Figure 1A and 1B).

The study was complemented with a chest computed tomography (CT), which showed a large multilocular, intraparenchymal cyst with no solid component, which caused compression of the chest wall tissues and structural alterations. (Figures 2A and 2B).

When the study was completed, the patient was one year old. An elective resection of the lesion was planned. At the admission, the patient was eupneic, for the first time with subcostal retraction, but without oxygen requirements.

A right thoracotomy approach was selected. Immediately upon entering the pleural cavity, a large cystic mass located in the upper lobe was found. It did not affect the parietal pleura and had no pleural effusion associated; there were not mediastinal adenopathies identified. The right upper lobe was decompressed and the lobectomy completed without complications, achieving lung re-expansion. The patient was extubated on the first postoperative day and discharged four days later, without presenting morbidity.

In the biopsy, a cystic, multiloculated, subpleural lesion was described, with cavities covered by cuboidal epithelium, immature fibrous stromal tissue, with necrotic areas and hemorrhages. No solid component was observed (Figure 3A and 3B); it was concluded a type I PPB in the diagnosis.

In the oncology consultation, the parents preferred not to accept chemotherapy and chest imaging follow-up was decided.

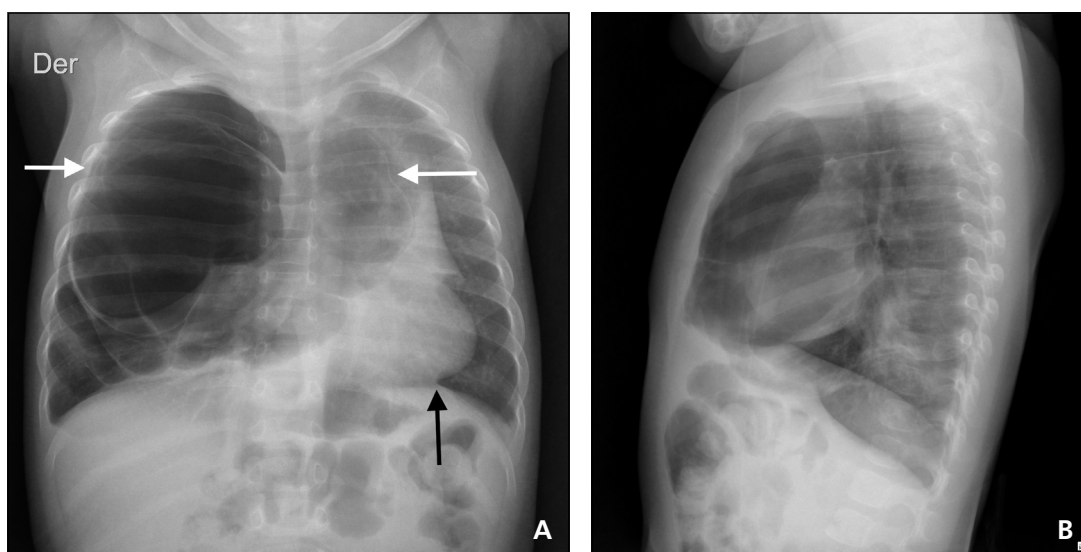


Figure 1. Antero-posterior (A) and lateral (B) Chest radiography. Thoracic asymmetry is evidences in relation to multiple thin walled cysts (white arrow) in the right hemithorax, which determines left mediastinal deviation (black arrow). The lateral projection shows both diaphragms descended, and a barrel shaped thorax, secondary to the increase in lung volumen. A secondary atelectatic phenomenon is observed in the retrocardiac region and the right lung appears herniated to the mediastinum.

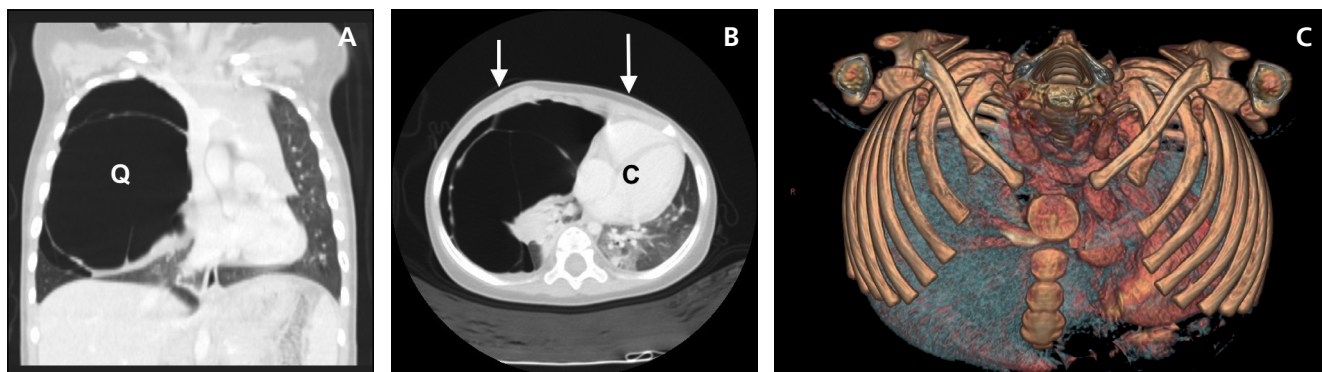


Figure 2. Computed tomography. **A:** Coronal reconstruction. A large intraparenchymal lung cystic formation displaces the mediastinum (Q). **B:** Axial cut. Thoracic asymmetry secondary to a multiloculated, right intraparenchymal cystic formation (arrows) is observed, it measures 9.7x9.4x10.2 cm and impresses to originate in the upper right lobe, raising partial atelectasis of the rest of the lung. No solid component is observed. Contralateral deviation of the heart and mediastinum (C) is also shown. Pneumothorax is not visualized.

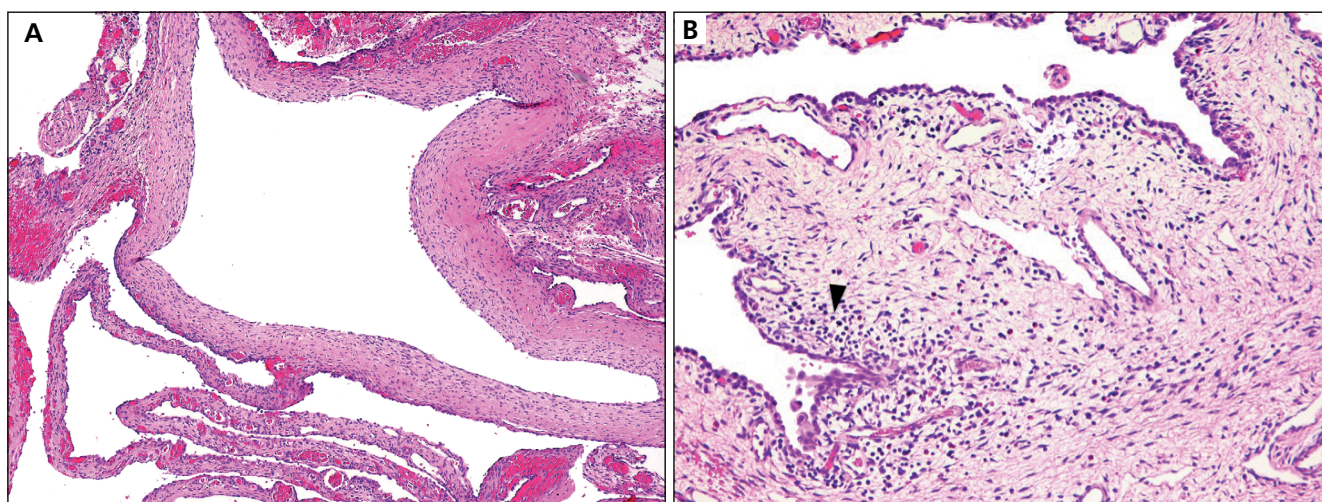


Figure 3. Microscopy (**A and B**), hematoxylin-eosin staining of the resected mass. In figure 3B it is possible to observe neoplastic cells under the epithelium. This transition zone (arrow) contains mesenchymal tumor cells with moderate atypia; small, round and with hyperchromatic nucleus. No solid component is observed.

Discussion

PPB represents 15% of pediatric lung tumors, up to 25% occur in children with family history⁶. 25-38% are associated with other neoplasms⁷: intestinal polyps, thyroid or gonadal tumors, cystic nephroma, and medulloblastoma⁸. The age at diagnosis is usually less than four years, with no gender preference and it mostly occurs on the right side⁶.

Genetic studies have allowed associating mutations with the development of PPB, including mutations in the p53 tumor suppressor gene, germline DICER1 alterations, chromosome 8 and trisomy 2 gains, and unbalanced X;1 translocations⁶.

A recent article that gathered 350 cases showed

that type I PPBs occur more frequently in men during the first year of life, earlier than types II or III. Most of them were unilateral, larger than five cm and only half were multiloculated¹.

The differential diagnosis of cystic pulmonary masses should consider both congenital airway malformation (*cystic adenomatoid malformation*) and type I PPB. Suggestive findings of PPB include cystic lesions (especially peripheral) without clear etiology, lack of prenatal diagnosis with normal anatomical study of the second trimester, rapid growth, pneumothorax and/or family history.

The treatment of asymptomatic lung injuries in childhood is still controversial⁹. In favor of early resections is the risk of malignancy¹⁰. We believe that all as-

ymptomatic lung lesions in children should be studied with chest CT at around two months of age, preferring resections between three to nine months given the risk of recurrent infections or malignancy^{7,11,12}. Elective surgery is safe, prevents the development of symptoms and reduces complications^{13,14}.

PPB usually produces respiratory symptoms, but it may be an incidental diagnosis¹⁵. Although between diagnosis and the definitive treatment the patient developed respiratory distress manifested as a subcostal retraction, according to our search, this is the first report of a PPB that was initially presented as a thoracic deformity.

The treatment is multimodal. Complete surgical resection is essential: it gives the best chance of long-term survival. Radiotherapy has a role in recurrence or residual disease^{15,16}. Current recommendations for type I include adjuvant chemotherapy, but it is not yet clear if it prevents progression¹⁵. Follow-up with chest CT is required until four or five years of age. In lung-limited disease, the complete resection is associated with good results; the rigorous follow-up should detect recurrences and may be an alternative^{1,11,12}.

In conclusion, the case of a PPB presented as thoracic deformity is reported, a form of presentation not previously described in the literature. PPB is a rare neoplasm that should be considered in the differential diagnosis of cystic lung lesions in children. Recognizing it as a neoplasm with malignant potential rather than a developmental abnormality is crucial for the

patient to receive appropriate surgical treatment and follow-up care.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Financial Disclosure

Authors state that no economic support has been associated with the present study.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

References

- Messinger YH, Stewart DR, Priest JR, et al. Pleuropulmonary blastoma: a report on 350 central pathology-confirmed pleuropulmonary blastoma cases by the International Pleuropulmonary Blastoma Registry. *Cancer*. 2015;121:276-85.
- Hashemi A, Souzani A, Souzani A, Keshavarzi S. Pleuropulmonary Blastoma in Children: A Case Report. *Iran J Cancer Prev*. 2012;5(2):105-7.
- Priest JR. Pleuropulmonary Blastoma. In: Schneider D, Brecht I, Olson T, Ferrari A. (eds) *Rare Tumors In Children and Adolescents*. Pediatric Oncology. Springer, Berlin, Heidelberg 2012.
- Brodowska-Kania D, Kotwica E, Paturej A, et al. What do we know about pulmonary blastoma? review of literature and clinical case report. *Nagoya Journal of Medical Science*. 2016;78(4):507-16.
- Manivel JC, Priest JR, Watterson J, et al. Pleuropulmonary blastoma: the so-called pulmonary blastoma of childhood. *Cancer* 1988;62(8):1516-26.
- Amjad Ali Khan, Ahmed Kamal El-Borai, Mohammad Alnoaiji. "Pleuropulmonary Blastoma: A Case Report and Review of the Literature", *Case Rep Pathol*. 2014;509086.
- Priest JR, Williams GM, Hill DA, Dehner LP, Jaffé A. Pulmonary cysts in early childhood and the risk of malignancy. *Pediatr Pulmonol*. 2009;44(1):14-30.
- Coleman A, Kline-Fath B, Stanek J, Lim FY. Pleuropulmonary blastoma in a neonate diagnosed prenatally as a congenital pulmonary airway malformation. *Fetal Diagn Ther*. 2016;39(3):234-7.
- Parikh DR, Rasiah S. Congenital lung lesions: Postnatal Management and outcome. *Seminars in Ped Surg*. 2015;24:160-7.
- Feinberg A, Hall N, Williams G, Schultz K, Miniati D, Hill A, Dehner L, Messinger Y, Langer J. Can congenital pulmonary airway malformation be distinguished from thype I pleuropulmonary blastoma base don clinical and radiological features? *J Pediatr Surg*. 2016;51:33-7.
- Miniati DN, Chintagumpala M, Langston C, Dishop MK, Olutoye OO, Nuchtern JG, Cass DL. Prenatal presentation and outcome of children with pleuropulmonary blastoma. *J Pediatr Surg*. 2006;41:66-71.
- Yong PJ, Von Dadelszen P, Carpara D, Lim K, Kent N, Tessier F, Delisle MF, Wong T, Blair G, Skarsgard ED. Prediction of pediatric outcome after prenatal diagnosis and expectant antenatal management of congenital cystic adenomatoid malformation. *Fetal Diagn Ther*. 2012;31:94-102.
- Kapralik J, Wayne C, Chan E, Nasr A. Surgical versus conservative Management of congenital pulmonary airway malformation in children: A systematic review and metaanalysis. *J Pediatr Surg*. 2016;51:508-12.
- Furukawa T, Kimura O, Sakai K, Higashi M, Fumino S, Aoi S, Tajiri T. Surgical

- intervention strategies for pediatric congenital cystic lesions of the lungs: A 20 year single institution experience. *J Pediatr Surg.* 2015;50:2025-7.
15. Christosova IR, Avramova BE, Drebov RS, Shivachev HI, Kamenova MA, Bobev DG, Brankov OG. Diagnosis and treatment of pleuropulmonary blastoma-single center experience. *Pediatr Pulmonol.* 2015;50:698-703.
16. Priest JR, Hill DA, Williams GM, et al. Type I pleuropulmonary blastoma; a report from the International Pleuropulmonary Blastoma Registry. *J Clin Oncol* 2006;24:4492-8.