A rare case report of simultaneous occurrence of a pediatric pleuropulmonary blastoma and an intralobar pulmonary sequestration

Marjeta Tanka, PhD, Anila Kristo, PhD, Dritan Alushani, Associate Professor, Irena Kasmi, PhD, Nikollaq Leka, Associate Professor

Pediatric Department, Imagery Service, University Hospital Center “Mother Tereza”, Tirana, Albania
Morphology Department, Faculty of Medicine, University of Medicine, Tirana, Albania
Pediatric Department, Surgery Service, UHC Mother Tereza, Tirana, Albania
Pediatric Department, UHC “Mother Tereza”, Tirana, Albania
Morphology Department, Faculty of Medicine, University of Medicine, Tirane, Albania

A B S T R A C T

We are presenting a rare case with the simultaneous occurrence of pleuropulmonary blastoma and an intra lobar pulmonary sequestration. Although there have been cases reported previously with pleuropulmonary blastoma associated with congenital pulmonary malformations, the association with an intra lobar pulmonary sequestration is very rare. The patient, a female, 6-month-old child arrived at our pediatric service with the clinic of cough, respiratory distress, and fever after being treated for 2 weeks for left lung bronchopneumonia according clinical signs and radiographic description but without clinical improvements. Contrast enhanced CT images showed the simultaneous presence of 2 different lesions in the left lung, a heterogeneous mass in the superior lobe without delineation with mediastinal structure compatible with a pleuropulmonary blastoma and a consolidation in the inferior lobe with bronchogram present and a systemic vessel feeding compatible with an intra lobar pulmonary sequestration, both confirmed by histologic examinations after the surgical intervention. Although it is very rare, the simultaneous presence of these distinct embryogenic lesions may occur and radiologist should be aware as the imaging diagnosis may be very helpful for the further management of the patient.

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Introduction

Pleuropulmonary blastoma (PPB) is the most common primary malignancy of the lung in pediatric age, an aggressive em-
I PPB progress to the more malignant types and these cystic cases are classified as type I regressed (type I) [4]. Signs and symptoms vary according to the different subtypes. The typical clinic sign of type I tumors is respiratory distress due to air-filled cysts associated or not with pneumothorax while the most frequent clinical signs of types II and III tumors are dyspnea, fever, and chest pain [4,5,6].

On the other hand, bronchopulmonary sequestration (BPS) is a rare tumor described firstly from Pryce in 1946 [7]. It is part of the group of congenital pulmonary airway malformations (CPAM). CPAM are a group of non–malignant developmental anomalies which include a varied group of disorders like congenital cystic adenomatoid malformation, bronchopulmonary sequestration, congenital lobar emphysema, bronchogenic cyst etc. CPAM are relatively rare, with a reported incidence between 1:25000 and 1:35000, while BPS account for 0.15%-6.4% of all congenital pulmonary malformations [8].

The main feature of this disease is that partial lung tissues separate from the main lung during the embryonic period, grow to generate a cystic mass without lung function and receive its blood supply from systemic circulation arteries, mainly from thoracic aorta and abdominal aorta. Its pathogenesis is not clear and is categorized into congenital and acquired BPS. Acquired theory was proposed by Pryce but an increasing number of studies support the congenital theory which explains that BPS is formed by the growth of the primitive foregut ventral side lung bud during embryonic development period [9,10].

BPS is usually classified as either intra lobar sequestration (ILS) which is located within a lung lobe and shares the visceral pleura with the corresponding lung lobe or extra lobar sequestration (ELS) which is a separate mass of lung parenchyma enclosed entirely by a separate pleural envelope. Intra lobar sequestrations account for 75% of pulmonary sequestrations, being more frequent within the lower lobe (98%) and more often in the left lung (55%) while extra lobar sequestrations constitute the remaining 25% and are typically found between diaphragm and the lower lobe [11].

We are reporting a rare case with the occurrence of these 2 distinct lesions simultaneously; a pleuropulmonary blastoma associated with an intra lobar pulmonary sequestration.

Case report

A female patient, 6-month-old child was presented at the Emergency Pediatric Department of University Hospital Center “Mother Teresa” with the clinic of cough, respiratory distress, and fever. The patient was treated for a period of 2 weeks for a left lung pulmonary bronchopneumonia according clinical signs and radiographic description. The Chest X-ray showed a consolidation in the inferior lobe of left lung with fine bronchogram present and also a small consolidation in the superior lobe of left lung. Contrast-enhanced CT images revealed a heterogeneous lesion in the superior lobe of left lung without delineation with mediastinal structure and minimal left second foramina dilatation (Fig. 1). The radiological diagnosis of this lesion was in favor of pleuropulmonary blastoma, confirmed later by the histopathologic examination after surgical intervention.

CT images revealed also the presence of a consolidation in the inferior lobe of left lung with the presence of bronchogram. The systemic vessel feeding this mass was an accessory branch of thoracic aorta (Fig. 2). The lung consolidation was compatible with pulmonary sequestration confirmed also by the histopathologic examination after surgery.

Discussion

Pleuropulmonary blastoma and pulmonary sequestration represent 2 distinct entities with different prognosis. Although there are case reports of suspected malignant degeneration of CPAM (the group of which BPS account) into sarcomas or other tumor types, in general CPAM and PBB are now regarded as separate entities [12,13,14]. In general, the most frequent reported radiographic features associated with a high likelihood of PBB are heterogeneous low attenuation mass, pleural effusion, contralateral mediastinal shift, and lack of chest wall invasion [15]. In children the lesions which may be confused are type I purely cystic PBB with one of several congenital cystic lesions of the lungs part of congenital pulmonary airway malformations (CPAM) and in these cases a careful pathologic examination is required [5,12,16]. In our case the nature of lesion, a heterogeneous mass, without delineation with mediastinal structure and the presence of minimal left second foramina dilatation were strong radiological features suggesting the diagnosis of PBB type II. In fact, prior to 12 months’ age, type I PBB lesions are more frequent while type II and III lesions are rarely seen [16] as in our case.

The importance of making the differential diagnosis between PBB and CCAM including pulmonary sequestrations is the high grade of malignancy of the PBB lesions especially type II and III which require surgical resection and subsequent chemotherapy [17]. Also, it is well known that PBB are part of DICER-1 gene syndrome where germline mutations of this gene are associated with increased risk of a wide variety of neoplastic conditions, including pleuropulmonary blastoma, cystic nephroma, nasal chondromesenchymal hamartoma, ovarian Sertoli-Leydig cell tumors, pituitary blastoma, nodular thyroid hyperplasia or thyroid carcinoma etc. Patients should be evaluated very carefully to exclude the presence of a concomitant pathology and surveillance strategies are recommended too [4,18]. So pediatric patients with cystic lung masses should be the subject of DICER1 germline testing especially when the appearance of cysts is with septa, multiple, bilateral, or identified in infancy. In addition, patients who are asymptomatic and being considered for a non–operative management strategy of CPAM should undergo DICER1 testing to exclude a PBB lesion [19].

The second lesion in the inferior lobe of left lung was compatible with a bronchopulmonary sequestration type ILS. The cystic lesions in ILS type may communicate with normal
bronchus which easily leads to repeated infection and produce the clinical manifestations of fever, cough, expectoration, cyanosis as in our case while in ELS type the sequestration lung is outside the lung lobe and its cystic lesions do not communicate with normal bronchial tree so it may be asymptomatic for a long time but in infancy may produce severe acute respiratory distress syndrome [20,21].

Radiographic features strongly associated with pulmonary sequestration include any area of pulmonary hyperinflation and the presence of a systemic feeding vessel which arise mostly from the thoracic aorta (75%) as in our case or abdominal aorta (15%-20%) [22].

Our patient clinical symptoms were cough, respiratory distress, and fever. Patients with similar clinic and ILS type lesion in imagery are often misdiagnosed with pneumonia, lung abscess, pulmonary tumor, and should be evaluated carefully during imagery examinations.

Regarding the treatment of pulmonary sequestration, surgery is preferred to avoid the recurrent infections, pulmonary fibrosis or hemoptysis and careful preoperative identification of systemic arterial supply and venous drainage from a pulmonary sequestration is necessary to avoid hemorrhage during surgery [23]. In our case due to the clinic of the patients and the presence of another lesion of malignant nature like PPB type II, the surgical intervention was the only option of treatment.

According to a large systematic review which assessed the simultaneously occurrence of PPB and CPAM, pleuropul-
Fig. 2 – Contrast-enhanced CT images show a lesion in the inferior lobe of the left lung compatible with a pulmonary sequestration (white arrow); the systemic vessel feeding the lesion is an accessory branch of thoracic aorta (red arrow) (Color version of the figure is available online.)

Pleuropulmonary blastoma has been reported most frequently with congenital cystic adenomatoid malformation (CCAM), CPAM type IV, bronchogenic cyst, congenital lobar emphysema and extra lobar type of sequestration (ELS) while the association between PPB and ILS type is rare [24].

**Conclusion**

The simultaneous presence of pleuropulmonary blastoma with pulmonary sequestration is very rare especially with ILS
type. In addition to clinical evaluation imaging examinations are very important in the correct diagnosis of these rare embryogenic chest tumors and very helpful for the future management of the patient.

**Patient consent**

The personal details of patient included in the article has been removed before submission.

**REFERENCES**

[1] Dehner LP. Pleuropulmonary blastoma is the pulmonary blastoma of childhood. Semin Diagn Pathol 1994;11(2):144–51.

[2] Manivel JC, Priest JR, Watterson J, Steiner M, Woods WG, Wick MR, et al. Pleuropulmonary blastoma. The so-called pulmonary blastoma of childhood. Cancer 1988;62(8):1516–26.

[3] Priest JR, McDermott MB, Bhatia S, Watterson J, Manivel JC, Dehner LP. Pleuropulmonary blastoma: a clinicopathologic study of 50 cases. Cancer 1997;80(1):147–61.

[4] Dehner LP, Schultz KA, Hill DA. Pleuropulmonary blastoma: More than a lung neoplasm of childhood. Mo Med 2019;116(3):206–10.

[5] Hill DA, Jarzemowski JA, Priest JR, Williams G, Schoettler P, Dehner LP. Type I pleuropulmonary blastoma: pathology and biology study of 51 cases from the international pleuropulmonary blastoma registry. Am J Surg Pathol 2008;32(2):282–95.

[6] Zhang H, Xu CW, Wei JG, Zhu GJ, Xu S, Wang J. Infant pleuropulmonary blastoma: report of a rare case and review of literature. Int J Clin Exp Pathol 2015;8(10):13571–7.

[7] Pryce DM. Lower accessory pulmonary artery with intralobar sequestration of lung: a report of seven cases. J Pathol Bacteriol 1946;58(3):457–67.

[8] MacSweeney F, Papagiannopoulos K, Goldstraw P, Sheppard MN, Corrin B, Nicholson AG. An assessment of the expanded classification of congenital cystic adenosomatoid malformations and their relationship to malignant transformation. Am J Surg Pathol 2003;27(8):1139–46.

[9] Van Raemdonck D, De Boeck K, Devlieger H, Demedts M, Moerman P,oosemans W, et al. Pulmonary sequestration: a comparison between pediatric and adult patients. Eur J Cardiothorac Surg 2001;19(4):388–95.

[10] Reiss I, van de Ven CP, Tibboel D. Congenital lung malformations. Intensivmed 2008;45:12–18.

[11] Savic B, Birtel FJ, Tholen W, Funke HD, Knoche R. Lung sequestration: report of seven cases and review of 540 published cases. Thorax 1979;34(1):96–101.

[12] 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.

[13] Lezmi G, Verkarre V, Khen-Dunlop N, Vihushan S, Hadchouel A, Rambaud C, et al. FGFR10 Signaling differences between type I pleuropulmonary blastoma and congenital cystic adenomatoid malformation. Orphanet J Rare Dis 2013;8:130.

[14] Leblanc C, Baron M, Desselas E, Phan MH, Rybak A, Thouvenin G, et al. Congenital pulmonary airway malformations: state-of-the-art review for pediatrician’s use. Eur J Pediatr 2017;176(12):1559–71.

[15] Naffaa LN, Donnelly LF. Imaging findings in pleuropulmonary blastoma. Pediatr Radiol 2005;35(4):387–91.

[16] Messinger YH, Stewart DR, Priest JR, Williams GM, Harris AK, Schultz KA, et al. Pleuropulmonary blastoma: a report on 350 central pathology-confirmed pleuropulmonary blastoma cases by the International Pleuropulmonary Blastoma Registry. Cancer 2015;121(2):276–85.

[17] Indolfi P, Casale F, Carli M, Bisogno G, Ninfo V, Cecchetto G, et al. Pleuropulmonary blastoma: management and prognosis of 11 cases. Cancer 2000;89(6):1396–401.

[18] Schultz KA, Yang J, Doros L, Williams GM, Harris A, Stewart DR, et al. DICER1-pleuropulmonary blastoma familial tumor predisposition syndrome: a unique constellation of neoplastic conditions. Pathol Case Rev 2014;19(2):90–100.

[19] Schultz KAP, Williams GM, Kamihara J, Stewart DR, Harris AK, Bauer AJ, et al. DICER1 and Associated Conditions: Identification of At-risk Individuals and Recommended Surveillance Strategies. Clin Cancer Res 2018;24(10):2251–61.

[20] Ou J, Lei X, Fu Z, Huang Y, Liu E, Luo Z, et al. Pulmonary sequestration in children: a clinical analysis of 48 cases. Int J Clin Exp Med 2014;7(5):1355–65.

[21] Wei Y, Li F. Pulmonary sequestration: a retrospective analysis of 2625 cases in China. Eur J Cardiothorac Surg 2011;40(1):e39–42.

[22] Long Q, Zhu Y, Yang Z. Evaluation of pulmonary sequestration with multidetector computed tomography angiography in a select cohort of patients: A retrospective study. Clinics (Sao Paulo) 2016;71(7):392–8.

[23] Zhang N, Zeng Q, Chen C, Yu J, Zhang X. Distribution, diagnosis, and treatment of pulmonary sequestration: Report of 208 cases. J Pediatr Surg 2019;54(7):1286–92.

[24] Casagrande A, Pederiva F. Association between congenital lung malformations and lung tumors in children and adults: A systematic review. J Thorac Oncol 2016;11(11):1837–45.