JOURNAL OF SCIENCE & DISEASES

Pleuropneumoblastoma, a rare pediatric tumor about a case at Ngaoundéré Regional Hospital-Cameroon

Pleuropneumoblastome, une tumeur pédiatrique rare à propos d'un cas à l'hôpital régional de Ngaoundéré-Cameroun

Neossi Guena M[1,2], Alapha Zilbinkai[1] F, Neossi Ngonga RL[3], Mohamadou A[4], Boutche Yaouba L[2], Awana A[2].

Cas clinique

1. Ngaoundere regional Hospital, Service of Radiology
2. Department of Biomedical Sciences, Faculty of Sciences, University of Ngaoundéré, Ngaoundéré, Cameroon
3. Tamale Teaching Hospital, Service of Internal Medicine, Tamale, Ghana
4. Faculty of Medicine and Biomedical Sciences, University of Garoua, Cameroon

Corresponding Author: Neossi Guena Mathurin, Ngaoundere Regional Hospital BP 45, Email: mneossiguena@yahoo.fr

Key words:
Pleuropneumoblastoma, pediatric, tumor, Chest CT scan

Mots clés :
Pleuropneumoblastome, tumeur pédiatrique, scanner thoracique

Date de soumission: 24/07/2025
Date d'acceptation: 25/09/2025

ABSTRACT

Pleuropneumoblastoma (PPB) is a rare and aggressive neoplasm that affects children almost exclusively under the age of 5 or 6 years. About 500 cases have been reported to date. This case report presents a case of 10 years old female who was admitted at Emergency Department of Ngaoundere Regional hospital with fever, non productive cough and dyspnea. Aa chest X-ray was immediately performed and showed a complete opacification of her left hemithorax. Then she underwent a chest CT scan suggested the diagnosis of PPB, which was been confirmed by the histo-pathological examination of a biopsy taken from the tumour. This case contibute to the knowledge of this rare event and when a large hemithorax mass is identified in a young child, pleuropneumoblastoma should be considered as the differential diagnosis.

RESUME

Le pleuropneumoblastome (PPB) est une tumeur rare et agressive qui touche presque exclusivement les enfants de moins de 5 ou 6 ans. Environ 500 cas ont été rapportés à ce jour. Nous présentons ici le cas d'une fillette de 10 ans admise aux urgences de l'hôpital régional de Ngaoundéré pour fièvre, toux sèche et dyspnée. Une radiographie thoracique a été immédiatement réalisée et a révélé une opacité complète de son hémi thorax gauche. Un scanner thoracique a ensuite suggéré le diagnostic de PPB, confirmé par l'examen histopathologique d'une biopsie prélevée sur la tumeur. Ce cas contribue à mieux connaitre cette maladie rare et suggère d'évoquer un pleuropneumoblastome comme diagnostic différentiel, lorsqu'une volumineuse masse d'un hémi thorax est identifiée chez un jeune enfant.

Citation as **J Sci Dis 2025, Vol 3 (3): 117 - 120**

Introduction

Primary lung cancer is the one of the most common adult cancers and is the leading cause of cancer-related death world-wide, it's incidence in children is rare, estimated to be 1 in 2 million, or 0.2% of all childhood malignancies [1]. Among these, pleuropneumoblastoma (PPB). It is a rare and aggressive neoplasm that affects children almost exclusively under the age of 5 or 6 years [2,3]. About 500 cases have been reported to date. It is the tumor most suggestive of DICER1 syndrome. Among patients with PPB, 66% have constitutional alteration of DICER1 [4], gene involved in lung development [5]. Multidetector CT (with injection) is the reference and most used cross-sectional imaging technique for the thorax in children. It allows rapid and precise analysis of all anatomical compartments. Histologically, the tumor is made up of blastematous cells and mesenchymal and/or epithelial contingents. There are three types of PPB: type I is a purely cystic, bullous lesion; type II combines solid and cystic areas and type III is exclusively solid. Each of these types very likely corresponds to different stages of successive evolution of the same malignant process. Type I, considered as the initial lesion, has a much better prognosis than types II and III which are highly aggressive tumors [6]. We described a case of type II PPB discovered in our hospital during a routine chest CT scan in 10-year-old female patient referred for recurrent respiratory infection.

Case report

A 10 years old female was admitted at Emergency Department of our hospital with fever, non-productive cough and dyspnea evolving since the age of 2. The diagnosis of recurrent respiratory infection was always made. Physical examination revealed a reduction in vocal vibrations and an absence of breath sounds, sensitivity during palpation of the left hemithorax and a deviation of heart sounds towards the right side. The biological assessment showed a significant inflammatory syndrome with moderate anaemia at 8 g/dl of haemoglobin. There were no reported anomalies in the antenatal ultrasound (US). Chest X-ray was immediately performed and showed a complete opacification of her left hemithorax with contralateral midline shift (Figure 1).

The patient underwent Computed Tomography (CT) of chest that showed heterogenous large solid-cystic mass lesion (approximately 16,5 × 13,8 × 12,5 cm) with inhomogeneous enhancement filling almost the entire left hemithorax with significant mass effect and displacing the mediastinum to the right side. The CT scan did not show progressive nodular lesions or mediastinal adenomegaly but partial lysis of the posterior costal arch of the 9th left rib.

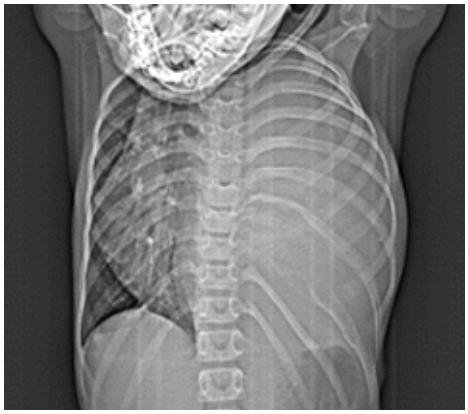

Figure 1: Chest X-ray showing a large, rounded homogeneous opacity in the left hemithorax with shifting of the mediastinum to the opposite side

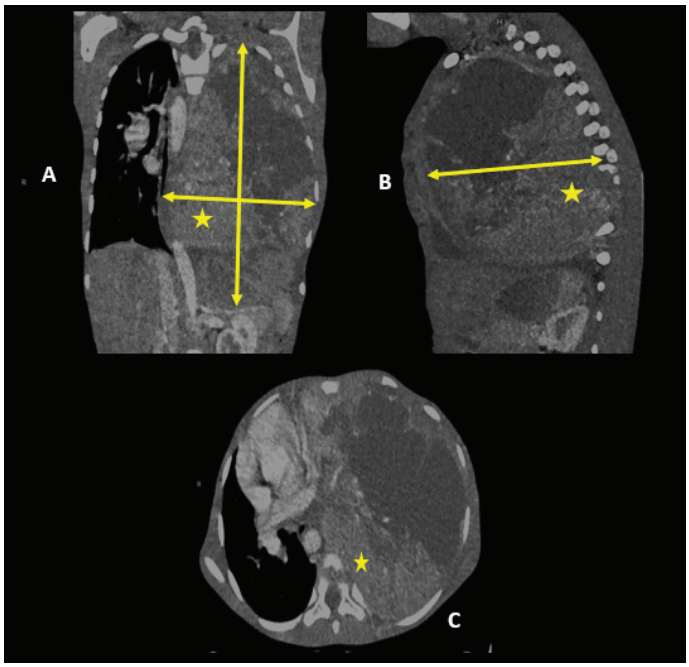

Figure 2: Contrast-enhanced coronal (A), sagittal (B) and axial (C) images of chest CT in the mediastinal window. showing a solid-cystic mass lesion with inhomogeneous enhancement filling the entire left hemithorax and displacing the mediastinum to the right side (double arrow). Solid portion was localized mainly along the postero-inferior part of the lesion (star).

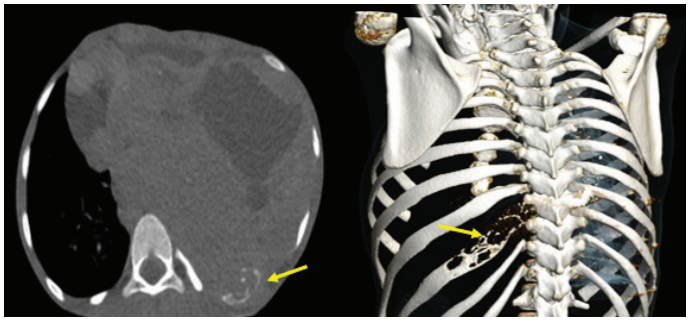

Figure 3 : Contrast-enhanced axial (A) in the bone window and posterior 3D VRT view (B) images of CT chest showing partial lysis of the posterior costal arch of the 9th left rib.

Citation as **J Sci Dis 2025, Vol 3 (3): 117 - 120**

The patient underwent thoracoscopy and the biopsies carried out, the microscopic study revealed double contingent tumor proliferation; a contingent spindle cell mesenchymal with anisokaryotic nuclei, hyperchromatophilic and containing cystic changes, a epithelial contingent with atypical epithelial cells at the nuclei pseudo-stratified basophils in places ; Histologic analysis confirmed the diagnosis of type II PPB because it contained both solid and cystic components, without the use of complementary immunohistochemical techniques. The patient was referred to the Yaoundé General Hospital for better care but died a month later.

Discussion

Pleuropneumoblastoma (PPB) is a rare and aggressive malignant tumor of children which was first described in 1952 [7], it represents approximately 0.25-0.5% of all lung tumors [8]. It can be extrapulmonary, pleural or mediastinal. It occurs in both boys and girls before the age of 4 [9]. It can be seen prenatally during prénatal ultrasound. This case also shows that it can occur in young children, although cases have been described in adults in some studies [10,11,12]. It is a primary tumor which is composed of blastematous elements and variably differentiated sarcomatous elements. There are 3 types, depending on the macroscopic appearance of the tumor: typeI (cystic), type II (cystic and solid) and type III (solid). A new subgroup of type I is individualized: regressive/non-progressive type I (type Ir) which only differs from type I by the absence of the blastematous contingent; the late age of onset and the non-progression to the aggressive forms (type II and III) which are the most frequent [9]. Its aggressive potential is great and the prognosis is poor: patients with type I tumors have 80% to 90% 5-year, disease-free survival while those with types II and III tumors present a 5-year, disease-free survival inferior to 50% [13]. The definite causative agent is not clearly determined, the association of DICER1 mutations and PPB is reported in approximately 66% of the cases [4,14]. No malignancy developement were found in other organs in our case. Some associations with other clinical situations have been reported as type IV congenital pulmonary airway malformations, dysplasia syndrome in 33% of the cases, multilocular cystic nephroma in 30% of cases and very rarely, Wilms tumor [14, 15,16]

The symptoms of pleuropneumoblastoma are not very specific and often cause delay in diagnosis [17]. Some authors suggest that symptoms present only in 60% of the cases and when present, may include fever, dyspnoea, respiratory distress, cough. Rarely, hemoptysis, chest pain, weight loss, anorexia, fatigue, even neurologic symptoms [14]. It generally appears as a solitary, well-demarcated, peripheral mass that can be large enough to extend beyond 10 cm in size [14]. Type I PPB reveals itself in the first two years of life, discovered on a chest x-ray in the event of a pulmonary infection or pneumothorax. Cysts can also be discovered by antenatal ultrasound.

The appearance of PPB on CT scan is variable depending on the histological type of the tumour. In type 1, the CT scan reveals bullous lesions without a solid component, making the diagnosis difficult because nothing distinguishes it on imaging from other much more common bullous lesions such as congenital pulmonary malformations. Type II or III PPBs are often revealed by a picture of pneumonia, sometimes accompanied by general signs. The chest x-ray shows an intrathoracic opacity, most often large, which suggests pleuropneumopathy. The CT scan highlights the at least partly solid nature of the lesion, specifies the degree of possible necrosis and possibly shows mediastinal, diaphragmatic and/or pleural invasion, factors for poor prognosis [17]. Typically, there are no adjacent rib erosions or calcifications, but there was a partial lysis of the posterior costal arch of the 9th left rib found in our case. The definitive diagnosis is based on the pathological examination, supplemented by the cytogenetic study of the tumor. Patients diagnosed with type 1 PBB are treated with surgical resection. While some experts suggest adjuvant chemotherapy, there is no unanimous agreement on this approach. For patients with type 2 and 3 PBB, it is advisable to consider surgical resection if possible followed by chemotherapy and/or radiation therapy [18].

Conclusion

Pleuropneumoblastoma is a rare intrathoracic mesenchymal tumor of young children. Its diagnosis is rarely made in the first instance, the radiological images often being confused with those of a congenital pulmonary malformation. Anatomopathological examination will help to correct the diagnosis. Chest CT scan is helpful and can lead to the diagnosis of a cystic, mixed or solid formation. When a large hemithorax mass is identified in a young child, Pleuropneumoblastoma should be considered in the differential diagnosis.

Conflicts of interest: The authors declare no conflict of interest.

Authors contributions: NGM conceptualized the study, designed the methodology. AZF collected, the data and drafted the initial manuscript. NNRL, MA, BYL and AA assisted with the literature review and critically reviewed the manuscript. All authors read and approved the final version of the manuscript.

References

1. Ben W. R. Balzer, Christine Loo, Craig R. Lewis, Toby N. Trahair, Antoinette C. Anazodo. Adenocarcinoma of the Lung in Childhood and Adolescence: A Systematic Review. Journal of Thoracic Oncology Vol. 13 No. 12: 1832-1841.
2. Cinzia Orazi, Alessandro Inserra, Paolo M. S. Schingo, Luigi De Sio, Renato Cutrera, Renata Boldrini, Saverio Malena. Pleuropulmonary blastoma, a distinctive neoplasm of childhood:

Citation as **J Sci Dis 2025, Vol 3 (3): 117 - 120**

report of three cases. Pediatr Radiol (2007) 37:337–344. DOI 10.1007/s00247-006-0402-0.

3. Dolores Ferrara, Francesco Esposito, Eugenio Rossi, Parvin Gholami Shangolabad, Vittoria D'Onofrio, Delfina Bifano, Diana Baldari, Carmela Brillantino, Raffaele Zeccolini, Massimo Zeccolini. Type II pleuropulmonary blastoma in a 3-years-old female with dyspnea: a case report and review of literature. Radiology Case Reports 16 (2021) 2736–2741. doi:10.1016/j.radcr.2021.06.022
4. 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(2):276–85.
5. Hill DA, Ivanovich J, Priest JR, et al. DICER1 mutations in familial pleuropulmonary blastoma. Science 2009; 325:965)
6. F. Dommange-Romero1, S. Collardeau-Frachon2, F. Hameury3. Pleuropneumoblastome de l'enfant. Bull Cancer vol. 97 • N° 9 • septembre 2010. doi: 10.1684/bdc.2010.1172
7. Travis WD, Bambilia E, Muller-Hermlink HK, Harris. Pathology and genetics of tumours of the lung, pleura, thymus and heart, Edited by CC. Lyon: IARC Press. 2004.
8. Smith RJ, Fabre A, Dodd JD, Bartosik W, Charles G. pulmonary
9. blastoma; a case report and review of the literature. BMC
10. Research Notes. 2014; 7: 294. PubMed | Google Scholar
11. Rabia Yasmine Namaoui, Wissam Hadjaoui, Asma Zerabib, Leila Belbekri, Fadéla Zar, Hemana Berrah, Ali Mouats, Abdessamed Bessaïh. Pleuropneumoblastome: une tumeur pédiatrique rare (à propos d'un cas. Pan African Medical Journal. 2022;43(8). 10.11604/pamj.2022.43.8.33823.
12. He Q, Xue LJ, Deng XH, Liu Y, Chen GH. [Pleuropulmonary blastoma in adult: report of a case]. Zhonghua Bing Li Xue Za Zhi. 2009 Feb;38(2):134-5. Chinese. PMID: 19573366.
13. Hill DA, Sadeghi S, Schultz MZ, Burr JS, Dehner LP. Pleuropulmonary blastoma in an adult: an initial case report. Cancer. 1999 Jun 1;85(11):2368-74. PMID: 10357407.
14. Mustapha Azzakhmam, Fouad Zouaidia, Ahmed Jahidd, Zakia Bernoussi, Kaoutar Znati, Asmae Lakhdissi, Mohamed Bouchikh, Najat Mahassini. Pneumoblastoma in adults: a new case report and literature review ; Pan African Medical Journal. 2017;28:198. doi:10.11604/pamj.2017.28.198.13907
15. Sáez B J, Pattillo S JC, Vuletin S JF. [Pleuro-pulmonary blastoma presenting as a chest wall deformity: a case report]. Rev Chil Pediatr. 2018 Apr;89(2):231-235.
16. Gbande P, Abukeshek T, Bensari F, El-Kamel S. Pleuropulmonary blastoma, a rare entity in childhood. BJR Case Rep 2021; 7: 20200206
17. Taylor B, Rice A, Nicholson AG, Hind M, Dean CH. Mechanism of lung development in the aetiology of adult congenital pulmonary airway malformations. Thorax 2020; 75: 1001–3. doi: https://doi.org/10. 1136/thoraxjnl-2020-214752.
18. Abbo O, Pinnagoda K, Brouchet L, Leobon B, Savagner F, Oliver I, et al. Wilms tumor, pleuropulmonary blastoma, and DICER1: case report and literature review. World J Surg Oncol 2018; 16: 164. doi: https://doi.org/ 10.1186/s12957-018-1469-4
19. M. David, E. Bosdure, P. de Lagausie, A. Rome, G. Gorincour, B. Chabrol, J.-C. Dubus. Pleuropneumoblastome kystique de découverte fortuite chez un nourrisson. Archives de pédiatrie 14 (2007) 1424–1426 doi:10.1016/j.arcped.2007.08.015.
20. Mlika M, Anjum F, El Mezni F. Pleuropulmonary Blastoma. 2023 Apr 27. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan–. PMID: 30480950

Citation as **J Sci Dis 2025, Vol 3 (3): 117 - 120**