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Case Report

Pleuropulmonary Blastoma in a 12-Year-Old Girl: A Case Report

Blastome Pleuropulmonaire chez une Fille de 12 ans : À Propos d'un Cas

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ABSTRACT

Pleuropulmonary blastoma (PPB) is a very rare, malignant aggressive primary lung tumor, which occurs mainly in children of both sexes less than 5 years old. The management is multidisciplinary and prognosis depends on the type at the time of diagnosis. We report a case of PPB where diagnosis was made thanks to chest CT and histopathology. This 12-year-old girl presented with a painful left hemithorax mass evolving for a year, associated with dry cough and fever. She had no prior vaccination and has always suffered severe malnutrition. Clinical examination revealed an altered general state and a large left chest mass. Chest CT scan showed a large cystic mass occupying the whole left chest. Histopathology results revealed a type II pleuropulmonary blastoma. The patient died two weeks after biopsy.

RÉSUMÉ

Le pleuropneumoblastome (PPB) est une tumeur pulmonaire primaire maligne agressive très rare, qui survient principalement chez les enfants de moins de 5 ans et atteint également les deux sexes. La prise en charge est multidisciplinaire et le pronostic dépend du type de tumeur au moment du diagnostic. Nous rapportons un cas de PPB dont le diagnostic a été posé grâce au scanner thoracique et à l'histopathologie. Cette jeune fille de 12 ans s'est présentée avec une masse douloureuse de l'hémithorax gauche évoluant depuis un an, associée à une toux sèche et à de la fièvre. Elle n'avait jamais été vaccinée et avait toujours souffert de malnutrition sévère. L'examen clinique a révélé une altération de l'état général et une volumineuse masse thoracique gauche. La tomodensitométrie thoracique a montré une volumineuse masse kystique occupant toute la partie gauche du thorax. Les résultats de l'histopathologie ont révélé un pleuropneumoblastome de type II. La patiente est décédée deux semaines après la biopsie.

INTRODUCTION

Pleuropulmonary blastoma (PPB) includes three main subtypes. Type I is a cystic lesion, type II is mixed cystic and solid, and type III is a solid multi-patterned sarcoma [3]. PPB is unique among pediatric solid tumors as it may progress from type I to type II to type III [3]. Type Ir PPB is a cystic lesion lacking a primitive malignant component. Nearly all PPBs have biallelic DICER1 pathogenic mutations; more than 70% arise in the setting of germ-line pathogenic variants.2[4]. In types II and III, the malignant cells are more likely to have acquired additional mutations, commonly involving TP53,3 and are associated with more aggressive clinical behavior, requiring more intensive treatment [4].

OBSERVATION

A 10-year-old girl presented with left hemithorax mass evolving for one year. The tum our was accompanied by cough, dyspnea and fever. She has never been vaccinated and has suffered from severe malnutrition for years without being followed up. She lived with her parents and denied any family history of lung cancer.

Physical examination revealed altered general appearance, GCS:15\15, HR: 141 bpm. Weight :19 kg Spo2: 91%. The chest wall was deformed with an arch like mass on the left hemithorax, measuring 12 cm× 7cm (second intercostal space to the left hypochondrium). The mass was soft, firm posteriorly (see fig C), hard anteriorly

(see figure B). Heart sounds were present on the right side and were irregular.

A: Lateral view **B: Anterior view** **C: Posterior view**

Figure 1: Left hemithorax chest mass

A diagnosis of bronchopulmonary cancer was retained. Chest CT Scan (see figure 2) revealed a cystic mass in the entire left pleuropulmonary space.

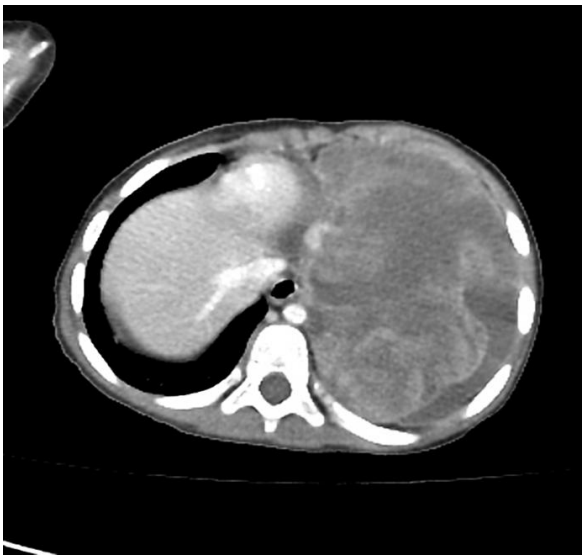

Figure 2: CT scan of the patient with pleuropulmonary Blastoma (mass in the entire pleuropulmunary space)

A minimal left posterolateral thoracotomy within the 5th and 6th intercostal space was done, well tolerated and sample sent for histopathology. Days post biopsy were favorable and patient came back two weeks later with severe respiratory distress syndrome. She was admitted in the intensive care unit but unfortunately, she died while awaiting for histopathology results which 1 month later on, revealed a type II pleuropulmonary blastoma.

DISCUSSION

Pleuropulmonary blastoma (PPB) is a rare cancer occurring mainly during early childhood and often associated with germline DICER1 mutations [5]. It is classified by the macroscopic appearance into 3 interrelated clinico-pathologic entities on a developmental continuum [5] Messinger et al in the USA reported that males and females with type II and type III pleuropulmonary blastoma are equally affected. Our case was a female with type II pleuropulmonary blastoma. We can then conclude that sex is less likely to be a risk factor for pleuropulmonary blastoma.

Type II and type III tumors present at an older age than type I. Type II patients and type III patients were diagnosed at median ages of 35 and 41 month [6]. None of the patients with type III and only 1 (6-month-old) patient with type II were younger than 1 year; 95% were diagnosed by 6.8 years and rare cases were found in adolescents [6]. Bisogno et al in France reported that Pleuropulmonary blastoma (PPB) was mostly diagnosed in children less than 6 years of age [5]. Our patient was diagnosed at the age of 12 which is much greater than the 35 and 41 months reported in the USA and the 6years in France. This could be due to factors such as limited financial resources to consult at the onset of symptoms and absence of adequate plateau technique in many hospitals in our setting which will delay the definitive diagnosis. We can then conclude that young age is a risk factor for pleuropulmonary blastoma in sub-Saharan Africa, Europe and in the USA since 35, 41-month ,6 years and 12 years are all pediatric patients.

Pleuropulmonary blastoma in children is characterized by symptoms often mistaken for respiratory infection, pneumothorax or lung malformation [5]. The tumor is usually located in the lung, but it may extend to the mediastinum, diaphragm and/or parietal pleura [5]. PPB should be suspected when a young child presents with a pulmonary lesion that can be completely cystic, cystic with a solid component or completely solid [5]. The clinical presentation and radiologic findings of pleuropulmonary blastoma are less differentiated and

depends on histologic type [3]. The diagnosis of pleuropulmonary blastoma is confirmed by histopathology which helps in classifying the cancer into the different types and guide treatment with follow up of the patient. Our patient presented with respiratory symptoms associated with altered general state. This oriented us toward the diagnosis of a lung cancer for which a chest CT scan (see fig 2) was requested and came in favor of a cystic and solid mass. We did a biopsy of the mass and sent for histopathology analysis. Results came back three weeks later with the diagnosis of type II pleuropulmonary blastoma.

Complete tumor resection is a main prognostic factor and can be performed at diagnosis or after a neo-adjuvant treatment that includes chemotherapy, and in some cases radiotherapy.

There are three different histological types:type I, cystic, type II, cystic and solid and type III,solid exclusively[4]. Type I is less aggressive and its treatment is essentially surgical. Types II and III are highly aggressive and require surgery associated to polychemotherapy. Type II and type III PPB have historically been associated with a poor prognosis [4].In our context, our patient could not be treated due to delayed in making the definitive diagnosis caused by prolonged awaiting time of histopathology results. Thus, the necessity to improve in diagnosis making time by habilitating hospitals to do extemporaneous histopathology and increase the number of pathologists.

CONCLUSION

Pleuropulmonary blastoma is an aggressive pediatric chest cancer. It has no gender predominance. It is diagnosed at a more advanced age in our context. It may be mistaken to respiratory tract infections and delay the definitive diagnosis. The final diagnosis of Pleuropulmonary blastoma is histopathology which classifies into various types (I,Ir, II and III). Its management and prognosis vary from one type to another with type II and III having the worst prognosis.

DECLARATIONS

Conflicts of interest

The authors declare no conflict of interest

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Ethical considerations

All stages of the work were carried out in compliance with the Declaration of Helsinki. The approval of the institutional ethics committee was obtained prior to the start of the study.

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