Vision of Pediatrics

Case Report

Pleuropulmonary blastoma: A case report from the department of pediatrics at Gabriel Touré University Hospital

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Abstract

Pleuropulmonary blastoma (PPB) is a primary malignant tumor of the lung and pleura that occurs in children. This case report concerns a 25-month-old male infant with no significant prior medical history. He was admitted for evaluation of a thoracic mass detected on a chest X-ray. According to the disease history, he became ill around the age of 7 months. Initial symptoms included a clear nasal discharge and a dry cough. He developed respiratory distress, which required hospitalization. A chest CT scan revealed a large solid-cystic pulmonary mass in the left apical region, measuring 73 x 64 x 53 mm. Pathological examination of the biopsy specimen showed that the tumor cells exhibited the following immunohistochemical profile: CD56-positive, chromogranin-negative, synaptophysin-negative, and desmin-negative.

The morphological and immunostaining characteristics were consistent with type III pleuropneumoblastoma. He received 3 courses of VAC (vincristine, actinomycin D, cyclophosphamide). The tumor progressed during chemotherapy. Palliative treatment consisting of analgesics and other supportive care was initiated. He died on 06/01/26 at home.

Keywords: Pleuropulmonary blastoma; Child; Mali

Introduction

Pleuropulmonary blastoma (PPB) is a primary malignant tumor of the lung and pleura that occurs in children [1]. It is a dysembryoplastic and blastematous tumor, analogous to nephroblastoma, hepatoblastoma, or neuroblastoma.
PPB generally occurs before the age of 5. It usually develops in the lung but rapidly invades the pleura, mediastinum, or diaphragm [2,3]. Treatment consists of complete oncological surgery and adjuvant chemotherapy in some cases [4]. Very few cases of pleuropulmonary blastoma in children have been reported in the literature in general and in sub-Saharan

Africa in particular [5]. We report a clinical observation of a case managed at the pediatric oncology unit of Gabriel Touré University Hospital.

Case Report

We report the case of a 25-month-old boy referred by a secondary care hospital on June 29, 2024, for the management of a thoracic mass detected on a chest X-ray. The prenatal period was uneventful. The patient is the fifth of five children. He is up to date on his vaccinations according to Mali's immunization schedule. The parents are in good health. The marriage is consanguineous (first degree). There is no history of cancer in the family. Socioeconomic conditions are favorable.
According to the medical history, he became ill around the age of 7 months (mid-June 2023). The initial symptoms included a clear runny nose and a dry cough. The family reportedly visited several health centers in the city for these symptoms. The clear nasal discharge improved, but he continued to cough.

A week later, the child developed respiratory distress, leading to his hospitalization at a healthcare facility. He received ceftriaxone 1 g IV twice daily for 4 days. A full-face chest X-ray taken on June 27, 2024, revealed a shadow suggestive of a mass in the upper left lung field. A chest CT scan performed on June 28, 2024, revealed a large solid-cystic pulmonary mass in the left apical region, measuring 73 x 64 x 53 mm, with regular contours and clear margins, exerting a mass effect on the left pulmonary artery and the homolateral main bronchus, pushing the heart toward the right side (Figure 1-2). In thoracic surgery, a pulmonary tumorectomy with partial resection of the lobes adjacent to the tumor was performed on July 11, 2024. Histological analysis revealed a pleuropneumoblastoma. The patient was referred to the oncology unit at Gabriel Touré University Hospital for further treatment.



Figure 1: Chest CT scan in pulmonary view, coronal section showing a mass in the left lung



Figure 2: Chest CT scan in pulmonary mode, axial section showing a mass in the left lower lobe of the lung

At admission, the anthropometric parameters were as follows: weight = 10 kg, height = 86 cm, head circumference = 46 cm. The weight-to-height ratio was greater than a -3 Z-score. Vital signs were as follows: heart rate 110 beats/min, respiratory rate 28 breaths/min, temperature 36°C, and oxygen saturation 100%. He did not appear pale.

On chest examination, there were no signs of respiratory distress. Left-sided dullness was noted with absent vesicular breath sounds. Vocal vibrations were increased on the left. The cardiac examination was normal. The remaining physical examination was also normal. The patient's case was discussed in a multidisciplinary team meeting.

It was agreed to treat him with carboplatin at 15 mg/kg/day, administered every 2 weeks. He received three courses of treatment from July 14, 24, to August 11, 24. The digestive and hematological tolerance of the treatment cycles was good. The evaluation CT scan showed that the pulmonary lesions were stable. The child's father requested a second opinion abroad. The child was seen at a private hospital in Tunis (Tunisia) on September 20, 2024. A chest X-ray and a chest computed tomography (CT) scan were performed; however, the results of these tests are not available. A biopsy of the mass was performed under general anesthesia. The child suffered a cardiopulmonary arrest. He was admitted to the intensive care unit for 5 days.

The histopathological examination, completed by an immunohistochemistry assessment of the specimen, showed that the tumor cells displayed the following pattern: CD56-positive, chromogranin-negative, synaptophysin-negative, and desmin-negative. The morphological characteristics, in conjunction with the immunohistochemistry profile, were typical of a type III pleuropulmonary blastoma. However, no evidence of cellular anaplasia was found.

The parents decided to return to Mali. The child was re-examined in the pediatric oncology department at Gabriel Touré University Hospital on 12/02/25. He was asthenic and anorexic. He did not present with respiratory distress, and the pulmonary examination revealed decreased vesicular murmur at the left lung apex and dullness. A thoraco-abdomino-pelvic CT scan, along with laboratory tests, was performed. The test results from December 4, 2025, showed: blood type AB positive, white blood cell count 10,600/mm³, neutrophil count 7,700/mm³, hemoglobin 11.4 g/dl, platelet count 270,000/mm³, with negative retroviral serology. The CT scan revealed tumor progression in the left pleuropulmonary region, associated with locoregional infiltration and a secondary homolateral pleural metastasis, with no evidence of abdominal or bone metastases at this stage. The bone marrow sample was normal. He received 3 cycles of VAC (vincristine, actinomycin D, cyclophosphamide) with good hematologic and digestive tolerance of the treatment. The tumor was still progressing on the evaluation CT scan performed on 02/11/26 following the third VAC cycle. Palliative treatment consisting of analgesics and other supportive care was initiated. He died on 06/01/26 at home due to respiratory distress.

Comments

pleuropulmonary blastoma (PPB) is an extremely rare and aggressive embryonic intrathoracic malignant tumor that occurs almost exclusively in infants and young children. It accounts for approximately 1% of primary lung tumors in children. Its exact incidence remains difficult to assess globally due to its rarity [6]. Few cases have been reported in Africa [7]. The median age at diagnosis is generally around 2 years. Approximately 60 to 70% of PPB cases are associated with a germline mutation in the DICER1 gene. This mutation predisposes the child and their family to other rare tumors [8,9]. The clinical epidemiology correlates with the tumor's structural characteristics, which are classified into three major types. Type I (purely cystic) occurs in younger children (median age at 10 months). This is the initial, least aggressive form. Type II (mixed: cystic and solid) is the intermediate form (median age at 34 months). Type III (purely solid) occurs in slightly older children (median age at 44 months). This is the most aggressive form, with a high risk of metastasis (brain, bone) [10,11]. Symptoms depend primarily on the tumor volume and histological type (cystic or solid). Respiratory distress and chest pain are common signs [12]. General symptoms were more noticeable in our patient at the onset of the disease.

Dyspnea occurred late in our patient. Chest CT and MRI are key imaging studies for determining the nature of the lesion. They allow for the differentiation between purely cystic forms (Type I), mixed forms (Type II), and large solid masses (Type III) [13,14]. The imaging also assesses the extent of involvement of the major vessels, the diaphragm, and the pleura. Histological examination of the tumor specimen is the only way to establish a definitive diagnosis (Image 1). It reveals a malignant stroma of mesenchymal origin (often rhabdomyosarcomatous) associated with trapped benign epithelial structures [15].

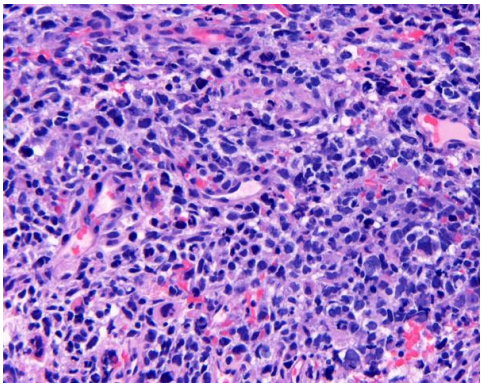


Image 1: Type III pleuropulmonary blastoma consisting of solid clusters of high-grade unusual malignant cells

Treatment of PPB requires an intensive multidisciplinary approach. Surgical resection is the cornerstone of treatment and requires excellent surgical facilities [16]. Following a multidisciplinary team discussion of the case, our team opted to perform only a biopsy to confirm the diagnosis.

The primary surgical procedure involves complete resection of the tumor (often via pulmonary lobectomy) with clear margins [17,18]. The main goal of surgery is complete resection, which is a major prognostic factor for survival. Neoadjuvant chemotherapy aims to reduce tumor size and limit the risk of intraoperative rupture. Adjuvant chemotherapy is essential to consolidate treatment and target micro metastases. The VAC regimen—vincristine, actinomycin D (dactinomycin), and cyclophosphamide—is the most commonly used. The prognosis ranges from an 80% 5-year survival rate for Type I (benign or less aggressive) to less than 40% for the more aggressive Types II and III [19,20]. Our patient was unable to undergo complete oncological resection; unfortunately, he passed away at the end of the second year of treatment.

Conclusion

Type III pleuropulmonary blastoma is a rare and aggressive tumor that is life threatening to the patient. Surgery for this tumor constitutes a challenge in sub-Saharan Africa. Improvements in surgical tools and resection techniques are key to its treatment.

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Authors' contributions

Traoré Fousseyni wrote the original manuscript text and conducted conceptualization, design, methodology, and interpretation of the data. Abdoul aziz Diakite conducted the data collection, formal data analysis and the interpretation of the data. All authors revised the manuscript and approved the final version to be published.

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Data availability

The database analyzed during the current study is available from the corresponding author.

Declarations

Conflicts of interest: the authors have not declared any conflicts of interest.

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