Case Report

Pleuropulmonary blastoma: Cystic tumour misdiagnosed as an emphysematous bulla

A. Chadli-Debbiche a, E. Ben Brahim a,*, R. Jouini a, N. Labbene a, S. Sayed b, M. Ben Ayed a, N. Kaâbar b

a Department of Pathology, Habib Thameur Hospital, 8, Avenue Ali Ben Ayed, Tunis 1008, Tunisia
b Department of Paediatric Surgery, Habib Thameur Hospital, Tunis, Tunisia

1. Introduction

Pleuropulmonary blastoma (PPB) is an uncommon solid and/or cystic intrathoracic childhood tumour, which belongs to the group of dysontogenetic, neoplasms such as wilms’ tumour, hepatoblastoma and neuroblastoma. It also affects adults and appears as pulmonary and/or pleural-based mass, histologically characterized by a primitive variably mixed blastematous and sarcomatous appearance.1 It presents a pattern of rapid growth and is associated with a poor prognosis.2

2. Case report

A 13-month-old girl was admitted to the paediatric surgical department when she presented with a one-week history of wheezing and dyspnea accompanied by loss of appetite. For 3 days, she had vomiting, diarrhoea and fever. Physical examination revealed a respiratory rate at 38/min with nasal flare, intercostal retraction and asymmetric chest. Pulmonary auscultation revealed decreased breath sound in the right middle and lower chest, without sibilant rale. Cardio-vascular examination was normal. There was neither hepatosplenomegaly nor peripheral lymphadenopathy and no abnormal neurological symptoms. Chest X-ray revealed a mediastinal deviation with a clear appearance of the right lung. Computed tomography showed a voluminous emphysematous bulla of the right lung and atelectasis of the lower lobe. The final diagnosis was made on the basis of histological features of a cystic tumour, showing blastematous elements associated with sarcomatous compound confirming the diagnosis of pleuropulmonary blastoma type I. Subsequent chemotherapy was performed. Four years after the operation, the child is well with no evidence of disease recurrence or metastasis.
Holoxan, Actinomycin and Oncovin. At present, 4 years after treatment, the child is well with no evidence of disease recurrence or metastasis.

3. Discussion

PPB is a rare primary malignant lung tumour, accounting for 0.25–0.5% of primary lung tumours.\(^1\) It was first described by Barnard in 1952 who named it embryoma of lung and was subsequently redefined by both Spencer in 1961 and Manivel in 1988. Since then, approximately 150 cases have been reported in the literature.\(^3,4\) While 60% or more of cases occur in adults over 40 years, 20% occur in patients under 20 years and 75% of these are under 10 years.\(^3\) An exceptional case of PPB has been diagnosed prenatally, at 32 weeks of gestation.\(^5\) Most series report a female predominance.\(^6\) Symptoms can be unspecific, such as dry cough, fever, chest or abdominal pain and infections that persist despite medical therapy. The uncommon symptoms are dyspnea and pneumothorax or respiratory distress. Our patient was a 13-month-old girl, presenting with wheezing, dyspnea and loss of appetite. In 20% of the cases, the tumour is discovered fortuitously.\(^7,8\) In 38% of children with PPB had radiologically identified pulmonary cysts, prior to surgery. In more than one-third of these instances, the cysts were supposed to be developmental or infectious and the children were just watched over for several months often until a suspicious, solid component had been developed. A computed tomography scan examination confirms the cystic lesion and shows possible zones of necrosis, solid component or pleural involvement.\(^9,10\)

Based on radiological features, several other diagnoses were suspected like hydatid or bronchogenic cyst, pulmonary abscess, cystic adenoma and congenital adenomatoid malformation. Imaging study showed in the present case an emphysematous bulla, and the diagnosis of PPB was done by histological examination. This tumour can be divided into 3 morphologic types, based on its cystic, cystic and solid, or solid character, as determined by the gross and microscopic findings. The exclusively cystic or type I PPB is the least complex and is presented at an earlier age than either type II or type III PPB.\(^1\) The cystic lesion is usually located in the periphery of

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**Fig. 1.** Chest X-ray: mediastinal shift with a clear appearance of the 2/3 of the right lung associated to a liquid level.

**Fig. 2.** Computed tomography: giant emphysematous bulla of the right lung and atelectasis of the lower lobe associated to an abundant pleural effusion.

**Fig. 3.** Macroscopic appearance: multi-loculated cystic tumour measuring 13 × 10 × 7 cm.

**Fig. 4.** Microscopic examination: the cystic walls were lined with a high cylindrical epithelium, often ciliated, standing on a stroma with a cellular cambium layer, composed of blastematous cells (Hematoxylin and eosin, ×250) at the upper right: mesenchymal cells with a rhabdomyoblastic differentiation (Hematoxylin and eosin, ×400).
present in about 25% of the cases. In our case report, the tumour bizarre, pleomorphic, multinucleated mesenchymal cells present features and areas resembling rhabdomyosarcoma with large, hematoxylin foci are consistently seen and contain numerous mitotic component and potential for sarcomatous differentiation. Blas-

like blastema and stroma, with the absence of a carcinomatous logically, childhood PPB, unlike the adulthood one has embryonic-

may be congenital in origin and thus malignant from the begin-

these two entities are said to be curiously associated. However, PPB was a PPB type I, diagnosed during histologic examination. Type I lesions must be histologically differentiated from cystic adenoma-
toid malformation and bronchogenic cyst. Type III must be differ-

-4 years later, well with no evidence of disease. 

4. Conclusion

Congenital cystic lesions should be followed up and never underestimated because they may conceal a PPB. Diagnosis is usually histologic and treatment should be submitted to a joint analysis from a pulmonologist, an oncologist and a thoracic surgeon. Early definitive surgery in PPB may have a positive impact on survival. After resection, postoperative chemotherapy is strongly recommended to prevent distant spread of the disease or transition from PPB type I to PPB type III.

Conflict of interest statement

None of the authors have a conflict of interest.

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