Pleuropulmonary blastoma type III extended into the left atrium in a 16-month old boy

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INTRODUCTION

Pleuropulmonary blastoma (PPB) is an uncommon neoplasm that arises in the lung, mediastinum or pleura of children and is characterized by a primitive blastema and malignant mesenchymal stroma that may show multidirectional differentiation [1, 2]. PPB occurs in three pathological types, cystic type I, cystic/solid type II and solid type III, and evidence indicates progression from type I to type II or III. This tumour is known to be part of the spectrum of tumours related to the mutation of the Dicer-1 gene, which includes Sertoli-Leydig cell tumours, ciliary-body medulloepithelioma, cystic nephroma and other tumours [3, 4].

Types II and III occur typically at ages 3 or 4 years and are aggressive mixed-pattern sarcomas. Among them, 11 cases with vascular extension or embolism were reported; in 7 tumours, extension involved the left heart [1, 3, 5]. We present a very young child with extensive thoracic PPB, occluded pulmonary veins and huge (thrombus-like) left atrial tumour. The vascular components did not respond to chemotherapy, embolic complications did not occur and complete resection was only possible in a ‘second-look’ surgery. We consider this worthy to report because of the very unusual and extensive disease in a 16-month old child.

CASE REPORT

A 16-month old male weighing 10 kg was hospitalized with a diagnosis of pneumonia. Chest computerized tomography (CT) scan revealed a mass extending into the right hemithorax from the posterior mediastinum and propagating inside the left atrium through the right pulmonary veins. Echocardiography showed a huge flexible mass occupying almost all of the left atrium cavity and intermittently prolapsing through the mitral valve. Two preoperative chemotherapy courses of ifosfamide, vincristine, actinomycin D and doxorubicin were administered at 3-week intervals. The patient was placed on cardiopulmonary bypass, cardiac arrest and the left atrium was opened. The tumour and orifice of the inferior right pulmonary vein were resected and the normal free borders reconstructed. A large mass occupying the right pleural space was removed. Seven additional courses of chemotherapy were given. In a ‘second-look’ surgery 6 months later, through a right thoracotomy, the residual mass at the posterior mediastinum was resected. The child received four additional courses of chemotherapy. After 12 months of the initial resection, there is no evidence of recurrence.

Keywords: Pleuropulmonary blastoma • Chemotherapy • Cardiac tumour

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cardiopulmonary bypass, a large mass occupying the right pleural space without infiltrating the lung was resected, but a portion of tumour in continuity with the posterior mediastinum was not accessible. No enlargement of mediastinal, hilar or peribronchial lymph nodes was observed.

The postoperative period was uneventful. Seven additional courses of IV ADo were given. Chest CT scan revealed a residual mass in the posterior mediastinum (Fig. 1C). The patient underwent a second-look’ surgery 6 months after the first attempt to remove the tumour. Through a right thoracotomy, all of the mass was resected, including a very small portion of infiltration at the right bronchus. There were no infiltrations to lung or oesophagus. Subsequently, the child received four courses of the same chemotherapy. The follow-up after initial cardiac surgery is 12 and 6 months after the second surgical intervention, with no evidence of metastasis or recurrence (Figs 1D and 2B).

**DISCUSSION**

Pleuropulmonary blastoma (PPB) is a very rare, highly aggressive childhood tumour. Approximately 300 cases have been reported in the literature and the International Pleuropulmonary Blastoma Registry [1, 3].

PPB type III tumours often have a poor prognosis, with a 5-year overall survival rate between 45 and 57.5% [2, 4]. This is linked to higher metastasis and recurrence rates, with the most common sites being the brain, bone and liver [1, 3, 4]. Tumour extensions into large thoracic vessels and the heart are a rare but important complication of PPB and occur in ~3% of types II and III. Because the detection of cardiac tumours is difficult due to the generally vague cardiovascular symptoms or signs, early suspicion is very important. A thorough cardiovascular evaluation by meticulous echocardiographic examination should be done in every suspicious case [5]. Recognition of vascular involvement is important in planning the initial surgical approach to PPB [1].

Systematic use of ifosfamide- and doxorubicin-based regimens for type II/III PPB is suggested with the aim of enabling delayed complete tumour resection and improving survival [1, 4].

Surgery is generally performed at diagnosis if complete resection is considered feasible. If not, a biopsy is obtained and chemotherapy is administered before surgery, aiming for complete resection [1, 4]. Initial surgery is completed in <33% of
reported cases, including those with delayed intervention after chemotherapy [4]. There are no established recommendations for radiotherapy, but it should be limited to high-risk patients to minimize the likelihood of long-term cardiac damage [1, 4].

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REFERENCES