PLEUROPULMONARY BLASTOMA A RARE PRIMARY PULMONARY TUMOR OF CHILDHOOD

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INTRODUCTION

Pulmonary neoplasms of all types are rare in children and pulmonary metastases are more common than primary tumors. Pleuropulmonary blastoma (PPB) is the least common malignant primary neoplasm. One quarter of cases occur in children and are often associated with congenital lung disease.[1,2]

PPB is a unique neoplasm of the lung and the pleura, containing both mesenchymal and/or epithelial elements that mimic the embryonal tissues of the developing lungs. Pediatric PPB differs from adult pulmonary blastoma in clinical presentation, gross and microscopic anatomical features, and outcomes. Patients usually have a poor prognosis and rarely survive more than 10 years.[3]

The clinical presentation is usually nonspecific. Symptoms and signs include respiratory difficulty, fever, chest pain and cough. Plain radiographs may demonstrate multicystic transformation of a lobe, pneumothorax, opacification of a hemithorax, pleural effusions, a pleural or diaphragmatic mass and shift of the mediastinum. Sometimes chest radiographs may simulate pleural effusion or empyema and fail to demonstrate a mass.

Two cases of pleuropulmonary blastoma are presented in this report.

CASE 1

A 3-year-old girl was referred from an outside hospital to our institution. She presented with high fever, chronic cough and anemia.

Chest radiograph at the outside hospital revealed a left pleural effusion. Thoracentesis yielded 800 cc of unclotted blood. A blood transfusion was performed, and the patient was transferred to our hospital.

On physical examination, the patient was tachypneic. There were decreased breath sounds on the left and decreased vocal resonance noted at the anterior left chest. Hepatomegaly was noted. Routine laboratory values were within normal limits.

A chest radiograph demonstrated near total opacification of the left hemithorax. (Figure 1)

CT scan showed a large cystic mass in the left hemithorax, measuring 9 x 6.5 cm in the axial plane. There were hyperdense areas within the mass which were thought to represent hemorrhage. Rim enhance-

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ment was present. The mass compressed adjacent lung parenchyma and caused a slight shift of the mediastinum to the right (Figures 2a-d).

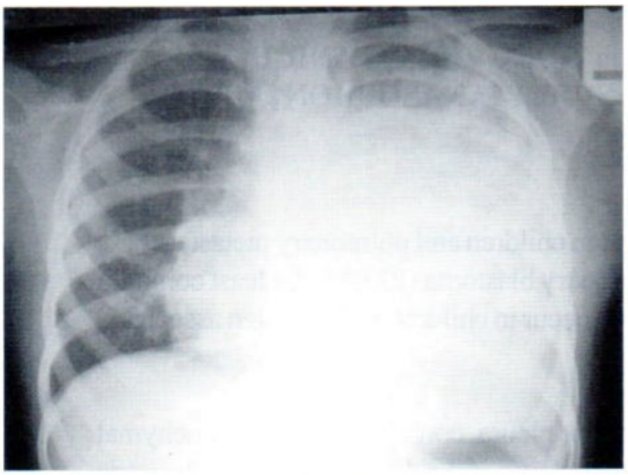

Fig.1 Chest radiograph of a 3-year-old girl (case 1) who presented with symptoms of respiratory tract infection. Near complete opacification of the left hemithorax is seen. A large left pleural effusion is suggested.

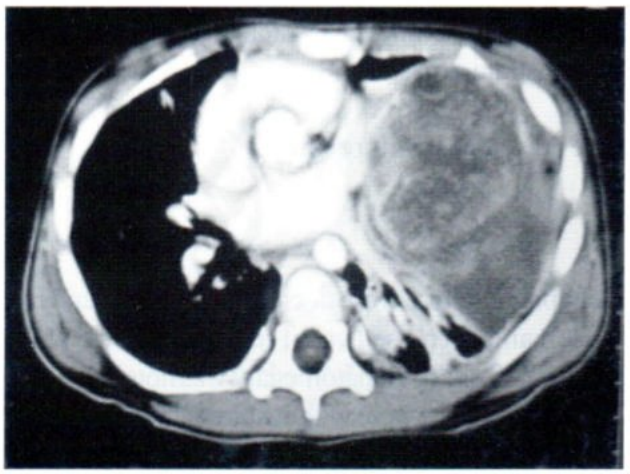

Fig.2A

Fig.2A-D Contrast-enhanced CT of the chest in case 1. There is a large cystic mass in the left hemithorax. There is compression of adjacent lung parenchyma and shift of the mediastinum.

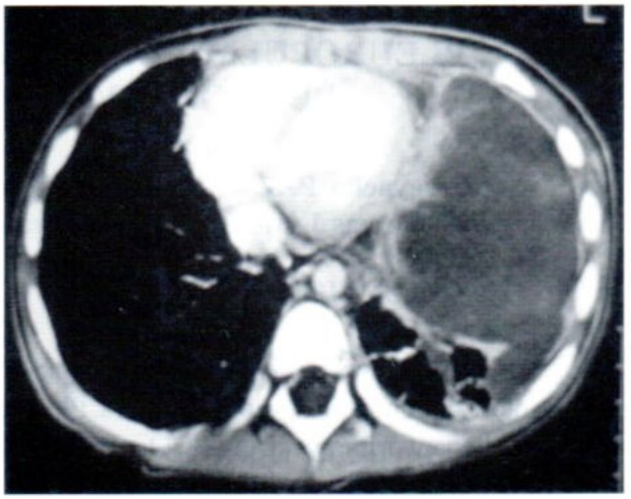

Fig.2B

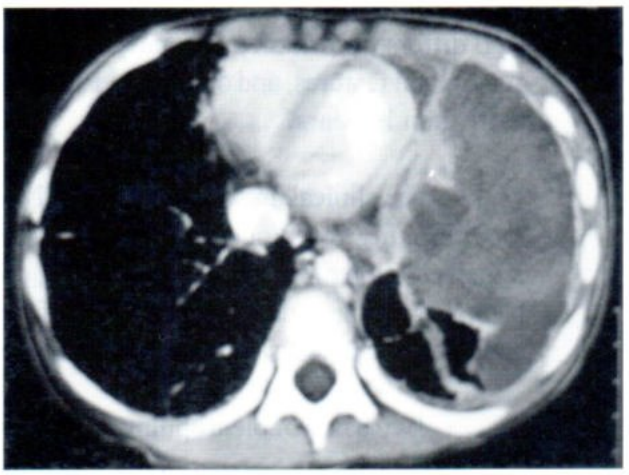

Fig.2C

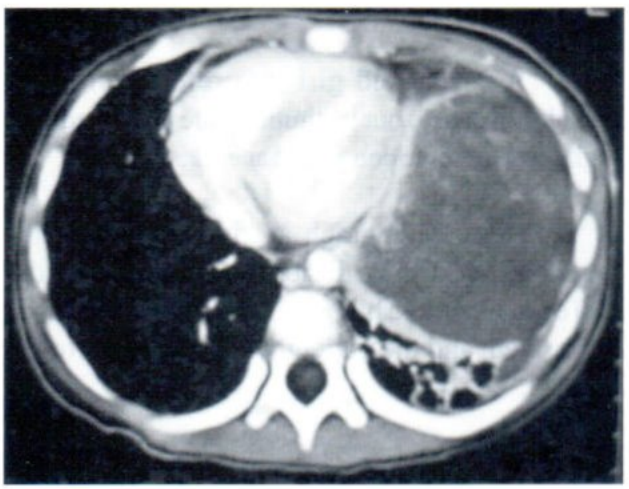

Fig.2D

CASE 2

A 4-year-old boy was referred to our institution from an outside hospital. He had presented with fever and dyspnea for 1 week.

A chest radiograph at the outside hospital showed a left lung mass. He underwent left thoracotomy with excision of the lung mass. Histologic findings were consistent with an embryonal rhabdomyo sarcoma with cartilageneous differentiation. He received postoperative chemotherapy. One week after chemotherapy, he developed a massive pleural effusion and was referred to our institution for further management.

Physical examination revealed a tachypneic patient. Breath sounds were decreased on the left. The abdomen was distended with no palpable mass. Routine laboratory values were within normal limits.

Chest radiograph showed opacification of the left hemithorax. There was marked mediastinal shift to the right. An underlying mass was felt to be likely. (Figure 3)

CT showed a large cystic mass with internal septations occupying the entire left hemithorax. The mass measured approximately 14 x 13 cm in the greatest axial dimensions. Herniation of the mass across the midline with shifting of the heart and mediastinal structures to the right was noted. (Figure 4a-c)

After contrast injection, peripheral and septal enhancement were demonstrated. (Figure 5a-c)

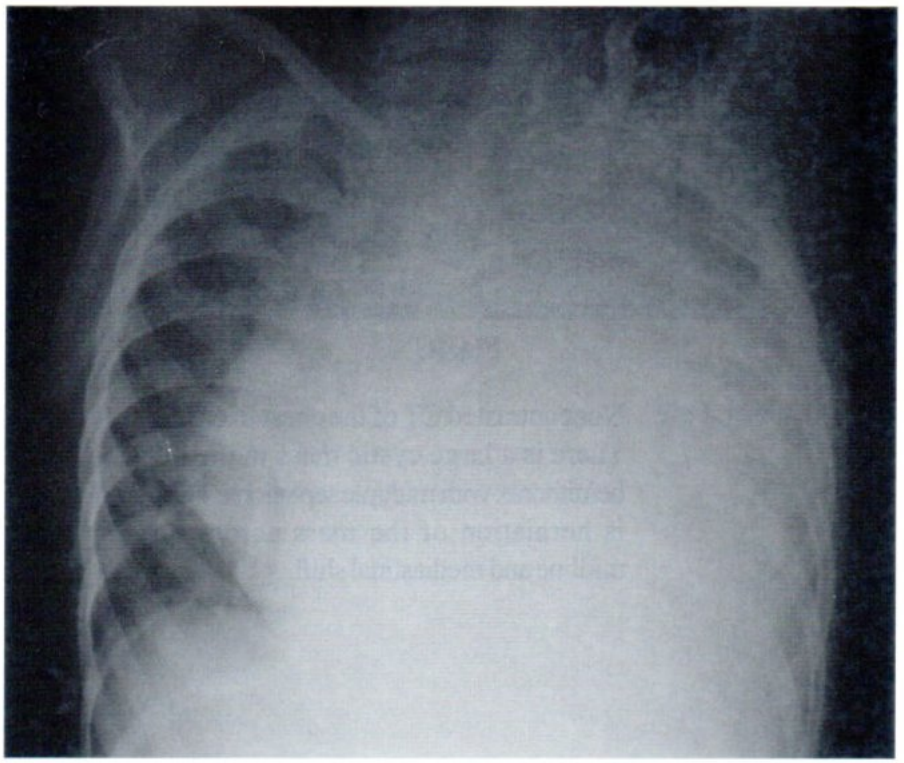

Fig.3 Chest radiograph of a 4-year-old boy (case 2). Opacification of the left hemithorax is seen with shift of the mediastinum to the right.

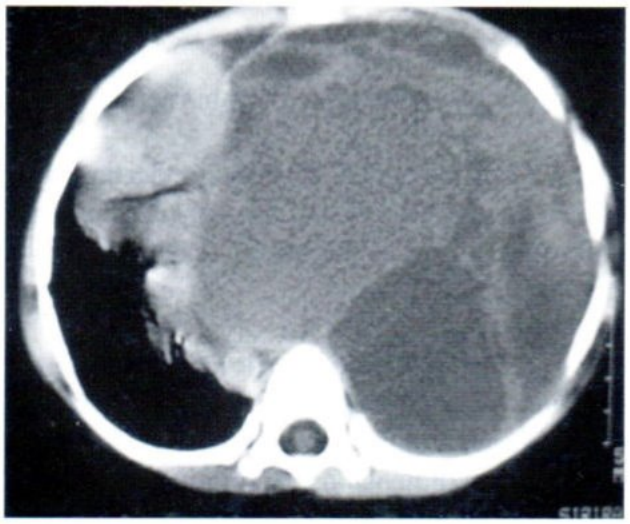

Fig.4A

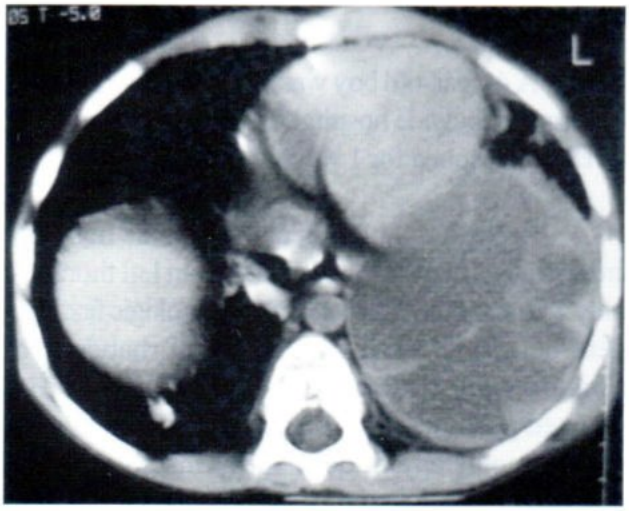

Fig.4B

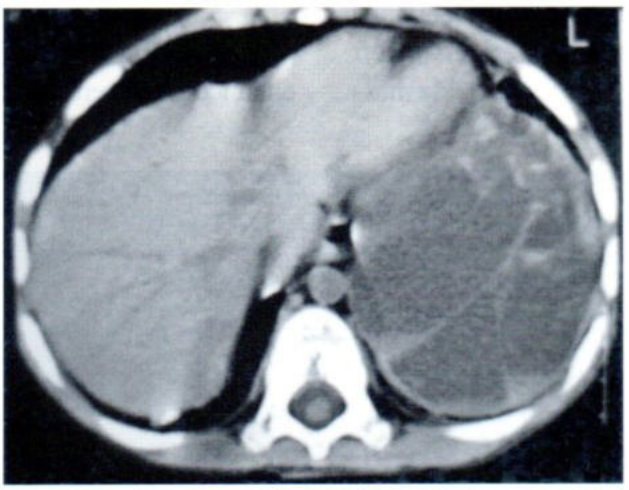

Fig.4C

Fig.4 a-c Noncontrasted CT of the chest in case 2. There is a large cystic mass in the left hemithorax with multiple septations. There is herniation of the mass across the midline and mediastinal shift.

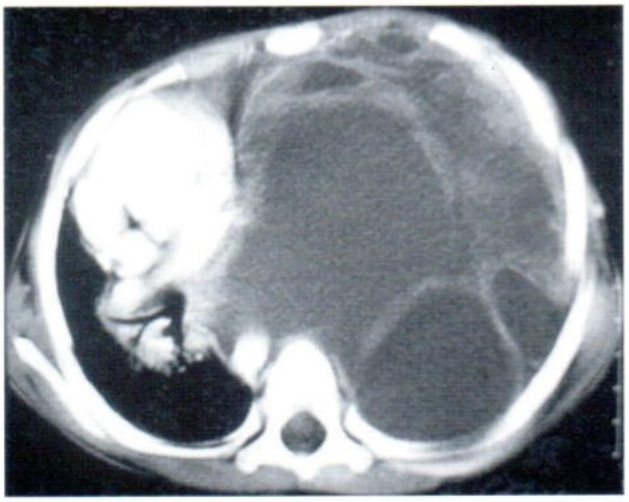

Fig.5A

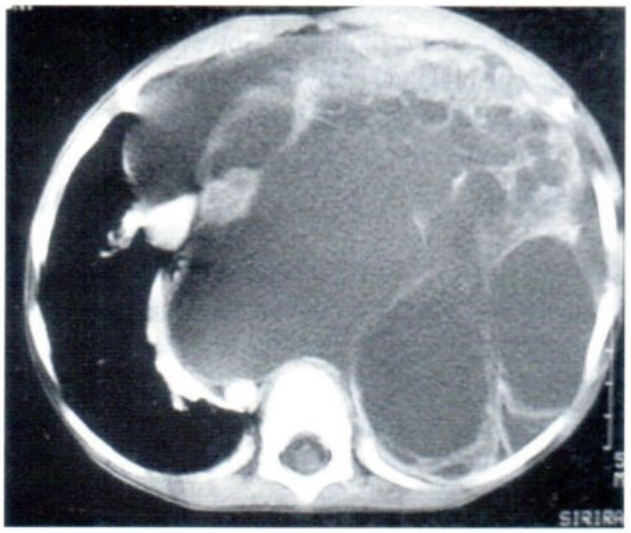

Fig.5B

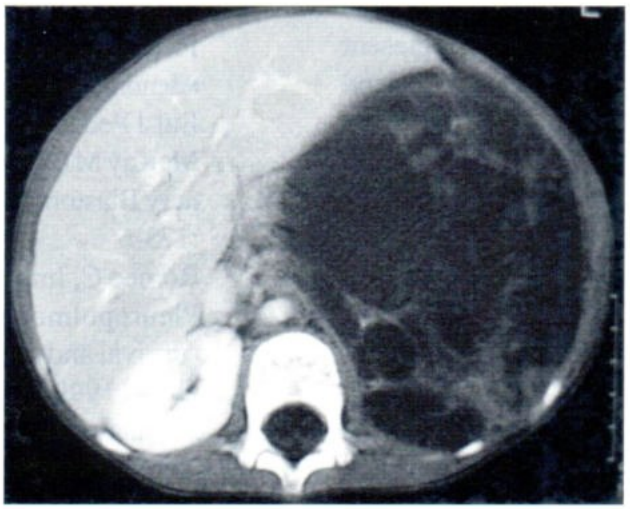

Fig.5C

Fig.5 a-c Contrast-enhanced CT in case 2 shows a mass with peripheral and septal enhancement

DISCUSSION

Plain radiographs in both of these cases showed near total opacification of the involved hemithorax. Such hemithoracic opacification with contralateral mediastinal shift could result from many causes such as a large pleural effusion or other space occupying mass. Consolidation alone usually does not cause marked mediastinal shift. In children presenting with respiratory symptoms and opacification of a hemithorax, a parapneumonic effusion should be considered. Pneumonia is the most common cause of pleural effusions in childhood. Other causes include congestive heart failure and renal failure. Malignancy is relatively rare in this age group.

If the etiology of a large pleural effusion appears fairly certain from history, physical examination and supporting laboratory data, thoracocentesis is usually avoided. Most patients recover without

having thoracotomy drainage.[4] However in complicated cases, pleural fluid analysis will help to identify the cause, and with large pleural effusion, thoracocentesis may be needed to relieve pressure in the affected hemithorax.

If patients fail to response to initial treatment, further investigation should be done. Ultrasound or computed tomography of the chest will help to determine whether the fluid is simple or complex, free-flowing or loculated, and identifying associated pathology such as an underlying mass lesion.

In our two cases, there were large cystic masses in the left hemithorax. No internal calcification was identified. In the first case, there was some internal hyperdensity which was thought likely to represent focal hemorrhage or a solid component. In the second case, internal septations were noted. Peripheral enhancement was demonstrated in both cases. According to these findings, an empyema could not be excluded. Due to the complexity of the visualized mass and the previous history of a histologically diagnosed tumor in case 2, a biopsy was performed. Biopsy results for both cases were consistent with pleuropulmonary blastoma (PPB).

Several cases of PPB have been previously reported.[5-8] Many of these cases involved cystic masses without a significant solid component. In such cases, approaching a specific diagnosis was very difficult and biopsy was necessary for definitive diagnosis.

Although the clinical and radiologic findings of this disease are not very specific, the histopathological appearance is quite distinctive, with a mixture of both epithelial and mesenchymal elements. PPB can be classified into three broad histologic categories.

Type 1 is purely cystic with smooth and glistening cyst linings. The adjacent pulmonary parenchyma is unremarkable. Twenty five percent of PPB is exclusively cystic.

Type 2 consists of multiple cysts and coalescing nodules with cystic degeneration, hemorrhage and necrosis.

Type 3 is a predominantly solid tumor with cystic areas only secondary to hemorrhage and necrosis.

In our two patients, surgical resection was performed and postoperative chemotherapy was administered. The tumors are classified in the type 2 category. These patients are still followed by pediatric oncologists at our institution.

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