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Pleuropulmonary Blastoma in a Young Adult Presenting as a Ruptured Cystic Teratoma in Radiology

Pleuropulmonary blastoma (PPB) is a rare malignant dysontogenetic neoplasm primarily affecting children and is characterized histologically by a variably mixed blastematous and sarcomatous patterns. We herein report a very exceptional adult case of PPB. A 21-yr-old male patient presented with a left chest pain of two weeks' duration. A computed tomography scan revealed a large, multicystic tumor occupying the left lower hemithorax, leading to the impression of a ruptured mediastinal cystic teratoma. A thoracotomy for resection of the tumor was performed. On histologic examination, the tumor consisted of cystic walls and associated solid lesions which showed undifferentiated blastemal tissues with focal fibrosarcomatous and rhabdoid features. Immunohistochemically the tumor cells only showed diffuse strong positivity for vimentin. The histologic findings corresponded to a type II PPB. The authors suggest that PPB, especially of type I or II, should be included in the radiologic differential diagnosis of mediastinal cystic neoplasms in a young adult.

Key Words : *Mediastinal Neoplasms; Pulmonary Blastoma; Teratoma; Adult*

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INTRODUCTION

In 1988, Manivel et al. (1) reported a distinctive intrathoracic pulmonary neoplasm of pleuropulmonary blastoma (PPB) in 11 children younger than 6 yrs. They suggested that PPB was a rare, distinct entity from classic pulmonary blastoma on the basis of its exclusive clinical presentation in childhood and its pathologic features of primitive blastema and sarcoma without a carcinomatous component. Since then, additional individual cases and small series of similar tumors in children have been reported under the diagnosis of PPB. Three pathological types of PPB have been described (2). Type I is exclusively cystic, type III is predominantly solid, and type II is an intermediate lesion with both solid and cystic elements. The cystic form is significant in that it may be misdiagnosed and managed as a benign cystic lesion. Generally the clinical course depends on the pathological type.

We herein report a case of PPB occurring in a 21-yr-old male patient, which presented with the radiological features of mediastinal cystic teratoma. To our knowledge, the current case is the third description in the English literature of PPB diagnosed in an adult since the initial report by Hill et al. (3) in 1999.

CASE REPORT

A 21-yr-old male patient was referred from a hospital with a left chest pain and mild dyspnea of two weeks' duration. Laboratory findings at admission were unremarkable. Sputum culture showed no growth for acid-fast bacilli or fungal organisms. There was no history of tumors in his close relatives and his past medical history was unremarkable. A chest radiograph showed an apparent left pleural effusion, and a computed tomography (CT) scan revealed a large, multicystic tumor occupying the left lower hemithorax (Fig. 1). The radiologic impression was a ruptured cystic teratoma. On echocardiogram, a left cardiac border was deviated to the right side. A thoracentesis was performed and removed about 1 liter of non-bloody, exudative fluid with differential count of polymorphs 40% and lymphocytes 60%. Pleural fluid cytology showed the appearance of malignant small round cells with mesothelial reaction. On November 11, 2001, he underwent a left anterior thoracotomy for the removal of the tumor. On operative field, it appeared to originate from a broad pedicle attached to the infracardiac mediastinal soft tissue, and showed adhesion with the lingular segment of the left lung. The tumor was removed in large fragments, totally weighing about 140 g, together with the left lingular segment adhered.

The resected tumor consisted of membranous cystic walls, associated with solid parts of fleshy, gray white masses including focal necroses and hemorrhages. The lingular segment of the left lung was submitted together and demonstrated a round, gray white, solid tumor nodule, about 2.1 cm in diameter. Histologically, the main tumor showed cystic and solid

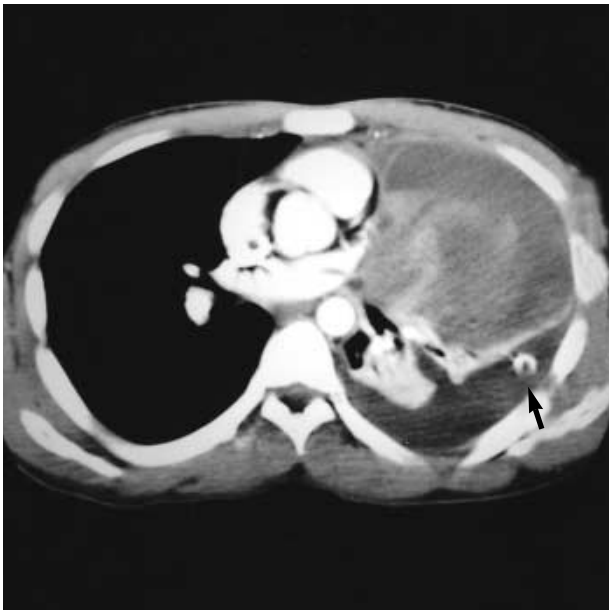

Fig. 1. Contrast-enhanced chest CT scan reveals an inhomogeneous, non-enhancing mass in the left hemithorax, associated with pleural effusion. The arrow indicates a thoracotomy tube located in the left hemithorax.

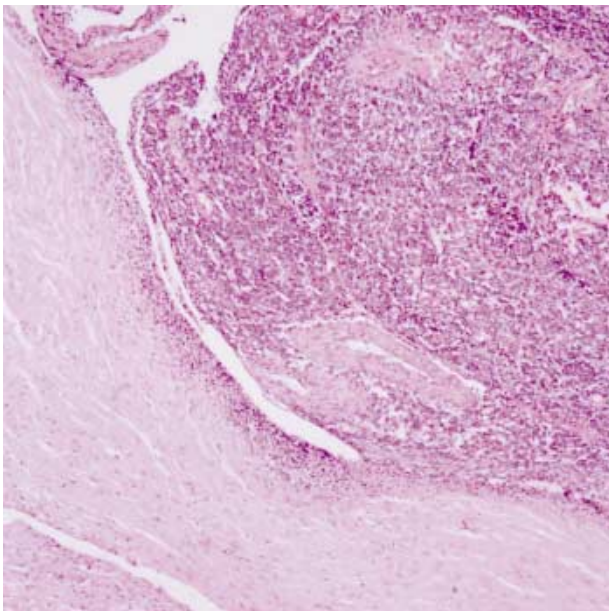

Fig. 2. The tumor shows a cystic structure, associated with nodular or plaque-like solid areas formed by the proliferation of primitive malignant mesenchymal cells (H&E, ×20).

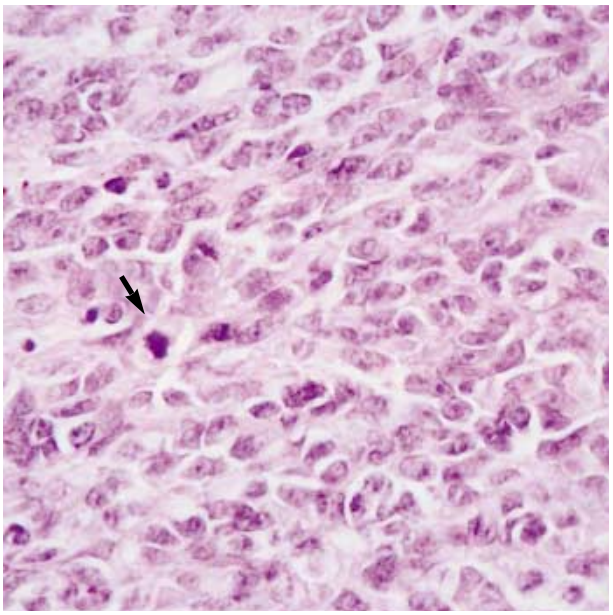

Fig. 3. The solid portion of the pleuropulmonary blastoma is composed of small primitive malignant cells (blastema) without features of differentiation. The arrow indicates a mitotic figure of tumor cell (H&E, ×400).

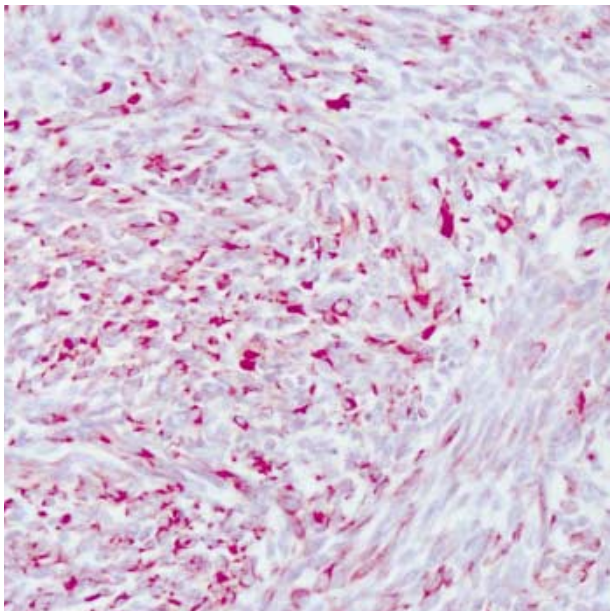

Fig. 4. Immunohistochemical staining for vimentin shows diffuse strong positivity in the blastemal cells seen in the solid portions of the pleuropulmonary blastoma (×200).

lesions with areas of thickening and/or nodularity with a relationship to the cysts (Fig. 2). The solid portion in large parts consisted of undifferentiated blastemal tissues, but focally they also demonstrated a complex pattern with fibrosarcomatous and rhabdoid features. Compactly packed blastematous cells revealed round-to-ovoid hyperchromatic nuclei and scanty cytoplasms with indistinct cell borders (Fig. 3). Mitoses of the tumor cells were frequently noted. Focally a few epithelial-lined spaces also occurred, and were regarded as entrapped mesothelial linings.

Immunohistochemically the tumor cells showed diffuse strong positivity for vimentin (Fig. 4). However, other mark-

ers such as cytokeratin, smooth muscle actin, desmin, neuron-specific enolase, S-100 protein, MIC-2 gene product (CD99), and CD34 were all negative in the tumor cells including rhabdoid cells. The histologic diagnosis was PPB, which would belong to the type II category.

After operation, follow-up radiologic examination showed no evidence of recurrence or distant metastasis. We planned postoperative chemotherapy with vincristine, actinomycin, and cyclophophamide, but he refused the treatment. Despite the refusal of the chemotherapeutic regimens, he remained well 6 months after operation. He was then lost to further follow-up observation.

DISCUSSION

PPB, which is a rare dysontogenetic neoplasm of the thoracopulmonary region, has been reported in the pediatric age between 2 and 5 yrs (1). Its occurrence in the adulthood is exceptional. On the clinicopathologic grounds, cases of adult PPBs including the present one have shown similar findings with childhood tumors in many aspects (3).

There are three pathological types (type I, II, and III) of PPB based on gross and microscopic features (2). The solid areas of the types II and III PPB are composed of undifferentiated blastemal tissue which may overlap with spindle cell sarcomatous, rhabdomyosarcomatous, anaplastic, and chondrosarcomatous foci. Our case was morphologically consistent with type II PPB. The three types of PPB are presumed to have histogenetic linkage with the potentials of progression into other forms. Priest et al. (4) suggested that the three types of PPB had median ages at diagnosis of 10, 34, and 44 months for types I, II, and III, respectively. The pathogenetic linkage among them was further supported by the report of recurrence of a type I PPB as a type II PPB. Because of the histologic variability in the solid component of the PPB, the differential diagnosis can be broad. However, the unique qualities of the PPB, such as cystic architecture, blastemal tissue, and other mesenchymal components including cartilage, justify its designation as a distinct entity. For its development, the tumor may arise from the lung or it may be extrapulmonary, involving the mediastinum or parietal pleura. This has raised the possibility that PPB might originate from the splanchnopleural or somatopleural mesoderm (1, 5). Our case showed predominant anterior mediastinal location with the partial involvement of lingular segment of the left lung.

Clinically, the tumor equally affects both sexes. Shortness of breath in the absence of other respiratory tract symptoms is the main symptom in most cases of PPB reported. A suspected pulmonary infection is the most frequent clinical impression in these patients. The tumor is known to have no characteristic findings on imaging studies, but it is important to bear in mind that type I PPB can not be differentiated from other benign cystic lung lesions on imaging studies (4). Our patient complained of an insidious chest pain with mild dyspnea, and a ruptured cystic teratoma was suspected on radiological evaluation. For the metastatic propensity, the CNS metastasis rate was 44% of all recurrences in the study of 50 cases of PPB by Priest et al. (4), and is distinctly higher than in most other types of solid malignant neoplasms in childhood (6). The skeletal system is the second most common site of the metastasis. Therefore, the two sites should be investigated in newly diagnosed patients. Our case showed neither metastasis nor recurrence about 6 months after operation, although he was lost to follow-up observation since then. The treatment of PPB is primarily surgical excision followed by chemotherapy (4). Types II and III PPBs are clearly aggressive malignancies with projected overall survival of 62% at 2 yrs and 42% at 5 yrs, even after multimodality therapy (4). The adult PPB case reported by Hill et al. (3) received aggressive chemotherapy with cyclophosphamide, doxorubicin, vincristine, ifosfamide, and etoposide. The therapy yielded substantial palliative benefit for some time. However, with the tumor metastasized to the right hip, he developed progressive respiratory dysfunction leading to death approximately 10 months after diagnosis of the tumor. In our case, we planned postoperative combination chemotherapy with vincristine, actinomycin, and cyclophosphamide, but the patient refused any further treatment.

A significant feature of patients with PPB is the extraordinarily high prevalence of other tumors in close relatives, which has been reported to be as high as 25%. Associated conditions include PPB, medulloblastoma, malignant germ cell tumor, thyroid neoplasia, and others (7). Additionally several cytogenetic studies have shown the detection of trisomies 2 and 8 in isolated cases (8-10). Thus, once the diagnosis of PPB is made, a thorough search for tumors should be initiated with the close relatives. Unfortunately, we could not perform cytogenetic studies in the current case.

Taken together, despite some questions still unanswered, we think that the current case is consistent with type II PPB occurring in a young adult.

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