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Pleuropulmonary Blastoma: Case Report

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ABSTRACT

Pleuropulmonary blastoma (PPB) is a rare and aggressive tumor that is emerging as a distinct entity of childhood disease .It is characterized by mesenchymal elements (including undifferentiated blastoma and often cartilaginous, rhabdomyoblastic, or fibroblastic differentiation) and epithelium-lined spaces. PPB may be exclusively cystic (type I), solid (type III) or both solid and cystic (type II).

A 5-month-old boy presented with a history of fever and respiratory distress. Chest radiograph and subsequent CT scan showed a large soft-tissue density occupying the left chest cavity. Radical resection of the mass was achieved by lobectomy. Histologic examination revealed PPB (type II). ***(Tanaffos 2004; 3(11): 71-76)***

Key words*: Pleuropulmonary blastoma, Pulmonary blastoma, Lungs*

INTRODUCTION

Pleuropulmonary blastoma (PPB) is a rare and highly aggressive intrathoracic malignancy in childhood and less than 100 cases have been reported in the literature.

In 1961, Spencer first used the term and suggested that PPB arose from mesodermal blastoma because of its similarities to nephroblastoma. In the year 1988, Manivel et al. described PPB in children as an entity that was distinct from the biphasic epithelial-stromal morphology of the classic adult type. Unlike pulmonary blastoma, PPB lacks the malignant epithelial component and entirely consists of primitive blastoma showing varying levels of sarcomatous differentiation (1,2).

We present here a case of this rare tumor.

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CLINICAL SUMMARY

A 5-month-old boy presented with respiratory distress and fever. He had a 20-day history of persistent cough and low fever. Chest radiograph showed opacity of the left hemithorax. Thoraco-abdominal CT-Scan revealed a large soft-tissue density occupying the left chest. The patient underwent left thoracotomy and a large friable and hemorrhagic tumor in the left upper lobe which was adherent to the visceral pleura was resected completely.

The patient recovered uneventfully and was discharged on the third postoperative day. The parents refused postoperative chemotherapy and until this time (24 months after surgical resection) the patient is well and there is no evidence of recurrent tumor or metastasis.

Pathologic findings:

Macroscopy:

The specimen was left superior lobe that was tumoral measuring 8×7×3 cm and weighing 90 gr. The cut surface of the mass showed grayish-white, fleshy tissue with foci of necrosis and hemorrhage as well as small cystic spaces measuring up to 1.5 cm in maximum dimension containing serosanginous fluid.

Microscopy:

The tumor consisted of blastomal stromal cells with round or spindle shaped nuclei arranged in short fasicles or loose cells in a myxoid matrix with numerous mitoses and large areas of necrosis. There were storiform areas as well as foci of chondroid differentiation. The stromal cells also were condensed beneath the attenuated and unremarkable lining of cystic spaces. There were also benign epithelial components like entrapped alveolar ducts in tumor. Immunohistochemical staining demonstrated a diffuse strong positivity for vimentine and a focal positivity for actin and desmin.

Pr- S100 was expressed in chondroid areas and P53 was completely negative. These findings suggested the diagnosis of pleuropulmonary blastoma (type II).

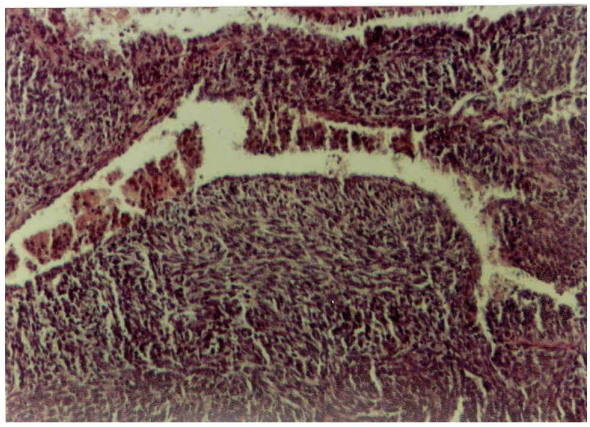
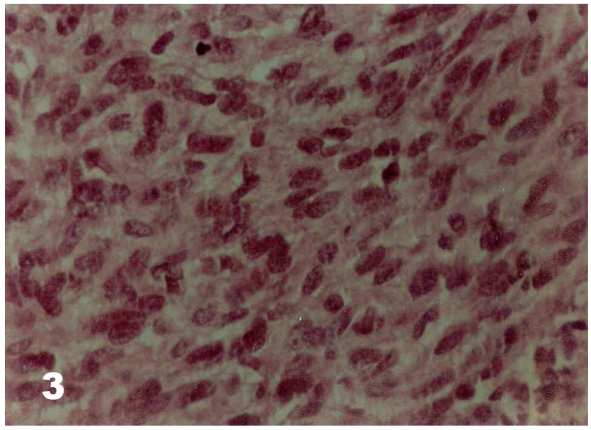


Figure 2. An entrapped air space with flattened epithelioma at the peripheral part of tumor.



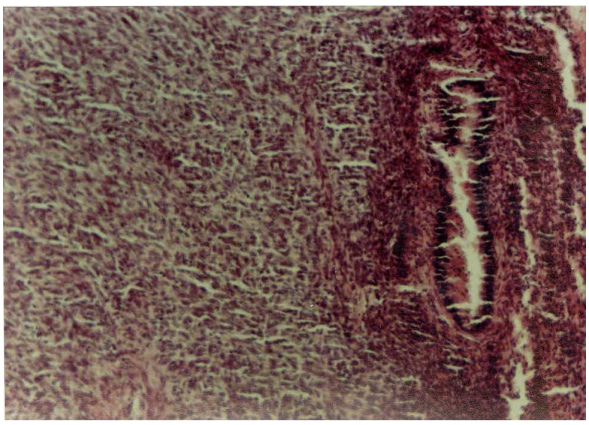


Figure 1. Bronchiolar structure adjacent to tumor which is composed of round to oval cells.

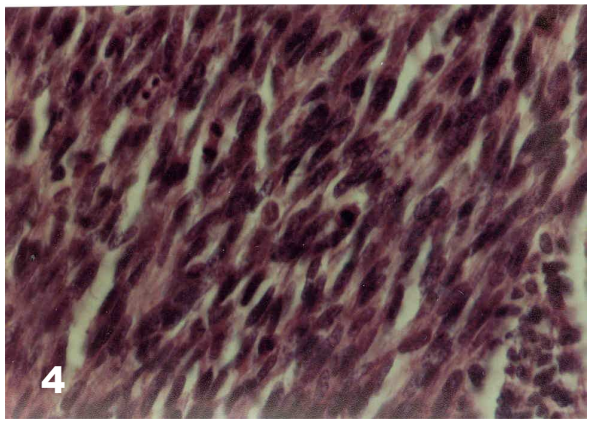


Figure 3, 4. Oval to spindle shaped cells with nuclear atypia and mitotic activity.

DISCUSSION

Pleuropulmonary blastoma is a primary intrathoracic malignancy that occurs mainly in early childhood. It is composed of immature mesenchyme, often differentiating toward skeletal muscle, cartilage, fibrous tissue, and sometimes fat, and most often includes epithelium. The mesenchymal elements are regarded as malignant. Since PPB was recognized as clinicopathologic entity distinct from adult pulmonary blastoma, which is characterized by malignant glands and malignant stroma, the epithelial elements in PPB have been described as benign (2). In the past, they have been termed pulmonary sarcoma arising in mesenchymal cystic hamartoma, embryonal sarcoma, or rhabdomyosarcoma arising in congenital cystic adenomatoid malformation or bronchogenic cysts (3).

An excellent clinicopathologic review of the condition has been carried out by Priest et al (4).

The age onset of presentation was between two weeks to 96 months. Although respiratory difficulty with or without fever is the most common clinical symptom, PPB can present with spontaneous pneumothorax (5,6), or empyema (7).

There are few reports of bilateral PPB (8,9). A significant feature of patients with PPB is the extraordinary high prevalence of other tumors in close relatives, which has been reported by Priest et al. to be as high as 25% (1,4).

In general, there are no characteristic findings on imaging studies.

Dehner et al. have proposed a classification scheme for PPB that divides these lesions into predominantly cystic (type I), cystic and solid (type II), and predominantly solid (type III) types. Each type is characterized by increasing histologic evidence of malignancy (10). There is a report in which the progression of PPB from type I to type III has been documented over the time (11).

Grossly, cystic tumors are single or multiloculated and may show nodular, thickened walls or pedunculated nodules. Solid tumors are multi lobulated white-gray with focal hemorrhage (3).

Microscopically cystic lesions consist of one or more spaces lined by benign alveolar or ciliated columnar epithelial cells, beneath them, there is a layer of primitive oval and spindled rhabdomyoblasts in a loose or dense fibrovascular stroma. Solid tumors consist of blastomal stromal cells, arranged in alternating bands of compact and loose cells in myxoid matrix. There may be anaplastic and pleomorphic mesenchymal cells with numerous mitoses. Areas of chondrosarcoma, rhabdomyosarcoma, and smooth muscle-like spindle cells, storiform areas, and rarely foci of lipoblastic differentiation may be found. There are also small cysts, which have a lining of histologically benign epithelial cells that are probably entrapped bronchiolar, alveolar, or mesothelial cells (3).

Immunohistochemical staining mirrors a range of differentiation, with vimentin, histiocytic markers, or myoid antigens being common (3,12).

Ultrastructural examination demonstrated a similarity between the proliferating cells in PPB and those seen in malignant fibrous histiocytoma which are mainly composed of mixture of primitive, fibroblastic, myofibroblastic, histiocytoid, and fibrohistiocytoid cells (3, 12).

The diagnosis is made only on histologic evaluation of the excised mass; however, fine- needle aspiration cytology has been used to diagnose it (13,14,15,16). Several cytogenetic changes such as trisomy 2, trisomy 8, and recently P53 mutation have been reported that the last one had fatal outcome (17, 18, 19, 20).

Vargas et al. demonstrated that gains in chromosome 8 are a consistent finding in PPB and are confined to mesenchymal components in this neoplasm (21).

Because of histologic variability in the solid

component of PPB, the differential diagnosis has a broad range. The most important differential diagnoses are rhabdomyosarcoma, malignant peripheral nerve sheath tumor, extrarenal Wilms' tumor, mesenchymal chondrosarcoma, malignant germ cell tumor and malignant fibrous histiocytoma (4).

In the case of type-I PPB, the diagnostic pitfall is failure to sample or recognize the immature mesenchymal cells, often with a rhabdomyoblastic immunophenotype, beneath the epithelial surface of the cysts. Thickened or plaque- like areas within such a cyst may yield the presence of blastematous and/or sarcomatous foci and if so, the neoplasm is a type II PPB (4).

The rarity of PPB has allowed only slow elucidation of its clinical features according to prognosis and its response to therapy (22). The treatment is primarily complete excision of the tumor (23,24), followed by intense chemotherapy (25). Although there is disagreement in the literature, local radiotherapy also has been applied to PPB (1).

Metastatic spread can also affect the ipsilateral lobes of the lung, the central nervous system including the spinal cord, and skeletal system. The relative frequency of CNS deposits occurring even 60 months after diagnosis suggests that long term monitoring may be prudent (1,4).

The prognosis depends largely on the staging at the time of diagnosis and the grading of the sarcomatous elements, but in general these are aggressive neoplasms with a 5-year survival probability of less than 50% of the cases with a solid component (26).

Our patients presented with type II lesions had intermediate prognosis (73% survival rate according to Priest et al.) (4).

The favorable outcome of our patient is probably due to the radical resection of the neoplasm.

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