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PLEUROPULMONARY BLASTOMA - A RARE PEDIATRIC MALIGNANCY

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ABSTRACT

Pleuropulmonary blastoma (PPB) is a rare dysontogenetic and aggressive intrathoracic malignant pulmonary neoplasm of early childhood. We present a case of pleuropulmonary blastoma in 2.5 year old female child who was referred to our hospital due to right side pleural effusion & right hydropneumothorax. Chest scan showed a large lytic lesion occupying most of the right hemithorax. Right posterolateral thoracotomy with excision of cyst was performed; the tumor was arising from the lower lobe and invading the chest wall. Histologically the lesion was pleuropulmonary blastoma type 2 with primitive looking round to oval blastematosus cells & sarcomatous differentiation. Prognosis of this type of tumor is grave with 5 year survival is 42%.

Keywords: Lung, malignancy, Pleuro pulmonary blastoma

INTRODUCTION

Primary pulmonary neoplasms are uncommon in children. One such tumor, pleuropulmonary blastoma (PPB), is very rare, highly aggressive and malignant. It originates from either the lungs or pleura. It contains both malignant mesenchymal and benign epithelial elements that resemble fetal lung and occurs mainly in children aged less than five years (1).

CASE REPORT

A 2.5-year-old female child was referred to our institute for evaluation of pleural effusion on a chest X-ray. She had respiratory distress, persistent cough and low grade fever since 1 month. She had received medical treatment for pediatric TB for 4 months but there is little clinical improvement. The abnormal chest x-ray showed no improvement.

She presented with a picture of pneumonia with pleural effusion. CT scan showed a large cystic mass occupying most of the right hemithorax. (Figure 1).

A thoracentesis was performed and about 50 ml of serous fluid was aspirated which shows no growth on culture.

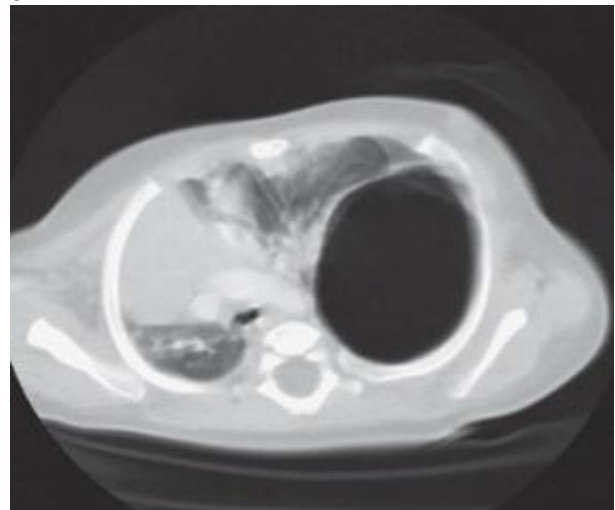


Figure 1: CT scan chest mediastinal window showing a large tumor mass occupying most of the right side of the chest; the mass is mainly cystic with some solid areas.

Patient underwent a right thoracotomy. There was large solid cystic mass. There was no hilar or

mediastinal lymph nodes; freeing the mass from the chest wall by extra pleural dissection was successful and right lower lobectomy was completed. The resected tumor was cystic with associated solid parts of fleshy, gray white masses. They sent it for histopathological examination.

On Histopathological examination, grossly, multiple large greyish white soft tissue fragments aggregate measuring approximately 16x12cm. The cut surface of the tissue fragments showed brownish, fleshy appearance with foci of necrosis and haemorrhage as well as small cystic spaces. Multiple sections were taken from the received specimen and stain with haematoxylin & eosin. Histologically, Sections show highly cellular tumor area, separated by widely open branching, congested, thin walled vasculature. Tumour cells are primitive looking monomorphic, with round to oval hyper chromatic nuclei & scant ill defined cytoplasm, with mitosis. Areas of Spindled tumor cells with bipolar elongated eosinophilic cytoplasm arranged randomly in loose myxoid background is seen suggesting rhabdomyoblastic

nature of tumor cells. There is also loose myxoid mesenchymal area and anastomosing sheet of well defined glandular & cuboidal epithelial area. There is also chondromyxoid nodule surrounded by strands of squamoid epithelial cells.

So collectively,

- Pleura based mediastinal solid cystic tumor in pediatric age group with following predominant microscopic findings :
 1. Primitive looking large stromal cell component
 2. Chondromyxoid areas with foci of well defined benign cartilage component
 3. Sarcomatous element (spindle cell area) resembling rhabdomyosarcoma
 4. Well defined benign epithelial element in the form of cuboidal to columnar epithelial lined cyst & papillae.

These findings correspond to a type II PPB. Postoperative course was uneventful.

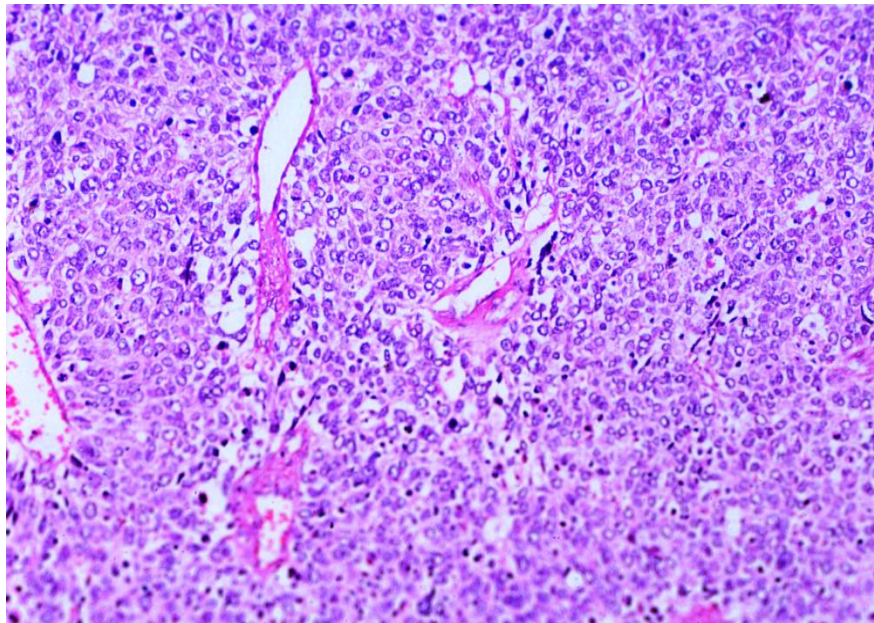


Figure 2: The primitive looking cellular area showing increase mitotic activity

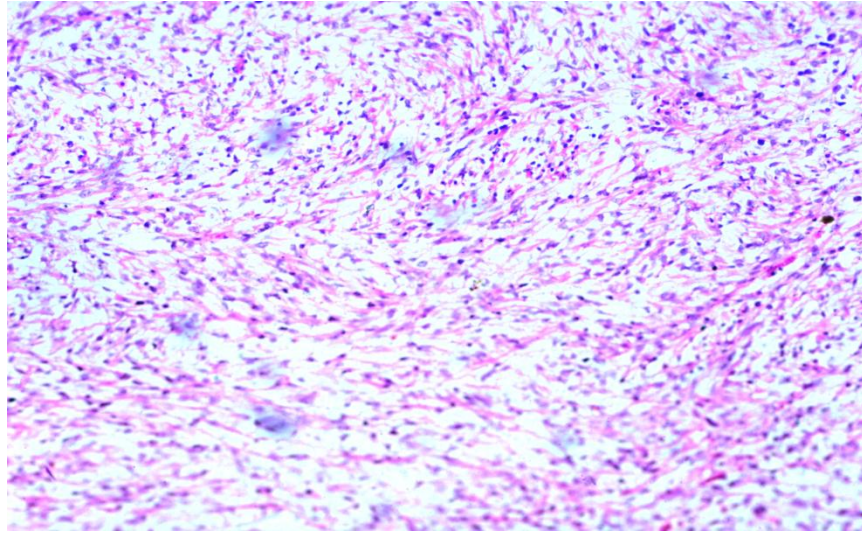


Figure 3: Spindled tumor cells arranged randomly in loose myxoid background

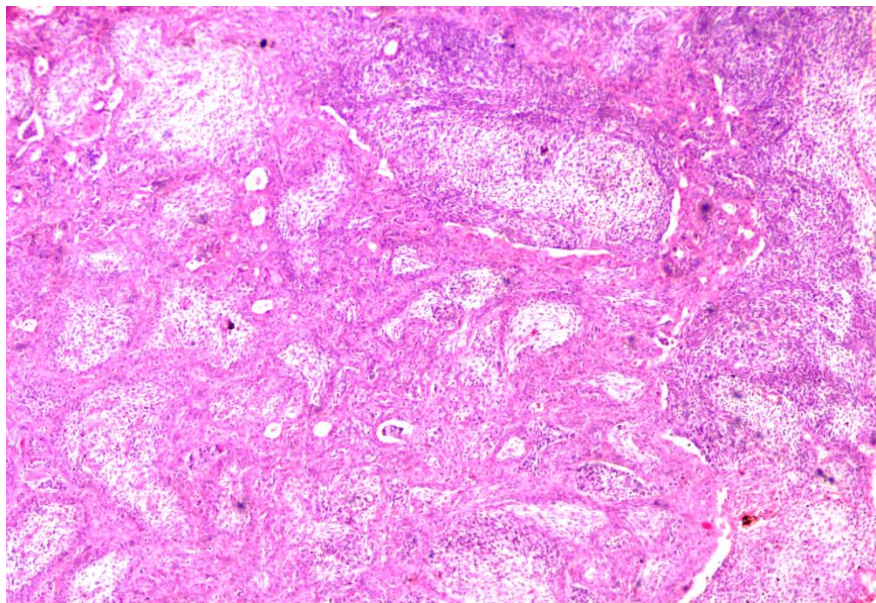


Figure 4: Loose myxoid mesenchymal & anastomosing sheet like epithelial area



Figure 5: Chondromyxoid nodule surrounded by strands of squamoid epithelial cells

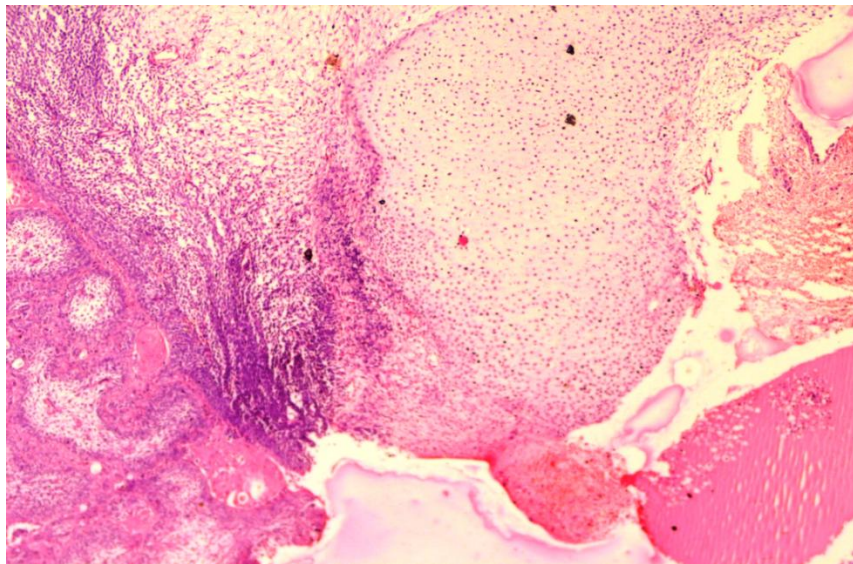


Figure 6: Benign chondroid lobule & anastomosing epithelial strands surrounded by myxoid mesenchymal component.

DISCUSSION

Pleuropulmonary blastoma is an aggressive tumor accounting for less than 1% of all primary malignant lung tumors in the pediatric population (2). Manivel and associates coined the term

Pleuro pulmonary blastoma to describe a specific subtype of pulmonary blastoma on the basis of its exclusive clinical presentation in childhood and its pathologic features of variable anatomic location, primitive embryonic-like blastema and

stroma, absence of a carcinomatous component, and potential for sarcomatous differentiation (3). It arises from Thoracopulmonary mesenchyma. There is a Proliferation of primitive mesenchymal cells which initially form Air-filled cysts lined by benign-appearing epithelium. Then Cells outgrow the cysts with formation of cystic solid area and finally only solid mass.

Accordingly Denher and associates classified PPB into three groups: type 1 with purely cystic tumors, type 3 with predominantly solid tumors, and type 2 as an intermediate type (4).

A progression from type I to type III over time may occur and each type is characterized by increasing histologic evidence of malignancy (5). The histologic appearance is variable - the tumor is characterized by primitive blastema and a malignant mesenchymal stroma often showing multidirectional differentiation as rhabdomyosarcomatous, chondrosarcomatous or liposarcomatous. The cystic component is lined by benign metaplastic epithelium (6).

Vargas et al. demonstrated with cytogenetic analysis that the polysomy of chromosome 8 is a constant feature of pleuropulmonary blastoma and the clonal proliferation in pleuropulmonary blastoma is restricted to the malignant mesenchymal elements, supporting the notion that the epithelial components are non-neoplastic. (7)

Immunohistochemically, the neoplastic cells can stain positive for vimentin. CD117 (c-kit) and alpha-1-antitrypsin are focally positive in tumor cells. CD99 is weakly positive. Other immunostains including EMA, myogenin, S100, GFAP, neuron specific enolase, TTF-1, alpha-fetoprotein, chromogranin, and synaptophysin are negative. The pneumocytes lining the cysts and small airspaces are highlighted by cytokeratin. Muscle specific actin and desmin are expressed in rhabdomyoblasts and primitive cells in the subepithelial regions of cystic lesions. (11)

This neoplasm occurs not only in lung, but it may arise from mediastinum, diaphragm and/or

pleura. This has raised the possibility that PPB might originate from the splanchnopleural or somatopleural mesoderm. Common metastatic sites include the brain, bone, lymph nodes, liver, pancreas, kidney, and adrenal glands (8).

PPB may be associated with cystic pulmonary lesions, which may be evident at the time of diagnosis or predate the appearance of the tumor; there are contradictory reports about the value of prophylactic resection of the pulmonary cysts in protecting patients from developing PPB (9).

The occurrence of PPB appears to herald a constitutional and heritable predisposition to dysplastic or neoplastic disease in approximately 25% of cases. Associated conditions include PPB, medulloblastoma, malignant germ cell tumor, thyroid neoplasia, and others. Thus, All patients with PPB and their families should be investigated carefully (2).

This patient, like most reported cases, presented with a picture of pulmonary infection and respiratory difficulty. The tumor has no characteristic findings on imaging studies, but it should be considered in the differential diagnosis of other benign cystic lung lesions on imaging studies (6).

As complete tumor ablation is essential to prevent local recurrence and allow any chance of survival, the main goal of therapy should be radical surgery, followed by chemotherapy. Because the response to chemotherapy is poor, some authors suggest that chemotherapy should be given with local radiotherapy in the majority of patients (10).

The prognosis for these patients is grave 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. Patients with pleural, mediastinal or extrapulmonary involvement at the time of diagnosis have worse prognosis than those without such involvement (6, 10)

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REFERENCES

1. Calabria R, Srikanth MS, Chamberlin K, Bloch J, Atkinson JB. Management of pulmonary blastoma in children. *Am Surg* 1993; 59:192-196.
2. Priest JR, Watterson J, Strong L, Huff V, Woods WG, Byrd RL. Pleuropulmonary blastoma: a marker for familial disease. *J Pediatr* 1996; 128: 220-4.
3. Manivel JC, Priest JR, Watterson J, Steiner M, Woods WG, Wick MR, Dehner LP. Pleuropulmonary blastoma. The so-called pulmonary blastoma in childhood. *Cancer* 1988;62: 1516-1526.
4. Dehner LP, Watterson J, Priest J. Pleuropulmonary blastoma. A unique intrathoracic pulmonary neoplasm of childhood. *Perspect Pediatr Pathol* 1995;18: 214-226.
5. Wright JR Jr. Pleuropulmonary blastoma: A case report documenting transition from type I (cystic) to type III (solid). *Cancer* 2000 ;88(12):2853-8
6. Priest JR, McDermott MB, Bhatia S, Watterson J, Manivel JC, Dehner LP. Pleuropulmonary blastoma: a clinicopathologic study of 50 cases. *Cancer* 1997; 80: 147-61.
7. Vargas SO, Nose V, Fletcher JA, Perez-Atayde AR. Gains of chromosome 8 are confined to mesenchymal components in pleuropulmonary blastoma. *Pediatr Dev Pathol* 2001 ;4(5):434-45
8. Kelsey AM, McNally K, Birch J, Mitchell EL. Case of extrapulmonary, pleuropulmonary blastoma in a child: pathological and cytogenetic findings. *Med Pediatr Oncol* 1997; 29: 61-4.
9. Papagiannopoulos K A, Sheppard M, Bush AP, Goldstraw P. Pleuropulmonary blastoma: is prophylactic resection of congenital lung cysts effective? *Ann thorac surg* 2001;72(2):604-605
10. Indolfi P, Casale F, Carli M, Bisogno G, Ninfo V, Cecchetto G, Bagnulo S. Pleuropulmonary blastoma: management and prognosis of 11 cases. *Cancer* 2000 ; 89(6):1396-401
11. Yeh YA, Edelman MC. Lung: Pleuropulmonary blastoma. *Atlas Genet Cytogenet Oncol Haematol*. July 2010.