

# PLEUROPULMONARY BLASTOMA: A CASE REPORT

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## ABSTRACT

*A 2 years old girl was managed for pulmonary complaint of cough, dyspnea, and wheezes. Computerized Topographic scan of the chest showed a mixed solid and cystic mass that was proved histopathologically as Pleuropulmonary blastoma (PPB) after excision by thoracotomy which is a rare primitive aggressive malignant neoplasm of the thorax occurring in childhood. The patient was treated by chemotherapy and she was quite well on her follow up after one year.*

**Key Words:** *Pleuropulmonary blastoma, Pulmonary blastoma, Pleural effusion, Hemothorax.*

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## INTRODUCTION

Pleuropulmonary blastoma (PPB) is a rare primitive aggressive malignant neoplasm of the thorax occurring in childhood<sup>1</sup>. This was first described by Barnard in 19522 but the term PPB was first used by Manivel et al<sup>3</sup>. Three histo-pathological types are described; Type I is exclusively cystic, type II mixed (cystic and solid) and type III is a solid tumor without epithelial lined cystic spaces<sup>4</sup>. PPB can occur within a family environment but has not been described as a well-defined component of a familial childhood cancer<sup>5,6</sup>. It originates from the pleura, lung parenchyma and mediastinum. Extrathoracic metastases can occur (especially in type I and II) and the common metastatic sites include the brain, bone, lymph nodes, liver, pancreas, kidneys and adrenal glands<sup>7</sup>. Despite the introduction

of multimodal therapy (surgery, chemotherapy and occasionally radiotherapy), the prognosis for PPB patients remain poor. In this report, a case of PPB is presented whose management was carried out successfully with surgical resection followed by chemotherapy, and the patient was well with no evidence of recurrence after one year of follow up.

## CASE REPORT

A 2 year old girl was admitted to the hospital with the complaints of cough, progressive dyspnea and wheeze. Her previous medical and family history was unremarkable. Breath sounds were diminished in left lung zones. Percussion revealed dullness in the same regions. Chest X-ray showed an opacity filling the left hemithorax and shifting of mediastinum to the right side (Figure 1).

Left thoracocentesis was performed with drainage of 50 ml of hemorrhagic fluid. Pleural fluid analysis showed exudative effusion. The cytology was non conclusive. Pleural fluid and serum level of alpha-fetoprotein (AFP) and beta human chorionic gonadotropin (b-HCG) were within normal limit as were other biochemical and hematological investigations. Chest computed tomography (CT-scan) revealed a large mass in the left hemithorax containing both solid and cystic components without pulmonary invasion (total collapse of the left lung) {Figure 2}.

The patient underwent left thoracotomy, which demonstrated a little amount of hemorrhagic pleural effusion with large solid mass with multiple thin wall cysts containing hemorrhagic fluid, and loosely adhered to the chest wall (but not the lung)

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and arise from anterior mediastinum. Total excision of the mass was done and two chest drains were placed.

The histopathological report showed one friable mass in two halves measuring 13×10cm. On cut section, it showed predominantly solid mass which was whitish-brown in color and soft in consistency with hemorrhagic pleural fluid. Microscopical Examination revealed predominately solid tumor showing biphasic components; primitive blastomatous elements composed of sheets of small round cells with finely granular chromatin of round/ oval nuclei and scanty cytoplasm admixed with sarcomatous spindle cells and tumor giant cells with myxoid stroma.

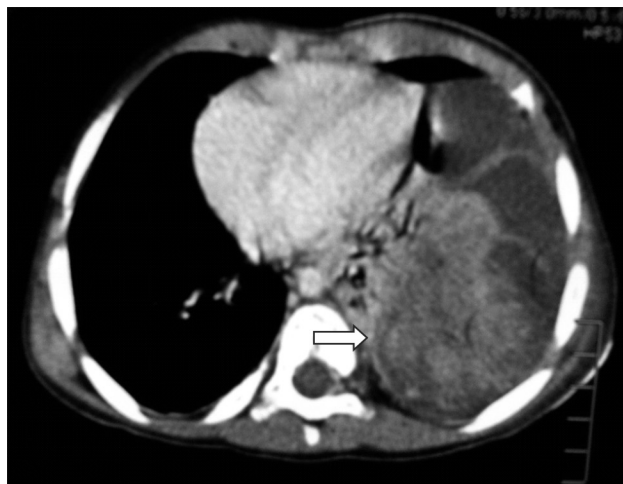
The tumor showed frequent abnormal mitoses with extensive areas of haemorrhage and tumor cells necrosis and the final diagnosis of pleuropulmonary blastoma was made (Figure 3).

The postoperative period passed smoothly and the patient was transferred to the paediatric oncology department to be started on chemotherapy in the form of IE/VAC (ifosfamide, and etoposide/ vincristine, actinomycin D and carboplatin). After two weeks, the patient developed mild left pleural effusion and thoracocentesis was done. No more complications occurred and no radiological evidence of recurrence was seen after one year of follow up (Figure 4).

**Figure 1: Plain chest X-ray: Total opacification of the left hemithorax with mediastinal shift to the right side**



**Figure 2: Chest CT-scan with contrast: Mixed solid and cystic mass involving the whole left hemithorax with total collapse of the lung (white arrow)**

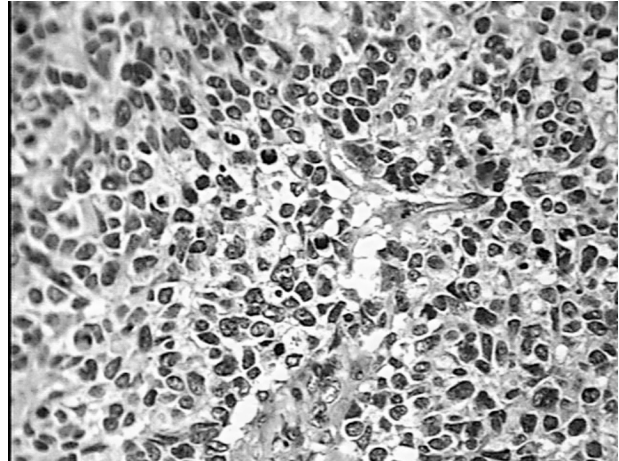


**Figure 3: Histopathological characteristics of Pleuropulmonary Blastoma**

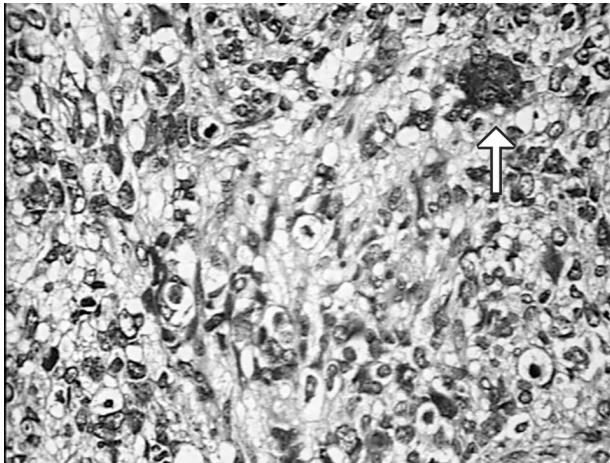
**A: Gross picture showing solid mass**



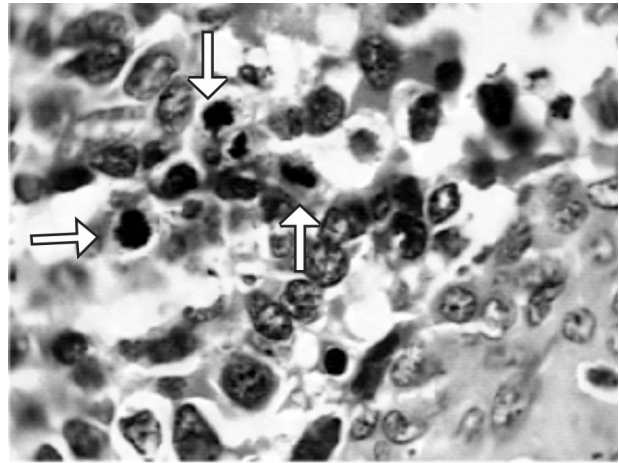
**B: Blastomatous components of Hyperchromatic round cells with scanty cytoplasm**



**C: Sarcomatous components of spindle cells with tumor giant cells (arrow) and myxoid background**



**D: High grade tumor with marked pleomorphism and frequent mitoses (arrows)**



**Figure 4: Plain chest X-ray: Taken 6 months postoperatively (Normal)**





## DISCUSSION

PPB is a rare embryonal neoplasm accounting for less than 1% of all primary malignant intrathoracic tumors in young children<sup>6</sup>. It arises not only within pulmonary parenchyma but also in association with pleura and mediastinum, like this case, and is in contrast to pulmonary blastoma that arise only within the lung parenchyma. PPB has two histological components; the blastemal cells and the mesenchymal cells. In many instances, mesenchymal differentiation is recognized as chondrosarcoma, liposarcoma and rhabdomyo-sarcomatous elements. Novak et al report that trisomy 8 is a karyotypic abnormality associated with PPB<sup>8</sup> but karyotyping was not done in our case.

The preoperative diagnosis is difficult in spite that the chest CT-scan is sensitive in detecting and delineating the lesion; and that is because the wide range of differential diagnoses including lymphoma, mediastinal tumors, metastases in the lung or pleura, congenital pulmonary malformations, inflammatory pseudotumor, and even tuberculous. Fine needle aspiration cytology still has a role in preoperative diagnosis<sup>9</sup>. In 25% of patients, there is a familial association with other dysplasia and neoplasm, especially the cystic nephroma, medulloblastoma and germ cell tumors<sup>6,10,11</sup>. Congenital pulmonary malformations (congenital bronchogenic cyst and congenital cystic adenomatoid malformation) are important predisposing factors for development of PPB<sup>12-14</sup>, thus early resection of congenital lung anomalies is advisable<sup>15</sup>. This anomaly was not present in our case.

PPB is usually unilateral, although bilateral lesion have been reported<sup>16</sup>. Transition from type I cystic to type III solid has also been reported<sup>17</sup>. The total surgical resection is the cornerstone of the treatment, as was done in the case of this patient.

## CONCLUSION

PPB is a rare aggressive malignant neoplasm with poor prognosis especially in type II and III. Despite the total resection done and chemotherapy given to our patient and no evidence of recurrence or metastases noticed, 12 months postoperatively, we still suspect poor prognosis for our patient as it was type II PPB, and had de novo origin from the pleura and the mediastinum<sup>18</sup>.

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