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Pleuropulmonary Blastoma in Pediatric Age Group: A Very Rare Case Report and Review of Literature

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Abstract

Pleuropulmonary blastoma (PPB) is a very rare and very aggressive malignant tumor that affects children and adults. Though its presentation in children has been reported in literature, incidence is very low as compared to adults. This neoplasm is characterized on histology by primitive blastema and a malignant mesenchymal stroma that often demonstrates multidirectional differentiation. Even though availability of multimodal therapy, the prognosis of patients with PPB remains poor.

Keywords

Pleuropulmonary Blastoma, Rare Pediatric Lung Cancers, Rare Causes of Solid Lung Lesions

1. Introduction

Dysontogenic neoplasms are a unique category of tumours observed exclusively in childhood such as Hepatoblastoma, Wilms' tumour, Neuroblastoma [1].

Pleuropulmonary blastoma is a very rare and highly aggressive pulmonary malignancy in children. In 1961 first time Spencer coined the term and suggested that PPB arose from mesodermal blastema because of its similarities to nephroblastoma [2]. In 1988 Manivel *et al.* described pleuropulmonary blastoma in children as an entity that was distinct from the biphasic epithelial-stromal morphology of the classic adult type [3]. Mainly in pediatric patients, the lesion is supposed to be a truely dysembryogenic neoplasm of thoraco-pulmonary mesenchyma without malignant epithelial cells.

Which is characterized histologically by primitive blastema and a malignant mesenchymal stroma that often shows multidirectional differentiation (a

rhabdomyosarcomatous, chondrosarcomatous, or liposarcomatous pattern). Pleuropulmonary blastoma in pediatric age is symptomatically mistaken for respiratory tract infection as pneumonia. Tumor is usually located in lung periphery as extrapulmonary mass, occasional involvement of mediastinum, diaphragm or pleura. Brain, bone, lymph nodes, liver, pancreas are common metastatic sites. Even though availability of multimodal therapy, the prognosis of patients with PPB remains poor. Herewith reporting a rare case of 2 years old baby girl presented with plueropulmonary blastoma of lung treated with multimodal approach and no recurrence till follow up being labeled as highly aggressive tumor with uncertain outcome in cases reported in literature.

2. Case Report

2 years baby girl was admitted for respiratory tract infection with left lower zone pneumonia under pediatric care. Baby was treated with intravenous antibiotics. Serial chest x rays were taken but there was no resolution of patch on left side in view of which case was referred to pediatric surgery. There was no significant antenatal history and no medical history from early neonatal period till date. This was first time baby presented with fever, cough and upper respiratory tract infection. On respiratory examination there was decreased the air entry on left side lower zone significantly. On chest x ray there was complete white patch seen on left lower zone lung shifting to mediastinum on opposite side slightly with prominent bronchial marking seen (**Figure 1**). On High-resolution computed tomography of thorax there was huge completely solid lesion seen on left side of lower zone in lung parenchyma with cut off sign of bronchus and involving pericardium, diaphragm (**Figure 2**). Hematological investigations showed normal range except increased white blood cells count. Patient was planned for open thoracotomy with excision of mass under general anesthesia. Preoperative parental counseling was done for lower lobectomy of left lung depending upon the intra-operative findings and postoperative ventilator support. On left sided open thoracotomy through fifth intercostal space, there was evidence of intra-parenchymal solid mass in left side lower lobe lung involving diaphragm and pericardium medially. Complete in toto dissection was done all around, shaving pericardium partially near the adhered part with lesion (**Figure 3**). Lower lobectomy was done. Bronchus entering into the lesion was transfixed and closed with non absorbable sutures. Lymph nodes clearance was done along the hilar region. Thoracotomy wound was closed after confirming haemostasis and putting inter costal drainaige tube in situ. Baby got extubated without any efforts on table, shifted to intensive care unit for further monitoring. On postoperative day 5, inter costal drainage tube was removed. Post operative chest x ray showed clear and well inflated left lung (**Figure 4**). Baby was maintaining all parameters without suppourt so discharged and kept on follow up. Specimen was sent for histopathology and immunohistochemistry study (**Figure 5**). On histopathology report there was evidence of tumour composed of an admixture of epithelial & stromal components. The epithelial component is mature and shows cystically

Figure 1. Preoperative X ray.

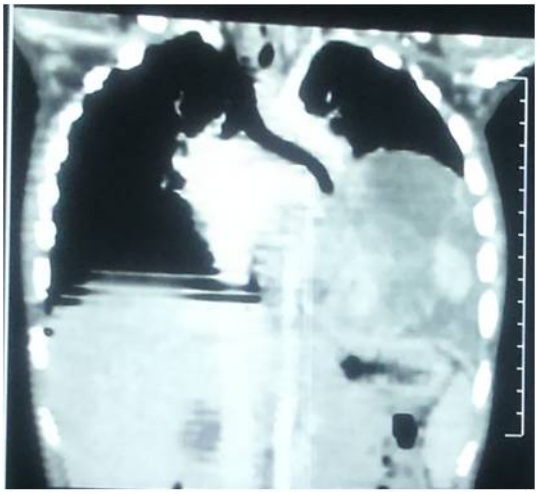

Figure 2. CT Scan thorax image.

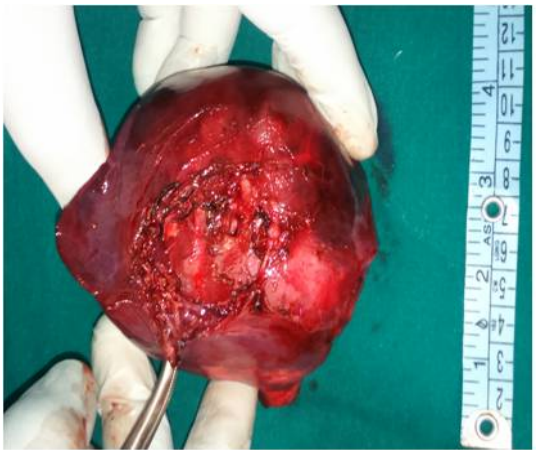

Figure 3. In-toto Excised specimen.

dilated spaces most of which are lined by respiratory epithelium with underlying mucin secreting glands. The stromal component consists of whorls & fascicles of spindle cells with sheets & islands of immature cartilage (Figure 6, Figure 7). Areas of necrosis & hemorrhage are seen. Mitotic activity is 6 - 8/10 Hpf. The adjacent parenchyma shows hemorrhages & changes of interstitial pneumonia.

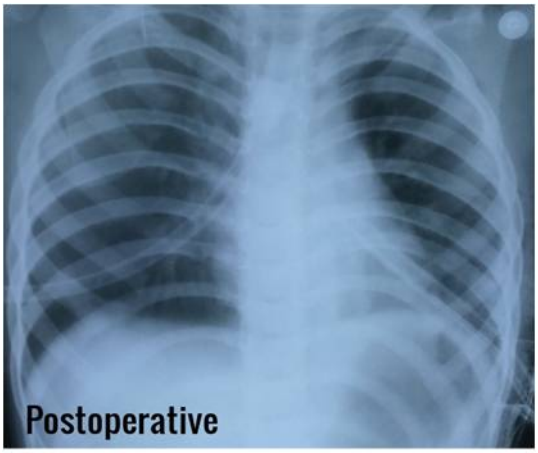

Figure 4. Post operative X ray.

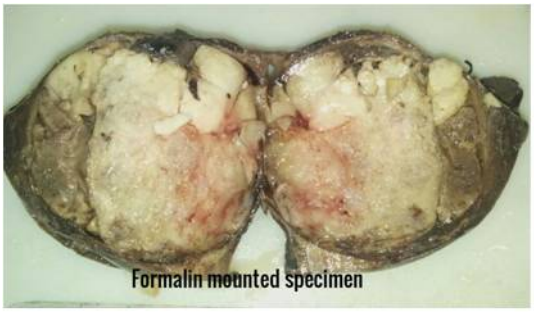

Figure 5. Formalin mounted cut open specimen.

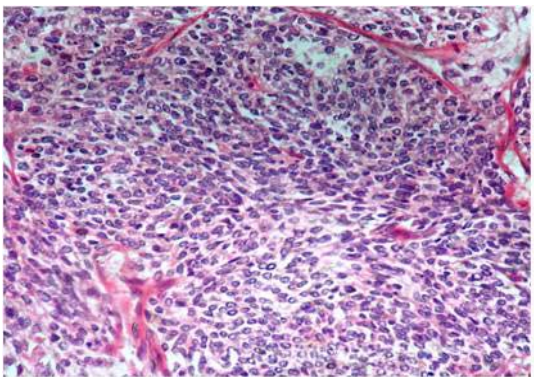

Figure 6. Tumor cells with High Power view.

Lympho-vascular invasion not seen. Impression being type 3 pleuropulmonary blastoma which is a very rare and very aggressive solid neoplasm in childhood with unfavorable prognosis due to predominant solid component. Post operative baby was investigated with PET scan revealing no any signs of distant metastasis. Baby was put on chemotherapy immediately under pediatric oncology unit treated with chemo-regimen including drugs carboplatin, epirubicin, vincristine, actinomycin D, ifosfamide, and etoposide. After completion of 5 cycles, repeat High-resolution computed tomography of thorax and PET scan was done to confirm further spread or recurrence of tumor. As there were no signs

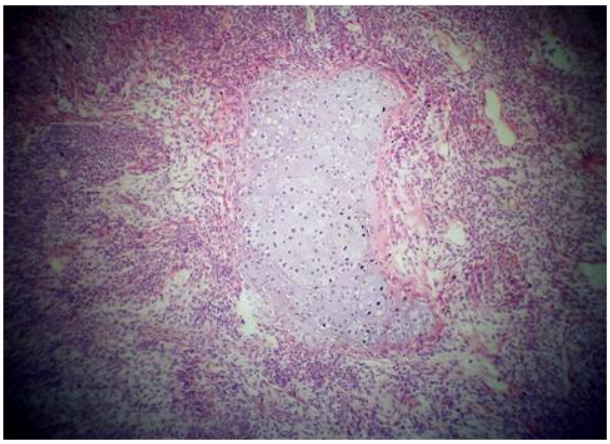

Figure 7. Tumor cells with cartilaginous differentiation.

of recurrence. Currently on 4 years of follow up post chemotherapy, baby is doing good without any signs of infection and recurrence/metastasis.

3. Discussion

Pleuropulmonary blastoma (PPB) is a very rare tumor of unclear histiogenesis. Being highly aggressive tumor in children with overall survival rate of 45% at 5 years and an event free survival rate of 49% at 2 years in the population as per study by Priest *et al.* [4], the rarity of PPB and very few literature reviews have kept restricted to know more about its clinical features and presenatations.

The clinical presentation of the patient in this case report was misinterpreted as an upper respiratory tract infection. For this reason the diagnosis of PPB was delayed for up to 21 days. During investigation, mediastinal or pleural involvement was found to be correlated with a poorer survival so baby was kept under closed follow up till date and reevaluated for recurrence in keeping with the findings of Priest *et al.* [4]. PPB usually develops in younger children without congenital pulmonary malformations. Delahunt *et al.* and Joshi *et al.* described a familial association between PPB and cystic nephroma [5] [6]. Many cases have been reported with formation of lung cysts arising 4 months to 3 years before the development of blastomatous solid lesion, considering to be a predisposing factor for the development of malignancy [7]-[11]. For the same reason surgical resection is always advisable for any lesions with a solid mass in lung parenchyma [12] [13]. Complete excision biposy along with a lobectomy or pneumonectomy, if necessary, is advocated as a preferred treatment in such cases. Literature mentions that patient with initially unresectable stage should be treated with neoadjuvant chemotherapy to reduce the size of lesion and then to be resected out. Although there are huge controversies about chemotherapy regimen, duration of chemotherapy and local radiotherapy to prevent recurrence, considering the very aggressive nature of tumor it is advisable to start chemotherapy immediately in post operative period [14]. Here in this case report, we have used combination of carboplatin, epirubicin, vincristine, actinomycin D, ifosfamide,

and etoposide drugs and showed good tolerance with no recurrence after 4 years of follow up.

Distant metastasis and local recurrence can occur anytime during or after the therapy. Brain metastasis is most common with fatal output. Local recurrence and distant metastasis may occur after or during therapy. Brain metastases are the most common distant lesion and usually are fatal [15].

4. Conclusion

More number of case studies are required to sort out an important issue of duration of chemotherapy to be continued. Herewith this case report, I would suggest having an international cooperative study of pleuropulmonary blastoma to get the answers of due queries and better outcomes in this disease.

Disclosure

An Informed consent has been taken from parents to publish this case report for academic purpose. I declare no potential conflict of interests, real or perceived.

Conflicts of Interest

The author declares no conflicts of interest regarding the publication of this paper.

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