Pleuropulmonary Blastoma in Children: A Case Report

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Abstract

Pleuropulmonary Blastoma [PPB] is a very rare, highly aggressive and malignant tumor that originates from either the lungs or pleura. It occurs mainly in children aged less than five or six years. It has poor prognosis with three different subtypes: cystic [type I], combined cystic and solid [type II] and solid [type III]. PPB is treated with aggressive multimodal therapies including surgery and chemotherapy. We present a case of 3.5 years old boy with PPB type II successfully treated with complete surgical resection followed by neoadjuvant chemotherapy.

Keywords: Pulmonary blastoma; Child; Neoplasms


Introduction

Primary pulmonary neoplasms are uncommon in children. One such tumor, Pleuropulmonary Blastoma (PPB), is very rare, highly aggressive and malignant, and originates from either the lungs or pleura [1]. It occurs mainly in children aged less than five or six years [1, 2]. PPB was classified into three groups by Dehner in 1995 as cystic (type I), mixed (type II), or solid (type III) [3]. Type I occurs in infants (median diagnosis age, 10 months) in contrast to types II and III (median diagnosis ages, 34 and 44 months), respectively[4]. Type I has a more favorable prognosis than type II and III [3]. Type II is similar to Wilms tumor morphologically and therefore, it is sometimes incorrectly called “extra-renal Wilms tumor” [5].

The treatment of children with type I PPB has been a surgical resection with or without adjuvant chemotherapy. Ideally, surgeons should consider the diagnosis of PPB and plan, when possible [6]. PPB is very rare in children and there is no defined optimal treatment regimen for it to date.

In this report, a case of PPB is presented whose management was carried out successfully with surgical resection followed by neoadjuvant chemotherapy.

Case Report

A 40 month old male child was admitted to the hospital with cough, and wheezing. His previous medical and family history was unremarkable. His breath sounds were diminished in the right lung zone. Chest X-ray showed an opacity filling the right lower lobe and leading to mediastinal shift to the opposite side.

Computerized Tomography (CT) of thorax revealed a large mass in the right hemithorax (7x5x3cm) containing solid and cystic components. The patient underwent right thoracotomy, which demonstrated a solid and cystic mass in that partial resection. Histological diagnosis of the tumor was PPB type II because it contained both solid and cystic components. Bone scintigraphy and abdominal USG (Ultrasonography) revealed no abnormality. We scheduled adjuvant chemotherapy with VIE (Vincristine 1.5mg/m2 on day 1, Ifosfamide1 gr/m2 on day1 to3, Etoposide 150mg/m2 on day 1to3) alternated with VAC (Vincristine 1.5 mg/m2 on day 1, Actinomycin-D 15 gamma/kg on day 1, and Cyclophosphamide 750mg / m2 on day 1)]. The ICE and VAC courses were alternated every three weeks. After the completion of the first course of chemotherapy, dyspnea resolved completely, and the third course of chemotherapy revealed a 90% reduction in mass size and the child was well at the 6 month follow up.

Discussion

Pleuropulmonary blastoma is an aggressive tumor accounting for less than 1% of all primary malignant
lung tumors in the pediatric population [7]. Manivel and associates coined the term PPB 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 blastoma and stroma, absence of a carcinomatous component, and potential for sarcomatous differentiation [8].

Denher and associates classified PPB into three groups; type 1 with purely cystic tumors, type 2 as an intermediate type, and type 3 with predominantly solid tumors [9]. A progression from type I to type III may occur over time [10]. The histologic appearance is variable - the tumor is characterized by primitive blastoma and a malignant mesenchymal stroma often showing multidirectional differentiation as Rhabdomyosarcomatous (as in our case), Chondrosarcomatous or Liposarcomatous. The cystic component is lined by benign metaplastic epithelium [11]. 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 [12]. This neoplasm occurs not only in the 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 [13].

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 [14].

The occurrence of PPB appears to herald a constitutional and heritable predisposition to dysplastic or neoplastic disease in approximately 25% of the 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 [7].

The tumor has no characteristic findings on imaging studies; however, it should be considered in the differential diagnosis of other benign cystic lung lesions on imaging studies [11]. The baby in this case like most reported cases presented with a picture of pulmonary infection and respiratory difficulty.

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 the patients [15].

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 extra pulmonary involvement at the time of diagnosis have worse prognosis than those without such involvement [11, 15].

**Conclusion**

PPB is an aggressive tumor having poor outcome and it usually occurs in young children. It needs a multimodal therapy regimen including aggressive surgery and chemotherapy. Though radiotherapy is an alternative treatment modality, especially in cases with high risk of recurrence, it causes severe morbidity, especially in younger children. We concluded that despite the presence of poor prognostic factors such as presence of solid tumor or those factors that increase the probability of local recurrence such as chest tube insertion and microscopically residual disease after surgery, chemotherapy and aggressive surgery may provide remission and a long-term disease-free period. Complete surgical resection of the tumor at the time of diagnosis is the cornerstone of PPB management, but in the majority of patients, initial surgery is incomplete as a large tumor may involve vital structures. Thus, chemo-reduction followed by complete excision may be the satisfactory treatment of this highly aggressive tumor in childhood [16].

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**Conflict of Interest**

The authors have no conflict of interest in this study.

**Authors' Contribution**

Azamsadat Hashemi diagnosed disease, treated and followed up the cases. Azadeh Souzani collected the data and wrote the paper. Amineh Souzani and Sara Keshavarzi helped for data collection.

**References**


