

A Rare Case of Pleuropulmonaryblastoma: A Case Report and Review of Literature

Dr Anandita Milind Desai¹, Dr Ajay Naik²

¹PG Resident MS General Surgery

²Professor and Head of Department General Surgery

Abstract: *Pleuro pulmonary blastoma is a rare form of potentially aggressive pulmonary malignancy seen in children less than 6 years of age. We have studied a case of a 2 year old child who presented to our surgical out patient department with complaints of fever, cough and breathlessness since 8 days and after thorough evaluation was diagnosed with Pleuro pulmonary blastoma and was treated for the same. Pleuro pulmonary blastoma is classified into three subtypes: biphasic pleuro pulmonary blastoma which consists of an epithelial and mesenchymal component, well differentiated foetal adenocarcinoma which consists of well differentiated epithelium and a mesenchymal component and malignant pleuro pulmonary blastoma. The prognosis of the malignant variant is extremely poor and it is an aggressive type of cancer.*

Keywords: griscelli syndrome, hemophagocytosis, Lymphohistiocytic proliferation, Silvery gray hair, hepatosplenomegaly

1. Case Report

- A 2 year old male child presented with complaints of fever, cough and breathlessness since 8 days.
- On examination, there was evidence of a right parasternal bulge; a dull note over the entire right hemi thorax with air entry absent on the right side of the chest.
- Chest x-ray–showed complete haziness in the right hemi thorax.



- USG thorax–revealed thick loculated pleural collection in right hemi thorax with collapse of underlying lung suggestive of empyema.
- HRCT thorax - well defined, heterogenous, soft tissue density occupying the right hemi thorax, most likely neurogenic in origin – Neuroblastoma.

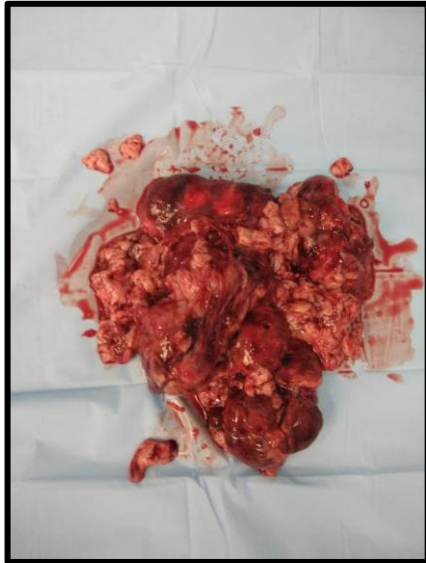
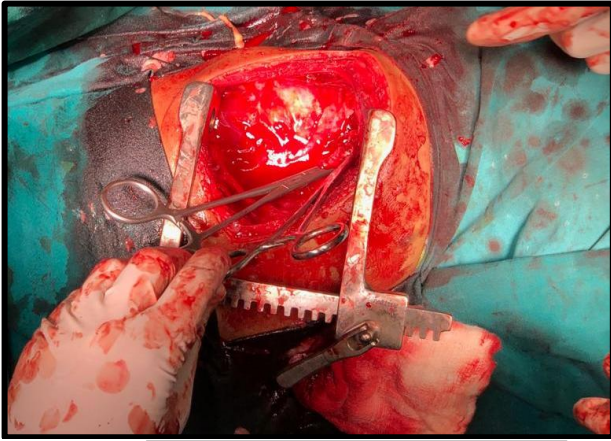


- After complete preoperative work up the patient was posted for surgery
- The patient underwent right open thoracotomy with intra operative findings suggestive of a soft solid tumor arising from the right middle and lower lobes of the lung with upper lobe of the lung normal. Right middle and lower lobectomy was done.

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survival rate post diagnosis. Long-term disease free survival is extremely rare. The tumour typically metastasises to the brain, mediastinum, pleura, diaphragm, liver and soft limb tissue submandibular glands, scrotum and ovaries. Relapses are common within the first 12 months of the surgical excision of the tumour. Relapse, the presence of metastasis and a tumour size exceeding 5 cm are reported to be unfavourable prognostic factors.

Pleuro pulmonary blastoma almost always presents as a unilateral, large, well-circumscribed, solitary mass on chest radiograph. Given the peripheral location of these tumours, tissue diagnosis by bronchoscopy only occurs in 25% of cases but they are well visualized on thoracic ultrasound with the diagnosis being confirmed on HRCT Thorax. The tumour consists of areas of haemorrhage and necrosis. Surgery remains the mainstay of treatment. But surprisingly recurrences occur within the first year post surgery or not at all.

4. Treatment

Because of the well localised nature of the tumour, surgery is the treatment of choice for pleuropulmonaryblastoma. The range of surgical excision should be determined individually and depending up on the tumour size, lymph node metastasis, pleural invasion and other comorbidities. The average survival rate among operated patients is 33 months, as compared to 2 months' survival in non-operated patients. The survival rate after partial lobectomy was found to be higher than compared to total lung resection. Retrospective analysis from a single center reports the longest pleuropulmonaryblastoma survival to be 9 years. Post operatively the Two year and five year survival rates were 85.7% and 71.4% respectively. Radiotherapy is used in unresectable tumours resistant to other forms of treatment and as an adjuvant therapy when the tumour has metastasized to the brain. Randomized controlled trials on the effectiveness of chemotherapy are lacking. Due to complex histological tumour structure, chemotherapy must be effective against both components. Response was only achieved with multidrug treatment including alkylating agents, antibiotics and mitotic inhibitors. One of the first therapies suggested in 1984 included cyclophosphamide, vincristine, doxorubicin and dactinomycin, therapy effective in histologically similar neoplasia to pulmonary blastoma: nephroblastoma and rhabdomyosarcoma. Partial response in a postsurgical pleuropulmonaryblastoma relapse was achieved using ifosfamide combined with doxorubicin and radiotherapy. The combination of cisplatin with etoposide has shown efficacy in cases of relapse and allowed reoperation. Treatment with an adjuvant platinum-based chemotherapy comprising of ifosfamide, carboplatin, and etoposide (ICE protocol) has shown effectiveness. Chemotherapy using etoposide and cisplatin have been reported to induce remission. He survived one year. Other regimens include carboplatin, paclitaxel and bevacizumab therapy. Other chemotherapeutic agents being used are Bevacizumab, an IgG monoclonal antibody against vascular endothelial growth factors and Sorafenib. The data regarding chemotherapy and radiotherapy is largely based on case reports in literature. Randomized clinical trials are

- The postoperative course was uneventful.
- The histopathological report was suggestive of **pleuro pulmonary blastoma type III**
- IHC markers revealed- **Vimentin positive**, SMA S - 100 negative and Desmin negative.
- The patient has currently undergone 5 cycles of chemotherapy with Vincristine, Ifosfamide, Etoposide, Adriamycin and Cyclophosphamide.

2. Review of Literature

- 1) Pleuropulmonaryblastoma(PPB) is a rare childhood cancer occurring in lungs and pleura. It is accountable for 0.25–0.5% of primary pulmonary malignancies.
- 2) A total of 350 cases have been reported in the pleuro pulmonary blastoma registry.
- 3) Three subtypes exist
 - a) Type 1 PPB – (birth to 2yrs) Multicystic, thin walled type.
 - b) Type 2 PPB – (> 2yrs of age) Mixed solid and cystic tumor.
 - c) Type 3 PPB – (2 to 8 yrs) mucoid solid tumor.
- 4) The prognosis is strongly correlated with the histological type.
- 5) Mutation is seen in DICER 1 gene

3. Discussion

Prognosis is very poor, with a mortality rate of 60% within 2 years of diagnosis, only 16% 5 year survival and 8% 10 year

needed to determine recommendations for the treatment regimen in pulmonary blastoma.

5. Conclusion

We have presented a rare case of pleuropulmonary blastoma, a rare lung cancer occurring at an earlier age and having poor prognosis as compared to other more common lung cancers. Recurrence after resection is high and regular surveillance is recommended especially within the first year. Our patient opted to undergo adjuvant chemotherapy and remains disease free at 2years.

Given the small number of cases and recent re-classification, interpreting the published epidemiology and clinical features of this disease is difficult. Many earlier reports may have included fetal adenocarcinomas (in particular high grade variant), which needs to be considered when discussing treatment and prognosis with newly diagnosed patients.