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CASE SERIES

Pleuropulmonary Blastoma - A Single Centre Experience

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Abstract

Background: Pleuropulmonary blastoma is a primary mesenchymal origin tumor of childhood with its origin in the thorax. It is considered a highly aggressive tumor because of its late diagnosis, potential for metastasis and recurrence and resistance to treatment. With very few series reported in literature and no standardized protocol, this tumor has become hard to treat. **Materials and Methods:** This series aims to study the cases that presented at our institute in terms of clinical presentation, work up, management and identify factors that can be associated with prognosis. It is a retrospective case series from a single centre over a period of 5 years. **Results:** Type II and III tumors were encountered in all the 4 children, who were all less than 3 years of age. Three children underwent neoadjuvant chemotherapy followed by surgical resection, two of whom also completed adjuvant chemotherapy and are on close follow up. The other child relapsed during follow up and the fourth child died during neoadjuvant chemotherapy. **Conclusion:** The prognosis of this rare tumor has direct association with degree of resection and completion of adjuvant chemotherapy. However, this could be statistically significant only if we could extrapolate it to a study with larger sample size.

Keywords: Pediatric Lung Tumor, Pleuropulmonary Blastoma, Plombage

1. Introduction

Pleuropulmonary blastoma is a highly aggressive and rare primary malignant intrathoracic tumor, with maximum incidence in the pediatric population. It can either arise from the lungs, from the pleura or both.

In children, the incidence of primary pulmonary tumors when compared to secondaries in lungs is of the ratio 1:5. The incidence of non- neoplastic lung lesions in children is however 60 times as common as a primary lung tumor[1]. Pleuropulmonary blastoma is noted to have an incidence of 0.5% of all pediatric malignant tumors. The ExPERT group calls this tumor to be a rare pediatric tumor, since its incidence is less than 1 in 2 million children in the world every year[2].

Histologically, the tumor has blastemal and sarcomatous components, hence making it a dysembryonic or dysontogenic neoplasm like other childhood tumors namely Wilm's tumor, neuroblastoma and hepatoblastoma which contain embryonic (blastemal) remnants[3].

The rarity of the tumor, its non- specific presentations, a high rate of misdiagnosis, poor prognosis and sparse literature adds to the challenge of treatment.

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2. Aim and Objectives

This series aimed to study:

- The presentation, workup, management and follow up of the cases diagnosed with pleuropulmonary blastoma.
- The factors which can affect outcome of this disease.

3. Review of Literature

This aggressive tumor presents with metastasis in 20% of cases. It has been described into three types based on their imaging appearance - Type I, purely cystic, Type II mixed- cystic and solid, Type III purely solid. Type I lesions may be mistaken for Congenital Pulmonary Airway Malformations, which have even reported to be precursors, synchronous and metachronous lesions[4]. Majority of them present before 4 years of age. The most common sites of metastasis are liver, brain and spinal cord and hence screening these parts is mandatory of primary evaluation. The characteristic clinical presentation is cough, respiratory distress, accompanied by fever; the child whose 'pneumonia' appears to be refractory to treatment. These cases are commonly misdiagnosed, the space occupying lesion mistaken for consolidation on chest x-rays. Computed Tomography of the chest reveals the lesion arising from the lung parenchyma or pleura or mediastinum, with few cases of extrapleural sequestrations also reported. Bronchoscopy and biopsy help clinch the diagnosis. Histopathology shows mesenchymal components along with blastemal elements and spindle cells. Multimodality therapy with chemotherapy, radiation therapy and surgery help survival against this tumor. The most common chemotherapeutic agents used are vincristine, actinomycin D, cyclophosphamide-based regimens with addition of doxorubicin and cisplatin[4].

4. Material and Methods

This is a retrospective case series over a period of 5 years from a single centre. There were 4 cases of pleuropulmonary blastoma, who presented to our Institute over the past 5 years (2019-2024). Data was collected from Medical Records section. We analysed their presentation, age, type of pleuropulmonary blastoma, imaging findings and management and arrived at the following results.

5. Results (Including Observations)

Our study of 4 cases had a mean age of presentation at 30 months, with equal male and female distribution and the universal presentation being cough with progressive respiratory distress (and no metastasis). The time period between onset of symptoms and initiation of chemotherapy, we felt, played a major role in prognosis. The first case underwent 9 cycles of IVADo chemotherapy (Ifosfamide/ Vincristine/ Actinomycin D and Doxorubicin) followed by left thoracotomy and left extrapleural pneumonectomy with intercostal drain (Figure 1). Child relapsed during his second year of follow up with left thoracic lesion and vertebral metastasis and succumbed. The second case was a right lower lobe mass, and was started on VAC (Vincristine/Actinomycin D/Cyclophosphamide); she didn't improve symptomatically even after 6 weeks of VAC, and hence stepped up to VDC/IE (Vincristine/Doxorubicin/Cyclophosphamide/Ifosfamide/ Etoposide), during which she succumbed to death. The third case was started on VAC, for which he showed tremendous response over 10 weeks. He underwent extra pleural left pneumonectomy followed by plombage of the empty left hemithorax with ping pong balls (Figure 2), to prevent the dreaded post pneumonectomy syndrome (cardiac herniation into empty left hemithorax can result in arrhythmias). Child tolerated surgery, plombage well, completed adjuvant chemotherapy. Two years follow up shows well accommodated ping pong balls within the thorax. The fourth case was started on IVADo 1 cycle, followed by Ifosfamide and Adriamycin for 3 cycles. She underwent a left thoracotomy and the pleural based tumor was excised in toto (Figure 3), parenchyma was normal. All the 3 operated case specimens proved histopathology of pleuropulmonary blastoma,

Table 1. Results of our series

	Case 1	Case 2	Case 3	Case 4
Age at diagnosis	36 months/ M	35 months/ F	30 months/ M	18 months/ F
Presentation	Respiratory distress	Respiratory Distress	Respiratory Distress	Respiratory Distress
Underlying lung Lesion	Nil	Nil	Nil	Nil
Initial imaging	Entire left hemithorax 12x8x7 cm mass, no mets	Right lower lobe 4x5x4 cm mass, no mets	Entire left hemithorax 12x10x9 cm, no Mets	Left hemithorax 9.5x9.5x8 cm mass, no mets
Type	Type II	Type III	Type II	Type III
Treatment	NACT + Surgery	NACT	NACT + Surgery + ACT	NACT + Surgery + ACT
Interval between symptoms and chemotherapy	30 days	45 days	15 days	35 days
Dicer 1 status	Not done	Not done	Negative	Negative
Current follow up	Relapse after 2 years - death	Death during chemotherapy	Alive – 2 years follow up, no relapse	Alive – 8 months follow up

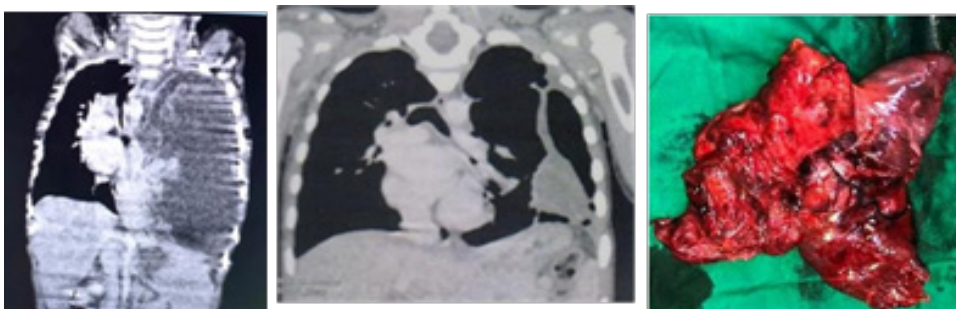


Figure 1. Case 1 - Tumor at presentation, tumor after NACT, pneumonectomy specimen.

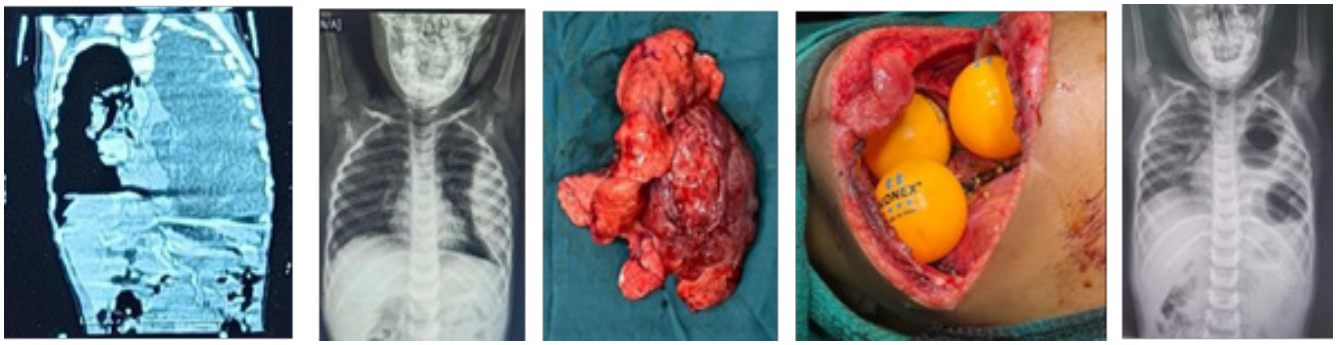


Figure 2. Case 3 - Tumor at presentation, tumor after NACT, pneumonectomy specimen, plombage, 2 years follow up Xray.

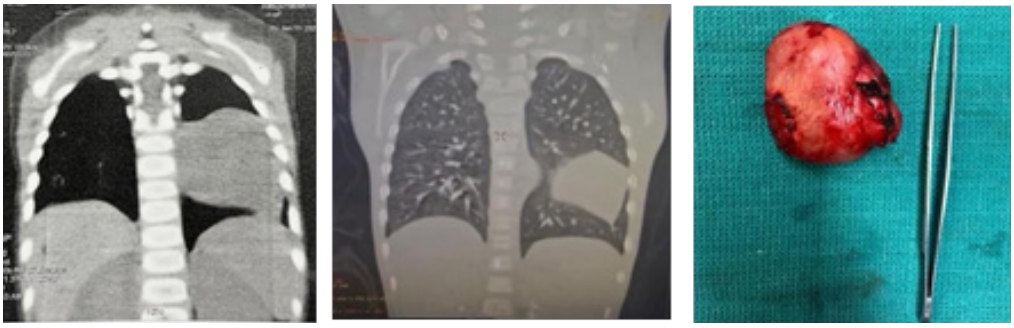


Figure 3. Case 4 - Tumor at presentation, tumor after NACT, tumor specimen.

confirmed by IHC markers vimentin and desmin positivity. Table 1 summarises our results.

6. Discussion

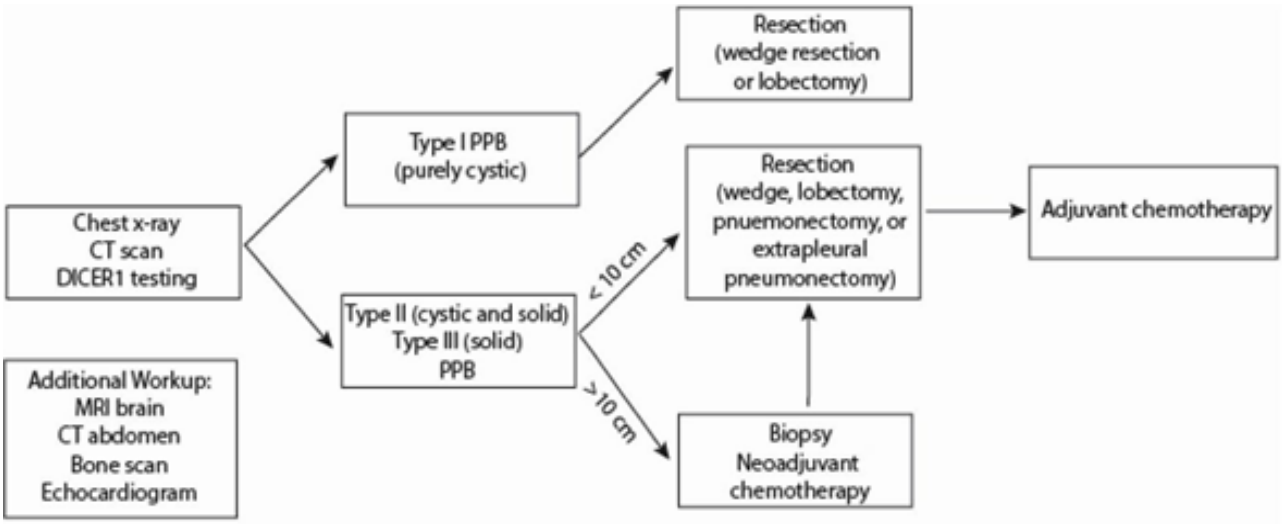
Prognosis depends on a number of factors and the type of lesion plays an important role. Type I lesions have favorable prognosis, Type II have 71% survival rate with 16% relapse rate and Type III have a 53% survival rate with 27% relapse rate[5].

Histopathologically, they show a mixed pattern of embryonal type rhabdomyosarcoma, nodules of chondrosarcoma, islands of primitive tumor cells with blastemal features, spindle cell foci resembling infantile fibrosarcoma and individual and groups of large, pleomorphic and anaplastic cells[6].

70% of children with this tumor express biallelic mutation in DICER1 with five specific hotspot mutations. DICER1 is a tumor suppressor gene encoding endoribonuclease - generating snrRNA. DICER1 positivity is also associated with other mesenchymal tumours such as Sertoli Leydig cell tumors, cystic nephroma, embryonal RMS, genitourinary sarcoma, differentiated thyroid cancers, pinealoblastoma and neuroendocrine tumors[7]. However, DICER1 mutations have not been associated with prognosis in pleuropulmonary blastoma.

Surgical measures to prevent post pneumonectomy syndromes include mediastinal repositioning, mediastinal fixation, thoracoplasty and muscle flap transposition. Plombage with silicone inserts have also been attempted, however plombage with ping

Table 2. Discussion: Algorithm by Knight *et al*



pong balls have not been reported for pleuropulmonary blastoma in pediatric age group[8].

Knight *et al*[9] gave the following algorithm for management of pleuropulmonary blastomas (Table 2).

7. Summary and Conclusion

From this series, we have drawn the following points of conclusion. The rarity of the tumor has contributed to the absence of a universal standardized protocol to adhere to. Locoregional control of the disease is directly associated with prognosis with additional chemotherapy adding on to long time survival. All cystic lung lesions should be addressed surgically in infancy so as to not to miss a type I pleuropulmonary blastoma.

8. References

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