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ABSTRACT

Introduction

Pleuro-Pulmonary Blastoma is a rare solid and or cystic progressive and aggressive malignant intra-thoracic tumour of childhood which such as Wilm’s tumour, Neuroblastoma and Hepato-blastoma belongs to a group of dysontogenic neoplasms. The incidence of pleuro-pulmonary Blastoma ranges between 0.25 and 0.5% of primary lung tumours making this entity a rare primary malignant lung tumour.

Case Report

A 2 month old male patient was referred from a regional hospital with a 3 days history of cough, fever and shortness of breath. No previous medical history reported. The patient was delivered at term through a normal vaginal delivery which was uneventful. Birth weight=3 kg, length=48 cm, Apgar score=10/10 and the head circumference was 35cm. The patient’s retro-viral disease status was negative. On percussion there was dullness and bilateral crepitations. There was bilateral decreased air-entry. No signs of cardiac failure. The rest of the systems were within normal limits. The arterial blood gases showed respiratory acidosis with hypoxia. Chest x-ray showed a white out of the right hemithorax and CT scan of the chest showed a large soft tissue mass in the right hemithorax associated with free fluid in the right hemithorax and the biopsy was performed which confirmed the diagnosis of a Pleuro-pulmonary blastoma. The Pleuro-pulmonary blastoma was a solid type (Type 3).

Conclusion

Pleuro-pulmonary blastoma poses a diagnostic challenge because it does not have specific clinical or radiological features. The Late diagnosis and delayed treatment of pleuro-pulmonary Blastoma are associated with a deadly prognosis.
Keywords: Pleuro-pulmonary blastoma, Types, Computed Tomography, Biopsy
INTRODUCTION

The incidence of pleuro-pulmonary Blastoma ranges between 0.25 and 0.5% of primary lung tumours making this entity a rare primary malignant lung tumour [1]. Pleuro-pulmonary blastomas have been described from the peri-natal period until adulthood and symptoms are generally non-specific and include fever, chest or abdominal pain, dry cough, persistent infection despite medical therapy, shortness of breath and pneumothorax [1]. The differential diagnosis of congenital pulmonary adenomatoid malformation poses a diagnostic challenge and also a definitive diagnosis of pleuro-pulmonary Blastoma cannot be secured only based on clinical and Radiological investigations [2].

CASE REPORT

A 2 month old male patient was referred from a regional hospital with a 3 days history of cough, fever and shortness of breath. No previous medical history reported. The patient was delivered at term through a normal vaginal delivery which was uneventful. Birth weight=3kg, head circumference = 35 cm, birth length= 48 cm, Apgar score=10/10. The patient’s retro-viral disease status was negative. On examination: Normal growth and development. On admission the body weight was 4.5kg, head circumference was 38cm and the length was 57 cm. No pallor and no cyanosis. The patient was in severe respiratory distress with use of the accessory muscles (respiratory rate=22 b/minute). The patient was pyrexial (temperature=38 degrees Celsius). On percussion there was dullness and bilateral crepitations. There was bilateral decreased air-entry. No signs of cardiac failure. The rest of the systems were within normal limits. Chest x-ray was performed and it showed a dense opacification of the right hemithorax. The Computed Tomography (CT) scan of the chest was performed following the chest x-ray findings for further characterization of the chest x-ray findings. CT scan of the chest showed a large right hemithorax mass associated with free fluid in the right hemithorax.
The patient was intubated and then admitted in Paediatric Intensive care unit. He was ventilated for 9 days and later discharged to the Paediatric Pulmonology ward. Ultrasound guided biopsy was performed when the patient’s condition had stabilized. The biopsy results were as follows:

Microscopy: Histological examination of section received shows sheets of oval to spindle shaped cells forming fascicles. The cells lack an epithelial component (Figure 3,4).

Immuno-Histochemistry: In the presence of an adequate control: CD 99 is diffusely positive and stains with moderate intensity. BCL-2 is also diffusely positive and stains with a strong intensity. Both cytokeratin stains, CK5/6 and MNF116 are negative. CD 34 is negative. The above mentioned features are those of a Pleuro-pulmonary Blastoma.

The Pleuro-pulmonary blastoma was a solid type (Type 3). Following the histo-pathology findings the patient was then referred to the Paediatric Surgical department for further management, however; the patient’s condition deteriorated and he demised before the Paediatric surgical intervention could be performed.

**DISCUSSION**

Pleuro-Pulmonary Blastoma is a rare solid and or cystic progressive and aggressive malignant intra-thoracic tumour of childhood which such as Wilm’s tumour, Neuroblastoma and Hepato-blastoma belongs to a group of dysontogenic neoplasms [1, 2]. Congenital pulmonary adenomatoid malformation is a rare developmental congenital abnormality of the lower respiratory tract whose aetiology is unknown [9]. The incidence of pleuro-pulmonary Blastoma ranges between 0.25 and 0.5% of primary lung tumours making this entity a rare primary malignant lung tumour [1]. Barnard was the first person to describe pleuro-pulmonary blastomas in year 1952 which he named them as Embryomas and the tumours were re-defined by Spencer and Mavivel in 1961 and 1988 respectively and since then only 150 cases have been reported in the English literature as of year 2010 [1]. Pleuro-pulmonary blastomas have been described from the peri-natal period until adulthood and symptoms are generally non-specific and include fever, chest or
abdominal pain, dry cough, persistent infection despite medical therapy, shortness of
breath and pneumothorax [1].

The differential diagnosis of congenital pulmonary adenomatoid malformation poses
a diagnostic challenge and also a definitive diagnosis of pleuro-pulmonary Blastoma
cannot be secured only based on clinical and Radiological investigations [2].

Late diagnosis and delayed treatment of pleuro-pulmonary Blastoma are associated
with a deadly prognosis [2].

The classification of pleuro-pulmonary Blastoma according to histological features by
Priest et al is as follows [2]

1. Type 1 (Pure cystic)
2. Type 2 (Cystic and solid)
3. Type 3 (Purely solid)

The recurrence rate of type 1 pleuro-pulmonary Blastoma is lower compared to type
2 and type 3 PPB and there is no metastatic focus associated with type 1 [2].

Progression of pleuro-pulmonary Blastoma from type 1 to type 3 is speculated as a
natural history of PPB and this tumour can occur in the lungs, pleura, mediastinum
and the diaphragm [3, 4].

According to Priest et al a 5 year survival rate for type 1 PPB is 83% while for type 2
and type 3 the survival rate is at 42% [4].

Association of a pleuro-pulmonary Blastoma with the familial cancer syndrome raises
suspicion that the PPB may have a genetic basis [5].

Type 2 and type 3 PPB metastasize to the brain, liver and the bone in 30% of cases
while type 1 PPB rarely metastasizes [6].

The histological characteristics of PPB are that of blastomatous component that is
variable associated with sarcomatous areas that have rhabdomyoblastic or
chondroblastic differentiation [7].

The ultimate treatment for children with pleuro-pulmonary Blastoma is complete
surgical resection with clear margins usually requiring a lobectomy [6, 8].
CONCLUSION

The diagnosis of a Pleuro-pulmonary Blastoma needs a multi-disciplinary approach which involves the Paediatricians, Radiologists and the Histo-pathologists. The ultimate diagnosis is secured through biopsy and the ultimate treatment is surgical.

CONFLICT OF INTEREST

Authors declare no conflict of interest

AUTHOR’S CONTRIBUTIONS

Luvo Gaxa

Group 1- Substantial contributions to conception and design, acquisition of data, analysis and interpretation of data,

Group 2- Drafting the article, revising it critically for important intellectual content,

Group 3- Final approval of the version to be published

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Group 1- Substantial contributions to conception and design,

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REFERENCES


FIGURE LEGENDS

Figure 1: (a b) Frontal and lateral views of the chest radiograph show a diffuse radio-dense opacity of the right hemi-thorax with mediastinal deviation to the left side
Figure 2: (a, b & c) Axial, Coronal and sagittal views of the chest CT scan confirm a large soft tissue mass in the right hemi-thorax.

Figure 3: Figure 1: H&E section of a tumor, x20 magnification: showing sheets of small round blue cells.

Figure 4: Figure 1: H&E section of a tumor, x40: showing darkly staining nuclei.

FIGURES

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