

# Solid (Type 3) pleuropulmonary blastoma

Luvo Gaxa, Bafana Hlatshwayo, Aletta Lefentse Motene

## ABSTRACT

**Introduction:** Pleuropulmonary blastoma is a rare solid and/or cystic progressive and aggressive malignant intra-thoracic tumor of childhood which belongs to a group of dysontogenetic neoplasms such as Wilms tumor, neuroblastoma and hepatoblastoma. The incidence of pleuropulmonary blastoma ranges between 0.25 and 0.5% of primary lung tumors making this entity a rare primary malignant lung tumor. **Case Report:** A two-month-old male neonate was referred from a regional hospital with a three-day 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 35 cm. 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 pleuropulmonary blastoma. The pleuropulmonary blastoma was a solid type (Type 3). **Conclusion:** Pleuropulmonary blastoma poses a diagnostic challenge because it does not have specific clinical or radiological features. The late diagnosis and delayed treatment of pleuropulmonary blastoma are associated with a deadly prognosis.

**Keywords:** Biopsy, Pleuropulmonary blastoma, Solid, Type 3, Types

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## INTRODUCTION

The incidence of pleuropulmonary blastoma ranges between 0.25 and 0.5% of primary lung tumors making this entity a rare primary malignant lung tumor [1]. Pleuropulmonary blastomas have been described from the perinatal 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 pleuropulmonary blastoma cannot be secured only based on clinical and radiological investigations [2].

## CASE REPORT

A two-month-old male neonate was referred from a regional hospital with a three-day 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, head circumference 35 cm, birth length 48 cm, Apgar score 10/10. The patient's retro-viral disease status was negative.

On examination the neonate has normal growth and development. On admission the body weight was 4.5 kg, head circumference was 38 cm and the length was 57 cm. There were no pallor and no cyanosis. The patient was in severe respiratory distress with use of the accessory muscles (respiratory rate 22 beats/minute). The patient was pyrexial (temperature 38°C).

On percussion there was dullness and bilateral crepitations. There was bilateral decreased air-entry and no signs of cardiac failure. The rest of the examination were within normal limits. Chest X-ray showed a dense opacification of the right hemithorax (Figure 1).

The computed tomography scan of the chest was performed following the chest X-ray findings for further characterization of the chest X-ray findings. Computed tomography scan of the chest showed a large right hemithorax mass associated with free fluid in the right hemithorax (Figure 2).

The patient was intubated and then admitted in pediatric intensive care unit. He was ventilated for nine days and later discharged to the pediatric pulmonology ward. Ultrasound guided biopsy was performed when the patient's condition had stabilized.

The biopsy results were as follows:

**Microscopy:** Histological examination showed sheets of oval to spindle shaped cells forming fascicles. The cells lack an epithelial component (Figures 3 and 4).

**Immunohistochemistry:** 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 pleuropulmonary blastoma. The pleuropulmonary blastoma was a solid type (Type 3). Following the histopathology findings the patient was then referred to the pediatric surgical department for further management. However; the patient's condition deteriorated and he demised before the pediatric surgical intervention.

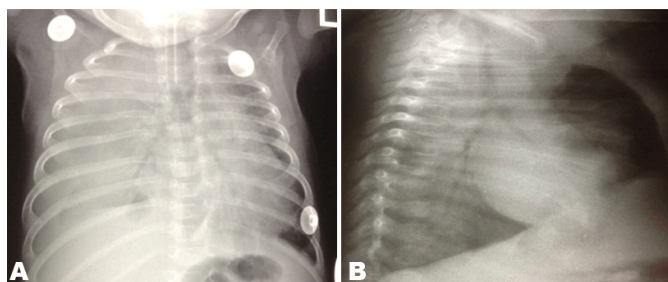


Figure 1: (A, B) Frontal and lateral views of the chest radiograph showing a diffuse radiodense opacity of the right hemithorax with mediastinal deviation to the left side.

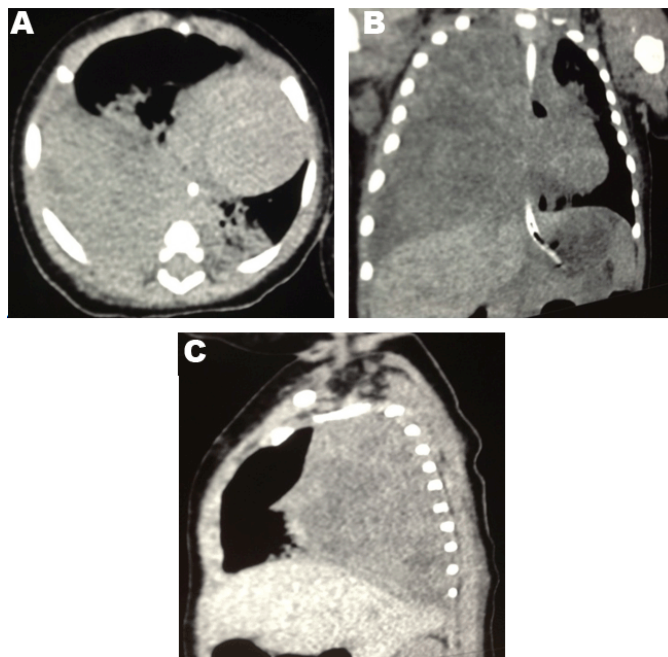


Figure 2: (A–C) Axial, coronal and sagittal views of the chest computed tomography scan confirm a large soft tissue mass in the right hemithorax.

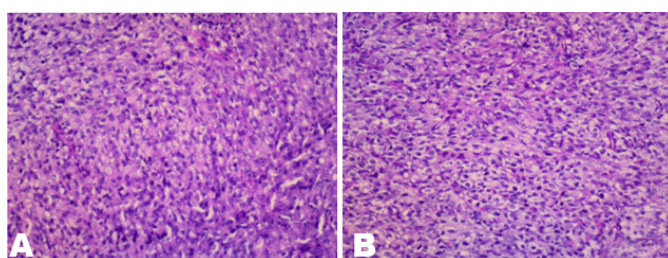


Figure 3: Sheets of small round blue cells (H&E stain, x200).

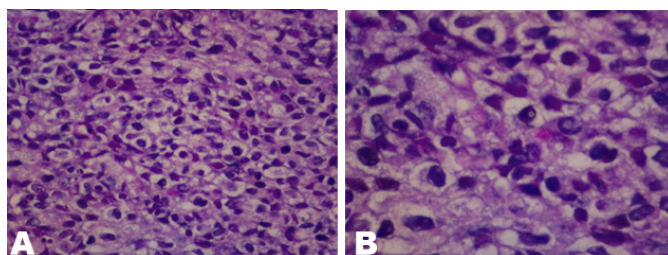


Figure 4: Darkly staining nuclei (H&E stain, x400).

## DISCUSSION

Pleuropulmonary blastoma is a rare solid and or cystic progressive and aggressive malignant intrathoracic tumor of childhood which such as Wilms tumor, neuroblastoma and hepatoblastoma belongs to a group of dysontogenetic neoplasms [1, 2]. Congenital pulmonary adenomatoid malformation is a rare developmental congenital abnormality of the lower respiratory tract whose etiology is unknown [3].

The incidence of pleuropulmonary blastoma ranges between 0.25 and 0.5% of primary lung tumors making this entity a rare primary malignant lung tumor [1].

Barnard was the first person to describe pleuropulmonary blastomas in 1952. He named them embryomas and the tumors were re-defined by Spencer and Mavivel in 1961 and 1988 respectively. Since then only 150 cases have been reported in the English literature until 2010 [1].

Pleuropulmonary blastomas have been described from the perinatal 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 pleuropulmonary blastoma cannot be secured only based on clinical and radiological investigations [2].

Late diagnosis and delayed treatment of pleuropulmonary blastoma are associated with a deadly prognosis [2].

The classification of pleuropulmonary 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 pleuropulmonary blastoma is lower compared to type 2 and type 3 PPB and there is no metastatic focus associated with type 1 [2]. Progression of pleuropulmonary blastoma from type 1 to type 3 is speculated as a natural history of PPB and this tumor can occur in the lungs, pleura, mediastinum and the diaphragm [4, 5].

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

Association of a pleuropulmonary blastoma with the familial cancer syndrome raises suspicion that the PPB may have a genetic basis [6].

Type 2 and type 3 PPB metastasize to the brain, liver and the bone in 30% of cases while type 1 PPB rarely metastasizes [7].

The histological characteristics of PPB are that of blastomatous component that is variable associated with sarcomatous areas that have rhabdomyoblastic or chondroblastic differentiation [8].

The ultimate treatment for children with pleuropulmonary blastoma is complete surgical resection with clear margins usually requiring a lobectomy [7, 9].

## CONCLUSION

The diagnosis of a pleuropulmonary blastoma needs a multidisciplinary approach which involves the pediatricians, radiologists and the histopathologists. The ultimate diagnosis is secured through biopsy and the ultimate treatment is surgical.

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## Author Contributions

Luvo Gaxa – Substantial contributions to conception and design, Acquisition of data, analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Bafana Elliot Hlatshwayo – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Aletta Lefentse Motene – Substantial contributions to conception and design, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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## REFERENCES

1. Chadli-Debbiche A, Brahim EB, Jouini R, et al. Pleuropulmonary blastoma: Cystic tumor misdiagnosed as an emphysematous bulla. *Respiratory Medicine CME* 2010;3(2):120–2.
2. Higashidate N, Asagiri KS, Fukahori S, et al. An infant with pleuropulmonary blastoma type 2 detected

- during the prenatal period. *J Ped Surg Case Reports* 2014;2(5):264–7.
3. Zhang ZJ, Huang MX. Children with congenital cystic adenomatoid malformation of the lung CT diagnosis. *Int J Clin Exp Med* 2015 Mar 15;8(3):4415–9.
  4. Al-Backer N, Puligandla PS, Su W, Anselmo M, Laberge JM. Type 1 pleuropulmonary blastoma in a 3-year-old male with a cystic lung mass. *J Pediatr Surg* 2006 Nov;41(11):e13–5.
  5. Libretti L, Ciriaco P, Casiraghi M, Arrigoni G, Zannini P. Pleuropulmonary blastoma in the area of a diagnosed congenital lung cyst. *Ann Thorac Surg* 2008 Feb;85(2):658–60.
  6. Boman F, Hill DA, Williams GM, et al. Familial association of pleuropulmonary blastoma with cystic nephroma and other renal tumors: A report from the International Pleuropulmonary Blastoma Registry. *J Pediatr* 2006 Dec;149(6):850–4.
  7. Miniati DN, Chintagumpala M, Langston C, et al. Prenatal presentation and outcome of children with pleuropulmonary blastoma. *J Pediatr Surg* 2006 Jan;41(1):66–71.
  8. Picaud JC, Levrey H, Bouvier R, et al. Bilateral cystic pleuropulmonary blastoma in early infancy. *J Pediatr* 2000 Jun;136(6):834–6.
  9. Simon R, McIntyre E, Khandhar P, Macknisd J, Fischere AC. Pleuropulmonary blastoma (PPB) in an infant: Is the timing of an elective resection of neonatal lung lesions challenged?. *J Ped Surg Case Reports* 2014;2(10):465–7.

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