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## Case report

# Pleuropulmonary blastoma (type III) in a two-year-old: A case report <sup>☆,☆☆</sup>

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## ABSTRACT

Pleuropulmonary blastoma is a rare, aggressive, malignant tumor of the lungs or pleura that primarily affects children. Pleuropulmonary blastoma is classified into 3 types based on morphology, including cystic (type I), mixed (type II), or solid (type III). These morphological types correlate with prognosis. In this article, we present a case of type III pleuropulmonary blastoma in a 2-year-old girl. The patient was treated with tumoral resection and chemotherapy; however, she experienced local recurrence and spinal metastasis after 5 months of treatment.

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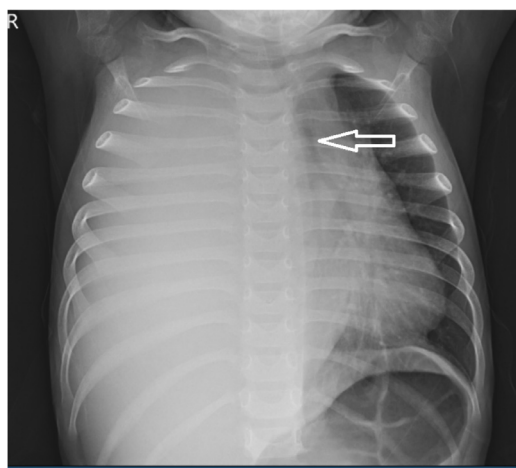
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## Introduction

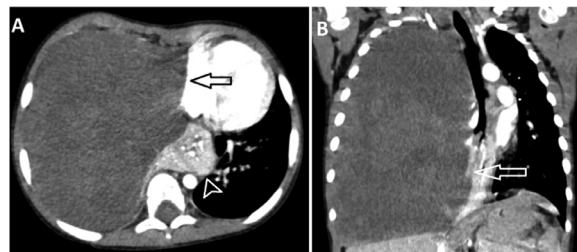
Pleuropulmonary blastoma (PPB) is a malignant mesenchymal tumor of the lungs and pleura that tends to occur in children before 6 years of age [1,2]. PPB was first reported by Manivel et al. [3] in 1988. Patients with mutations in *DICER1* have an increased risk of developing PPB [4]. Children with PPB often present with non-specific symptoms, and the primary treatment approaches include surgery and chemotherapy [5]. Here, we report a case of a large PPB in a child with spinal metastasis.

## Case report

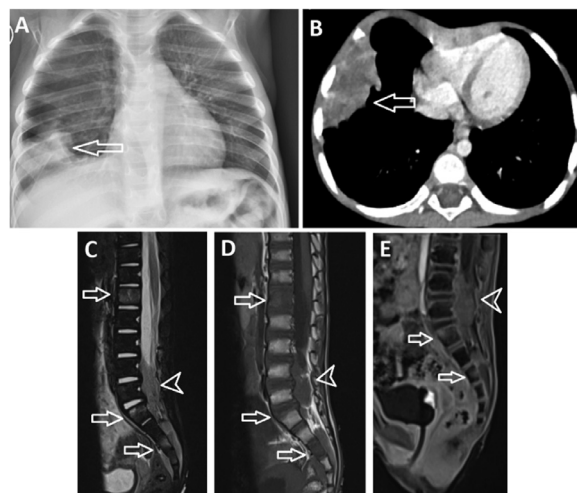
A 2-year-old girl presented to the hospital with increasing breathlessness for 1 month. On admission, she had a temperature of 37°C, a respiratory rate of 60 breaths/min, and oxygen saturation of 88% on room air. No focal neurological deficit was noted. Breath sounds were absent in the right lung. Radiography demonstrated the complete opacification of the right hemithorax, the trachea, and the left and right main bronchi, and the mediastinum was shifted to the left (Fig. 1). A chest computed tomography (CT) scan revealed a large, heterogeneous mass (measuring 10 × 12 × 14 cm), pushing the heart to the left. No costal erosion was identified (Fig. 2). Spine and brain magnetic resonance imaging (MRI) appeared normal. The patient underwent tumor biopsy, and the histopathology result suggested a PPB. The patient underwent tumoral resection, and the histological result confirmed a solid PPB. The patient received adjuvant chemotherapy with vincristine, ifosfamide, etoposide, actinomycin-D, and cyclophosphamide. However, 5 months later, the patient appeared paraplegia. Chest X-ray showed opacity on the right lung, and CT scans revealed a heterogeneous enhancing mass in the periphery of the right lung (Fig. 3). Spinal MRI showed multiple vertebral



**Fig. 1** – Chest X-ray showed complete opacification of the right hemithorax and the trachea, and the mediastinum was shifted to the left (arrow).



**Fig. 2** – Computed tomography imaging revealed a large heterogeneous mass (A and B, arrows), causing the heart to shift to the left side and the complete collapse of the entire right lung (A, arrowhead). No costal erosion was identified.



**Fig. 3** – Imaging after 5 mo of chemotherapy. Chest X-ray showed opacity on the right lung (A, arrow). Chest computed tomography imaging showed a heterogeneous lesion on the right lung (B, arrow). Spinal MRI showed multiple lesions in the vertebral bodies that were hyperintense on T2-weighted imaging (C), hypointense on T1-weighted imaging (D), and showed enhancement post-contrast (E, arrows). Lesions were also detected in the spinal canal (C, D, and E, arrowheads).

body lesions, which extended into the spinal canal (Fig. 3). Her parents declined treatment after that.

## Discussion

PPB is a malignant neoplasm that accounts for fewer than 1% of all primary pulmonary malignant tumors in children [2]. Most type I PPB patients are diagnosed during their first year of life (62%); however, type II and type III cases tend to be detected later but under the age of 3 years [4]. Type I PPBs have a slight male predominance; however, an equal male-to-female ratio has been reported for types II and III [4,6].

Type I PPBs appear as multiloculated air-containing cysts and often appearing unilateral, unifocal, peripheral, and larger

than 5 cm [6]. Type II PPBs contain both solid and cystic components, whereas type III PPBs present as a solid mass without cystic spaces [6].

Breathlessness is the most common symptom of PPB, and others include pulmonary infection, cough, and abdominal pain [7]. Pneumothorax is present in up to 30% of type I cases [4]. Imaging features are variable, depending on the tumor type. Type I PPB presents as multiloculated, air-filled lung cysts with thin septa often located in the periphery of the lung [6,7]. However, type I PPB can be difficult to differentiate from congenital pulmonary airway malformations. Type II PPB presents as a solid-cystic tumor, and type III PPB presents as a solid tumor that may invade the adjacent organs [7]. The patient in this report also presented with breathlessness, and the tumor was quite large. On imaging, the tumor presented as a solid mass suggesting type III PPB, and these results were confirmed histologically after surgery.

Type I PPB can be managed with complete resection; in cases of intraoperative tumor spill, incomplete resection, or local invasion, type I PPB is often treated with adjuvant chemotherapy [6]. For type II and III PPBs, surgical resection and chemotherapy are both required [6]. Chemotherapy includes both neoadjuvant chemotherapy and adjuvant chemotherapy. Neoadjuvant chemotherapy is typically performed in an attempt to reduce the tumor size before surgery [1].

PPB can metastasize, and the brain is the most common target organ [8]. Type I PPBs have a 5-year overall survival of 91% [4]. Type II and III PPBs are more aggressive than type I with 2-year and 5-year overall survival rates are 62% and 42%, respectively, even after multimodal therapy [2]. In this case, the patient had type III PPB and received surgery and adjuvant chemotherapy; however, she experienced local recurrence and spinal metastasis after 5 months of treatment.

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## Conclusion

PPB is an uncommon malignant neoplasm of the respiratory system that typically occurs in children. PPB can be divided into 3 types based on morphology, of which types II and III have a poor prognosis. PPB can widely metastasize. Imaging modalities, such as CT and MRI, can suggest PPB diagnosis and assess the extent of the tumor.

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## Patient consent

Informed consent for patient information to be published in this article was obtained.

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## Ethical statement

Appropriate written informed consent was obtained for the publication of this case report and accompanying images.

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## Author contributions

Le AD, Le TV, Tran H, and Nguyen MD contributed equally to this article as co-first authors. All authors have read the manuscript and agree to the contents.

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