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# RADIOLOGIC DIAGNOSIS OF A TYPE-III PLEUROPULMONARY BLASTOMA

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**Pleuropulmonary blastoma (PPB) is a rare and aggressive dysontogenic neoplasm, occurring in children under the age of 6 years in most cases. CT and MRI findings are well-known, a mixed solid and cystic lesion with variable contrast enhancement and a necrotic centre. We report the radiologic features of type III PPB case.**

**Key-words: Infants, respiratory system – Blastoma.**

Primary pulmonary neoplasms account for 0.27% of all cases of pediatric cancer and pleuropulmonary blastoma (PPB) is the most frequent type (1, 2), and most cases (94%) present in children younger than 6 years. PPB is a dysontogenic tumor and composed of immature malignant epithelial and/or mesenchymal tissues whose features may resemble early embryological lung tissues. There are 3 types of PPB: Type I: predominantly cystic; type II: cystic and solid; type III: purely solid. Malignancy risk increase from type I to type III (3). As the prognosis depends closely to the type of the tumor, recognizing this neoplasm earlier is vital. Clinically, majority of the patients present with non-productive cough and fever. Cases with predominant pleural involvement often present with pleuritis and empyema refractory to medical treatment. PPB should be suspected when symptoms do not respond to proper treatment (2). Radiologic findings of a patient diagnosed with type III PPB is reported here.

## Case report

A 21-month-old boy who has an unremarkable personal medical history was brought to the hospital with fever and tachypnea. On arrival his breathing frequency was 48 breaths/min, heart rate 132 beats/min, oxygen saturation with pulse oxymeter 99%, and body temperature 38.2°C. His breath sounds were diminished at the middle and lower zone of the right chest with intercostal retractions and his liver was palpable 3 cm at the costal margin. The white-blood-cell count was 17.5 $\times$ $10^3$ cells/μL, with 50% neutrophils, the hemoglobin level was 11.1 g/dL, and the platelet count was 509 $\times$ $10^3$ cells/μL. His serum C-reactive protein (CRP) level was 5.8 mg/dL (normal, 0-0.5 mg/dL), but his serum electrolyte levels and renal and liver function tests were normal. On chest X-ray there was a large homogeneous soft tissue opacity within the the right lung (Fig. 1). He was hospitalized with diagnosis of nosocomial pneumonia and put on intravenous sulbactam-ampicillin and clarythromycin treatment. Thoracic ultrasonography (US) was done to confirm the presence of pleural effusion and a large consolidation with odd lobulation was observed associated with a small pleural fluid. Examination of pleural effusion obtained with thoracentesis revealed exudative characteristics. After switching the antibiotics to meropenem and vancomycin, and insertion of chest tube to the right pleural space all his clinical and laboratory findings improved except chest X-ray. On computerized tomography (CT) (Fig. 2) of chest a

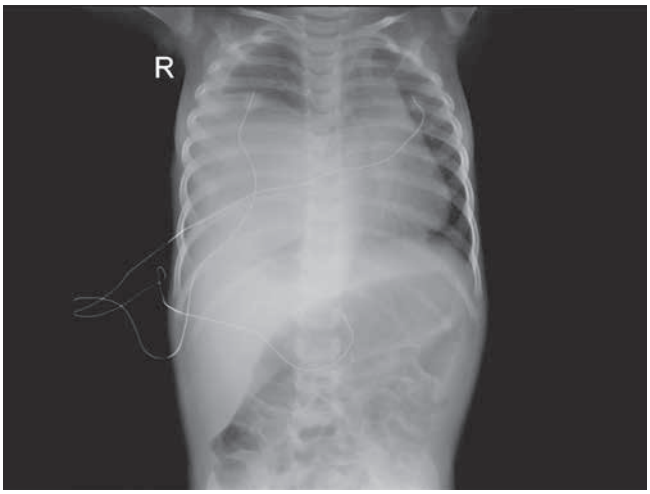


*Fig. 1.* — Chest X-ray. Opacification of the right hemitorax with pleural effusion is noted.

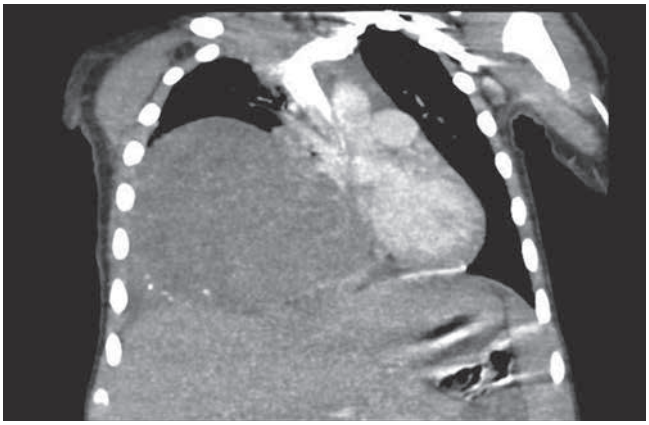

*Fig. 2.* — Contrast enhanced CT. A huge, homogenous, hypodense and round mass, without enhancement is seen. Note small peripheral calcifications.

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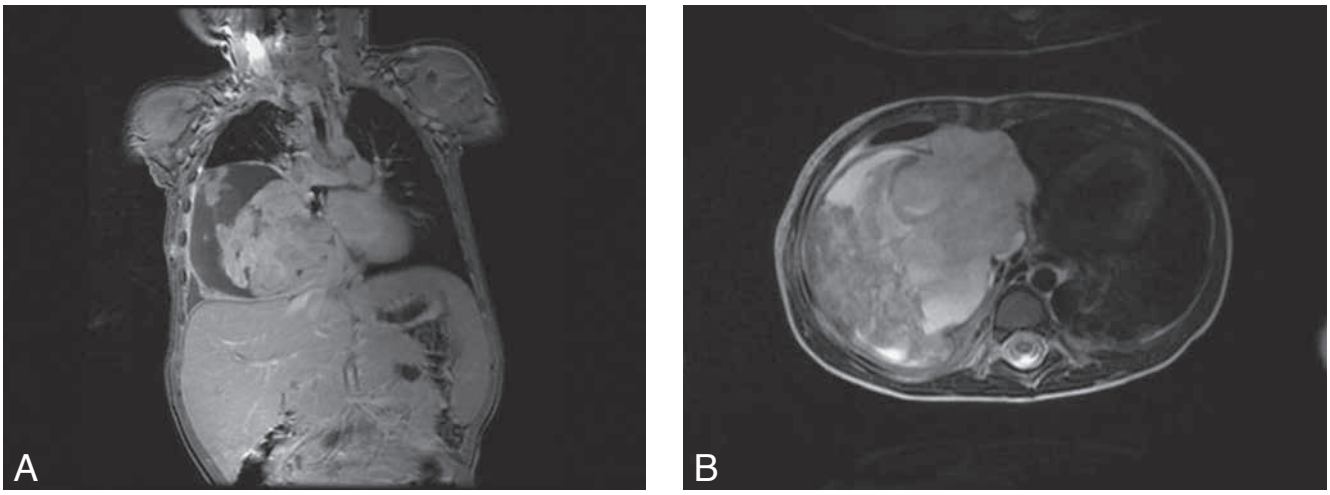


*Fig. 3.* — T2-weighted axial (A) and postcontrast fat saturated T1-weighted coronal (B) scans. Heterogeneous solid mass is seen, nonenhancing haemorrhagic areas of the tumour is noted on postcontrast scan. Broad pleural base of the tumour is distinct with small entrapped pleural effusion. The diaphragm, the heart and the hilar bronchopulmonary structures were displaced. There is no evidence of rib or soft tissue invasion.

huge pleura broad based, hypodense homogeneous round tumor measuring 10 × 9 × 8 cm in size in the right hemithorax was seen. The tumor was located predominantly on costal and diaphragmatic pleura causing inferior displacement of the right hemidiaphragma; extending medially causing displacement of the broncho-vascular structures. The heart also was shifted leftwards. There was not contrast enhancement of the tumor; only a thin focal peripheral and a few internal calcification were observed. Small entrapped pleural fluid next to the tumor was seen. The middle lobe was collapsed and the lower lobe was underdeveloped; whereas upper lobe was overinflated causing mediastinal shift; some ground glass opacity were observed within the upper lobe. Left lung parenchyma was normal. The mass was very heterogeneous on MRI scans (Fig. 3) the periphery of the tumor was in crescentic configuration without contrast enhancement suggesting haemorrhage, whereas the medial part of the tumor was also heterogeneous with avid enhancement and diffusion restriction. Pleural broad based location of the tumor suggested pleura; the diaphragm, the heart and the hilar broncho-pulmonary structures were displaced, however mediastinal invasion was not suggested. There was not any rib or soft tissue invasion, and mediastinum and spine were normal. A few mediastinal small lymph nodes were observed.

Distant metastasis was excluded with cranial and abdominal MRI, bone scintigraphy, and bone marrow smear and biopsy. Right lower lobectomy with negative surgical margins was performed.

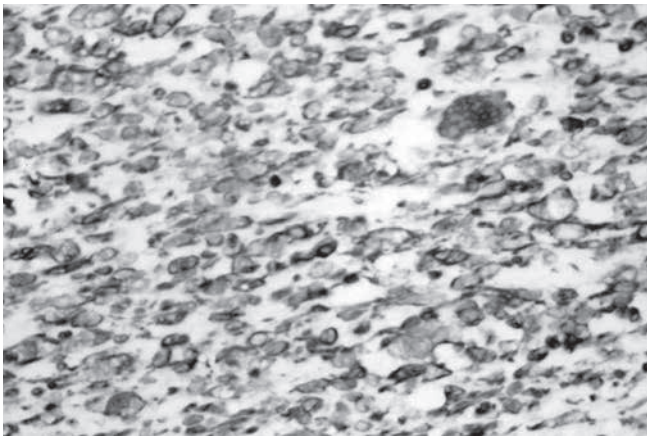

*Fig. 4.* — Histopathologic appearance. Strong cytoplasmic vimentin positivity (× 200).

In macroscopic examination, a hard, mostly solid, but in some areas cystic seeming whitish yellow tumoral mass, measuring 7.5 × 7 × 7 cm in diameters, with well demarcated but irregular margins and lobulated contours was observed in right caudal lung lobe. Immunohistochemistry was performed on sections of the mass for the detection of Pancytokeratin (clone AE1/AE3:20/1), Vimentin (clone SRL33), Desmin (clone DE-R-11), CD99 (clone HO36-1.1), Epithelial membrane antigen (EMA, clone GP1.4), Carcinoembryonic antigen (CEA, clone II-7), Smooth muscle actin (SMA, clone asm-1), Thyroid transcription factor (TTF1, clone SPT24). Immunohistochemically, the cells were vimentin, desmin and CD99 positive (Fig. 4). Final diagnosis was pleuropulmonary blastoma (PPB) type III (solid).

## Discussion

CT and MRI features of the tumor were highly suggestive for PPB. Histopathology of the tumor was compatible with the radiologic diagnosis. In our case, CT revealed a very homogeneous and hypo dense round mass with thin peripheral and internal calcifications. On MRI the internal texture of the tumor was very heterogeneous and there was a large haemorrhage and solid enhancing areas with diffusion restriction. It was

difficult to determine the origin of such a large tumor, but pleural broad base location pointed out the pleural origin. In spite of the large size of the tumor, there was not any invasion or infiltration of the adjacent structures. Sparing of the ribs and thoracic wall excluded a primitive neuroectodermal tumor or a sarcoma. Underdevelopment of the lower lobe and over inflation of the upper lobe gave rise to thought that the tumor has a fetal origin. Finally, according to the clinical and radiological findings the pre-diagnosis was a PPB. Absence of internal air-filled cysts, large solid component with diffusion restriction and haemorrhage of the tumor suggested type III PPB (4-6]. The tumor was in the right hemithorax such as most of the reported cases (3).

Although the tumor was misinterpreted sonographically, US could be preferred before CT or MRI scan because of it's low cost, lack of radiation, contrast injection or sedation. when both pulmonary and pleural lesions are present, distinction between these two lesions is not always easy at chest radiographies, US enables differentiation of pleural pathologies from pulmonary parenchymal lesions (4).

Prognosis of type I PPB is very good (> 90% cure), whereas type II and III PPB are not so good (approximately 50%). Its invasiveness is determined by the high rate of local recurrence and distant metastasis. In the largest series of PPB published, 5-year survival rates were 83% for type I PPB and 42% for type II and type III PPB (3). Pleural and mediastinal involvement at diagnosis are indicative of a poor prognosis, as does extra-pleural involvement (7, 8). The most frequent sites of metastasis are the central nervous system, bone, bone marrow and the liver, and metastasis occurs in aggressive forms of PPB: Types II and III PPB. Cerebral metastases occur in 11% of type II and 54% of type III patients and may appear when thoracic disease is under control (3).

PPB is a unique childhood malignancy that is often misdiagnosed. As the prognosis depends closely to the type of the tumor, recognizing this neoplasm earlier is vital. When a large pleural-based mass is identified in a young child, PPB should be considered. To the best of our knowledge, this is the first report about the sonographic pattern of PPB. Sonographic 'wave' sign of a large consolidation without sonographic air bronchogram finding could also be a clue for PPB cases. It's detection should alert the radiologist of the probability that it could be a huge tumor.

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