**CASE REPORT**

**Pleuropulmonary blastoma, a rare entity in childhood**

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**INTRODUCTION**

Pleuropulmonary blastoma (PPB) accounts for 0.25 to 0.5% of all primary lung neoplasms. They are malignant tumours that can be pleural, pulmonary or both. Despite being rare, they are considered the most common primary cancers in the lung during childhood, especially in those who are under 5 years of age. The first two years of age remain the highest risk for developing PPB, with about 90% of the cases occurring within this age group. PPB is divided into type I, II & III according to histological type of the tumour. Each of the subtypes correlates to the age at diagnosis and the overall prognosis of the patient. The main aim of reporting this case is to highlight the importance of both diagnostic and interventional radiology in helping other physicians to reach rare diagnosis correctly.

**CASE PRESENTATION**

A 3-year-old male child presented to our emergency department with a history of persistent dry cough of 4 days duration. He had a history of hospitalisation once for acute bronchiolitis at the age of 1 year. His family history was unremarkable. Upon clinical evaluation, he had tachypnea, intercostal retractions, absent air entry on the right lung and abdominal distention. Otherwise, the clinical examination was normal. Laboratory investigations revealed leukocytosis up to 14,900 mm$^{-3}$, an elevated C reactive protein (CRP) level of 108 mg/l, and anaemia with a haemoglobin level of 9.8 g dl$^{-1}$.

A chest X-ray (Figure 1) was performed and showed a large rounded homogenous opacity occupying the right middle and lower zones, with shifting of the mediastinum to the opposite side, alongside a mild (loculated) pleural effusion within the right upper zone. The radiograph showed no evidence of calcifications, cavitations, air bronchogram or underlying bony destruction. Chest ultrasound (Figure 2) was done and showed a large heterogeneous echogenic mass at the base of right pleural cavity with a mild pleural effusion within the upper lobe. No liver invasion was appreciated. CT scan with IV contrast (Figure 3) was done for further assement and revealed a well-defined heterogeneous soft tissue density lesion measuring 11 × 8 × 14 cm (AP × TR × CC) in the right lower lobe. The lesion showed enhancing areas and some non-enhancing areas, upon contrast administration, suggesting a presence of necrosis, in addition to a mass effect shifting the mediastinum to the opposite side and resulting in passive atelectasis of the right upper lobe associated with a small right upper lobe pleural effusion. No evidence of calcification within the lesion or chest wall invasion could be seen. There is no associated cystic lung disease in remaining lungs. No evidence of metastasis in chest or abdomen could be appreciated. A CT-guided biopsy was taken from the tumour. Histopathological analysis of the biopsy revealed a PPB type III. Therefore, the patient was referred for chemotherapy.

**DISCUSSION**

Lung malignant tumours during childhood are considered a rare entity, representing only 0.5–1% of primary lung cancers. Pulmonary blastomas are uncommon fast-growing lung neoplasms that account for approximately 0.25–0.5% of primary lung malignancies. The pathogenesis of these...
tumours remains uncertain, but due to their resemblance to fetal lung tissue, pluripotent pulmonary blastomas have been postulated to be the origin of the disease. They occur mostly in the infants and young children, however, similar cases had been reported in adults.5–7

Up to date, no definite causative agent has been determined, but some associations have been reported. PPBs are associated with type IV congenital pulmonary airway malformations.8 They are also associated with dysplasia syndrome in 33% of the cases.9 The association of DICER1 mutations and PPB is reported in approximately 66% of the cases.2 Therefore, all patients with PPB should be screened for DICER1 mutation. If DICER1 mutation is detected during screening, the possibility for the patient to develop malignancies in other organs must be considered. In 30% of cases, patients with PPB may also present with multilocular cystic nephroma, and, very rarely, Wilms tumor.9 Following the initial description by Barrett and Barnard in 1945 (originally termed embryoma of the lung), pulmonary blastomas were classified into three histological subtypes:10 I, II, and III. Type I is a purely cystic lesion (14%), type II tumours have both cystic and solid components (48%) and type III are purely solid (38%). The classification has also a prognostic value, type I being the least malignant and type III the most malignant lesion.

The clinical picture of blastomas is highly variable. There are no specific presenting signs or symptoms of biphasic pulmonary blastomas. Some studies suggest that symptoms present only in 60% of the cases, with the lesions for the rest of the cases found incidentally.10,11 Symptoms, when present, may include fever, dyspnoea, respiratory distress, cough. Rarely, hemoptysis, chest pain, weight loss, anorexia, fatigue, or even neurologic symptoms can occur. Currently, there is no specific serum marker for pulmonary blastomas.10

Imaging studies have enormous capability on identifying the characteristics of lung lesions. Usually, PPB appears as a solitary, well-demarcated, peripheral mass that can be large enough to extend beyond 10 cm in size.12 PPB is often found late on plain film radiographs as a unilateral lung whiteout with a mediastinal shift to the opposite side. Typically, there are no adjacent rib erosions or calcifications. In the majority of cases, the disease is unilateral, but bilateral disease has been reported. In some cases, pleural effusion may be seen.11 Ultrasound is a poor imaging modality for diagnosing PPB as findings are non-specific and may show a large region of consolidation without sonographic air bronchogram. The appearance of PPB on CT scan is variable depending on the histological type of the tumour. Type I lesions manifest as a single cyst or a multicystic lesion (often air-filled). Type II lesions show air- or fluid-filled cavities with possible air-fluid levels along with solid internal nodules. In type III lesions, neoplasms are solid and show low attenuation, with homogeneous or heterogeneous enhancement. On MRI, the tumor manifests as a heterogeneous mass, typically hyperintense on both T1W and T2W sequences.13 A FDG-PET CT scan reveals a hypermetabolic lesion. PET CT is helpful in evaluating
The differential diagnosis for a chest wall mass depends on the imaging findings. It is important to identify imaging findings that can make a specific diagnosis. For example, PPBs, particularly when they are locally aggressive, include pleuropericardial blastoma, pleuropulmonary blastoma, and DICER1 syndrome. The definitive diagnosis of PPB was made by cytogenic analysis of the tumor cells, which were biopsied under the guidance of CT.

**CONFLICTS OF INTEREST**

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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