CASE REPORT

Pleuropulmonary Blastoma in an Infant Presenting with Tension Pneumothorax

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ABSTRACT
Pleuropulmonary blastoma is one of the rarest tumours occurring in childhood. The pathogenesis and therapeutic management of this condition remain controversial. We report the case of an 8-month-old girl with cystic pleuropulmonary blastoma (type 1), presenting with respiratory distress and a tension pneumothorax. The clinical and radiological aspects of the pleuropulmonary blastoma are also discussed.

Key Words: Childhood, Pleuropulmonary blastoma, Pulmonary cyst, Tension pneumothorax

INTRODUCTION
Pleuropulmonary blastoma (PPB) is a rare childhood malignancy. The largest study of this condition, in which only 50 cases were described, was reported by Priest et al in 1997. Due to the rarity of this disease, pathogenesis and therapeutic management of this condition remain controversial. We report the case of a patient with cystic PPB (type 1), presenting with tension pneumothorax. The clinical and radiological aspects of the disease are also discussed.

CASE REPORT
An 8-month-old girl was admitted to the paediatric unit due to shortness of breath for 3 days, along with vomiting and decreased feeding. The infant had previously enjoyed good health. Physical examination revealed a respiratory rate of 40 breaths per minute with subcostal insucking. Reduced breath sounds on the left side were noted. Heart rate was 150 beats per minute. Oxygen saturation was 94%. Chest radiography showed a left pneumothorax, with mediastinal shift to the right side (Figure 1). A thin-walled cystic lesion was also noted at the left lower zone of the lung (Figure 2). Following the diagnosis of left tension pneumothorax, left chest drainage was performed. The condition of the patient improved and the chest drain was removed three days after insertion.

Computed tomography (CT) scanning of the thorax was performed for further delineation of the cystic lesion. A 3 cm solitary, air-containing, thin-walled cyst was noted at the left lingular segment, abutting the left heart border (Figure 3). No mural nodule or air-fluid level was seen. No pleural effusion was noted. In view of the potential risk of recurrence of the tension pneumothorax, surgical intervention was planned.
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A left thoracotomy was subsequently performed. A 4 cm x 3 cm cyst at the tip of the lingular segment of the left lung was located intraoperatively. The cyst was excised and sent for pathological studies. Gross specimen studies showed that the cyst, measuring 35 mm x 20 mm x 17 mm, had multiple, adjacent thin-walled cysts (Figure 4). Microscopic examination demonstrated that the cysts were lined by low cuboidal epithelium, resembling bronchiolar cells, with a cambium layer of immature round cells beneath. The round cells showed mitotic activity and a Ki67 index of approximately 40%. A few foci of bone formation were present. The overall features were compatible with PPB type 1.

The postoperative course was uneventful. No evidence of tumour recurrence or other associated abnormalities have been detected over a period of 18 months, with regular follow-up.

DISCUSSION

PPB is a very rare tumour, previously known as cystic rhabdomyosarcoma, pulmonary blastoma of childhood, or rhabdomyosarcoma arising in a cystic adenomatoid malformation. PPB is classified into three categories — type 1 (purely cystic), type 2 (cystic and solid), and type 3 (purely solid). Transition of type 1 lesions to type 3 lesions has been reported.2 The lower lobe of the lung is more commonly involved than the upper and middle lobes. The disease is usually unilateral, although bilateral lesions have been reported.3

According to the clinicopathological study conducted by Priest et al,1 the commonest clinical presentation of PPB is respiratory distress, as seen in the presentation of this patient. Other signs and symptoms include fever, chest or abdominal pain, cough, anorexia, and malaise. On radiological examination, pulmonary cysts, with or without pneumothorax, are seen in 38% of patients. Other presentations include pleural effusion,4 and posterior mediastinal mass.5 The radiological finding of a thin-walled lung cyst on chest radiograph should be evaluated thoroughly. Differential diagnoses include congenital cystic adenomatoid malformation, bronchogenic cyst, pulmonary sequestration, pneumatocele, hydatid cyst, and pulmonary abscess. CT scanning of the thorax is sensitive in detecting and delineating the lesion, and should be the procedure of choice for further investigation.

For PPB type 1, non-operative management, with close clinical and radiological follow-up, may be
considered. Radical surgery remains the primary therapy, especially for PPB types 2 and 3. There is no consensus regarding the use of chemotherapy and radiotherapy as adjuvant therapy. It has been suggested that as the response to chemotherapy is poor, chemotherapy should be given alongside local radiotherapy. The prognosis for PPB type 1 appears generally favourable. PPB type 2 and PPB type 3, however, are aggressive malignancies, with documented overall 5-year survival rates of only 42% with multimodality therapy. Case reports have suggested that up to 25% of patients with PPB or their young relatives have associated dysplastic lesions, such as cystic nephroma, pulmonary cyst, cystic adenomatoid malformation, or neoplastic lesions, such as sarcoma, medulloblastoma, and thyroid neoplasm. The molecular genetics of PPB is currently being investigated and in the future, screening of family members may help to detect the disease at an earlier stage.

REFERENCES