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Case Report

Extra pulmonary pleuropulmonary blastoma in two-year-old child: a rare cystic lesion of lung

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ABSTRACT

Pleuropulmonary blastoma (PPB) is a rare but fatal intrathoracic malignancy, which arises from the lung parenchyma and/or pleura. Despite being rare, PPB is the most common lung tumor in young children. Clinically, this rare malignancy is often mistaken for symptoms of respiratory tract infection or pneumothorax. The PPB require aggressive treatment which include surgical resection and chemotherapy. We report this case with the aim to raise awareness about this fatal and misleading malignancy. Knowledge of clinical presentation, imaging findings, histopathological findings, staging and association with other tumors is crucial for timely diagnosis of pleuropulmonary blastoma and subsequent adequate management.

Keywords: Pleuropulmonary blastoma, Cystic, Lung, Pediatric

INTRODUCTION

Congenital cystic lung lesions are a group of lung diseases which represent variable pathology ranging from benign congenital malformation to neoplasm. These lesions can present with overlapping clinical and radiological features. Several classifications have been proposed based on the radiological and pathological features.[1] Pleuropulmonary blastoma (PPB) is an extremely rare and potentially serious subtype of neoplastic cystic lung lesions, which can be misdiagnosed as a benign congenial cystic lesion. It is a dysembryonic neoplasms of thoracopulmonary mesenchyme. PPB accounts 0.5%-1% of all pediatric primary malignant lung cancers. Usually it arises from lung tissue, however rarely the parietal pleura may be the tissue of origin (extra pulmonary PPB) which are extremely rare.

The prognosis is poor with distant metastasis to central nervous system and bone with survival rate of approximately 42.9% at 5 years. Here we described a case report of multi-septate cystic pleural lesion in a 2-year-old child who was diagnosed to have pleuropulmonary blastoma type 1 based on histopathological examination and underwent successful resection.

CASE REPORT

A 2-year-old boy presented to pediatric emergency department, Fortis Hospital, Gurugram, Haryana with short history of dry cough, high grade fever and difficulty in breathing. No history of sudden choking episode or noisy breathing. The child was born via LSCS at 37-week gestation to a G3P1L1 non- consanguineously married mother with an uneventful perinatal period with birth weight of 2.7 kilo grams. On day 2 of life, he developed respiratory distress for which he required ventilator support. He was treated for *Acinetobacter baumannii* sepsis, fungal UTI. Chest X-ray was normal at that time. He had attained all milestones appropriate for age. On admission child had visible suprasternal, intercostal,

subcostal retractions and nasal flaring. Breath sounds were reduced on right side of chest. He had tachycardia and tachypnea. He was febrile but was hemodynamically stable. Pallor was present, there was no edema, cyanosis or lymphadenopathy. Saturation was 90% on room air. Other system examination was unremarkable.

Figure 1: Chest X-ray showing hyperlucent right hemithorax with septations and mediastinal shift to left.

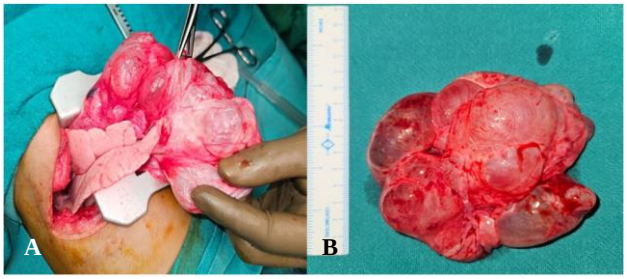

Figure 2: (A) Large cystic lesion arising from visceral pleura; (B) right lung appears pink and atelectatic.

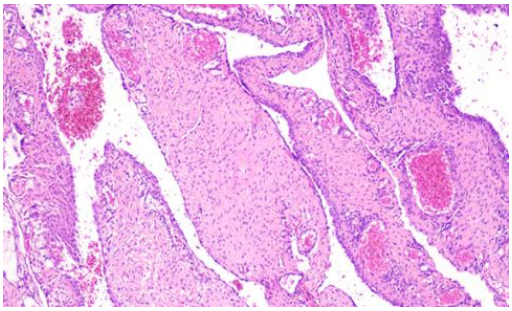

Figure 3: Expanded cyst like structure lined by bronchial to alveolar lining.

Cyst wall is fibromuscular with various sizes of blood vessels. At places, stroma showing subepithelial collection of mesenchymal cells. The wall showing normal lung alveoli at places. Findings suggestive of Pleuropulmonary blastoma type 1.

Initially chest X-ray was done which revealed large hyperlucent lesion occupying right hemi-thorax with septations and mediastinal shift to left. Blood investigations revealed hemoglobin of 9.6 g/dl, white blood cell count 8230 /mm^3, and platelets 3.74 lac/mm^3. Liver and kidney function tests were within normal limits. C-reactive protein was 5.0 mg/dl, procalcitonin was 0.05 ng/ml. Total IgE was 97.0 Ku/l. For evaluation of airways flexible bronchoscopy was done under sedation which revealed fixed obstruction of right main bronchus and right lower lobe bronchus, Gram stain of bronchoalveolar lavage showed no pus cells, aerobic culture was sterile, cytology showed few polymorphs and lymphocytes, without any hemosiderin and lipid laden macrophages and atypical cells. CECT thorax showed a complete collapse of right lung with replacement of right hemithorax with large air filled cystic spaces.

Child was admitted in PICU in view of respiratory distress and started with intravenous antibiotics, oxygen supplementation via mask, bronchodilators and other supportive measures. His tachypnea improved and pediatric surgeon opinion was taken in view of CECT finding of large emphysematous bulla. Right sided posterolateral thoracotomy and cyst excision was done under general anesthesia. Intraoperative findings revealed Large multi-lobulated cystic lesion of size around 15cmx 15×10 cm. Cyst was arising from visceral pleura, right lung appeared healthy and pink but was completely collapsed underneath the cyst. The cyst was excised and sent for histopathological examination. His respiratory distress improved and discharged in stable condition on 8th post-operative day.

Diagnosis of pleuropulmonary blastoma type 1 was confirmed by histopathological examination which revealed expanded cyst like structures lined by bronchial to alveolar lining. The cyst wall was fibromuscular with blood vessels of varying sizes. At places stroma showed sub epithelial collection of mesenchymal cells. The wall showed normal lung alveoli at places. Rhabdomyosarcomatous differentiation was not seen. Genetic testing showed that the patient had heterozygous pathogenic mutation in DICER1 gene.

DISCUSSION

PPB is a rare intrathoracic malignancy arising from the lung parenchyma and/or pleura. It is considered the most common primary intrathoracic malignancy of childhood. Manivel and associates coined the term PPB to describe a specific subtype of pulmonary blastoma based on its exclusive clinical presentation in childhood and its pathologic features of variable anatomic location, primitive embryonic-like blastoma and stroma, absence of carcinomatous component, and potential for sarcomatous differentiation.[2]

PPB was first described in 19883 as a dysembryonic neoplasm of pleuropulmonary mesenchyme.[4] Extrapulmonary PPBs are extremely rare pediatric primary neoplasms of chest and accounts for 25% of all PPB cases. These highly malignant and aggressive tumors, usually occur in the children <5-6 years of age. Type I pleuropulmonary tumors mostly occur in children younger than 2 years of age with a median age of 10 months, type II occurs at a median age of 35 months and Type III pleuropulmonary tumors: occurs at a median age of 41 months.[5]

PPB is classified into three subtypes: type I consists only of cystic lesions; type II consists of mixed cystic and solid components, whereas type III consists of only solid tumors. These subtypes represent a progression process based on reported cases of type I progression into type II and type III.[6,7] Type I tumors are seen to be associated with preexisting long-standing congenital cystic lesions of lung and can be more aggressive if inadequately managed. Microscopic findings vary according to the different subtypes.

Type I pleuropulmonary tumor is characterized by multicystic lesions lined by respiratory-type epithelium with the presence in the cystic wall of a population of small malignant cells that can lie as a continuous or discontinuous cambium layer zone. Areas of rhabdomyoblastic differentiation may be present such as immature cartilage.

Type II pleuropulmonary tumor is characterized by a complete overgrowth of the septal stroma by sheets of primitive small cells without apparent differentiation, areas of embryonal rhabdomyosarcoma or fascicles of a spindle cell sarcoma.

Type III pleuropulmonary blastoma is characterized by mixed sheets of blastematous and sarcomatous areas like chondrosarcoma -like, fibrosarcoma like, rhabdomyo-sarcoma-like or anaplastic areas.[8]

Most of the children are asymptomatic initially. The lesions are identified incidentally by chest X-rays obtained for different reasons. Delay in diagnosis, therefore, may occur until disease progresses to advanced stages, causing compressive symptoms, hemodynamic instability, and metastatic lesions, pneumothorax or neurological symptoms resulting from brains metastasis. This tumor is also known to metastasize to bone, liver, lymph nodes, kidney, pancreas, and adrenal glands.[9] Our case presented at 2 years of age corresponding gross and histological features suggestive that of type I PPB. Intrathoracic space occupying lesions (SOL) with septations in children may create a diagnostic dilemma with hydatid cyst or congenital cystic lung lesions often delaying the correct diagnosis. PPB remains a differential diagnosis in these radiological scenarios despite its rarity. Surgical excision remains the mainstay of treatment. Adjuvant chemotherapy is recommended to prevent recurrence. Most of the time, response to chemotherapy is poor, so combination of chemotherapy with local radiotherapy is also recommended by some authors. Ifosfamide, vincristine, actinomycin D, and doxorubicin regimen is the regime of choice. However, due to limited literature there is no consensus on duration of chemotherapy. Extra pulmonary PPBs have poor prognosis. Local recurrence and distant metastasis frequently occur after or during therapy. Patients with pleural, mediastinal or extra pulmonary involvement at the time of diagnosis have worse prognosis. Types II and III PPBs are highly aggressive malignancies with overall survival of 42.9% at 5 years, even after multimodality therapy. High-dose consolidation therapy with autologous stem cell rescue is recommended for recurrence of these tumors. For patients with unresectable or residual tumors post-surgery chemotherapy, radiotherapy is prescribed.[10]

CONCLUSION

Extra pulmonary PPBs are rare and highly aggressive primary intrathoracic tumors of childhood which require multimodality therapy including radical surgical excision, chemotherapy and rarely radiotherapy. Pulmonary cystic lesions in infancy need to be evaluated with caution to rule out serious disease entities like PPB.

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