Case Report

Pleuropulmonary blastoma in 3 year old child - A rare case report

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Abstract

Pleuropulmonary blastoma (PPB) is a uncommon, aggressive, dysontogenetic, and malignant tumor of originating from intrathoracic (pulmonary, pleural, or combined) mesenchyme.[1,2]The purpose of this case report is to increase the general awareness for this entity so as to recognize this rare entity early in its course, since type I tumors (resembling congenital lung cyst) can progress over time to aggressive type II and type III tumors. Therefore, early histological recognition and differentiation from congenital airway malformations like bronchogenic cyst and other benign cysts like duplication cyst, neurenteric cyst are very important.

Key Words: Lung mass, malignant.

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CASE PRESENTATION

A 3 year old child presented with fever, cough and difficulty in breathing. Chest radiograph (Fig.1) showed well-circumscribed mass in the left mid-lung zone silhouetting with left heart border. Contrast Enhanced Computerized tomography (CECT) scan showed well defined heterogeneous enhancing mass (Fig.2) measuring 9x5.5x4.3cm showing peripheral enhancement with focal enhancing soft tissue and thin septae (Fig.3) & minimal left sided pleural effusion .The patient underwent CT guided biopsy which revealed lung and bronchial epithelium with tiny bits of round to oval blastemal cells with hyperchromatic nuclei and scanty cytoplasm (Fig.4) and bits of immature cartilage (Fig.5). Therefore differential diagnosis of chordoma, chondroid hamartoma, and pleuropulmonary blastoma was raised and pneumonectomy was suggested. Left Thoracotomy was performed; the mass was found to be limited to left lower lobe. Left pneumonectomy was performed without complications. The excised mass was solid, circumscribed, firm, and measuring 7.8x4.2x2.8 cm surrounded by normal lung tissue. Its cut surface showed gelatinous firm with areas of necrosis, with intermingled greyish white fleshy areas. Microscopically, it was composed of growth septal stroma by clusters and fascicles of primitive small sheets, oval to spindle blastemal cells with hyperchromatic nuclei and scanty cytoplasm (Fig.4) and bits of immature cartilage (Fig.5) and a few areas of embryonal rhabomyosarcoma amidist nodules of chondrosarcoma like areas. Necrotic, haemorrhagic & fibrotic area were seen.

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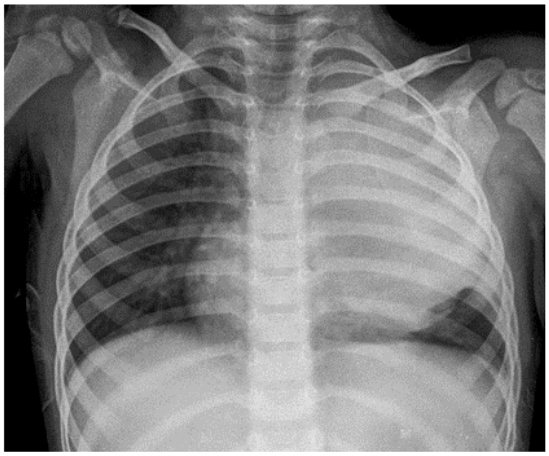
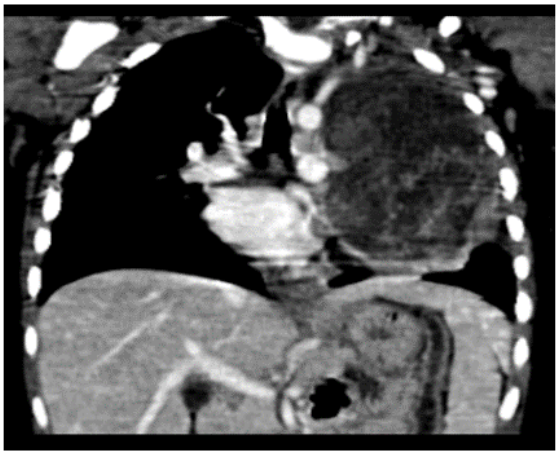


Figure 1:



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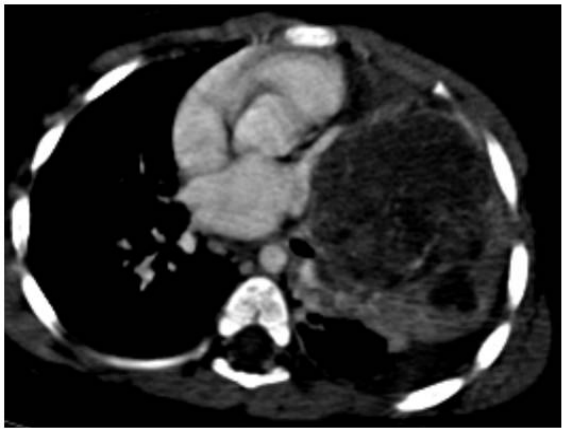

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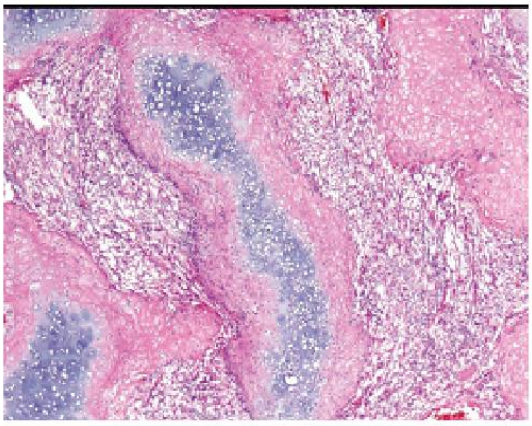

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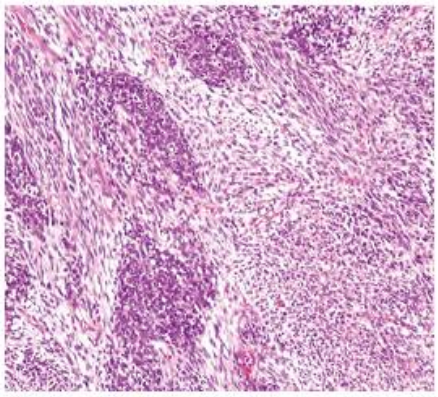

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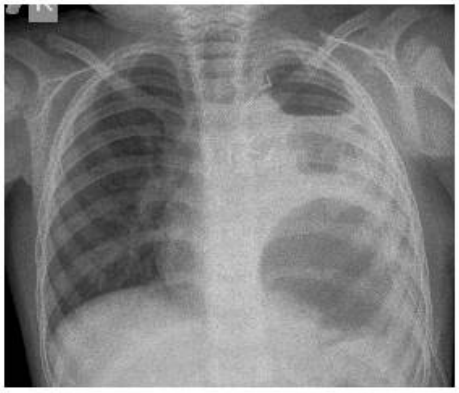

Figure 6:

Figure 7:

DISCUSSION

Pleuropulmonary blastoma is not a common intrathoracic tumor, however it accounts for 15% of all primary pulmonary tumors of paediatric age group and 25% of these cases occur in familial settings [4]; the associated malignancies in first and second-degree relatives include rhabdomyosarcomas, synovial sarcoma, pleuropulmonary blastoma, thyroid carcinomas, ovarian Sertoli- Leydig cell tumors, gonadal germ cell tumors, and certain types of leukemia. Pulmonary blastoma is histologically composed of both carcinomatous and sarcomatous components and a mixture of epithelial and mesenchymal tissues resembling embryonic lung tissue. Morphologically there are three types of PPB (I, II, and III).[3]

- **Type I** : Cystic , 14% (prenatal and <1year)
- **Type II** : Mixed , 48% (mean age 2.8-2.9 years)
- **Type III** : Solid , 38% (mean age 3.6-3.7 years)

The age of presentation is usually not more than 4 years. The median age of presentation depends largely on the tumor type. For type I tumors, the median age is less than 1 year; for type II the median age 2.8-3 years; and for type III the median age is 3.6-3.7 years.[5] There is no as such gender predilection and common tumor location noted, however it is more commonly seen on the right side.[1]Certain genetic mutations are associated with pleuropulmonary blastomas; these include germ line DICER1 mutation (loss of function) in familial cases, gains of chromosome 8 (most consistent chromosomal abnormality), trisomy 2, unbalanced translocation between chromosomes 1 and X, and p53 mutations or deletions.[2]

Denher and associates classified PPB mainly into three groups; type 1 being purely cystic tumors, type 2 as an intermediate type, and type 3 predominantly being solid tumors.[7] A progression from type I to type III may be seen over time.[7] The histologic appearance is variable - the tumor is characterized by primitive blastoma and a malignant mesenchymal stroma often showing multidirectional differentiation as rhabdomyosarcomatous, chondrosarcomatous or liposarcomatous. The cystic component shows benign metaplastic epithelium lining.[8] Vargas et al. demonstrated, with cytogenetic analysis, that the polysomy of chromosome 8 is a constant feature of pleuropulmonary blastoma and the clonal proliferation in pleuropulmonary blastoma is restricted to the malignant mesenchymal elements, supporting the notion that the epithelial components are non-neoplastic.[7] This neoplasm is not only limited to occur in the lung, but it may arise from mediastinum, diaphragm and/or pleura. This raises the possibility that pleuropulmonary blastoma may be even originate from the splanchnopleural or somatopleural mesoderm. Common metastatic sites include the brain, bone, lymph nodes, liver, pancreas, kidney, and adrenal glands.[9]This case is important reminder for consideration of this rare entity as one of the differential while dealing with primary pulmonary neoplasm of paediatric age group.

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