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Pleuropulmonary blastoma-A case report

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Pleuropulmonary Blastoma (PPB) is a rare primitive primary neoplasm of the thorax in children. It may arise in the pulmonary parenchyma, pleura and / or mediastinum. PPB arises from the primitive mesenchymal cell and is encountered in the first few years of life. Three subtypes (Type I, II, III) are a continuum from the least to the most malignant lesion. An international registry (ppbregistry.org) has been established. Treatment is multimodal (surgery and chemotherapy, rarely radiation therapy) and depends on the type and aggressiveness of the disease. We report a 3 year old child weighing, 15 kilograms, who presented to the pediatrician with history of cough and fever of 2 weeks duration. Contrast computerized tomography (CT) scan of the thorax displayed a large (100x55x53mm), well- defined, heterogeneous (80-120HU) mass, occupying two-third of the left hemithorax. The mass had a focal enhancing soft tissue component with thin septae. Lung parenchyma was found inferior to the mass. Trachea and main bronchi were normal. CT guided biopsy of the mass was reported as round cell tumor. The patient underwent surgery via a left posterolateral thoracotomy via the 4th intercostal space. A heterogeneous (predominately solid with few cystic areas) mass, 12x14cm was found occupying the upper two-thirds of left hemithorax, adherent to chest wall, pericardium and the left lobe of thymus . The mass had infiltrated into the lower lobe of lung. The upper lobe and Lingula were not identifiable. Three hilar lymph nodes which were 1cm in diameter and firm in consistency were present. Enbloc excision biopsy was done and tissue sent for histopathological examination (HPE). The HPE report was PPB type III involving the lymph nodes; the bronchial surgical margins were free of tumor. The adjacent lung parenchyma showed interstitial pneumonia. Chemotherapy was begun one month after surgery. The child succumbed after the third cycle of chemotherapy. PPB is an aggressive tumor accounting for less than 1% of all primary malignant lung tumors in the pediatric population. Manivel and associates coined the term PPB on the basis of its exclusive clinical presentation in childhood and its pathologic features of primitive embryonic-like blastoma, absence of carcinomatous component and potential for sarcomatous differentiation. Dehner and associates classified PPB into 3 types Type I-Purely cystic, Type II-intermediate or mixed (cystic & solid), Type III-predominantly solid. A progression from type-I to type-III may occur over time. It can arise from lung, pleura, mediastinum and/or diaphragm. This has raised the possibility that PPB may originate from splanchnopleural or somatopleural mesoderm. Bilateral occurrence is very rare. Common metastatic sites are brain, bone, lymph-nodes, liver, pancreas, kidney and adrenal gland. 30-40% children with PPB may have cancers such as multilocular cystic nephromas, ovarian tumors and thyroid tumors-this is known as PPB familial cancer syndrome or the DICER I syndrome. Many of these patients have mutation of the DICER I gene. The tumor has no characteristic findings on imaging studies. Biopsy is the main stay for diagnosis. Radical surgery followed by chemotherapy and/or radiation therapy is the treatment pathway followed the prognosis is grave. Type II & III PPB have a projected overall survival of 62% at 2 years and 42% at 5 years, even after multimodality therapy and those with pleural, mediastinal or extra pulmonary involvement have worse prognosis.

Biography

Rekha Matta has independently performed more than ten thousand cardiovascular and thoracic surgeries (adult & pediatric). She is currently working as a professor and HOD of Department of Cardiovascular and thoracic surgery at Krishna Institute of Medical sciences deemed university at karad, India.

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