Type II- Pleuropulmonary Blastoma: A case report

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Summary:
Pleuropulmonary blastoma is a rare and aggressive neoplasm typically presents in young children, younger than 5 years, as a pulmonary and/or pleural-based tumor. We reported a case of type-II pleuropulmonary blastoma (PPB) in an 8 month old infant who presented to Al-Khansa'a Maternity Hospital in Western Mosul with a history of repeated attacks of shortness of breath and signs of pulmonary infection unresponsive to treatment. A bronchoscopic examination showed a mass involved and obstructed the orifice of the main bronchial tree of the right upper lung lobe. A thoracotomy was performed; tumor involved the right upper lung lobe, the covering pleural surface and invading the mediastinal structures. Right upper lobectomy was performed. Tumor had cystic and solid components. It characterized histologically by a mixture of primitive blastematous and sarcomatous elements.

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Introduction:

Pleuropulmonary blastoma (PPB) is a unique neoplasm of childhood that appears as a pulmonary and/or pleural-based mass with cystic, solid, or combined cystic and solid features and is characterized histologically by a primitive, variably mixed blastematous and sarcomatous appearance (1). PPB occurs most often in children younger than four, sometimes in children 4-8 years old, and very rarely in older children, teenagers, and adults. PPB occurs in boys and girls approximately equally (2). It was first described in 1988 by Manivel and associates who coined the term PPB to describe a distinct entity from pulmonary blastoma on the basis of its exclusive clinical presentation in childhood and its pathologic features of variable anatomic location, primitive embryonic-like blastema and stroma, absence of carcinomatous component, and potential for sarcomatous differentiation (3).

Case Report:

An eight month old male presented with repeated attacks of shortness of breath and signs of pulmonary infection. Chest X-ray showed persistent homogenous opacity in the right upper lung lobe which failed to resolve by the treatment (Fig 1). The patient underwent bronchoscopic examination which showed a mass involved and obstructed the orifice of the main bronchial tree of the right upper lung lobe.

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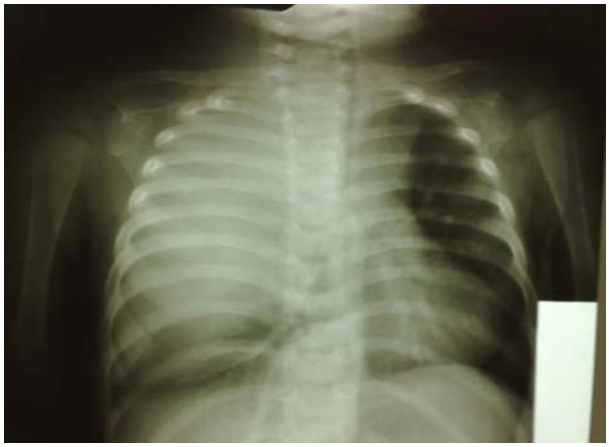


Figure 1: Large right upper lung consolidation

Under general anesthesia with endotrachial intubation, the patient underwent right thoracotomy. A solid lobulated mass involved right upper lobe, pleural surface and infiltrating the mediastinum was identified. A right upper lobectomy was performed after freeing of the mass from the chest wall, the vital blood vessels and surrounding structures (Fig 2). Postoperatively the patient remained on ventilator for one day then discharged home after a week. The patient developed Horner's syndrome as the tumor extended superiorly and posteriorly to the sympathetic chain.

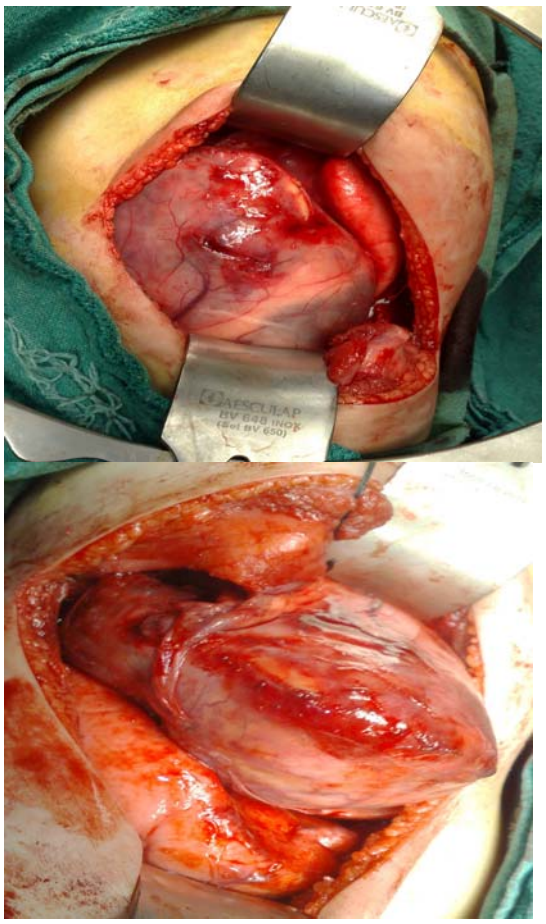


Figure 2: Solid large upper lobe mass involving the pleura & mediastinum

The resected tumor was large whitish-gray firm and lobulated measuring 13.0 X 6.0 x 5.0 cm & weighing 300gm. Cut sections showed variegated surface of predominant solid, cystic and necrotic areas (Fig 3).

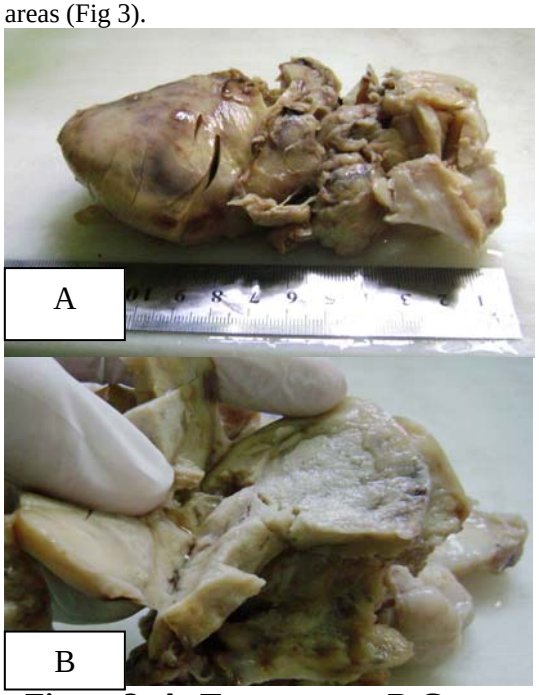
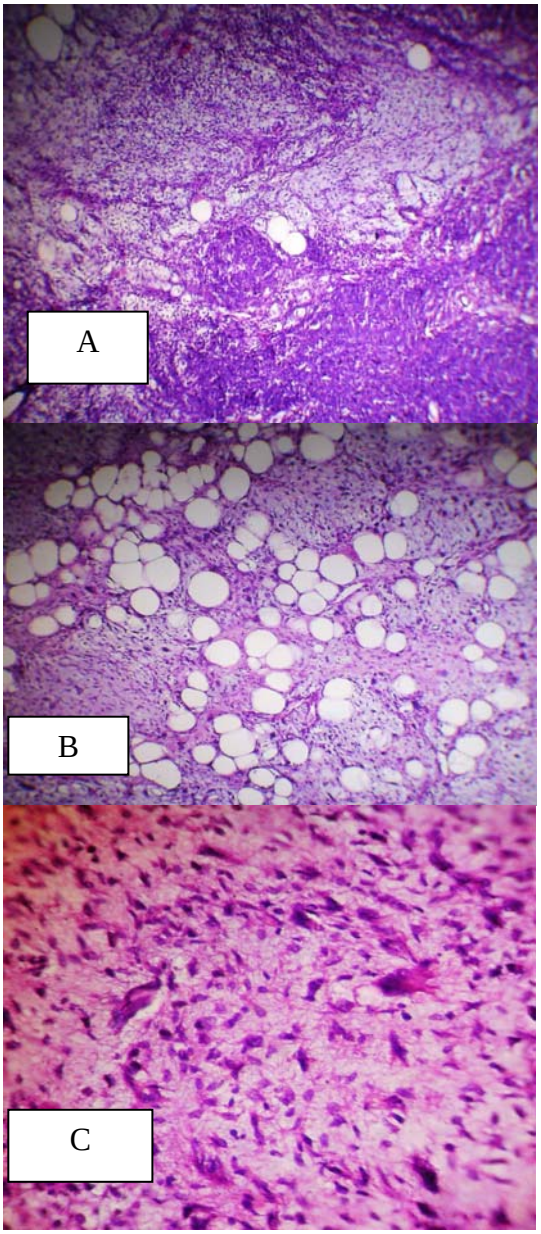


Figure 3: A- Tumor mass, B-Cut sections show solid, cystic and necrotic areas.

Histologically the solid portion was composed of combination of primitive blastema (clusters of undifferentiated oval/spindle cells with vesicular nuclei) and malignant mesenchymal component with a diversity of differentiation into liposarcoma (lipoblast with myxoid matrix), rhabdomyosarcoma (spindle and primitive rhabdomyoblast with eosinophilic cytoplasm), and undifferentiated MFH-like sarcoma (large multinucleated giant cells with spindle stroma). The cystic spaces were lined by the primitive blastomal cells. The tumor contained foci of necrosis (Fig 4). These findings were diagnostic of type II pleuropulmonary blastoma.



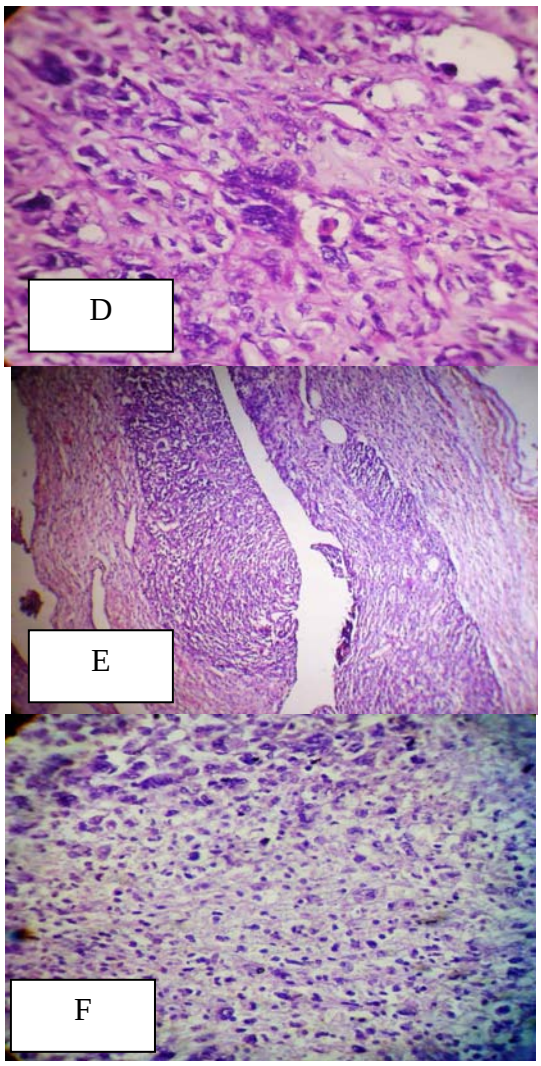


Figure 4: Microscopic features: A-Primitive blastema, B-Liposarcoma, C-Rhabdomyosarcoma, D-Undifferentiated giant cell (MFH-like), E-Cyst lined by primitive blastema, F-Necrosis.

Discussion:

Pleuropulmonary blastoma (PPB) is a rare primary neoplasm of pleuropulmonary mesenchyme. Fewer than 300 cases have been reported in literatures allover the world and few single institutions have reported more than several cases (4, 5). To the date, this case is the first to be reported in our locality and in Iraq (6, 7). Cytogenetic analysis has frequently reported polysomy of chromosome 8 or more generally, gains of chromosome 8 as a feature of pleuropulmonary blastoma (8, 9). A recent study showed multiple chromosomal abnormalities. However, the most consistent of which is the extra copies of chromosome 8, which has been found in approximately 75-80% of the cases (10). The occurrence of PPB may herald a constitutional and heritable predisposition to dysplastic or neoplastic disease in approximately 25% of cases. Associated conditions include lung cysts, kidney cysts, thyroid tumors (sometimes malignant), and various other cancers. This is called PPB family cancer syndrome. It is not known what are the causes the abnormalities in these children and families (11, 12). In the presented case there were neither other pathological or radiological findings, nor family history of relevant diseases. Our case, like most other reported cases, was presented with repeated attacks of shortness of breath and signs of pulmonary infection which failed to cure by the usual medical treatment (1, 2). Denher et al. classified PPB into 3 types on the basis of the cystic versus solid nature of the lesions as well as the histologic appearance (13). Type I PPB generally occurs in children younger than one year and is made up of mostly cysts, with almost no nodules. A thin layer of the wall of the cyst may appear cancerous. Type I PPB usually has the best prognosis of the three types. On the other hand, Type II PPB contains both cysts and cancerous nodules, and occurs most commonly in children around three years old. Type III PPB is a solid cancerous tumor, and occurs most often in children around four years old (13, 14). A progression from type I to type III had been reported over a short span of a few months (14, 15). In the present case, the tumor consisted predominantly of solid areas with cystic spaces and foci of necrosis. Histologically the solid foci are characterized by primitive mesenchymal or a mixture of undifferentiated blastematous and sarcomatous components with diversity of differentiation into chondrosarcoma, liposarcoma, rhabdomyosarcoma, or undifferentiated anaplastic sarcoma. While the cystic areas appears as multilocular lesion lined by discontinuous layer of rounded to spindle-shaped primitive tumor cells in the subepithelial stroma producing a cambium layer effect (3,4,16,17). These features correspond with the diagnosis of type-II pleuropulmonary blastoma, despite the younger age of the patient for this specific type of PPB. Type II and III PPB can metastasize and the common sites of metastasis are the brain, bones and the remaining parts of the lung (12, 18). As complete tumor ablation is essential to prevent local recurrence and allow any chance of survival, the main goal of therapy should be radical surgery, followed by chemotherapy. Because the response to chemotherapy is poor, some authors suggest that chemotherapy should be given with local radiotherapy in the majority of patients (18, 19, and 20). In our case a complete resection of the tumor were performed followed by chemotherapy. This regimen of treatment was similar to all other studies and reported cases (4, 18, 19, and 20). The 5 year survival is 83% for type I, 62% for type II, and 42% for type III PPB. Also patients with pleural or mediastinal involvement fared significantly worse than those without such involvement (1,19).

Conclusion:

Pleuropulmonary blastoma is a rare aggressive, intrathoracic neoplasm of early childhood which, in general, has unfavorable outcome. A complete tumor ablation followed by chemotherapy is the suitable type of treatment for this type of tumor.

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