A. Khatami MD[1]
A. Gharib MD[2]
M. Hadaie MD[3]
F. Hadaie MD[4]

Pleuro-Pulmonary Blastoma in a 4-Year-Old Child

Pulmonary blastoma is a rare malignancy with various histopathological appearances in children which present as an intra-thoracic mass. In this case report we present a 4-year-old boy with pleuro-pulmonary blastoma which was unrecognized for several weeks and was managed as pleural effusion and pneumonia. We decided to present this case because of the diagnostic difficulties and radiographic appearances.

Keywords: Pulmonary Blastoma, Child, Cancer, Chemotherapy

Introduction

Pulmonary blastoma is defined by the histological classification of lung neoplasm as a tumor consisting of immature or primitive epithelial and stromal components.[1] Pulmonary blastomas were sub-classified by Koss and colleagues into three categories: namely, classic biphasic pulmonary blastoma (CBPB), well differentiated fetal adenocarcinoma (WDFA) and pleuro-pulmonary blastoma (PPB) of childhood.[2]

PPB is defined as a distinctive pulmonary and/or pleural tumor of childhood with blastomatous and sarcomatous features without any epithelial components which differentiates it from the classical adult pulmonary blastoma.[3,4] It is pathologically divided into cystic, solid and mixed types.[5] The etiology and predisposing factors are unclear. The common presenting symptoms in PPB are respiratory distress, fever, chest pain, cough, anorexia and malaise.[6] This tumor is an extremely rare early childhood malignancy and has an unfavorable outcome.[7]
Bone erosion and adrenal metastasis were two relatively uncommon presentations of this tumor after chemotherapy in our case.

Case Presentation

A 4-year-old boy was referred to our center with a history of anorexia, weight loss, fever, cough, dyspnea and left chest pain since 6 months ago. He had multiple admissions in other centers and had several x-rays that were reported as opaque left hemithorax. However, significant mediastinal shift and downward displacement of the diaphragm were detected and diagnosis of pneumonia with pleural effusion was made (Fig. 1A). The patient underwent antibiotic therapy for pneumonia with chest tube placement in the left side for several days but no significant fluid was drained and there was no change in the patient's general condition. A chest CT-scan was taken with the administration of IV contrast. A heterogeneous enhancing solid mass with central hypodensities suggestive of necrotic areas in the left hemithorax with shifting of the heart and mediastinum to the right side and mild pleural effusion in the left side were seen (Fig. 1B). In abdominal sonography, no abnormal finding was detected. The patient was scheduled to undergo surgery to remove the tumor. At thoracotomy a large mass that adhered to the heart and mediastinal structures was detected, so complete

1. Assistant Professor, Department of Radiology, Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
2. Assistant Professor, Department of Pathology, Mofid Children's Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
3. Department of Radiology, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
4. Faculty of Medicine, Shaahed University, Tehran, Iran.

Corresponding Author:
Alireza Khatami
Address: Department of Radiology, Mofid Children's Hospital, Shariati Ave., Tehran, Iran.
Tel:+9821-2222-7021
Fax:+9821-2222-7030
Email:alireza_khatami31@yahoo.com.

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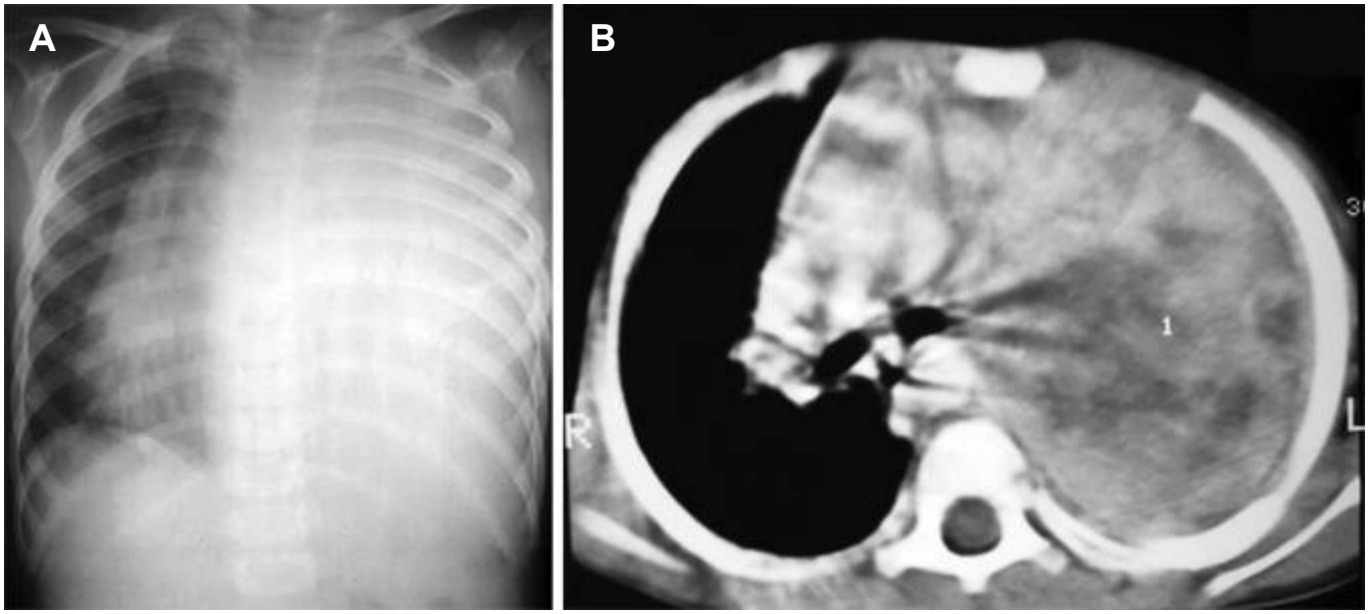

Fig. 1. A 4-year-old boy with pleuropulmonary blastoma.

A. Posteroanterior chest radiograph at presentation reveals left opaque hemithorax with mediastinal shift to the right side.

B. CT scan of the thorax with IV contrast at the same time reveals heterogeneous enhancing left thoracic mass with mediastinal shift to the right side.

removal of the mass was impossible.

Debulking of the mass was done and a chest tube was placed to drain the pleural effusion. The resected mass was histopathologically evaluated.

Microscopically, the tumor was composed of interlacing bundles of sarcomatous spindle cells mixed with primitive blastematous cells, showing high mitotic activity. Immunohistochemically, the neoplastic cells were negative for desmin, myogenin, muscle specific actin, cytokeratin and c-kit. The histological findings were consistent with the diagnosis of pleuropulmonary blastoma (Fig. 2).

The patient was referred for chemotherapy. After two weeks of chemotherapy the chest wall invasion was recognized and bone erosion of the rib cage which was confirmed by chest CT scan, was detected (Figs. 3A-C). Abdominal sonography was the next step to rule out other organ involvement. There was a 6.5 ×4.5 cm septated cystic mass in the RUQ, superior to the right kidney, (Fig. 3D) which was supposed to be metastasis or less likely hematoma of the adrenal gland. That mass also had heterogenous enhancement after IV contrast on the abdominal CT scan and was located in the right adrenal gland (Fig. 3E).

Despite two months of chemotherapy, the thoracic and adrenal masses did not reduce in size and the patient died. The patient had fever, respiratory distress, perianal abscess, ascitis and electrolyte disturbance at the time of death. We had no autopsy but based on the evidence, the adrenal mass appeared to be metastases to the adrenal gland.

Discussion

PPB may be diagnosed as massive pleural effusion with pneumonia like our case. Tumors such as rhabdomyosarcoma and primitive neuroectodermal tumors (PNET) are other differential diagnoses. Ac-

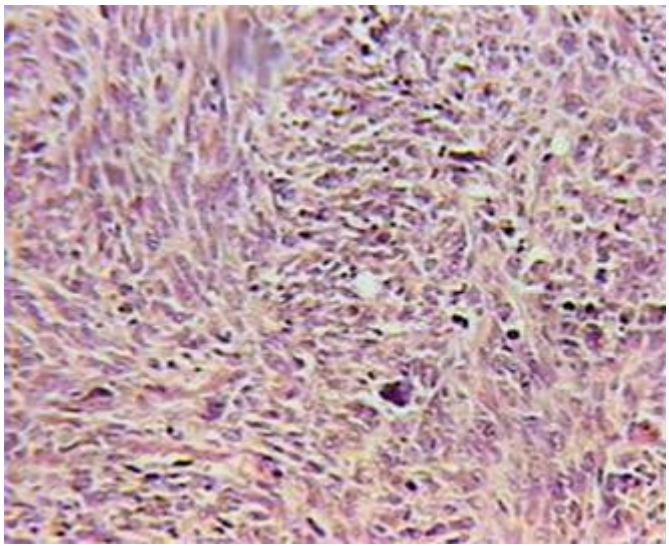

Fig. 2. Histopathologic view of pleuro-pulmonary blastoma in a 4-year-old boy.

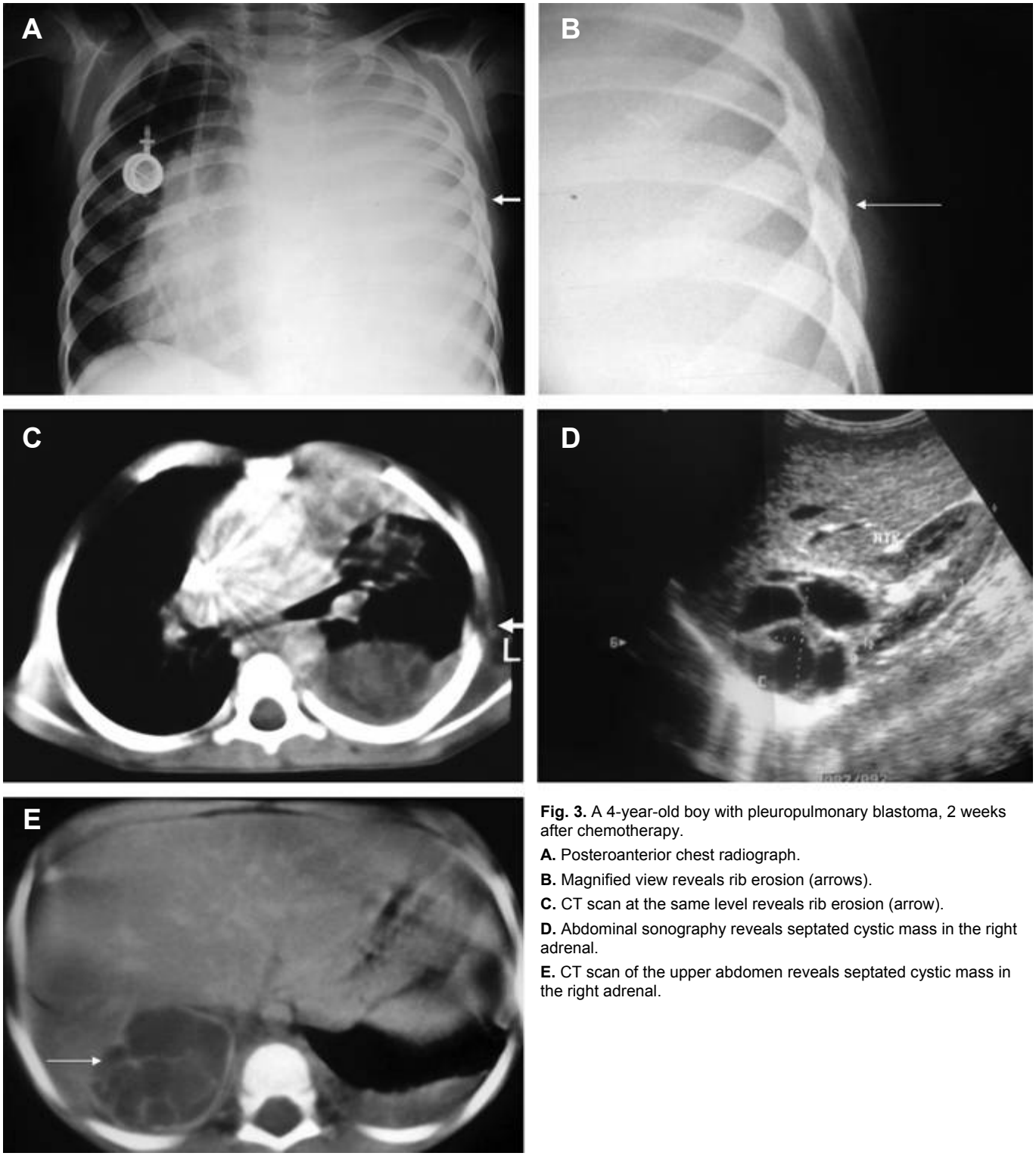

Fig. 3. A 4-year-old boy with pleuropulmonary blastoma, 2 weeks after chemotherapy.

A. Posteroanterior chest radiograph.

B. Magnified view reveals rib erosion (arrows).

C. CT scan at the same level reveals rib erosion (arrow).

D. Abdominal sonography reveals septated cystic mass in the right adrenal.

E. CT scan of the upper abdomen reveals septated cystic mass in the right adrenal.

cording to pathological characteristics, pleuropulmonary blastoma has been classified into 3 groups: type 1, purely cystic; type 2, intermediate cystic/ solid; and type 3, predominantly solid.[8,9]

Type 2 and 3 have relatively poor prognosis and present in 2 to 4-year-old children.[3] According to the pathological study, our case was categorized as type 3, which was seen in 19 of 50 cases in the Priest et al study.[6] Because of its rarity, most of the publications on PPB are case reports. The largest case series published by Priest includes 50 patients. These patients were 0-12 years old at diagnosis and had a median age

of 10 months at presentation.[6] In this study, older patients generally presented with a more advanced disease (type 2 and 3). Pleuro-pulmonary blastoma in children differs from the pulmonary blastoma observed in adults because of its variable anatomic location. It also presents most commonly with symptoms that are often mistaken for a lower respiratory tract infection. Clinical presentation of our patient was the same as other studies and similar to Priest's work which described 15 of 47 patients who were diagnosed as pneumonia cases. Our case was also treated for pneumonia for several weeks.[6] However, pathological characteristics and immunohistochemical studies had the characteristic appearance of this tumor. The tumor is usually located at the periphery of the lung, but it may also be located in extra-pulmonary locations, such as the mediastinum, diaphragm and/or pleura. Metastasis is rare at the presentation time. This case, just like the Priest et al. study, revealed bone and adrenal involvement after diagnosis.[6] Metastatic sites for PPB include the brain, bones, lymph nodes, liver, pancreas, kidney and less likely the adrenal glands.[8,10] Although bone metastasis is relatively common in this disease (12%), adrenal metastasis is a rare condition which is seen in only 2% of patients.[6] Despite aggressive treatment protocols, the prognosis of patients with PPB is not good. Priest et al. reported a 5 year survival rate of 83% for type 1 and 42% for types 2 and 3.[6]

Furthermore, it seems that type 2 and 3 lesions have a tendency to recur, even at remote or contra-lateral sites, despite presumed complete primary resection.[11]

The surgical procedure of choice for PPB is lobectomy because the limit between the lesion and normal parenchyma may be difficult to determine grossly.[11]

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