Pleuropulmonary Blastoma

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ABSTRACT

Pleuropulmonary blastoma (PPB) is a rare childhood neoplasm accounting for less than one percent of all primary malignant lung tumors of children less than six years of age. Metastasis to central nervous system (CNS), orbit, iris, bone, contralateral lung, liver, kidney, and pancreas and rarely to adrenal glands has been described. This report presents a case of pleuropulmonary blastoma with hemorrhagic pleural effusion and metastasis to the CNS at the time of presentation.

Keywords: Child, Metastasis, Pleuropulmonary blastoma, Tumor

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INTRODUCTION

Pleuropulmonary blastoma, a subtype of pulmonary blastomas (PB), is rare. It is a dysembryonic childhood neoplasm which was first described in eleven children by Manivel, et al. in 1988 [1]. The tumor is a dysembryonic neoplasm of thoraco-pulmonary mesenchyme, which arises from the lung, pleural surface, or from both. The exact histogenesis is unknown. Pleuropulmonary blastoma is classified into; type I (cystic), type II (cystic and solid) and type III (predominantly solid) lesions [2]. Herein, we report one case of infantile PPB, which presented to us with hemorrhagic pleural effusion; to our knowledge such a case has not been reported.

CASE HISTORY

An 8-month-old male infant was referred from a peripheral medical center with 10-day history of intermittent fever responding to antipyretic medications and cold with cough and 2-day history of labored breathing. On examination, infant had pallor, generalized lymphadenopathy and severe respiratory distress with cyanosis. Respiratory system examination revealed stony dull percussion note with decrease in air entry on the right side. Spleen and liver were moderately enlarged in size. Chest x-ray (Figure 1) revealed right sided consolidation with pleural effusion. In view of increasing respiratory distress, infant was intubated and mechanical ventilation was instituted at admission. Pleurocentesis showed hemorrhagic effusion, which on routine microscopy showed atypical cells with a suspicion of malignant cells. Computed tomography (CT) of the chest revealed a lobular heterogeneous mass in the posterior mediastinum encompassing the great vessels.

Tru cut biopsy was suggestive of round cell tumor, which raised suspicion of neuroblastoma, however, urinary vanillylmandelic acid (VMA) and homovanillic acid (HVA) were within normal and the bone marrow was negative for infiltrations. CT brain showed prominence of the third, lateral and fourth ventricles with gross acute hydrocephalus. Magnetic resonance imaging (MRI) of brain showed a cystic posterior 3rd ventricular tumor involving the tectum (Figure 2). Immunohistochemistry later revealed a mediastinal malignant, round cell tumor, positive for neuron specific enolase (NSE) antibody staining, chromogranin and focally positive for epithelial membrane antigen (EMA), synaptophysin, MIC 2 and Desmin. It was negative for creatinine kinase and myogenin; staining chanracteristics suggestive of pleuropulmonary blastoma. In view of the infant's deteriorating clinical condition, he was started on chemotherapy with vincristine, adriamycin and cyclophosphamide (VAC). The infant responded extremely well and was weaned off the ventilator within a week following the start of therapy. He received 2 cycles of chemotherapy and was then lost to clinical follow-up.

DISCUSSION

Pleuropulmonary blastoma (PPB) is an aggressive tumor accounting for less than 1% of

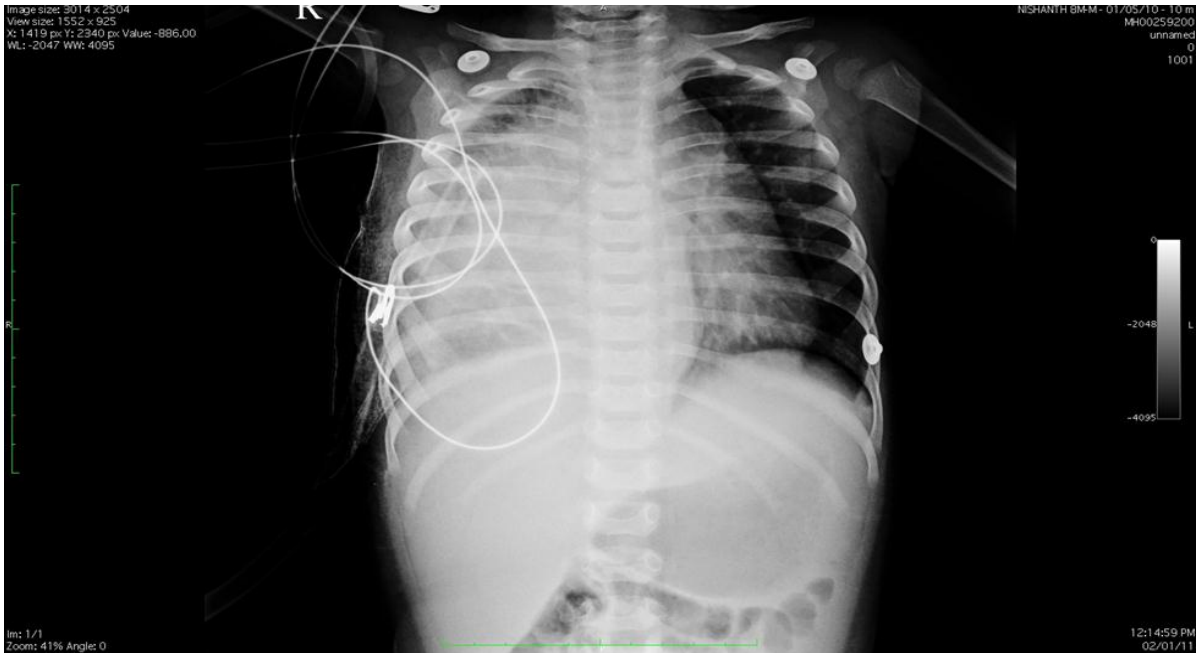

Figure 1: Figure shows the chest X-ray with right-sided pleural effusion, consolidation, and intercostal drainage tube in situ.

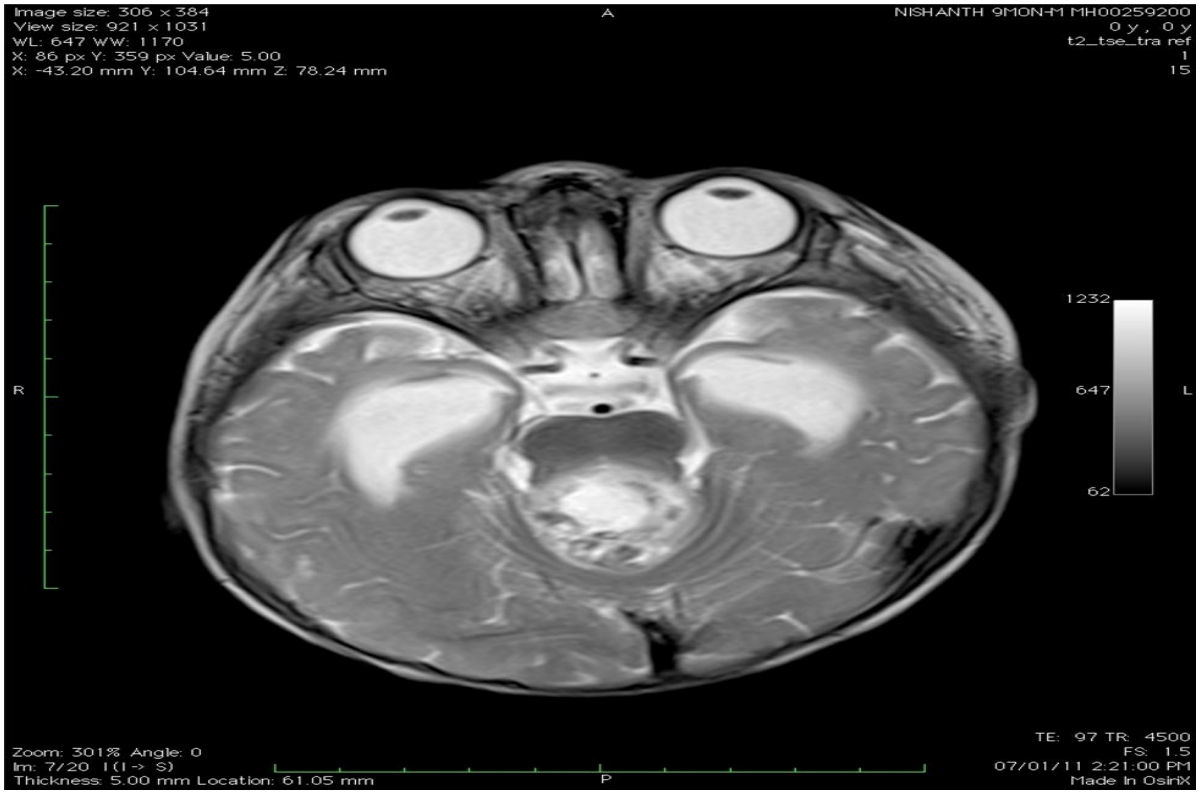

Figure 2: MRI of brain showing cystic tumor in the posterior fossa involving the tectum.

all primary malignant lung tumors in the pediatric population [3]. Manivel and associates coined the term PPB to describe a specific subtype of pulmonary blastoma on the basis of its exclusive clinical presentation in childhood and its pathologic features including variable anatomic location, primitive embryonic-like blastema and stroma, absence of a carcinomatous component, and potential for sarcomatous differentiation [2]. PPB is characterized histologically by primitive blastema and a malignant mesenchymal stroma and often shows multidirectional differentiation (rhabdomyosarcomatous, chondrosarcomatous, or liposarcomatous pattern).

Priest, et al. [4] studied the clinicopathological correlates between 50 patients with pleuropulmonary blastoma in which the most common presentation was respiratory distress, as was in our case. Other symptoms included fever, chest or abdominal pain, cough and malaise. Common sites for metastasis were central nervous system (11 patients), as seen in our case, orbit and iris. Other sites of metastasis included bones, adrenals, liver, kidneys, pancreas and rarely ovaries and lymph nodes. Two patients had recurrences in the contralateral chest.

Treatment for type I consists of surgery and possibly chemotherapy. For PPB types II and III, surgery, chemotherapy, and possibly radiation therapy is used. At present, about 50-60% of types II and III children are cured of the disease. Although several of the previously reported cases have received multi-agent chemotherapy, typically modeled after sarcoma treatment with vincristine, actinomycin, cyclophosphamide [5], the role of radiotherapy is more difficult to assess. Often, patients with great tumor bulk or with residual disease received radiotherapy with doses ranging from 4 to 55 Gy [5]. In a meta-analysis of 50 patients, reported by Priest and colleagues [4], 16 children received radiation therapy, the majority of whom had purely solid tumors. Total resection of the tumor at the time of diagnosis appears to be the preferred treatment followed by chemotherapy. Survival rates did not significantly differ among those treated with or without radiotherapy. Five year survival for type II and III lesions is approximately 42% after multimodality therapy [6]. Patients with pleural, mediastinal and extrapulmonary involvement have the worst prognosis. Total resection of the tumor at the time of diagnosis appears to be the preferred treatment followed by chemotherapy. Our patient received 2 cycles of VAC, and was thereafter lost to follow-up.

PPB is a highly aggressive tumor, which requires early and aggressive treatment. Complete surgical resection remains the primary goal of treatment in children with PPB. Subsequent chemotherapy and radiotherapy are also recommended by most researchers. Previous studies have shown that histopathological type, mediastinal or pleural involvement, and metastases were correlated to the outcomes, relapses are common and overall prognosis is bleak [7]. In summary, PPB is an extremely rare tumor, which presents most often with features that mimic acute respiratory infection with distress. Preoperative diagnosis is difficult, and often requires exploration and open biopsy to confirm the diagnosis. In spite of aggressive therapy, the overall prognosis remains guarded in these children.

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