Bilateral Spontaneous Hemothorax with PNET : a Case Report and Review

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Abstract:

Introduction:
Primitive neuroectodermal tumours (PNET) are malignant small cells neoplasm mainly occurring in children but can occur at any age. Extensive bleeding into the pleural cavity is rare in children. Hemothorax would be compatible with a variety of congenital anomalies including sequestration, patent ductus artreiosus, and pulmonary arteriovenous malformation.

Case Report:
A 2.5-year old girl who was known a case of PNET with dyspnea and tachypnea referred to our hospital. The chest X-ray revealed bilateral opacity. After consulting the general surgeon, the bilateral chest-tubes inserted and 200cc blood drained.

Discussion:
The diagnosis of a hemothorax can be made only by thoracentesis. The symptomatology of the peripheral primitive neuroectodermal tumors is related to size and their location.
We did not find any residue of cancer as a cause of hemothorax, and the hemothorax was not related to side effects of chemotherapy, infection and trauma. Therefore, PNET should be included as a cause of children hemothorax.

Keywords: Bilateral Spontaneous, Hemothorax, PNET.

Introduction

Peripheral primitive neuroectodermal tumors (pPNETs) are a group of highly cellular primitive round cell neoplasm's which occur extracranially in soft tissues and bones and are derived from embryonal neural crest cells. Askin tumours are malignant cell tumors of neuroepithelial origin appearing in the soft tissues of chest wall, occasionally in bone or rarely in the periphery of the lung (1).

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Case Report

A 2.5-year old girl who was known case of PNET referred to our hospital with dyspnea and tachypnea.

In physical examination, she had pallor appearance, pulse rate was 130/minutes and BP was 90/60 mm Hg. We decided to start chemotherapy regime. The chest X-ray revealed bilateral opacity (fig.1). After consulting with general surgeon, the bilateral chest-tubes were inserted and 200cc blood was drained. The hemoglobin was 4. 6gr/dl and platelet count 259000 cell/µl, PT: 12 and PTT: 25 sec.

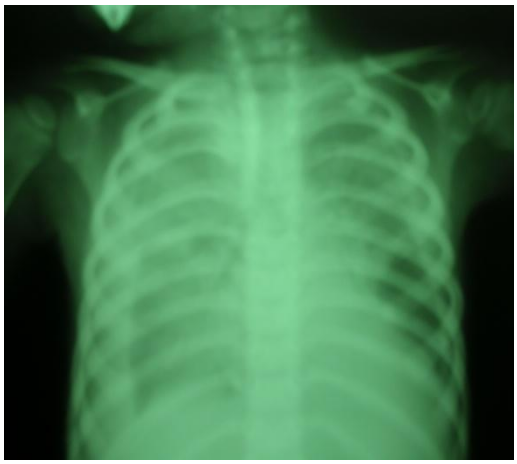

Fig 1: An AP view X- ray showing bilateral pleural effusion

In the plural tap, total cell count was 2110 cell/ml, neut:7, lymph:8, RBC:2095, LDH: 1974, protein:5/2gr/dl GLU:40mg/dl and concomitant BS was 65 mg/dl. Cytopathology of pleural fluid did not show malignant cells. Hemotherapy with packed Red Blood cells transfusion was done. She remained in hospital and received conservative management with broad spectrum antibiotic. After a few days the chest–tubes were clamped and then removed. Our patient discharged with good condition. The patient had been diagnosed 9 months ago with symptoms such as fever, anorexia, weight loss, gait disturbance and limping.

In Para-clinical study and Magnetic resonance imaging (MRI) of spinal cord and vertebra there was evidence of mass at postero lateral part of spinal canal which impressed cord at level T2 through T4 vertebral bodies (fig.2). The biopsy showed malignant round cell tumor compatible with primary nuroectodermal tumor (PNET) (fig.3) and immunohistochemistry was positive for mic2 (CD99) and chromo-granin A.

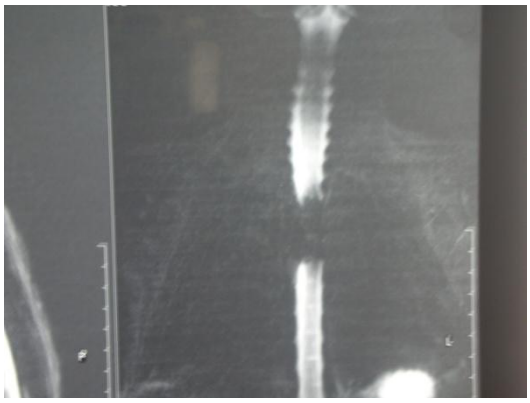

Fig 2: An MRI of spinal cord had evidence of mass at posterolateral

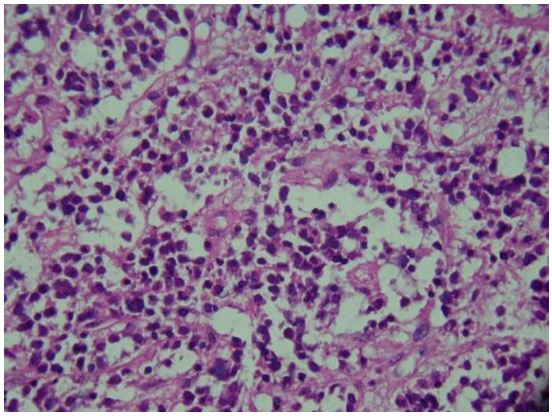

Fig 3: Pathology sample

Discussion

Ewing's family of tumors (EFTs) include Ewing's tumor of bone (ETB or Ewing's sarcoma of bone), extraosseous Ewing's (EOE), primitive neuroectodermal tumor (PNET or peripheral primitive neuroepithelioma), and Askin's tumor (PNET of the chest wall) (2). Studies using immunohistochemical markers,cytogenetics, molecular genetics, and tissue culture indicate that these tumors are derived

from the same primordial stem cell (3,4). Hemothorax is a well-recognized sequel of chest trauma, ruptured aortic aneurysm, inadvertent vessel damage, pulmonary infarct, intrapulmonary arteriovenous fistula, neoplasm, etc (1).

The diagnosis of a hemothorax can be made only by thoracentesis (5). Inadequate removal of blood in extensive hemothorax may lead to substantial restrictive disease secondary to organization of fibrin (5). Primitive neuroectodermal tumors (PNET) are malignant small cells neoplasm mainly occurring in children but can occur at any age (6).

PNETs have a common histological appearance of densely cellular masses of uniform small oval or round cells (7).

The symptomatology of the peripheral primitive neuroectodermal pumors is related to size and their location (1).

In our case, the location of the tumor was in vertebra (T2-T4) and symptoms were gait disturbance, limping, and fever. Drugs that were used for chemotherapy in our patient were vincristine, actinomycin-D, cyclophosphamide, adriamycin, and alternatively ifosfamide and etoposid. The adverse effect of hemothorax has not been reported in drug references (8,9) We did not find any residue of cancer as a cause of hemothorax, and the hemothorax was not related to side effects of chemotherapy, infection and trauma. Therefore, PNET should be included as a cause of children hemothorax.

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