Egyptian Journal of Bronchology

Vol 3, No 2, December, 2009

CASE REPORT

METASTATIC PERIPHERAL PRIMITIVE NEUROECTODERMAL TUMOR

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INTRODUCTION

A 22 years old single male patient living in Embaba, Giza with no special habits of medical importance presented to our outpatient clinic complaining of right sided pleuritic chest pain of one month duration together with progressive exertional dyspnea on moderate exertion, low grade fever all over the day, loss of appetite and loss of weight. On presentation the patient was under weight, febrile (37.8^{o}C), and local chest examination revealed diminished movement, tactile vocal fremitus (TVF) and intensity of breath sounds as well as stony dullness on right mammary, inframammary, lower axillary, scapular and infrascapular areas with shift of the trachea to the left side. The patient was admitted to our department where chest x-ray was done (Fig. 1) revealing picture suggestive of right sided pleural effusion. Tuberculin test was negative and routine laboratory investigations were normal except for elevated erythrocyte sedimentation rate (95-1st hour). Thoracocentesis and Abrams closed pleural biopsy were performed in which one liter of serosanginous fluid was aspirated. Results revealed exudative pleural effusion with few mature lymphocytes along with fibrotic pleura and no evidence of malignancy. The patient was advised to undergo diagnostic thoracoscopy, but he refused and was discharged upon his own will.

Three months later, the patient was readmitted after progression of his of right sided chest pain which became severe enough to interfere with his daily activity as well as his sleep rhythm and was not responding to analgesics. Dyspnea progressed to be on mild exertion. On presentation the patient was febrile (38.5^{o}C) and abdominal examination revealed hepatomegaly (lower border felt four fingers below costal margin, firm in consistency with rounded border and irregular surface). A localized, oval shaped swelling about 5×7cm, over the 10th, 11th, 12th ribs at the mid axillary line, firm, fixed to the underlying tissue. The skin overlying was normal and does not give expansible impulse on cough. There were diminished movement, TVF and intensity of breath sounds as well as stony dullness on the right inframammary, lower axillary & infrascapular areas while the trachea became central. New chest x-ray was done revealing picture suggestive of right sided pleural effusion similar to previous study. CT scan chest (Fig. 2) was done revealing right sided pleural effusion along with area of consolidation at the right lower basal area in addition to huge abdominal mass seen elevating the right copula of the diaphragm. Pelviabdominal CT Scan (Fig. 3) showed abdominal mass (heterogenous pattern cystic & solid), elevating the right copula of diaphragm and destructing the last rib with shift of the liver and right kidney along with paraaortic lymph node.

CT-scan guided core biopsy from abdominal mass revealed small rounded cell tumor-Primitive Neuroectodermal tumor.

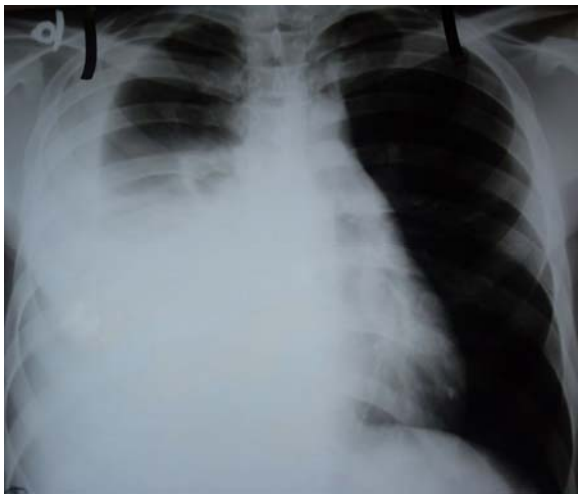

Fig 1. Chest x-ray on admission showing right sided pleural effusion.

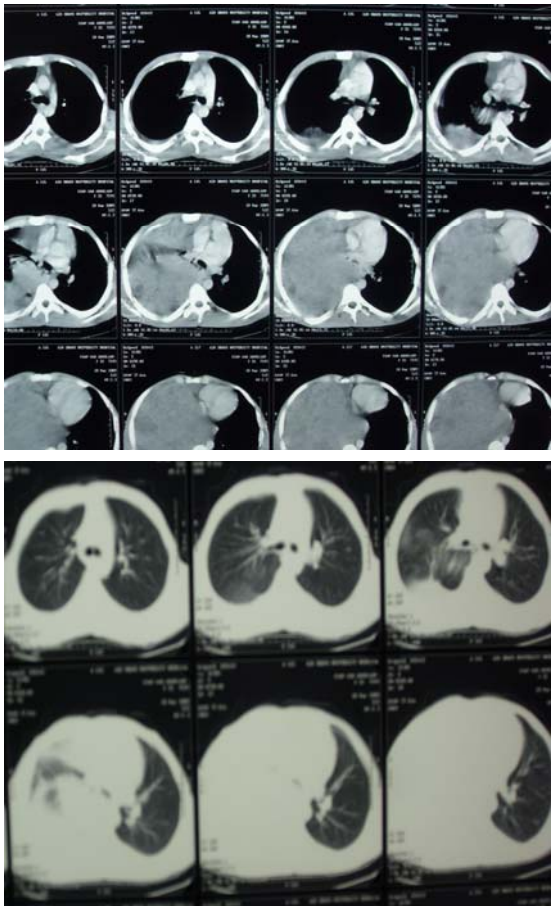

Fig 2. CT scan chest showing right sided pleural effusion along with area of consolidation at the right lower basal area in addition to huge abdominal mass seen elevating the right copula of the diaphragm.

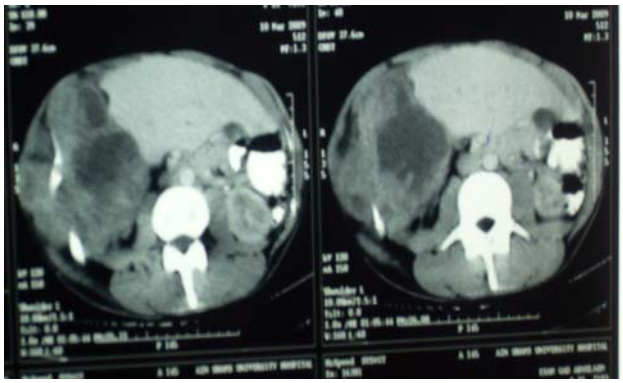

Fig 3. Pelviabdominal CT Scan showing abdominal mass destructing the last rib with shift of the liver and right kidney along with paraaortic lymph node.

Final diagnosis: Peripheral primitive neuroectodermal tumor arising from the retroperitoneal space and extending to the abdomen destructing the right last rib with right sided metastatic malignant pleural effusion.

Neuroectodermal Tumor:

Introduction: The Peripheral primitive neuroectodermal tumors (PPNET) are rare but highly aggressive tumor of the peripheral non-central nervous system tissue. The overall incidence is 1% of all sarcomas. Thoracopulmonary region is the most common site; however they have been described in other sites as well e.g. the retroperitoneal paravertebral soft tissues, the soft tissues of the head and neck and the intraabdominal and intrapelvic soft tissues & extremities.

Historical Background: PPNETs were 1st reported by Stout in 1918. James Ewing 1st described Ewing sarcoma in 1921 after observing radiosensitivity in a subgroup of bone tumors. In the early 1980s, Ewing sarcoma & PPNETs were both found to contain the same reciprocal translocation between chromosomes 11 & 22. These tumors were categorized as the Ewing sarcoma family of tumors because of the shared translocation & the similar cellular physiology.

Epidemiology: The annual incidence of Ewing sarcoma family tumors from birth to age 20 years in USA is 2.9 cases/million population. Approximately 10% of patients are aged 20-30 years. The survival of patients highly depends on the initial manifestation of the disease. Approximately 80% of patients present with localized disease, whereas 20% present with clinically detectable metastatic disease, most often to the lungs, bone, and/or bone marrow. The overall survival rate is 60%; however, for patients with localized disease, the survival rate approaches 70%. Patients with metastatic disease have a

long-term survival rate < 25%. The incidence of Ewing sarcoma family tumors in male individuals is more than in female individuals being 3.3 & 2.6 cases/million population, respectively. Incidence peaks in the late teenage years. Overall, 27% of cases occur in the 1st decade of life, 64% of cases occur in the 2nd decade of life, and 9% of cases occur in the 3rd decade of life. The incidence in whites is at least 9 times higher than that in blacks.

Pathophysiology: Tumors in the Ewing sarcoma family are thought to derive from cells of the neural crest, possibly postganglionic cholinergic neurons. The exact cell of origin of the Ewing sarcoma family of tumors is unknown.

Classification: The Ewing sarcoma family of tumors includes Ewing sarcoma, peripheral primitive neuroectodermal tumor, neuroepithelioma, atypical Ewing sarcoma, & Askin tumor (tumor of the chest wall). The tumors in the Ewing sarcoma family are treated similarly on the basis of their clinical presentation (e.g. metastatic or localized) rather than their histologic subtype.

Staging: Staging includes both local imaging to reveal the full extent of tumor prior to therapy & evaluation for distant metastases. Local imaging usually includes both MRI & CT scanning. The evaluation for metastases should include bilateral bone marrow biopsies, chest CT scanning and radionuclide total body scanning, such as technetium-99 & FDG-PET scanning.

Diagnosis:

- ***Imaging studies:*** Evaluation of the 1ry lesion e.g. CT scan, MRI & skeletal imaging.

  Evaluation of metastasis e.g. CT scan, U/S & PET.

- ***Procedures:***

  Biopsy.

  Cytogenetic & molecular studies.

Prognosis: The only significant factor that determines the prognosis is the presence or absence of metastatic disease.

Treatment:

- ***Chemotherapy:***

  It is part of the treatment for all patients. It is usually given 1st to shrink the tumor before treatment with surgery or radiation therapy.

- ***Surgery:***

  It is usually done to remove cancer that is left after chemotherapy or radiation therapy.

- ***Radiation therapy:***

  Radiation therapy may be used to shrink the tumor before surgery. It may also be used to kill tumor cells that are left after surgery or chemotherapy.