ORIGINAL RESEARCH PAPER

Pulmonary Medicine

A CASE OF PRIMITIVE NEUROECTODERMAL TUMOR (PNET) PRESENTING AS ANTERIOR MEDIASTINAL MASS

KEY WORDS:
mediastinal mass, tumor, neuroectodermal tumor, soft tissue tumor

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ABSTRACT

Peripheral primitive neuroectodermal tumors (pPNET) are a group of highly malignant tumors composed of small round cells of neuroetodermal origin that affect soft tissue and bone ,most commonly present in the thoracopulmonary region (Askin tumor), abdomen , pelvis and rarely in the head and neck. They usually present in the second decade of life, with slight male preponderance and account for 4-17% of all soft tissue tumors. Clinical symptoms depend on the site of presentation but invariably include pain and swelling of surrounding structure due to mass effect. Current recommendations advocate complete surgical resection whenever possible, adjuvant or neoadjuvant chemotherapy and radiotherapy. Here we present a 20 year old male patient who presented with chest pain and breathlessness and on investigation found to have an anterior mediastinal mass which further evaluation found to be a primitive neuroectodermal tumor.

INTRODUCTION

A 20 year old male patient presented with complaints of breathlessness on exertion, left sided chest pain since 2-3 days with no significant past medical or surgical illness.

General examination was normal. Respiratory system examination revealed fullness over left side with reduced breath sounds.

Radiology showed presence of anterior mediastinal mass lesion. Serological investigations done to rule out germ cell tumors were within normal limits.There were no enlarged peripheral lymph nodes.

CT guided biopsy of the lesion proved to be pNET after pathological and IHC examination. Patient received 2 cycles of combination chemotherapy with Etoposide and Ifosfamide and is planned for restaging for further management.

Investigations:

COMPLETE BLOOD COUNTS : Normal

LIVER FUNCTION TESTS : Normal

RENAL FUNCION TEST : Normal

Beta HCG: 45mU/ml

AFP: 1.39 ng/ml (0-8.0)

T3: 77.49 ng/dl (60-181)

T4: 14.70 mcg/dl (3.2-12.6)

TSH : 0.81 (0.55-4.78)

Sr LDH: 412 IU/L

USG THORAX:

Mass lesion seen over left anterior and lateral chest wall of varying echotexture with areas of necrosis seen within it.

USG ABDOMEN: within normal limits

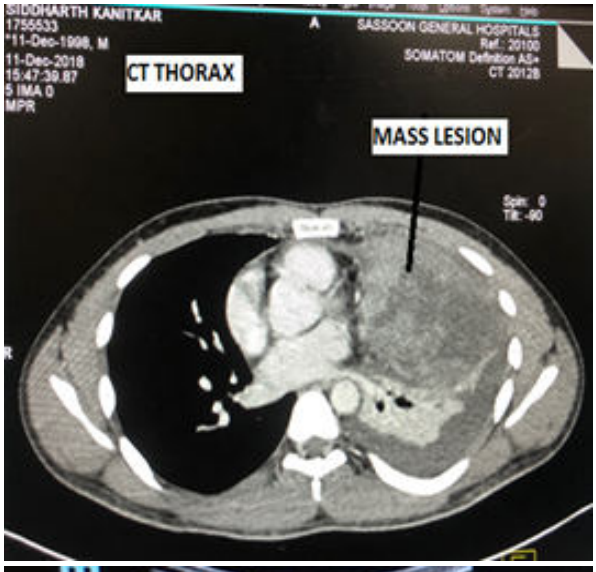
CT CHEST: well defined 7.9x8.6x9.2 cm heterogenously enhancing soft tissue lesion involving anterior mediastinum extending to left para cardiac location and left hemithorax along the anterior chest wall.

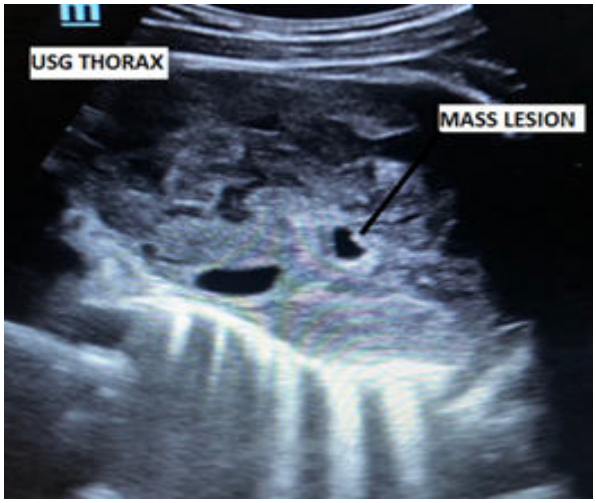
HPE: cores of fibro-connective tissue infiltrated by tumor composed of small round cells arranged in sheets and nests – Suggestive of malignant round cell tumor.

IHC: CK, CD99, FLI1 - Positive.

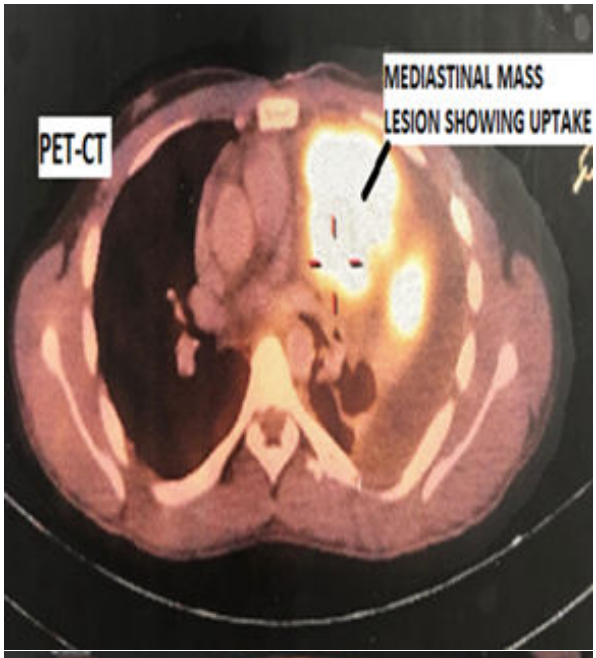
TTF1, PLAP, CD30, TdT, Desmin, S100, Vimentin – Negative -Suggestive of primitive neuroectodermal tumor (PNET)

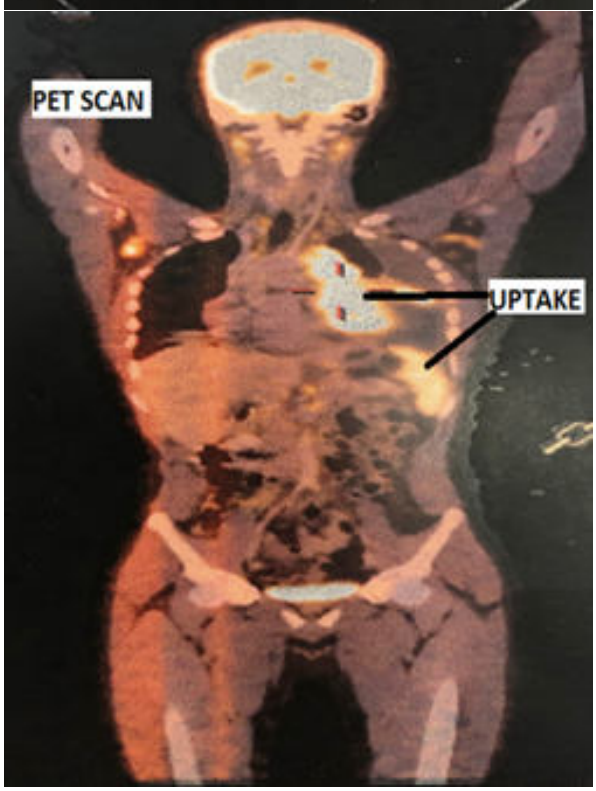
PET-CT: Metabolically active anterior mediastinal mass lesion measuring 110x88x116mm. Splenomegaly with increased metabolic activity. Diffuse metabolic activity involving axial skeleton.



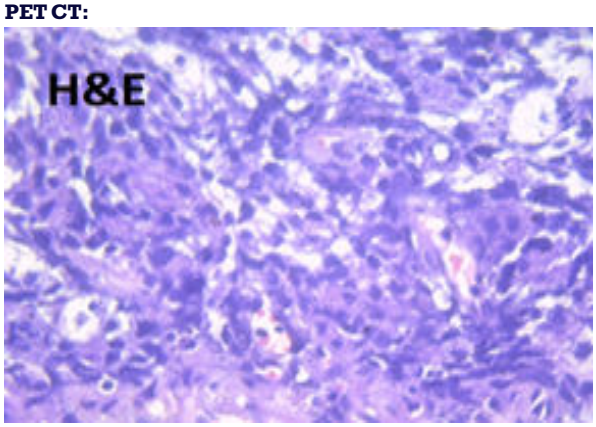


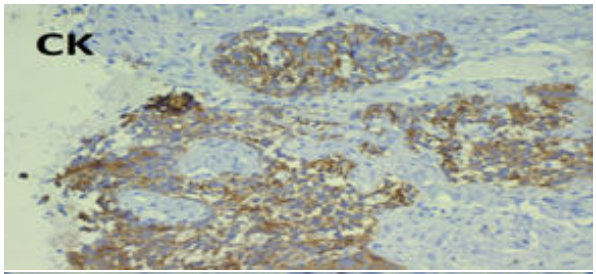
CT AND USG THORAX:

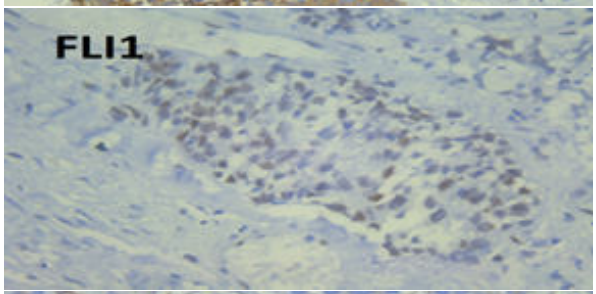


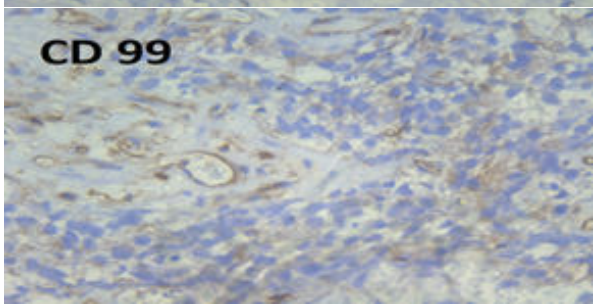


PET CT:









HISTOPATH AND IMMUNOHISTOCHEMISTRY MARKERS:

DISCUSSION:

PNET is a rare (1.8 % of all mediastinal tumours), highly malignant undifferentiated soft tissue sarcoma . It is believed to arise from embryonal cells migrating from the neural crest. Commonly seen in young females in second decade of life.

Thoracic involvement of PNET is characteristically painful due to tumour invasion into chest wall or mediastinum.

IHC plays important role in making a diagnosis. CK positivity suggestive of epithelial tumor and FLi1 and CD 99 positive suggestive of PNET although not specific.

Thoraco-pulmonary PNET is a highly aggressive neoplasm with a mean survival of eight months. Prognosis is poor despite multidisciplinary modalities of treatment, which includes neoadjuvant chemotherapy, localized radiotherapy and systemic chemotherapy. The overall 2 year survival is only 38% . But in case of localized disease, 5 year survival rate of 60% has been described.

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