Abstract:
Peripheral Primitive neuroectodermal tumor (pPNET) is very rare tumor in elderly age group. We report a case of 60 year old female who presented with right mid thigh swelling, diagnosed as pPNET by histopathological, immunohistochemistry and molecular methods. The case has been reported for its rarity and to highlight the importance of modern techniques in diagnosing unusual cases, which may have an impact on disease outcome.

Introduction:
Peripheral Primitive neuroectodermal tumor (pPNET) is Ewing’s family of tumors (EFT) (1). PNET is a malignancy of children, adolescents and young adults (2). PNET are malignant round cell tumor and unlike Ewing’s sarcoma, predominantly arises from soft tissue compared to bones. PNET is also associated with reciprocal translocation- EWS-FLI1, t(11;22)(q24;q12) (3). In young age nonmetastatic PNET, outcome is good with multimodality of treatment (4). Outcome of EFT in elderly patients is not known due to paucity of data. PNET is very rare in above 50 years population (5,6). We report a rare case of right thigh-extra osseous PNET in elderly female.

Case report:
A previously healthy 60 year old female, asthmatic was admitted to hospital with 6 months history of swelling of middle third of right thigh medial aspect. Patient was seen by a local general surgeon after 2 months of onset of swelling and underwent unplanned
excision presuming it to be of benign aetiology. Histopathological examination of excised mass was not done. After 1 month of excision, swelling reappeared and increased rapidly in size. At the time of presentation in our hospital, she had 11x8 cms swelling over medial aspect of right thigh; rest of the examination was unremarkable. Magnetic resonance imaging showed soft tissue mass in mid medial compartment of right thigh abutting and superficial to vastus medialis muscle. Histopathological examination of biopsy of this soft tissue mass showed malignant round cell tumor (Figure.1) and immunohistochemistry was suggestive of PNET [CD 99 positive (figure.1), LCA negative, Keratin negative, Vimentin negative, Synaptophysin negative, Chromogranin negative, Ki 67 – 40%]. EWS-FLI 1(RT-PCR) was positive. On staging evaluation, bone scan and bone marrow study were normal and high resolution CT chest showed 2 metastatic peripheral nodules on right side of chest and 1 metastatic nodule on left side. Patient was diagnosed as extra osseous metastatic pPNET of right thigh. Patient underwent wide local excision and split skin grafting and histopathological report showed multinodular PNET measuring 9x6x6 cms. Patient has been initiated on vincristin, adriamycin, cyclophosphamide based chemotherapy. She shall be reassessed for pulmonary nodules after 4-6 cycles of chemotherapy.

**Discussion:**
pPNET is very rare in elderly patients and very few case reports of pPNET in this age group are published (7,8). In this age group, these tumors may be confused with other soft tissue sarcomas. Other soft tissue sarcomas are chemoresistant unlike PNET and hence aggressive workup should be initiated upfront to discriminate such tumors as it may affect the ultimate outcome. However prognosis of this disease in elderly population is not clearly known due to scarcity of published data (9). Two studies (5,6) have demonstrated that age is a poor prognostic factor, but two other studies (10,11) have not shown the same. Age is also a concern for delivering optimal chemotherapy. The effects and adverse events of chemotherapy in elderly age group are not clearly defined because of their limited accrual in trials. The dosage of chemotherapy should be individualized according to patient’s tolerability. Stout first described PNETs in 1918, and these tumors were thought to arise directly from nerves (12). The pathologic and cytogenetic understanding of these tumors has significantly advanced over the last 25 years. Histopathologically, PNETs are undifferentiated, small, round-cell tumors with hyperchromatic nuclei and features of neural differentiation, which typically form Homer Wright rosettes. On immunohistochemical examination, the most useful antibody for the diagnosis of PNET is the monoclonal antibody CD99, directed against the cell surface protein MIC2. PNETs often have the same reciprocal chromosomal translocation, i.e. t(11;22)(q24;q12), which is the other key to the diagnosis of PNET (13). The case highlights the significance of detailed histopathological examination, immunohistochemistry and molecular studies in diagnosing “childhood” cancer in geriatric population.

In conclusion, pPNET is a rare malignancy in old age, but can be diagnosed with available new techniques. Such unusual diagnosis changes line of treatment and outcome also.

**References:**
1 Iwamoto Y. Diagnosis and Treatment of Ewing’s Sarcoma. Jpn J Clin Oncol. 2007 Feb 1;37(2):79–89.


