Case Report

Primary extraskeletal peripheral primitive neuroectodermal tumor of subcutaneous tissue neck in a young adult: A rare case report

Sumeet Aggarwal, Vivek Kaushal, Abhishek Soni, Sujata Singla[1]

Departments of Radiation Oncology and [1]Surgery, Pt. B D Sharma Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India

ABSTRACT

Primitive neuroectodermal tumors (PNETs) are poorly differentiated small round cell neoplasms which primarily affect childhood age and very rarely seen in adults. Peripheral PNET (pPNET) cases are very rare compared to central PNET, and most of them originate from neural crest cells located outside the central nervous system. We report a case of large extraosseous pPNET arising from subcutaneous tissue of left side neck in a young male patient. Despite aggressive inherent nature of histology, structural complexity of neck area and poor predictors like tumor size >6 cm, extraosseous nature, this case advocate that complete resolution is possible with aggressive multimodal treatment including surgery, radiation therapy, and chemotherapy. Other major concerns in such cases of pPNET are difficulty in diagnosis due to low incidence, unpredictable site involvement, histological similarity with other round cell tumors and lack of established treatment guidelines.

Key words: Chemotherapy, head and neck cancer, primitive neuroectodermal tumor, radiotherapy

INTRODUCTION

Primitive neuroectodermal tumors (PNETs) are rare small blue round cell tumors, first illustrated by Stout in 1918.[1] "Primitive neuroectodermal tumor" term was coined in 1973 by Hart and Earle.[2] Most of the patients present in first or second decades of life. Peripheral PNET (pPNET) are mostly aggressive and malignant in nature. There is a relative paucity of established guidelines and literature pertaining to outcome of extraosseous pPNET for an adult.

CASE REPORT

A 28-year-old young male patient presented with a complaint of swelling in the left side of the neck for 2 months in our outpatient department. On clinical examination, there was a nontender, firm swelling over left neck fixed to underlying structures. No other significant complaints or comorbidities were noted. The patient was nonsmoker and nonalcoholic, and no relevant family history was noted. The patient was further investigated to rule out tuberculosis, lymphoma, and primary in the head and neck region. Ultrasonography of neck was suggestive of large, well-defined hypoechoic mass at cervical lymph node level II/III/IV. Contrast enhanced computed tomography scan of neck suggested large, well-defined partially necrotic mass lateral to carotid sheath and medial to sternocleidomastoid muscle with maintained fat planes [Figure 1a-c]. Fine needle aspiration cytology from cervical swelling demonstrated a tumor composed of sheets of uniform small cells and atypical mitotic figures along with necrosis suggesting small round cell tumor. Immunohistochemistry (IHC) was positive for CD99 and synaptophysin and negative for

Address for correspondence: Dr. Sumeet Aggarwal, Department of Radiation Oncology, Pt. B D Sharma Post Graduate Institute of Medical Sciences, Rohtak - 124 001, Haryana, India.
E-mail: s.aggarwal43@gmail.com

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chromogranin, vimentin, leukocyte common antigen, and HMB-45. The patient was planned for radical excision of mass and subsequently left modified radical neck dissection was done. Histopathology confirmed the diagnosis of PNET with IHC positivity for CD99 [Figures 2 and 3]. All the lymph nodes excised were uninvolved by tumor and were reactive. The patient was further given adjuvant radiotherapy with 50.4 Gy in 28 fractions, 5 days a week on telecobalt machine. Whole body fluorodeoxyglucose positron emission tomography-computed tomography (^{18}F-FDG PET-CT) after 6 months of treatment suggested no evidence of any FDG avid at local site in neck or elsewhere in the body [Figure 4].

DISCUSSION

PNET is a broad group that includes poorly differentiated or undifferentiated round cell tumors and has morphological attributes of neuroectodermal origin.[3] All ages are affected but most of the patients presenting in either first/second decades of life. PNET can be further differentiated as pPNET and central PNET (cPNET). cPNETs develop from a precursor cells present in the external granular layer of the cerebellum/precursor cell of the subependymal matrix of the central nervous system, pinealocytes and the subependymal cells of the ventricular system, whereas pPNET occurs frequently in the thorax as Askin tumor, urogenital tract and very occasionally in the head and neck.[4-6] Most often these are grouped together for both prognostic and treatment factor analysis. pPNET comprises about <1% of all sarcomas, and are mostly aggressive and malignant in nature.[7] Radiological Imaging is essential for making diagnosis and then planning surgical excision of the target lesion. On CT, PNET mostly appears as hyperdense lesion. On magnetic resonance imaging, it appears hypo-to-hyperintense on T2-weighted sequences and iso-to-hypointense on noncontrast T1-weighted sequences and enhances with gadolinium.[8] FDG-PET scan

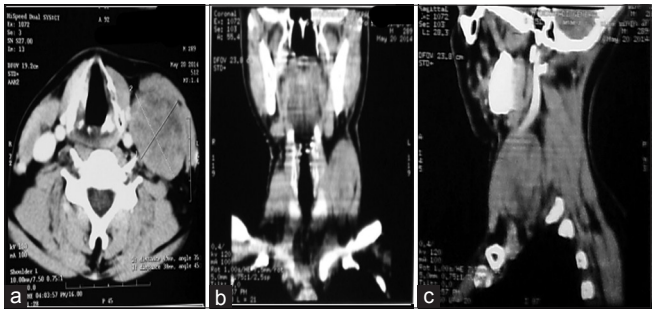

Figure 1: Computed tomography images in transverse (a), coronal (b) and sagittal (c) section showing well defined mass in left mid and lower deep cervical area measuring 66 mm (craniocaudal) by 43 mm (transverse) by 36 mm (anteroposterior) lateral to carotid sheath and medial to sternocleidomastoid muscle

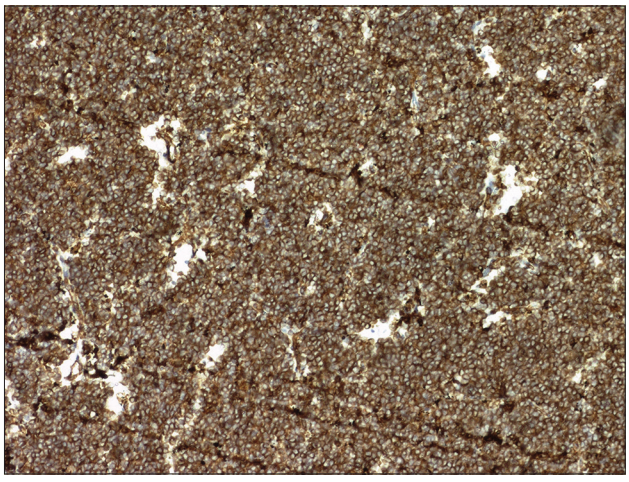

Figure 3: CD 99 diffuse, uniform positivity in small round cells (IHC, ×100)

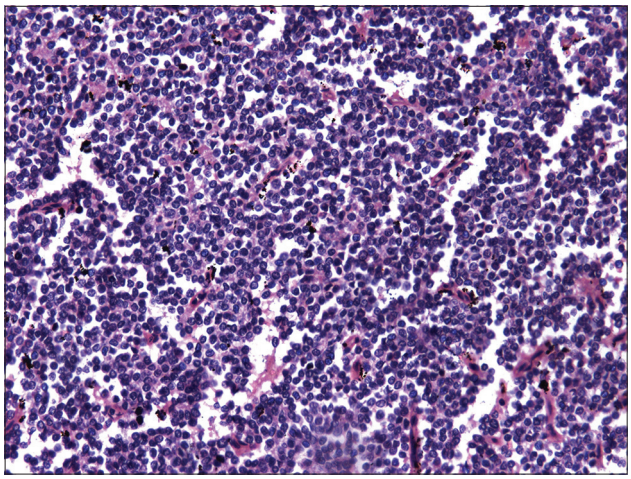

Figure 2: Solid sheets of uniform small round cells along with well-developed vascular network and intervening fibrous bands (H and E, ×200)

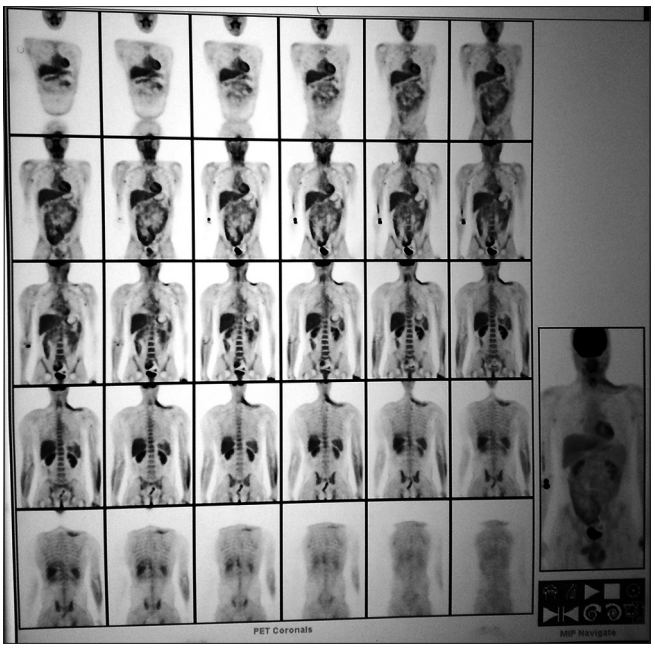

Figure 4: Whole body fluorodeoxyglucose positron emission tomography computed tomography suggested no evidence of any fluorodeoxyglucose avid residual disease

or bone scan optionally can be used for further systemic staging. Bone marrow biopsy should be performed as a staging protocol if bony involvement is suspected, as bone marrow involvement presage a worse prognosis. IHC plays a path-breaking role in establishing the diagnosis of PNET. Tumor cells often show positivity for MIC-2 (CD99) and vimentin and negativity for desmin and CK (cytokeratin). Local treatment usually consists of surgery alone, radiation therapy alone or combined modality therapy including surgery plus radiation therapy. PNET has a marked propensity for systemic spread, and therefore intensive multiagent chemotherapy is also recommended when indicated. Curative treatment for both the localized cPNETs and pPNETs involves adequate surgical wide resection/amputation/maximum debulking of tumor or resection plus postoperative radiotherapy with dose of 45 Gy or radical/definitive radiotherapy with 60 Gy at least (45 Gy to gross disease and further boost sparing organ at risk). Although definitive radiotherapy has comparable results as surgery in relapse-free survival and overall survival, the use of various altered fractionation such as hypofractionation, hyperfractionation, and hyperfractionated split-course irradiation schedules shows no benefit in terms of survival rates.[9] Significant adverse predictors for survival in various studies include metastatic disease at presentation, primary extraosseous tumor and older age (>26 years) and most favorable factors include localized disease at presentation, primary tumor origin in bone, size of the primary <6 cm, and response to chemotherapy. No study directly compared the outcomes for adults and children. Among many hypotheses for the fact that older patients with PNET have survival rates inferior to those of younger counterparts, most common are the difference in the level of differentiation, gene expression, biological differences due to age and low bone marrow reservoir with advancing age. Due to rare incidence of pPNET, decision of optimal treatment is exigent, particularly if the tumor site is in head and neck area. Currently, the most accepted strategy is multimodal treatment including surgery with adjuvant radiotherapy.[10] Structural complexity of tumor in head and neck region accounts for difficulty in complete excision. Multidisciplinary cooperation between on surgeons, medical oncologists, and radiation oncologists can improve outcome and survival rates of such rare diseases.

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Conflicts of interest
There are no conflicts of interest.

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