

Case Report

A Rare Case of Dumbbell-shaped Primary Intraspinal Peripheral Primitive Neuroectodermal Tumor Involving Thoracic Spinal Epidural Space

Abstract

Primary intraspinal primitive neuroectodermal tumor (PNET) is a type of round cell malignant tumor which is reported only above 100 in literature. We report a case of epidural thoracic peripheral PNET, discuss its pathological features, radiology, and treatment options.

Keywords: *CD99, EWS-FLI1 translocation, spinal epidural peripheral primitive neuroectodermal tumor, thoracic compressive myelopathy*

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Introduction

Malignant small round cell tumors are characterized by small, round, relatively undifferentiated cells in histopathology. These are also called small round blue cell tumors as the cells are blue, in the sense that they have large hyperchromatic nuclei and a thin rim of cytoplasm. Tumors that belong to this group are: desmoplastic small-round-cell tumor, Ewing's Sarcoma (ES)/primitive neuroectodermal tumors (PNET), neuroblastoma, medulloblastoma, rhabdomyosarcoma, synovial sarcoma, carcinoid tumor, mesothelioma, hybrid oncocytoma/chromophobe renal cell carcinoma, leiomyosarcoma, small cell lung cancer, Wilms' tumor, retinoblastoma, Small-cell lymphoma, hepatoblastoma, and Merkel cell carcinoma.^[1] Differential diagnosis of small-round-cell tumors is particularly difficult due to their undifferentiated or primitive character from their morphology alone. Therefore, a multimodal approach using fine-needle aspiration cytology, immunohistochemistry (IHC) and immunophenotyping by flow cytometry, reverse transcriptase polymerase chain reaction, fluorescence *in situ* hybridization, and electron microscopy are employed.

PNET is a type of round cell tumor, which is of neuroectodermal origin but has poor differentiation. It is classified into two types as follows: peripheral and central nervous system (CNS) PNET. Peripheral PNET (p-PNET) belong to Ewing's family of

tumor, since they have similar histological and immunohistochemical characteristics.

p-PNET sarcoma predominates in the second decade of life, and the pelvis and femur are most commonly affected sites. The term, "PNET" includes malignant small-round-cell tumors of the thoracopulmonary region (Askin's tumor), extraskelatal ES, peripheral neuroblastoma, and peripheral neuroepithelioma.

Primary spinal epidural p-PNET is rarely reported in the literature.

Case Report

An 18-year-old, healthy girl presented with the complaints of gradually progressive backache along with weakness of both lower limbs of 3 weeks duration. The progression of paraparesis to paraplegia was agile, over about a day. At the time of presentation, she was paraplegic, had absent sensation below D11 level bilaterally, bilateral plantar extensor, and bladder catheterized. Her magnetic resonance imaging (Siemens, Magnetom Essenza, Erlangen, Germany) thoracic spine showed lesion in anterior epidural space at the level of T9, 10 and 11 vertebral bodies, causing spinal canal stenosis, and compression of adjacent spinal cord [Figure 1a and b]. The lesion was extending into left T9–10 neural foramina [Figure 1c] and adjacent left prevertebral space. The lesion appeared heterogeneously hypointense on T2-weighted (T2W) fast spin echo (SE) [Figure 1b] and isointense on T1W SE

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Figure 1: (a) – T1 sagittal, (b) – sagittal T2 weighted fast spin echo, (c) - T1 axial showing extension into T9-T10 neural foramen, (d) - postcontrast sagittal T1

images [Figure 1a] to normal lumbar spinal cord with minimal heterogeneous contrast enhancement on contrast T1 image [Figure 1d].

She underwent D9, D10, and D11 left-sided hemilaminectomy and gross total excision of the lesion. Intraoperatively, a whitish, soft elastic, moderately vascular with necrotic areas in between extradural tumor compressing the spinal cord on the left side found. Tumor was found to be involving both D9, and D10 left intervertebral foramina. Tumor was dumbbell-shaped.

Tumor was removed completely in a piecemeal fashion, and there was multiple gray-brown soft tissue within on the left side of thecal sac which was extending along the T9-T10 foramen paraspinally. There was a severe mass effect on the cord. The lesion was dissected meticulously from the dura and nerve roots to achieve a gross total resection.

Histopathological examination showed cellular small round cell tumor arranged in sheets, rosettes and focally in the perivascular pattern. Cells had high nuclear-cytoplasmic ratio (N/C), round and hyperchromatic nuclei and exhibited brisk mitosis with large areas of necrosis. IHC was negative for myeloperoxidase and synaptophysin and positive for CD (Cluster of differentiation) 99. MIB-1 (cell proliferation marker) labeling indices were high. Findings were suggestive of PNET.

She was then referred to a radiation oncologist for further management. After 1 month of the review, she had only marginal improvement in symptoms.

Discussion

PNET is a small-round-cell malignancy arising in soft tissue and bone, predominantly in older children and adolescents with a male preponderance. They can arise either from central nervous system (cns- PNET) or from periphery (p-PNET). p-PNET has the histopathological characteristic

similar to ES. Osseous ES, extraskeletal ES, pPNET are nowadays generally known as ES family tumors.

p-PNET are tumors of adolescents, mostly in male in the second decade. They most commonly involve chest wall (askins tumor) pleura, pericardium, and soft tissues.

The incidence of these tumors in the spinal cord is rare.^[2] Review of previous cases reported in the literature suggests that p-PNET may arise from all levels of the spine and can be intradural-intramedullary or extramedullary (most common site being cauda equina), or extradural. Literature also suggests intradural extramedullary and intramedullary location to be almost equal in frequency.^[3] Although p-PNET/ES mostly arises from peripheral soft tissues and bone, they have also been reported from CNS. The tumor is most frequently located at lower spinal levels, in lumbar and lumbosacral regions. Their occurrence in thoracic spinal epidural space is very rarely reported in the literature.

On histopathology, the cytoplasm is scanty, eosinophilic, and usually contains glycogen, which is detected by periodic acid-Schiff stain and is diastase degradable. The nuclei are round, with finely dispersed chromatin, and one or more tiny nucleoli.

Depending on differentiation, tumor cells may also express neuroendocrine proteins (synaptophysin and chromogranin A), neural crest-derived protein (S 100), neuron-specific enolase. The mesenchymal markers such as intermediate filaments (cytokeratin, vimentin, neurofilament, desmin, and glial fibrillary acid protein) though nonspecific can be positive. These tumors share the chromosomal translocation, t(11:22)(q24:q12). This genetic anomaly leads to the creation of a fusion protein consisting of EWS and FLI-1 gene products. The FLI-1 protein, the gene product of FLI-1, t(11:22), is positive in 85% of all EWS/PNET cases.

The microneme protein 2 (MIC2) gene is a pseudoautosomal gene, located on the short arms of the sex chromosomes. ES and pPNET cells express glycoprotein CD99 in very high amounts and a highly selective manner, which help to differentiate from other malignant round cell tumor.^[3]

Surgery followed by craniospinal irradiation and chemotherapy with cyclophosphamide or ifosfamide, cisplatin or carboplatin, and vincristinebleomycin have shown to benefit.^[4] However, despite all treatment mean survival rate is around 3 years.

A very high index of suspicion with the good immunohistochemical analysis is required for the diagnosis of primary intraspinal p-PNET. Awareness about this condition is necessary for early diagnosis and appropriate management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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