Primary dorsal spine primitive neuroectodermal tumor in an adult patient: Case report and literature review

ABSTRACT
Primary spinal primitive neuroectodermal tumor (psPNET) is a rare entity with few cases reported in literature. We report a case of a 50-year-old female who presented to us with paraplegia and was diagnosed with extradural dorsal spine psPNET. The diagnosis was not suspected at presentation or on radiology but was established on histopathological examination. It is important to distinguish it from central nervous system primitive neuroectodermal tumors and from other spinal tumors since it follows a different clinical course and therapeutic outcome.

Keywords: Adult, dorsal spine, extradural, primitive neuroectodermal tumor

INTRODUCTION
Primitive neuroectodermal tumors (PNET) are rare tumors that arise from undifferentiated matrix or germinal cells.[1,2] Annual incidence of these tumors is around 0.2–0.4/100,000.[1,3,4] It is a highly malignant and invasive tumor with poor prognosis usually occurring in children and young adults.[1-4] Primary spinal PNETs (psPNET) are even rarer, limited to only a few case reports, especially in patients above 50 years of age.[1] We present a case of a 50-year-old female who presented to us with rapidly progressive paraparesis without bladder bowel involvement. Extensive search of English literature has revealed that our patient is only the second case of dorsal spine extradural tumor in patients aged 50 years or above. We also review the literature on the subject and discuss distinguishing features between central nervous system (CNS) PNET and psPNET.

CASE REPORT
A 50-year-old female patient presented to us with tingling sensation over both lower limbs which affected the right side more than the left side for the last 6 weeks. She then developed weakness of both lower limbs which initially affected the right side manifesting as heaviness in legs followed by stiffness and difficulty in walking. Weakness was insidious in onset but rapidly progressed to involve both lower limbs that patient was unable to stand without support in the next 4 weeks. When she came to us, both her lower limbs were severely spastic. Power in both her lower limbs was MRC 1/5. Tone in both upper limbs were unaffected. Reflexes in both lower limbs were exaggerated. Bilateral plantars were extensor. She had no sensory deficits. The patient underwent magnetic resonance imaging (MRI) of the cervicodorsal spine [Figure 1] which revealed an heterogeneously enhancing intraspinal tumor at D1–D2 level on the right side causing cord compression and pushing it to the opposite side. The tumor extended into the right D1–D2 foramen with extension into paravertebral area.

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Noncontrast computed tomography cervical spine showed mild degenerative changes with anterior osteophyte formation and right facet joint arthropathy from C4 to C7 level. The right side D1–D2 neural foramina was wider and the D1 pedicle thinned out as compared to the left side [Figure 2]. With a possible diagnosis of peripheral nerve sheath tumor with cord compression, the patient underwent D1-D2 laminectomy using posterior midline incision in the prone position. A large reddish pink color extra dural tumor extending from C7 to D2 level was seen on the right anterolateral side of the spinal canal pushing the cord to the left side. The tumor began to bulge out of the spinal canal even as the laminectomy was being performed. D1 and D2 right-sided facet joint was also excised to create a corridor to approach the foraminal and extraparaspinal part of tumor. There was a clear plane of cleavage between the tumor and adjacent tissues. The wound was then closed in layers. Power and spasticity in both lower limbs started to improve after surgery. At the time of discharge, power in the right lower limb was 2/5 and in the left lower limb was 3/5. Histopathological examination [Figure 3] revealed sheets of monomorphic small round cells with scanty cytoplasm and high nuclear/cytoplasmic ratio and hyperchromatic round nuclei consistent with diagnosis of PNET. It was positive for neuron-specific enolase and CD99. It showed focal expression of synaptophysin and cytokeratin. It was negative for leukocyte common antigen, CD3, and CD20. The Ki-67 proliferation index was 20%. MRI brain and whole body positron emission tomography (PET) scan showed no other tumor sites or distant metastases, and thus a diagnosis of psPNET was established. The patient was then referred to an oncologist for adjuvant therapy.

**DISCUSSION**

psPNET or extraskeletal Ewing’s sarcoma (ES) of the spine is a rare entity. It generally occurs in children and young adults (<35 years) in the lumbosacral region. Their incidence in patients >50 years of age is limited to a few case reports. It is a highly malignant and invasive tumor and presents as rapidly growing soft tissue mass. PNET belongs to a family of undifferentiated round cell tumors (i.e., neuroblastoma, non-Hodgkin’s lymphoma, rhabdomyosarcoma, and ES) and primarily affects CNS. It probably originates from matrix or germinal cells and/or neural crest cells of embryonic neural tube and is believed to have behavioral similarities to ES. Therefore, osseous ES, extraskeletal Ewing’s sarcoma (EES), Askin’s tumor, and peripheral PNET (pPNET) are grouped together as ES family of tumors.

The distinction between CNS/central PNET and EES/psPNET is important to establish a diagnosis of psPNET. CNS PNET usually occurs in infants and children and rarely in the elderly. It often presents as spinal intramedullary drop metastases from intracranial PNET whereas psPNET occurs in adolescents and young adults and is usually either extradural or intradural extramedullary. CNS PNET most likely originates from matrix or germinal cells while psPNET...
Table 1: Reported cases of primary spinal primitive neuroectodermal tumor in patients of age 50 years and above

| Author (et al.) | Year | Age/sex | Vertebral level | Location | Treatment | Follow-up (months) | Outcome | t(11:22) |
|----------------|------|---------|----------------|----------|-----------|--------------------|---------|----------|
| Kepes et al.   | 1985 | 56/male | Cauda equina   | IDEM     | STR/RT    | 36                 | Alive   | NA       |
| Isitalo et al. | 2000 | 52/male | Cauda equina   | IDEM     | STR/RT    | 12                 | Alive   | +        |
| Mawrin et al.  | 2002 | 69/male | C7-T3          | IM       | STR/RT    | 3                  | Dead    | NA       |
| Jain et al.    | 2006 | 54/female| C2-C5         | IM       | STR/RT    | NA                 | NA      | NA       |
| Fabre et al.   | 2006 | 70/male | Cauda equina   | IDEM     | STR/RT/CT | 12                 | Alive   | +        |
| Jingyu et al.  | 2009 | 58/male | D4             | ED       | GTR       | 25                 | Alive   | +        |
| Present case   | 2017 | 50/female| D1-D2          | ED       | STR       | 4                  | Alive   | +        |

ED - Extradural; IDEM - Intradural extramedullary; IM - Intramedullary; GTR - Gross-total resection; STR - Subtotal resection; RT - Radiotherapy; CT - Chemotherapy; C - Cervical; D - Dorsal; NA - Not available; + - Positive

Table 2: Differences between central nervous system primitive neuroectodermal tumor and primary spinal primitive neuroectodermal tumor

| CNS PNET | psPNET |
|----------|--------|
| Age group | Infants and children | Adolescents, young adults |
| Gender | No preponderance | Male preponderance |
| Spinal level | Mostly intramedullary | Mostly extradural or intradural extramedullary |
| Location | Mostly throughout | Mostly extradural or intradural extramedullary |
| Duration of symptoms | Shorter | Relatively longer |
| Metastases | More common | Less common |
| Site of metastases | Mostly within CNS | Extra CNS metastases common |
| CD 99 | Absent | Present |
| t(11:22) | Absent | Present |

CNS - Central nervous system; PNET - Primitive neuroectodermal tumor; psPNET - Primary spinal primitive neuroectodermal tumor

Figure 3: (a) Monomorphic tumor cells with vesicular chromatin, small nucleoli, and brisk mitotic karyorrhectic activity (H and E, ×40). (b) Diffuse membranous positivity for CD99 (×40). (c) Positivity for neuron-specific enolase (×20)

Histopathologic examination of PNET characteristically reveals sheets of poorly differentiated small, round, or spindle-shaped cells. Homer Wright rosettes may be sometimes present. On immunohistochemistry, CD 99 expression, which is a 30 or 32 kDa glycoprotein derived from MIC2 gene, is characteristic of psPNET. It also shows chromosomal translocation in t (11;22) (q24;q12) gene while CNS PNET is negative for both.

Management of these cases requires urgent operative decompression as they present with rapidly progressive neurological deficits. After confirmation of diagnosis on histopathology, adjuvant chemoradiotherapy is given. Adjuvant chemotherapy may reduce the incidence of distant metastases.
metastases. Radiotherapy is usually given for residual disease. Inspite of all measures, the prognosis of psPNET tends to be poor with median survival of 1–2 years.

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Conflicts of interest
There are no conflicts of interest.

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