Case Report

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 and power 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.

Satyashiva Munjal, Amit Srivastava, Shivya Tucker[1], Neha Bakshi[2], Sunita Bhalla[2], VS Mehta

Department of Neurosurgery, Paras Hospital, Gurgaon, Haryana, [1]Department of Radiology, Moolchand Medcity, [2]Department of Histopathology, Sir Ganga Ram Hospital, New Delhi, India

Address for correspondence: Dr. Satyashiva Munjal, Department of Neurosurgery, Paras Hospital, Sector 43, Gurgaon, Haryana, India.
E-mail: satyashivamunjal@gmail.com

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.JCVJS_70_17	

How to cite this article: Munjal S, Srivastava A, Tucker S, Bakshi N, Bhalla S, Mehta VS. Primary dorsal spine primitive neuroectodermal tumor in an adult patient: Case report and literature review. J Craniovert Jun Spine 2017;8:378-81.

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 extraforaminal/paraspinal part of tumor. There was a clear plane of cleavage between the tumor and adjacent tissues. Right D1 pedicle was thinned out due to compression by the tumor. Gross total excision of the intraspinal part of the tumor was done. The tumor was densely adherent to the right side D1 nerve root which was sacrificed. 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.

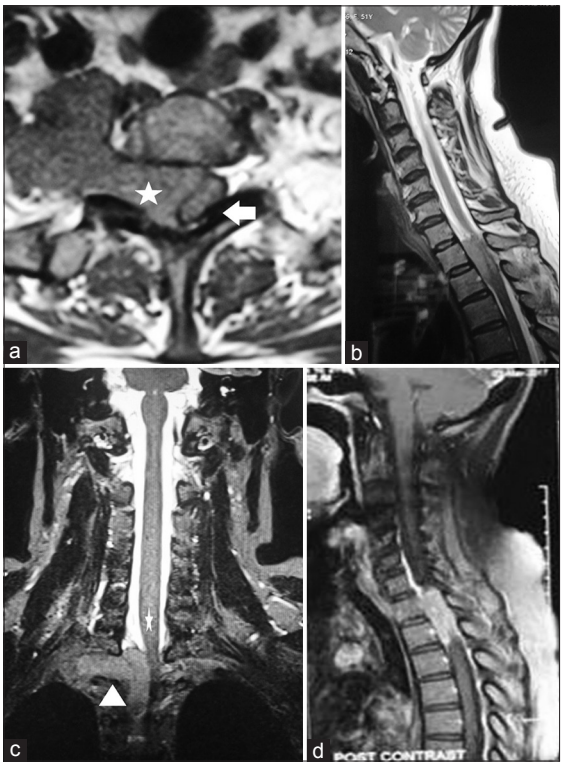

Figure 1: Magnetic resonance imaging of the cervicodorsal spine. (a) T1 axial showing isointense tumor (asterisk) pushing cord to left (b) T2 sagittal image showing hyperintense lesion causing cord compression. (c) Short tau inversion recovery coronal image showing tumor (arrow head) extending through the foramen, cord (asterisk) being pushed to left. (d) Tumor heterogeneously enhancing to contrast on sagittal and coronal images

DISCUSSION

psPNET or extraskeletal Ewing's sarcoma (ES) of the spine is a rare entity.[1-4] It generally occurs in children and young adults (<35 years) in the lumbosacral region.[1] Their incidence in patients >50 years of age is limited to a few case reports [Table 1]. It is highly malignant and invasive tumor and presents as rapidly growing soft tissue mass.[1,3] 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.[1] 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 [Table 2] is important to establish a diagnosis of psPNET.[1,3,4] 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

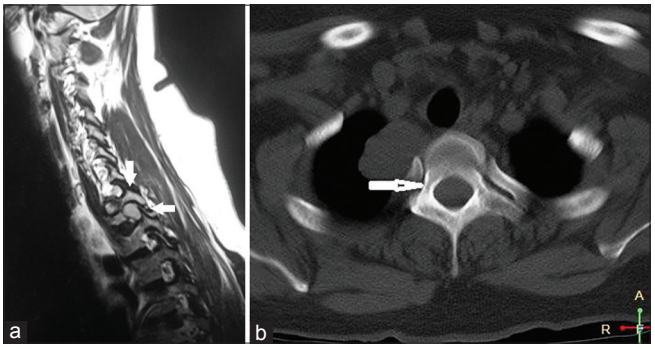

Figure 2: (a) Sagittal image showing enlargement of the foramen and thinning of pedicle (arrows). (b) Computed tomography image showing thinned out right side pedicle (arrow)

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	CD-99	t(11:22)
Kepes *et al*.[5]	1985	56/male	Cauda equina	IDEM	STR/RT	36	Alive	NA	NA
Isotalo *et al*.[6]	2000	52/male	Cauda equina	IDEM	STR/RT	12	Alive	+	NA
Mawrin *et al*.[7]	2002	69/male	C7-T3	IM	STR/RT	3	Dead	NA	NA
Jain *et al*.[8]	2006	54/female	C2-C5	IM	STR/RT	NA	NA	NA	NA
Fabre *et al*.[9]	2006	70/male	Cauda equina	IDEM	STR/RT/CT	12	Alive	+	+
Jingyu *et al*.[10]	2009	58/male	D4	ED	GTR	25	Alive	+	NA
Present case	2017	50/female	D1-D2	ED	STR	4	Alive	+	NA

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 predilection	Male preponderance
Spinal level	Occurs throughout	Mostly lumbar
Location	Mostly intramedullary	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

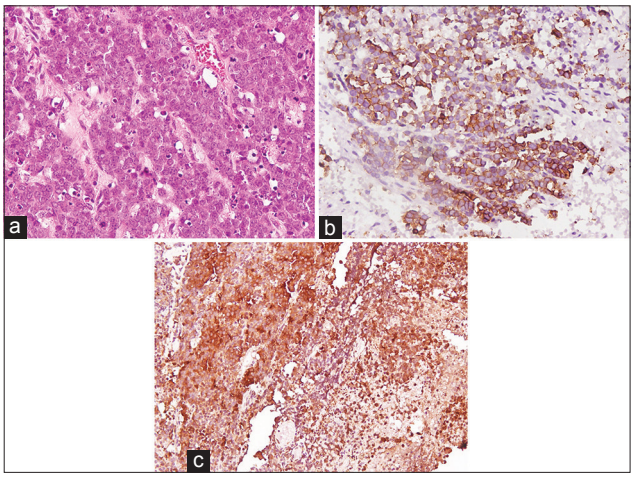

Figure 3: (a) Monomorphic tumor cells with vesicular chromatin, small nucleoli, and brisk mitotic karyorrhectic activity (H and E, ×40). (b) Diffuse membranous positivity for CD99 (×40). (c) Positivity for neuron-specific enolase (×20)

originates from neural crest cells. Both types occur rarely after 50 years of age.[1] CNS PNET is not associated with any gender predilection while psPNET has a male preponderance.[1,4] Our patient was a 50-year-old female. Clinical presentation in both groups tends to be according to the level of involvement, but pain, sensory symptoms, and radicular pain tend to be more common in psPNET.[1] Duration of symptoms tends to be shorter in patients with CNS PNET.[1]

CNS PNET tends to distribute equally throughout the spine while psPNET tends to occur more commonly in the lumbar spine.[1,4] Our case had extradural tumor in the upper dorsal spine. Clinical course of both these groups is different. CNS PNET tends to metastasize more commonly and it usually spreads to other sites in CNS while extra CNS metastases are more common in psPNET with lung being the most common site.[1,3] Fluorodeoxyglucose-PET scan must be done in all cases to rule out metastatic disease. PET scan done on follow-up in our case revealed no distant metastases.

Although MRI is an excellent imaging modality for evaluation of spinal tumors, it is difficult to establish a diagnosis of PNET based on imaging characteristics alone.[11] PNETs are usually hypointense-isointense on T1, hyperintense on T2, and enhance heterogeneously to contrast. In this regard, psPNET mimic most of the other spinal tumors such as lymphomas, metastases, chordomas, or schwannomas. Like in our case, sometimes they extend through the neural foramina and are even dumbbell shaped just like neurofibromas.[11] Needless to say, an intracranial lesion must be ruled out to establish a diagnosis of psPNET from "drop metastases" from CNS PNET.

Histopathologic examination[1,4] 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.[3] Adjuvant chemotherapy may reduce the incidence of distant

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Saeedinia S, Nouri M, Alimohammadi M, Moradi H, Amirjamshidi A. Primary spinal extradural Ewing's sarcoma (primitive neuroectodermal tumor): Report of a case and meta-analysis of the reported cases in the literature. Surg Neurol Int 2012;3:55.
2. Sharma BS, Khosla VK, Banerjee AK. Primary spinal epidural Ewing's sarcoma. Clin Neurol Neurosurg 1986;88:299-302.
3. Ghanta RK, Koti K, Ghanta VS, Teegala R. Intracranial metastasis from primary spinal primitive neuroectodermal tumor. Asian J Neurosurg 2013;8:42-7.
4. Ellis JA, Rothrock RJ, Moise G, McCormick PC 2nd, Tanji K, Canoll P, *et al.* Primitive neuroectodermal tumors of the spine: A comprehensive review with illustrative clinical cases. Neurosurg Focus 2011;30:E1.
5. Kepes JJ, Belton K, Roessmann U, Ketcherside WJ. Primitive neuroectodermal tumors of the cauda equina in adults with no detectable primary intracranial neoplasm – Three case studies. Clin Neuropathol 1985;4:1-11.
6. Isotalo PA, Agbi C, Davidson B, Girard A, Verma S, Robertson SJ. Primary primitive neuroectodermal tumor of the cauda equina. Hum Pathol 2000;31:999-1001.
7. Mawrin C, Synowitz HJ, Kirches E, Kutz E, Dietzmann K, Weis S. Primary primitive neuroectodermal tumor of the spinal cord: Case report and review of the literature. Clin Neurol Neurosurg 2002;104:36-40.
8. Jain A, Jalali R, Nadkarni TD, Sharma S. Primary intramedullary primitive neuroectodermal tumor of the cervical spinal cord. Case report. J Neurosurg Spine 2006;4:497-502.
9. Fabre E, Guillevin R, Chretien F, Le Guerinel C, Duffau H. Peripheral primitive neuroectodermal tumor of the cauda equina in an elderly patient. Case report. J Neurosurg Spine 2006;5:68-71.
10. Jingyu C, Jinning S, Hui M, Hua F. Intraspinal primitive neuroectodermal tumors: Report of four cases and review of the literature. Neurol India 2009;57:661-8.
11. Wu G, Ghimire P, Zhu L, Baral A, Su J. Magnetic resonance imaging characteristics of primary intraspinal peripheral primitive neuroectodermal tumour. Can Assoc Radiol J 2013;64:240-5.