Comparison of Primary Spinal Central and Peripheral Primitive Neuroectodermal Tumors in Clinical and Imaging Characteristics and Long-Term Outcome

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■ OBJECTIVE: **Primary spinal primitive neuroectodermal tumors are extremely rare entities. The purpose of this study was to analyze the differences in clinical and imaging characteristics and outcomes between primary spinal central PNETs (cPNETs) and peripheral PNETs (pPNETs).**

■ METHODS: **There were 25 consecutive patients with primary spinal primitive neuroectodermal tumors enrolled. The diagnosis was cPNET in 6 patients with negative CD99 expression and pPNET in 19 patients with positive CD99 expression. Gross total resection (GTR) was achieved in 12 patients, subtotal resection was performed in 9 patients, and partial resection was performed in 4 patients. Postoperative chemotherapy was given to 14 patients, and radiotherapy was given to 16 patients.**

■ RESULTS: **The age at diagnosis was significantly younger in the cPNET group (mean 12.8 years) compared with the pPNET group (mean 22.5 years) ($P = 0.040$); the 2 pathologies did not show a significant difference in prognosis. GTR ($P = 0.041$), radiotherapy ($P = 0.008$), and GTR with radiotherapy ($P = 0.009$) were significant factors leading to a higher 2-year survival rate. Kaplan-Meier analysis showed that radiotherapy ($P < 0.001$) and GTR with radiotherapy ($P = 0.040$) resulted in a longer median survival time. Patients who underwent GTR, chemotherapy, and radiotherapy all together had the highest 1-year (100.0%) and 2-year (71.4%) survival rates and the longest median survival time (32 months).**

■ CONCLUSIONS: **Patients with spinal cPNETs were younger compared with patients with pPNETs. The prognosis of spinal cPNETs and pPNETs was poor with no significant difference between the entities. The most beneficial treatment modality is GTR combined with adjuvant radiotherapy and chemotherapy.**

INTRODUCTION

Primary spinal primitive neuroectodermal tumors (PNETs) are poorly differentiated malignant tumors that usually affect children and adolescents.[1-3] PNETs include 2 categories: central nervous system (CNS) PNETs, or central PNETs (cPNETs), which chiefly arise in the CNS in the form of medulloblastoma, and Ewing sarcoma/peripheral PNETs (pPNETs), which usually occur in the chest wall (Askin tumor), paravertebral region, pelvis, and extremities.[4-9]

Primary spinal PNETs, including both cPNETs and pPNETs, are extremely rare entities.[5,6,9-11] Because of the paucity of cases of the disease, the epidemiology, clinical and imaging characteristics, and prognosis are unclear. In particular, the differences between primary spinal cPNETs and pPNETs are unknown. In this article, we review our series of 25 patients with primary spinal PNETs,

Key words
- Ewing sarcoma
- Microsurgery
- Multimodality therapy
- Primitive neuroectodermal tumors
- Spinal cord

Abbreviations and Acronyms
CI: Confidence interval
CNS: Central nervous system
cPNET: Central primitive neuroectodermal tumor
GTR: Gross total resection
KPS: Karnofsky performance scale
MST: Median survival time
OR: Odds ratio
PNET: Primitive neuroectodermal tumor
pPNET: Peripheral primitive neuroectodermal tumor
PR: Partial resection
STR: Subtotal resection

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Table 1. Clinical, Imaging, and Follow-Up Data of 25 Patients with Primary Spinal Primitive Neuroectodermal Tumors

Patient Number	Sex/Age (years)	Clinical Symptoms	DOS	Level	Location	Surgery	KPS		CD99*	Chemotherapy	Radiotherapy	Reoperation	Follow-up (months)
							Preoperative	Postoperative					
1	M/13	LE weakness	2 weeks	T2-5	Intramedullary	STR	70	80	–	—	50 Gy	—	18 (DOD)
2	M/15	LE weakness	3 days	T4-8	Epidural	GTR	40	70	–	VAC	50 Gy	1	38 (alive)
3	M/24	Neck pain, LE weakness	40 days	C6-T3	Epidural	PR	60	70	–	VAC	—	—	13 (DOD)
4	F/6	Right UE pain	2 months	C5-7	Epidural	STR	80	90	–	—	—	—	4 (DOD)
5	F/10	Back pain, LE weakness	7 days	T3-5	Epidural	STR	40	50	–	—	—	—	4 (DOD)
6	F/8	Right LE pain	1 month	L5-S2	Epidural	GTR	80	90	–	VIDE	50 Gy	—	28 (alive)
7	F/27	Back pain, LE weakness	2 months	T6-7	Intramedullary	STR	80	90	+	—	35 Gy	1	84 (DOD)
8	M/35	LE numbness	1 month	T11-12	Intradural	STR	80	90	+	VIDE	30 Gy	4	120 (DOD)
9	F/43	Neck and shoulder pain	3 months	C1-2	Epidural	GTR	90	90	+	VAC	35 Gy	2	86 (DOD)
10	M/26	LE weakness, incontinence	1 month	T9-10	Intradural	PR	60	70	+	VAC	50 Gy	—	18 (DOD)
11	F/26	LE weakness, incontinence	4 months	C4-T6	Intramedullary	PR	60	60	+	—	—	—	14 (DOD)
12	F/12	LE weakness, incontinence	3 days	C5-T1	Epidural	GTR	30	40	+	—	35 Gy	—	6 (DOD)
13	F/11	Right UE and LE pain and weakness	1 month	C6-7	Epidural	GTR	70	80	+	VIDE	35 Gy	—	25 (DOD)
14	F/4	Neck pain, left UE weakness	1 month	C2-5	Epidural	STR	50	50	+	—	35 Gy	—	18 (DOD)
15	F/8	Back pain, LE weakness	20 days	T10-12	Epidural	GTR	60	70	+	VAC	50 Gy	—	32 (DOD)
16	M/43	Right LE pain and weakness	3 months	L3-4	Epidural	GTR	90	90	+	—	50 Gy	—	70 (alive)
17	F/17	LE weakness, incontinence	20 days	L5-S1	Epidural	GTR	70	80	+	VIDE	50 Gy	—	15 (DOD)
18	F/16	LE weakness, incontinence	3 months	L5-S1	Intradural	STR	70	80	+	—	—	2	13 (DOD)
19	M/20	LE weakness	2 days	C5-T1	Epidural	GTR	40	40	+	—	—	—	18 (DOD)
20	F/17	Neck pain, right UE and LE weakness	14 days	C3-5	Epidural	GTR	80	90	+	VIDE	45 Gy	—	18 (DOD)
21	F/19	Back pain, LE weakness	1 month	L5-S1	Intradural	STR	80	50	+	VAC	—	1	13 (DOD)
22	F/43	LE weakness, incontinence	1 month	L5-S1	Intradural	PR	70	70	+	VAC	50 Gy	—	11 (DOD)
23	M/36	LE numbness	20 days	C7-T1	Intramedullary	STR	70	50	+	VAC	—	—	13 (DOD)
24	F/9	UE weakness	7 days	C2-4	Epidural	GTR	60	70	+	VIDE	—	—	9 (DOD)
25	M/7	Incontinence	2 months	S1-3	Epidural	GTR	80	80	+	—	50 Gy	—	38 (live)

DOS, duration of symptoms; KPS, Karnofsky performance scale; M, male; LE, lower extremity; STR, subtotal resection; DOD, died of disease; GTR, gross total resection; VAC, vincristine, doxorubicin (Adriamycin), cyclophosphamide; PR, partial resection; F, female; UE, upper extremity; VIDE, vincristine, ifosfamide, doxorubicin, etoposide.

*CD99 status: – = negative; + = positive.