

Primary Intraspinal Extradural Tumor Previously Termed Primitive Neuroectodermal Tumor: A Case Report

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ABSTRACT

Primitive neuroectodermal tumors are aggressive childhood neoplasms that primarily arise in the Central Nervous System and rarely occur in the spine, and are currently reclassified within embryonal tumor groups. We present a case of a 15-year-old male with lower extremity weakness and sphincter dysfunction. Spinal magnetic resonance imaging revealed a well-defined extradural mass at thoracic (Th) 11–L1 with extraspinal extension and spinal cord compression. The tumor was completely resected, and histopathology was consistent with Primitive Neuroectodermal Tumor (former classification) although additional markers were inconclusive due to technical limitations. Neurological recovery was favorable. Sphincter dysfunction resolved by the time of hospital discharge, and lower extremity weakness progressively improved, with near complete recovery at 3-month follow-up. However, seven years later, the tumor recurred and required repeat surgical treatment. This case highlights the importance of magnetic resonance imaging in the diagnosis, assessment of disease extent, and long-term follow-up of rare spinal embryonal tumors.



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INTRODUCTION

Primitive neuroectodermal tumors (PNETs) are aggressive childhood malignancies derived from the primitive neural crest and primarily arise in the central nervous system (CNS), rarely in the spine; Ma et al. reported only about 30 cases in 2020.¹ Historically, primitive neuroectodermal tumors (PNETs) were divided into central (cPNET) and peripheral (pPNET) forms, the latter belonging to the Ewing family of tumors. According to the current World Health Organisation (WHO) classification of CNS tumors, the term “PNET” has been discontinued, and these tumors are now classified within embryonal tumors or the Ewing sarcoma family.² They most commonly occur in the cerebellum.^{3,4} Spinal involvement is uncommon and may present as intramedullary, extramedullary, or extradural lesions.⁴ Children and young adults are typically affected, with most spinal cases resulting from subarachnoid spread within the neuraxis.⁵

CASE REPORT

A 15-year-old male presented to a neurosurgeon with lower extremity weakness as the main symptom progressive over several weeks. Neurological examination on admission showed pronounced lower extremity weakness, sensory deficit below Th12 level, hyperreflexia with positive Babinski sign and

sphincter dysfunction. Upper limb examination was normal.

The patient was referred for spinal magnetic resonance imaging (MRI) shortly after admission, which revealed a well-defined extradural oval mass measuring $6.5 \times 2 \times 1.5$ cm at Th11–L1. The lesion appeared isointense on both T1-weighted and T2-weighted images, with multifocal areas of high signal intensity. Post-contrast sequences demonstrated cyst-like areas within a heterogeneously enhancing mass. Imaging revealed extraspinal extension of the tumor through the right neural foramen, causing anterior cord compression and displacement to the left. Adjacent vertebrae and discs had normal signal intensity (Figure 1). The location and extent of the tumor correlated well with the patient's neurological symptoms, including lower extremity weakness and sphincter dysfunction.

Given the imaging findings, the differential diagnosis included neural sheath tumor, lymphoma, meningioma, and solitary metastasis. The patient had no significant past medical history and no family history of malignancy.

The patient underwent surgery three days after the MRI. The patient was placed in the prone position and

neuromonitoring was set up where motor evoked potentials and somatosensory evoked potentials were monitored.

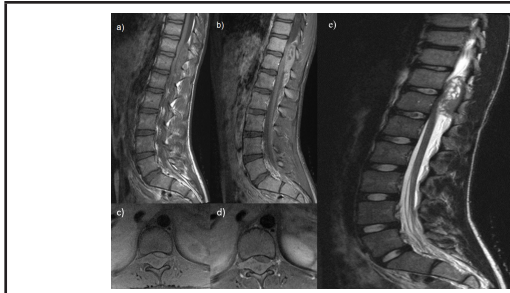


Figure 1. a) Non-contrast sagittal T1w MRI shows a round, well-defined isointense lesion at Th11-L1 with b) heterogeneous enhancement after the administration of contrast agents; c) and d) on axial tomograms, the mass displays extraspinal extension of the tumor through the right neural foramen with anterior cord compression; e) sagittal T2w sequence revealed isointense mass with multifocal high signal intensity cyst-like lesions.

Subsequently, a linear incision was made in the projection of the Th11-L1 spinous processes. This was followed by soft tissue preparation, laminectomy of the Th11, Th12 and L1 laminae, with a right-sided Th11–Th12 facetectomy. Extradural localisation was confirmed intraoperatively. A well-circumscribed pink-grayish tumor with moderate vascularity and gelatinous consistency was encountered. No infiltration of the surrounding nervous tissue was observed. The lesion was removed microscurgically en bloc, after microscurgical dissection of the tumor from the

right Th12 nerve root, which was successfully preserved, under intraoperative neuromonitoring, with an estimated blood loss of approximately 200 mL. Intraoperative ultrasound was used during the procedure.

Postoperatively, neurological deficits improved. Sphincter dysfunction resolved by the time of hospital discharge, and lower extremity weakness progressively improved, with near complete recovery at 3-month follow-up. The patient was discharged three weeks after surgery and subsequently underwent adjuvant radiotherapy, along with physical therapy, and was followed regularly by a neurosurgeon.

Histopathology showed that the tumor was composed of small round cells with a high nuclear-cytoplasmic ratio. Immunohistochemical evaluation of CD99 revealed strong diffuse membranous expression. In addition, high Ki-67 nuclear expression was detected. The tumor cells were also vimentin-positive (Figure 2). Additional markers, including cytokeratin AE1/AE3, epithelial membrane antigen, and synaptophysin, were inconclusive due to technical limitations. Based on the previously mentioned histopathological, immunohistochemical, and imaging findings, and in the absence of molecular analysis, the tumor was diagnosed as PNET (former classification), corresponding to an embryonal tumor, not otherwise specified (NOS), in the current classification. A limitation of this case is the absence of molecular analysis, which limits accurate classification within current WHO-defined embryonal tumor entities.

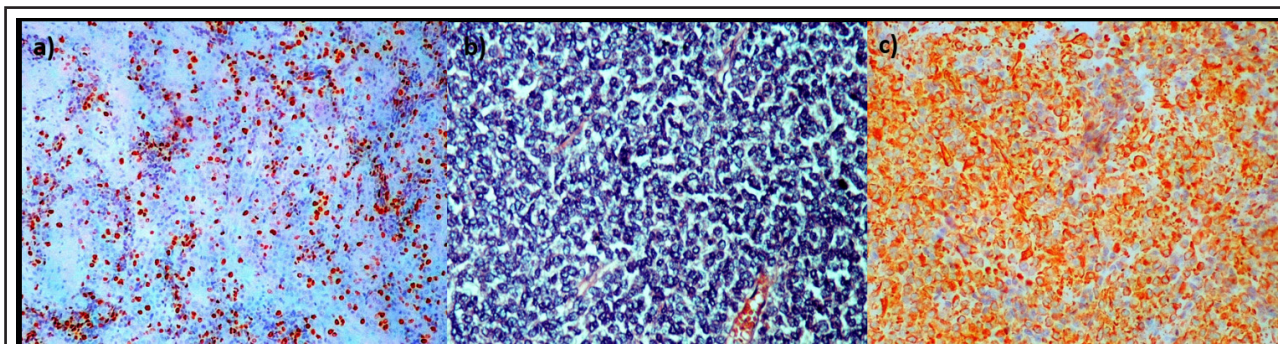


Figure 2. a) High nuclear expression of Ki-67. Obj. x10; b) Small, round cells, with high nuclear-cytoplasmic ratio. HE, obj. x20; c) Strong diffuse membranous expression of CD99. Obj. x20.

Seven years after initial treatment, the patient developed paraparesis. In this period following hospital discharge, the patient underwent regular neurosurgical follow-up with clinical assessment and MRI at 3 and 6 months postoperatively. Thereafter, follow-up visits, including clinical evaluation and MRI, were performed annually. On follow-up imaging, spinal MRI revealed a mass extending from Th9 to L1. The lesion was primarily epidural, invading the right neural foramen at Th11–12. Anterior cord compression was present, with the anteroposterior diameter of the spinal canal reduced to 3.5–4 mm. Additionally, an extraspinal component with paraspinal muscle involvement extending to the skin was observed along the surgically induced bone defect at Th10–L1 (Figure 3). No other lesions were identified in the remaining spine or intracranially.

During reoperation, the surgical access was extended to Th8 and Th9 levels. Laminectomy at Th8 and Th9 revealed a tumor firmly attached to the dural sheath and spinal cord. The tumor was excised by blunt dissection and separated from macroscopically normal tissue. Postoperatively, paraparesis improved, and the patient was able to ambulate with the assistance of a cane. However, follow-up MRI after three months demonstrated tumor recurrence. Histopathology again confirmed the diagnosis of PNET.

DISCUSSION

PNETs are rare tumors that mainly affect the pediatric population.^{1,3} Hart and Earle defined PNET as

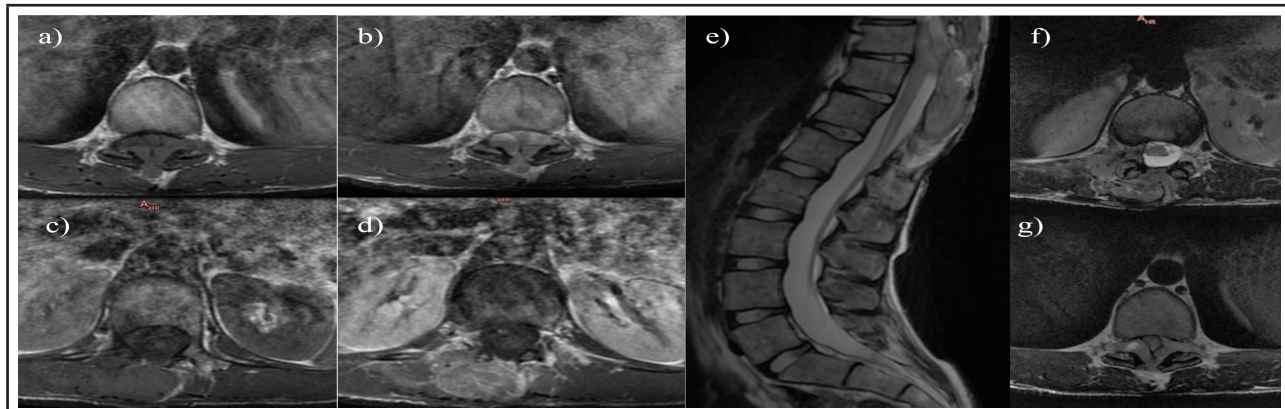


Figure 3. a) and b) pre and post-contrast axial MRI tomograms show a dominantly epidural lesion with anterior cord compression; c) and d) an extraspinal formation with paraspinous muscle damage extending to the skin located on the right side. e) Sagittal and, f) and g) axial T2w tomograms – at the vertebral level of Th9 to L1, there is a mass in the spinal canal compressing the spinal cord.

tumors composed of 90-95% of undifferentiated cells not fulfilling the diagnostic criteria for other pathological entities.³ Rorke established that all CNS tumors fundamentally composed of undifferentiated neuroepithelial cells should be referred to as PNETs.⁶ However, in the current WHO classification, these tumors are redefined into distinct molecularly characterized entities. In the absence of molecular testing, this classification may be limited and relies primarily on histopathological and immunohistochemical findings, which can pose a diagnostic challenge, particularly in low-resource settings.

Peripheral PNETs (pPNETs) often present with distant metastases, local recurrence, and poor prognosis.⁷ The imaging features of pPNET are generally non-specific. This lack of specificity often makes preoperative diagnosis challenging and requires correlation with clinical and histopathological findings. An ill-defined, aggressive mass and variable enhancement on CT and MR images may suggest the diagnosis of pPNET.⁷ These imaging characteristics, although non-specific, suggested an aggressive lesion with both intraspinal and extraspinal components, raising suspicion for a malignant process. The absence of vertebral involvement further narrowed the differential diagnosis. Our MRI findings showed slightly different enhancement patterns compared with those described by Rege et al., but were similar to those reported by Meng et al.^{8,9}

The clinical findings in our case are similar to those reported by Otero-Rodríguez, except for patient age.⁴ This highlights the variability in clinical presentation and the importance of considering this entity even in atypical age groups. Compared to other spinal tumors, there are no specific clinical features, making differentiation difficult.¹⁰ Clinical presentation depends on tumor location and involved structures, including pain, limb weakness, hypoesthesia, and bladder and bowel dysfunction.⁹ All of these were observed in our patient.

Regarding the cases described in the literature, a correlation can be observed with previous intraoperative surgical findings.⁴ Shimosawa et al. reported approximately 38 cases in the

literature, underscoring the rarity of this entity.⁵

Due to the rarity of this entity, there are no well-established, location-specific treatment guidelines.¹ Our patient received radiotherapy, as in most reported cases. Chemotherapy may also be considered.¹

In our case, tumor relapse occurred after seven years, representing a relatively long recurrence-free interval. Such delayed recurrence emphasizes the need for long-term follow-up in these patients. Surgery was again the primary treatment. Shimosawa reported that nearly 50% of patients with similar tumor localization have a survival of less than 24 months.⁵ Whenever feasible, gross total resection is preferred, as it has been associated with better local control and improved survival compared with subtotal resection. However, because of rarity and aggressive nature of spinal PNETs, multimodal treatment remains essential.¹¹

CONCLUSIONS

Spinal tumors previously termed PNETs, now reclassified within embryonal tumor groups, are rare and mainly affect young patients. MRI can be helpful in diagnosing these tumors. This is particularly important in low- and middle-income countries where molecular testing is not yet available. Additionally, MRI provides insight into disease extent and, as in our case, enables assessment of treatment response and detection of recurrence during follow-up. Unfortunately, survival rates in patients with these tumors remain poor.

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Conflict of Interest: None.

Consent: JNMA Case Report Consent Form was signed by the patient and the original article is attached with the patient's chart.

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