Primary intraspinal extradural primitive neuroectodermal tumor: A rare case

Shrikant V. Rege, Jitendra Tadghare, Harshad Patil, Sharadendu Narayan
Department of Neurosurgery, Sri Aurobindo Medical College and PG Institute, Indore, Madhya Pradesh, India

Address for correspondence: Dr. Harshad Patil, Department of Neurosurgery, Sri Aurobindo Medical College and PG Institute, Indore - 452 010, Madhya Pradesh, India. E-mail: dr.harshadpatil@gmail.com

ABSTRACT

Primitive neuroectodermal tumors (PNETs) are aggressive childhood malignancies and are difficult to treat. Primary intraspinal PNETs are rare. These patients have poor prognosis with short survival time even after surgery and chemoradiation. As there are no standard guidelines exist for the management of these tumors, a multidisciplinary approach has been employed with varying success. According to the review of literature, only few cases of primary intraspinal extradural PNETs have been reported. Herein, author has described a case of intraspinal, extradural PNET.

Key words: Intraspinal extradural, primitive neuroectodermal tumor, spinal cord

Introduction

The term primitive neuroectodermal tumors (PNETs) were coined by Hart and Earle in 1973. These are a group of malignant neoplasms derived from the primitive neural crest and are highly malignant and mainly exist in the central nervous system (CNS), chest wall, lower extremities, trunk, kidney, orbit, and rarely in the spine. The majority of childhood PNET tumors is located in the brain. In contrast, primary spinal cord tumors are very rare among children with an overall incidence rate of 0.26 per 100 person-years. Population-based data indicate that the most common histological types were meningiomas (29%), nerve sheath tumors (24%), and ependymomas (23%) in this age group. Although multimodality treatments are available, the standard treatment for PNET is complete surgical excision followed by craniospinal radiotherapy. In this article, we present a patient with primary intraspinal extradural PNET and describe its treatment regimen and outcome.

Case Report

An 8-year-old female child had been experiencing back pain and weakness of both lower limbs for 1 month. Fifteen days before admission, she also noticed difficulty in standing. Within days, the weakness progressed, and the patient was bedridden. Neurological examination revealed bilateral lower limb power 1/5 as per Medical Research Council scale. There was decrease in all sensations below D7 dermatome. Her knee and ankle reflexes were brisk bilaterally, and plantar reflexes were extensor. Magnetic resonance imaging (MRI) of the dorsal spine revealed a well-defined oval dorsal epidural lesion of size 6 cm × 1 cm at D7–D10 level with anterior compression of the cord. It was hypointense on T1-weighted image (T1-WI) and T2-WI and no obvious contrast enhancement [Figure 1]. MRI of the brain was normal, and metastatic workup was negative.

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Diagnosis was neural sheath tumor, meningioma, and solitary metastasis.

In view of her symptoms, the patient was taken up for surgery, and D6–D11 laminectomy was done. A grayish-white, friable, soft, nonvascular tumor was seen extradurally which was adherent to the surrounding arachnoid and pia mater of cord. The tumor was pushing the cord to the right side with extraforaminal extension. Tumor was completely excised, and cord decompression was achieved [Figure 2]. Histopathology revealed tumor composed of small round cells having scant cytoplasm forming diffuse sheets and islands separated by thin fibrous bands. Few pseudorosettes and occasional rosette formation seen along with areas of necrosis and vascular proliferation [Figure 3]. There was a high mitotic rate, and many karyorrhectic cells were present. On immunohistochemical (IHC) staining, we found diffuse, finely granular cytoplasmic material staining positive for synaptophysin throughout the tumor. The final pathologic diagnosis of PNET was made along with some neuronal differentiation. In the absence of a primary brain tumor, a diagnosis of primary PNET of the spinal cord was made.

The patient was planned for postoperative radiotherapy and was referred to radiation oncology for adjuvant radiotherapy. A treatment plan offering best therapeutic benefit both in terms of optimum dose to target tissue and maximum sparing of the uninvolved organs was selected. The patient was given a total of 30 Gy to the brain and spinal cord along with 8 G boost to involved area on DMX Varian Linear Accelerator. The patient tolerated the adjuvant radiotherapy well, and there was a marked improvement of pain and weakness symptoms 1 month after the radiotherapy.

Discussion

In 1973, Hart and Earle were the first to propose the term “primitive neuroectodermal tumor” for describing a neoplasm which was composed of 90%–95% of undifferentiated cells which did not fulfill the diagnostic criteria for other entities. Rorke and Becker and Hinton in 1983 proved that all CNS tumors composed of undifferentiated neuroepithelial cells are to be called PNETs. However, this concept is controversial even today. PNETs are more common in children as in our case and can also occur in adults. The mean age is between 5 and 77 years, and 80% of tumors occur in <15 years of age. There is a male predominance, being 1.4–4.8 times more common in males than in females. The use of maternal folate, iron, and multivitamin supplementation decreases the incidence.

Microscopic findings show predominantly undifferentiated tumor with occasional differentiation.
along glial or neuronal lines. PNET appears histologically as predominantly undifferentiated small, blue, round cell tumor with hyperchromatic nuclei, scanty cytoplasm, and frequent mitotic figures as was found in our case. On IHC, variable positivity may be noted depending on differentiation (neuronal, glial, or myogenic). In our case, it was positive for synaptophysin. The tumor usually presents with nonspecific symptoms such as paraparesis, paresthesias, gait disturbance, and low back pain as in our case.

MRI features of PNET are also usually nonspecific, with most of them being hyperintense on T2-WI and iso- to hypointense on T1-WI, with heterogeneous enhancement on postcontrast sequences. In our case, it was hypointense on T1-WI and T2-WI and had no obvious contrast enhancement. Intratumoral hemorrhage is highly uncommon. Duan et al. suggest that although imaging findings are not specific, the diagnosis could be suggested when MRI shows extradural large, well-circumscribed mass extending out from the intervertebral foramen and invading paraspinal soft tissues or vertebral bones in a young patient.

Treatment of PNETs consists of aggressive surgery followed by radiotherapy and chemotherapy. After adjuvant chemotherapy, survival rates may be as high as 82% at 5 years. The use of chemotherapy for treating patients with PNETs outside the cerebellum, particularly spinal PNETs, has been rather inconsistent apart from the standard use of surgical resection and radiotherapy. It is probably because such tumors are rare, and there is a lack of prospective protocol to address the benefits of specific chemotherapeutic regimens. Survival in spinal PNET ranges from 3 months to more than 3 years from the time of diagnosis.

**Conclusion**

Intraspinal and extradural PNETs are rare tumors affecting children and young adults. Survival rate is poor even after surgical resection and chemo-irradiation. PNET should be included in differential diagnosis of spinal tumors, especially in children and young adults having an intraspinal mass. As there is no standard protocol for the management of spinal PNETs, we advise complete resection with postoperative chemoradiation. Primary intraspinal PNET subtyping after histopathological and IHC studies helps us plan an optimal treatment strategy for these aggressive tumors.

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**Conflicts of interest**

There are no conflicts of interest.

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