Case Report

Primary primitive neuroectodermal tumor of spinal cord

Ashutosh Das Sharma, Jyoti Poddar, Ubrangala Kunikullaya Suryanarayan

Department of Radiotherapy, Gujarat Cancer and Research Institute, Ahmedabad, Gujarat, India

ABSTRACT

Primarily spinal primitive neuroectodermal tumors are rare neoplasm. A 28-year-old female presented with complaints of pain in lower back, radiating to both lower limbs. Magnetic resonance imaging scan of the lumbosacral spine showed an intradural extramedullary space-occupying lesion. The patient underwent L2–L5 laminectomy with excision of the lesion. Histopathology and immunohistochemistry reports confirmed the diagnosis of primitive neuroectodermal tumor while a thorough metastatic workup ruled out secondary to the spinal cord. The patient developed recurrence at local site within a month after surgery, even before the adjuvant treatment could be started. She is being treated with chemotherapy (human resources protocol).

Key words: Primary neuroectodermal tumor, rare, spinal cord

INTRODUCTION

With <100 cases reported till date, primary spinal primitive neuroectodermal tumors are rare neoplasm of children and young adults.[1] They are highly malignant with grave prognosis. These tumors were first described and named by Hart and Earle as tumors of neuroectodermal origin.[1] A definitive diagnosis becomes difficult at times due to the clinical and pathological characteristics they share with intracranial primitive neuroectodermal tumor (PNET). We report such a rare case with the intention of increasing the available data on the subject and hopefully adding to the development of a better management policy. The patient underwent L2–L5 laminectomy with excision of the lesion. The histopathology report was suggestive of PNET of the central nervous system (CNS). Immunohistochemistry (IHS) confirmed the diagnosis of primitive neuroectodermal tumor. Following the surgery, the patient developed local recurrence. She was then treated on human resources (HR) protocol of chemotherapy in view of the recurrent status of the disease.

Address for correspondence: Dr. Ashutosh Das Sharma, Department of Radiotherapy, Gujarat Cancer and Research Institute, Ahmedabad, Gujarat, India.
E-mail: sharmaashutoshdas@gmail.com

Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.197865

CASE REPORT

A 28-year-old female presented with complaints of pain in lower back, radiating to both lower limbs. The pain was insidious in onset and progressive. There was no associated fever, weight loss, bowel and bladder symptoms, or any sensory or motor deficit. Later, she developed weakness of left lower limb while the sensory system was intact in that limb. Magnetic resonance imaging (MRI) scan of the lumbosacral spine showed an intradural extramedullary space occupying lesion, 1.3 cm × 1.6 cm × 9.5 cm in size, extending from L3 to S1 vertebral region [Figure 1]. It caused displacement and compression of the lumbar nerve roots. The patient underwent L2–L5 laminectomy with excision of the lesion on February 27, 2016. The histopathology report was suggestive of PNET of the CNS [Figure 2]. The IHS report came out to be vimentin positive, CD99 positive, synaptophysin positive, leukocyte common antigen (LCA) negative, CD20 (pan B) negative, and desmin

Cite this article as: Sharma AD, Poddar J, Suryanarayan UK. Primary primitive neuroectodermal tumor of spinal cord. Clin Cancer Investig J 2016;5:454-6.

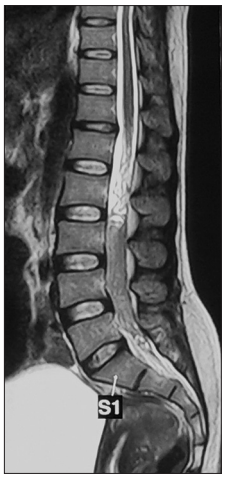

Figure 1: Magnetic resonance imaging dorsolumbar spine showing the mass lesion

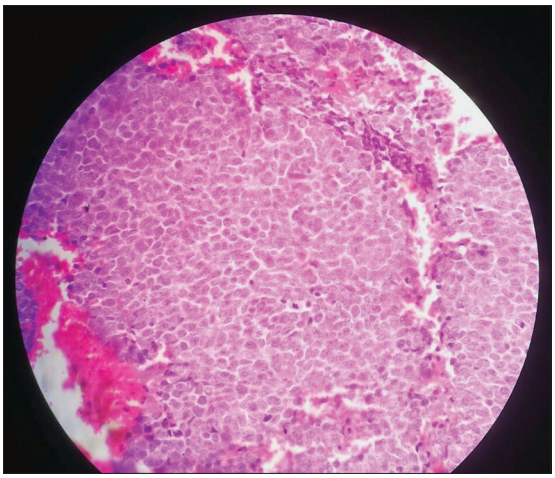

Figure 2: High power H and E stained histopathology slide

negative which was consistent with the diagnosis of PNET. A thorough metastatic workup was done. Computed tomography (CT) scan of thorax and abdomen, pelvis and MRI scan of brain and cerebrospinal fluid (CSF) cytology were found to be negative leading to a diagnosis of a primitive neuroectodermal tumor involving the spine as the primary organ, rather than a metastatic disease. The patient achieved symptomatic relief with the surgical intervention. She regained limb power and could walk on her own, but after a month of surgery, the pain in the back reappeared. A repeat postoperative MRI scan of lumbar spine showed a diffused heterogeneous meningeal lesion in lumbar spinal canal and neural foramen at L2–L5 with associated paraspinal soft tissue enhancement. In view of the recurrent lesion, the patient was started on HR protocol chemotherapy.

DISCUSSION

The primary spinal primitive neuroectodermal tumors are an extremely rare variety of malignancy with <100 cases reported in medical literature till date. PNET as a group was first described by Hart and Earle in 1973 in a series of 28 cases of cerebral tumors with undifferentiated cell of neuroectodermal origin.[1] The WHO classifies them under embryonal tumors, with PNET used as a generic term for cerebellar medulloblastoma and all other CNS tumors with histopathology identical to medulloblastoma.[2] Central PNET arises from the primitive neuroectodermal cells present in the subependymal zone scattered throughout the CNS.[3,4] However, for reasons unknown, as a primary, they are uncommon in the spinal cord as compared to the brain. The primary spinal PNET is rarer as compared to those arising elsewhere throughout the body, are more common in children and young adults, equally in both sexes. The segment of spine involved is the thoracic and cauda equina. Intracranial PNET has a propensity to metastasize via the CSF to subarachnoid spaces throughout the CNS.[5] This pattern of spread makes the finding of a spinal PNET more probably a drop metastasis from an intracranial primary than a primary PNET of spinal origin.[6] A normal CT/MRI scan of the brain can differentiate the two. Detection of MIC2 gene glycoprotein by IHC (CD99) can confirm the diagnosis of spinal PNET.[7] Due to the limited data available on the disease, the optimal treatment is not yet established. Surgery is the primary modality of treatment for lesions amenable to excision. The lesion present eloquent areas of spine may block this avenue. Craniospinal irradiation and hyperfractionated radiotherapy to local site only have shown some benefits.[8] Radiation to growing spine in a prepuberty patient becomes a matter of serious concerns regarding the growth and development. Chemotherapy as various combinations of agents such as cyclophosphamide/ifosfamide, platinum compounds, vincristine, and peplomycin can be used in some cases.[6] Adaptive immunotherapy using lymphokine-activated killer cells has been tried as an adjuvant therapy for primitive neuroectodermal tumor.[5]

CONCLUSION

Primitive neuroectodermal tumor primarily involving the spinal cord is an extremely rare occurrence. Even though they share the same poor prognosis and aggressive behavior with cranial primitive neuroectodermal tumor, the treatment modalities are different for both of them, and that makes the identification of a primary spinal PNET and differentiating it from a drop metastasis a crucial step in the management.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Hart MN, Earle KM. Primitive neuroectodermal tumors of the brain in children. Cancer 1973;32:890-7.
2. Aydin MV, Sen O, Ozel S, Kayaselcuk F, Caner H, Altinors N. Primary primitive neuroectodermal tumor within the spinal epidural space: Report of a case and review of the literature. Neurol Res 2004;26:774-7.
3. Rorke LB. The cerebellar medulloblastoma and its relationship to primitive neuroectodermal tumors. J Neuropathol Exp Neurol 1983;42:1-15.
4. Crain Barbara J, Wilkins RH, editor. Primitive neuroectodermal tumours. Neurosurg Rengachary 1996;2:1707-13.
5. George, Richard E. *In vitro* cytolysis of primitive neuroectodermal tumors of the posterior fossa (medulloblastoma) by lymphokine-activated killer cells. J Neurosurgery 1988:69.3:403-9.
6. Deme S, Ang LC, Skaf G, Rowed DW. Primary intramedullary primitive neuroectodermal tumour of the spinal cord: Case report and review of the literature. Neurosurgery 1997;41:1417-20.
7. Kampman WA, Kros JM, De Jong TH, Lequin MH. Primitive neuroectodermal tumours (PNETs) located in the spinal canal; The relevance of classification as central or peripheral PNET: Case report of a primary spinal PNET occurrence with a critical literature review. J Neurooncol 2006;77:65-72.
8. Dorfmüller G, Würtz FG, Umschaden HW, Kleinert R, Ambros PF. Intraspinal primitive neuroectodermal tumour: Report of two cases and review of the literature. Acta Neurochir (Wien) 1999;141:1169-75.