

# Primary spinal peripheral primitive neuroectodermal tumor with acute presentation: A case report and review of literature



Abta Bachchan<sup>1</sup>, Kaushik Roy<sup>2</sup>, Suniti Kumar Saha<sup>3</sup>, Debajyoti Pathak<sup>4</sup>, Partha Ghosh<sup>5</sup>, Abhishek Nadkarni<sup>6</sup>

<sup>1,4,6</sup>Resident, Department of Neurosurgery, N.R.S. Medical College, Kolkata, <sup>2</sup>Associate Professor, <sup>3</sup>Professor and Head, Department of Neurosurgery, N.R.S. Medical College, Kolkata, <sup>5</sup>Institute of Neurosciences, Kolkata

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## ABSTRACT

Primary spinal primitive neuroectodermal tumors (PNET) is a rare occurrence and carries a poor prognosis. A 13-year old female patient acutely presented with pain in the thoracic region, bilateral lower limb weakness, bladder and bowel dysfunction. Clinically paraplegia with truncal weakness, lower limb deep tendon reflexes of both side were absent and planter reflexes equivocal bilaterally. Preoperative MRI of thoracic spine revealed D4-D6 extradural SOL. A D4-D5 Laminectomy and left Cortico transversectomy done. Pathological findings were consistent with PNET. The clinical, histopathological, and radiological findings of the patient are presented.

Key words: PNET; Extradural spinal SOL; Dorsal spinal SOL

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## INTRODUCTION

The term primitive neuroectodermal tumors (PNETs) was first proposed by Hart and Earle in 1973 to describe mainly for undifferentiated tumors of the cerebrum which further describe as central (cPNET) and peripheral (pPNET).<sup>1</sup>

Extracranial PNETs arising from the soft tissues or bones outside the central or the sympathetic nervous system are presumed to be of neural crest origin and are composed of differentiated small, round, and hyperchromatic tumor cells.<sup>2</sup> pPNETs rarely occur in the spine and CNS, and often in the chest wall, trunk, lower extremities, kidney, and orbit and belong to the Ewing sarcoma family (EWS) arise commonly in adolescents or young adults, with a slight male predominance.<sup>3</sup>

To the best of our knowledge, one hundred seven cases of primary EWS/pPNET (extradural/intradural extramedullary lesions) has been reported in the literature, of which 60% are extradural location and are most commonly located in lumbar spine.<sup>2</sup>

Here, we report a rare case of primary spinal pPNET, in the epidural space of the thoracic spine, presenting acutely clinically.

## CASE REPORT

A 13-year female, who was healthy previously presented to the OPD of our institute NRSMC&H with history of trivial trauma and weakness of both lower limbs associated with back pain. Her symptoms progressed rapidly to severe motor weakness and she became bed bound within a week with paresthesia and urinary retention.

Initially it was presumed to traumatic in nature and MRI thoracic spine showed an extradural spinal SOL at D4-D6 (Figure 1). CT guided FNAC was done, which was suggestive of small round cell tumour.

Patient was conscious & well oriented. Lower limb power was 0/5 bilaterally associated with truncal weakness while upper limb, neck muscles power was normal. Lower limbs

### Address for Correspondence:

Dr. Kaushik Roy, Associate Professor, Department of Neurosurgery, N.R.S. Medical College, 138, A.J.C. Bose Road, Kolkata -700014, West Bengal, India Tel: +91-9830317242. E-mail: dr.kaushik@rediffmail.com

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deep tendon reflexes were absent and planter reflexes were equivocal on both side. All sensations below D6 was absent.

Surgery was planned with the aim of spinal decompression and gross total excision of the lesion through posterior approach in prone position. D4, D5 Laminectomy and left D5 cortico-transversectomy done. In this case near total removal of tumor was possible. Intra-operative and Postoperative period was uneventful and patient's neurological status was improved partially following surgery.

Histopathological analysis of the tumour showed malignant small round cell tumour – Ewing sarcoma/PNET. Immuno-histological analysis was CD99 positive and CD45 negative.

## DISCUSSION

Primary spinal PNETs have been increasingly reported in recent years but there are still very few reports of PNETs originating in the spinal cord. The earliest recognized case report of a spinal PNET is from an article by Smith et al.<sup>4</sup> Their use of the term PNET was not properly defined. However, the nomenclature and criteria for diagnosing PNET were not formally introduced until 1973 by Hart and Earle. It was used to describe mainly for undifferentiated tumors of the cerebrum which were further sub-classified as central (cPNET) and peripheral (pPNET).<sup>1</sup> The difference between cPNETs and pPNETs are significant for clinicians, mainly because the location and pathology of the tumor, treatment planning and prognosis differ. cPNET are heterogeneous group of embryonal tumors and according to the World Health Organization (WHO) classification of tumors of the human central nervous system (CNS) have been referred to as CNS PNETs to avoid confusion. pPNETs are a type of soft-tissue peripheral malignant round cell tumors, like rhabdomyosarcoma or Ewing's sarcoma (EWS). Expression of glycoprotein p30/32 (CD99), which is encoded by the *MIC2* gene and EWS-FLI1 chimeric gene is strongly associated with EWS and pPNET.<sup>2</sup> Differentiation between pPNET and EWS is possible on the basis of conventional light microscopy and immunohistochemistry, the presence of Homer-Wright rosettes and/or the expression of at least two neural markers indicating pPNET. The absence of Homer-Wright rosettes and none or only one neural marker indicates EWS.<sup>5</sup>

Those tumors currently understood as PNETs are further divided into two categories: central nervous system PNETs (CNS-PNET) and Ewing sarcoma/PNETs (EWS/pPNET).<sup>1</sup> Various differences between central and pPNET are described below<sup>6</sup> (Table 1).

**Table 1: Comparison of cPNET and pPNET**

| Variable                      | cPNET                                   | EWG/pPNET                               |
|-------------------------------|---|---|
| Age group                     | Infants and children                    | Adolescent/Young adult                  |
| Gender                        | No predilection                         | Male preponderance                      |
| Origin                        | Most likely germinal cells              | Neural crest cells                      |
| Spinal level Location         | Occurs throughout Mostly intramedullary | Mostly Lumbar Mostly extradural/ IDEM   |
| Duration of symptom           | Shorter                                 | Relatively longer                       |
| Metastases Site of metastases | More common Mostly within CNS           | Less common Extra CNS metastases common |
| CD99                          | Absent                                  | present                                 |
| T(11:22)                      | Absent                                  | Present                                 |



**Figure 1:** Pre-op MRI images of dorsal spine T1 sagittal (a), T2 sagittal (b), STIR sequence sagittal (c), and post-operative MRI scan (d)

The 2016 World Health Organization (WHO) CNS Tumor Update Classification Eliminate the term “Primitive Neuroectodermal Tumor” (PNET). PNETs have distinct immunohistopathology. These tumors are a group of “small round-cell” tumors which includes the Ewing’s sarcoma/PNET family group, neuroblastoma, rhabdomyosarcoma, and malignant lymphoma. CD45 is a transmembrane protein tyrosine phosphatase, its immunoreactivity is highly specific for non-Hodgkin’s lymphomas. pPNETs have a tendency to metastasize outside neuroaxis with the most frequent sites being the lungs, bones, and lymph nodes, similar to cPNETs. The aggressive nature of the tumor is highlighted by its rapid recurrence rate.<sup>7</sup>

Primary spinal PNETs represent less than 1% of primary spinal tumors.<sup>8</sup> It is most prevalent in the adolescent and young-adult populations and are slightly more common in males than in females. Both primary intraspinal EWS/pPNET and CNS-PNET occur rarely after 50 years of age.<sup>9</sup> In our two cases, both patients were female.

The most common symptom in Spinal cPNET/pPNET tumour groups is muscle weakness. However, sensory

symptoms, local pain, and radiculopathy are more common in primary intraspinal EES/pPNET. The development of symptoms like pain, weakness, sensory alteration or urinary incontinence takes several weeks to months are commonly observed.<sup>8</sup> Both of our cases presented acutely within a week.

Magnetic resonance imaging with Gd contrast is the imaging modality of choice for evaluating the spinal tumors however it is difficult to establish the diagnosis of PNET on radiological basis.<sup>10</sup> Any intracranial lesion should be ruled out to diagnose primary spinal PNET from drop metastases. PNETs are typically hypo- to isointense on T1 and iso- to hyperintense on T2, there is often minimal contrast enhancement, but in both of our cases it shows edhomogenous enhancement, thus confusing with the nerve sheath tumor or meningioma preoperatively.

Due to rare occurrence and lack of large series studies, there are currently no standard clinical guidelines outlining their management. Still these cases require urgent operative decompression as they present with rapidly progressive neurological deficits followed by adjuvant chemo and radiotherapy is given as suggested by literature by Ghanta RK *et al* in 2013.<sup>11</sup> Both of our cases underwent gross total excision of the tumor followed by adjuvant radiation therapy.

Poor outcome of primary spinal PNET tumor have been reported with a median survival of 1 to 2 years.<sup>12</sup> The present cases did not show any evidence of dissemination or metastasis till 6 months follow-up following which they were lost in follow-up.

## CONCLUSION

The reported cases of undifferentiated small round cell tumors in the spine have increased in number in recent years. As per review and analysis suggested primary spinal CNS PNET and EWG/pPNET can be differentiated on tumor location, as CNS PNET mainly intramedullary and EWG/pPNET are extramedullary.

Due to rare occurrence and limited evidence of these tumors, there are currently no standard clinical guidelines outlining their management. However, different studies suggest total excision of tumor followed by chemoradiation as the best treatment procedure, which require further evaluation.

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### Authors Contribution:

**AB and KR**- Concept and design of the study, reviewed the literature, manuscript preparation and critical revision of the manuscript; **SKS**- Concept, collected data and review of literature and helped in preparing first draft of manuscript; **DP**- Conceptualized study, literature search, statistically analyzed and interpreted, prepared first draft of manuscript and critical revision of the manuscript; **PG and AN**- Concept of study, collected data and review of study.

### Work attributed to:

Department of Neurosurgery, N.R.S. Medical College, 138, A.J.C. Bose Road, Kolkata -700014

### Orcid ID:

Dr. Abta Bachchan- <https://orcid.org/0000-0002-1398-004X>  
 Dr. Kaushik Roy- <https://orcid.org/0000-0001-8317-9840>  
 Dr. Suniti Kumar Saha- <https://orcid.org/0000-0002-6217-9355>  
 Dr. Debajyoti Pathak- <https://orcid.org/0000-0002-8503-4618>  
 Dr. Abhishek Nadkarni- <https://orcid.org/0000-0003-2842-475>

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