MRI of Primary Spinal Epidural Primitive Neuroectodermal Tumor: a case report

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Primitive neuroectodermal tumor (PNET) is highly malignant and extremely rare in the spine. We present a case with primary spinal primitive neuroectodermal tumor (PSPNET) which involved epidural space of thoracic spine with cord compression. Magnetic resonance imaging (MRI) showed an isointense epidural lesion on T1WI and T2WI. Heterogeneous enhancement following the administration of gadolinium is also noted.

The prognosis of PSPNET is very poor. Even though PNET in spine is an extremely rare condition, it should be included in differential diagnoses for patients with spinal tumor.
round cells with round to void nuclei, single conspicuous nuclei, high nucleocyttoplasmic ratio with indistinct cell border and amphophilic cytoplasm, brisk mitotic activity, frequent apoptosis and tingible-body macrophages (Fig. 3). Pronounced fresh hemorrhage was noticed between the tumor cell aggregates. Immunohistochemically, the tumor cells revealed diffusely and strongly membranous staining for CD99 (Fig. 4a) and dot-like granular cytoplasmic staining for synaptophysin (Fig. 4b). The tumor cells were also focally reactive to vimentin but non-reactive to GFAP, EMA, cytokeratin, LCA, CD3, CD20, CD34, desmin, MyoD1, myeloperoxidase, lysozyme, and S-100 protein. This immunoprofile was confirmative of a PNET. The final pathological diagnosis was PSPNET.

The symptoms disappeared immediately after the operation and the patient was discharged one month later. He received radiotherapy and chemotherapy at another hospital with a smooth course. However, 14 months after the operation, he came back to our neurosurgery department with the complaint of right shoulder palpable painful mass. Series of CT and MRI studies showed multiple metastases at spine, lung and liver.

DISCUSSION

PNET was first introduced by Hart and Earle [2] in 1973 to describe a diverse group of malignant primitive and/or undifferentiated tumors occurring in children and young adults, which tend to have a
relatively poor prognosis. In the recently updated World Health Organization classification, PNET is defined as an embryonal tumor composed of undifferentiated or poorly differentiated neuroepithelial cells which have the capacity for or display divergent differentiation along neuronal astrocytic, ependymal, muscular or melanotic lines [3]. The 2000 WHO classification includes only supratentorial PNETs in the group of embryonal tumors and without mention of PSPNETs. In the revised 2007 WHO classification of tumors of the central nervous system, PSPNET was included in the entity of “CNS primitive neuroectodermal tumors”, which include similar tumors located in the brain stem and spinal cord in the more general term (PNET) [4]. The most frequent sites of PNET are the intracranial, chest wall, lower extremities, trunk, kidney and orbits; These tumors rarely originate from the spine. The histopathology and immunohistochemistry characteristics of PNET including: (1) Poorly differentiated small round/spindle-shaped cells. (2) Architecture: either densely packed or placed in sheets or nests. (3) Cells are generally positive for neuronal markers like neuron-specific enolase; it could be for precursor intermediate filaments like nestin, vimentin or microfilaments; synaptophysin or GFAP may be positive depending on the differentiation of the cells; S-100 is also commonly positive. (4) In case of undifferentiated cells, they may not stain positive for any analyses [5].

Only about 30 cases of PSPNET were previously reported[1, 5-9]. The mean age of these patients at diagnosis was 24 years (range, 3 months to 69 years) with slight male predominance. PSPNET may arise from all levels of the spine and can be intramedullary, extramedullary/ extradural. The intramedullary tumors might originate from spinal cord, while the extradural tumors might arise from vertebrae, soft tissue or spinal nerve roots. The commonest location is the cauda equina region. This might be due to the fact that the cauda equina

Figure 3. The tumor cells were growing in solid sheets with a picture of undifferentiated small blue round cell tumor with high nucleocytoplasmic ratio. The nuclei of the tumor cells were round to ovoid shaped with prominent nucleoli, brisk mitotic activity and apoptosis. (H&E stain, magnification x400)

Figure 4. a. Immunohistochemical stain showed diffusely and strongly membranous staining for CD99 of the tumor cells (magnification x400). b. A dot-like granular cytoplasmic staining for synaptophysin was discernible in the tumor cells (magnification x400).
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equina forms part of the peripheral nervous system, where the axons have become dependent on Schwann cells for the maintenance of their myelin sheaths [5].

MRI is very helpful in evaluation of the location and extent of the tumor. A review of the literature showed no specific image characteristics described for PSPNET [1, 5]. Most of them are hyperintense on T2WI but the tumor in our case is isointense on T2WI. One particular finding in our case is intra-tumoral hemorrhage which has not been described earlier in the literature of PSPNET. However, it is not uncommon in the supratentorial PNETs. Another finding in our case is the involvement of spinous process which has been described only in few cases with intraspinal relapse [5]. The other MRI findings including widening of intervertebral foramina, isointensity on T1WI and heterogeneous enhancement are not specific and can be confused with other spinal lesions including epidural hemangioma, neurofibromatosis, metastasis and lymphoma. The epidural hemangioma usually shows isointense signal to spinal cord on T1WI, hyperintense signal on T2WI and stronger enhancement with gadolinium [10]. Neurofibromatosis is usually located in intradural, extramedullary space; it is a well-defined mass with a dumbbell configuration and widening of intervertebral foramina. Neurofibromatosis shows homogeneous isointense to spinal cord on T1WI and hyperintense on T2WI. Metastatic tumors tend to enclose the spinal cord epidurally and are usually associated with erosion of the adjacent vertebral bone. Most lymphomas are hyperintense on T2WI, isointense on T1WI with homogeneous enhancement after gadolinium enhancement [11]. Another rare differential diagnosis is extra-osseous Ewing’s sarcoma with epidural extension. It is usually hyperintense on T2WI, low-to-isointense on T1WI and exhibits heterogeneous enhancement after gadolinium enhancement [12]. Extra-osseous Ewing’s sarcoma commonly affects the paravertebral regions and rarely involves the epidural space. But it can not be ruled out by MRI findings in our case.

Most of the PSPNET were impossible to resect en bloc, because it usually involved nerve roots, spinal cord and vertebrae. Radiotherapy and chemotherapy have been used in most of the patients. In view of the limited cases, the optimal management has not be defined yet. The prognosis of PSPNET is very poor and most patients had recurrence and metastasis. The average time from the operation to death was 18.1 months [1].

In conclusion, PSPNET is very rare and extremely malignant. The prognosis of this tumor is very poor and the treatment is far from satisfactory. Even though PNET in spine is an extremely rare condition, it should be included in differential diagnoses for patients with spinal tumor. 

REFERENCE
原發性脊樑硬膜上原始神經外胚層腫瘤之磁振造影影像：病例報告

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原始神經外胚層腫瘤為高度惡性且在脊椎相當罕見。我們報告一 18 歲男孩之原發性脊椎原始神經外胚層腫瘤病例，腫瘤侵犯胸椎硬膜上區域伴隨脊髓受壓。磁振造影在 T1 加權影像及 T2 加權影像上均顯示和脊髓相同訊號的病灶，並且在注射對比劑後顯示不均勻顯影。

原發性脊椎原始神經外胚層腫瘤之預後極差，雖然在脊椎發生原始神經外胚層腫瘤是極為罕見的情形，對於脊椎腫瘤的病人仍應該將它列入鑑別診斷。