Primary intraspinal primitive neuroectodermal tumors (PNETs) are rare. We report a case of sacral PNET with review the literature. A 15-year-old boy presented with progressive lumbar area pain, lower leg numbness and neurogenic bladder. Preoperative magnetic resonance imaging revealed a sacral tumor with epidural, presacral, and sacral bone extensions. An urgent operation was performed with gross tumor removal. The pathological findings were consistent with a PNET. This tumor is somewhat responsive to chemotherapy and radiotherapy thus far. A review of the English literatures showed that only some cases of primary intraspinal PNETs have been reported to date, and the present case is one of very rare cases reported in English literatures in which the tumors were of purely sacral origin sites. Most of the reported PNET patients survived less than 2 years. Primary intraspinal sacral PNETs are rare tumors and carry a poor prognosis.

CASE REPORT

A previously healthy 15-year-old boy presented complaints of low back pain radiating down to the right leg and then to the left leg of several weeks' duration. Frequency of micturition and a sensation of bladder fullness had been noted for a week. The patient was unable to void urine on the day of admission.

Neurological examination revealed weakness of both little toes (the right more than the left). The patient was areflexic in the lower extremities and a sensory level to pinprick was present at L5 bilaterally. There were no other abnormal findings on general physical examination.

Radiographic investigations

Plain films of L-spine and KUB showed enlarged right S1 neuroforamen without definite bony destructions. Contrast-enhanced magnetic resonance (MR) imaging of lumbar-sacral area disclosed a huge mass lesion showing hypointense on T1-weighted images (T1WI) (1.5T MR machine, TR/TE/Excitations=430/16/2), slightly hyperintense intensity on T2-weighted images (T2WI) (TR/TE/Excitations=3000/95/2), and abnormal contrast enhancement within the sacral canal, more on right side. The lesion was centered in the level of S1 with presacral, sacral bone extensions and local bony erosions (Fig. 1).

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**Operation**

The patient received emergent operation and laminectomy was performed at level of L5. A dorsal midline myelotomy was performed, exposing an intraspinal tumor that was grayish in color and well demarcated with epidural extension. Tumor removal was performed under the operating microscope.

**Pathological findings**

The paraffin sections revealed a very cellular tumor, consisting of small round cells with hyperchromatic nuclei and scanty cytoplasm (Fig. 2). Mitotic figures were frequent. No well-defined Homer-Wright or ependymal rosettes were noted. There were few cells with moderate eosinophilic cytoplasm and occasional cells with large vesicular nuclei and prominent nucleoli suggestive of neuronal differentiation.

Immunohistochemistry was performed using the following five markers and the results were as followings: (1) Vimentin positive; (2) Neuron-specific enolase (NSE) positive; (3) Leukocyte common antigen (LCA) negative; (4) S-100 positive; (5) Cytokeratin negative. According to the histopathological pictures and the result of immunohistochemical stain, it was compatible with primitive neuroectodermal tumor. The possibility of lymphoma and small cell carcinoma were excluded.

**Postoperative course**

Residual tumors were noted on CT scan after surgery. Post-operative radiotherapy and chemotherapy were then given with gradual improvement of the symptoms. The patient was able to walk with...
a cane and had normal bowel and bladder function 6 weeks after radiotherapy. His postoperative course was smooth and discharged in stable condition. Postoperative follow-up MR imaging of the brain and whole spine did not reveal any other tumor seed- ings, especially in the posterior cranial fossa.

DISCUSSION

PNETs are common tumors in children but can occur in any age and are mainly intracranial in location. They are composed of largely undifferentiated neuroepithelial cells. The original nomenclature of these tumors was based on location and/or differentiation, i.e., a pineal tumor was called a “pineoblastoma,” a cerebellar tumor “medulloblastoma,” and a cerebral tumor with neuronal differentiation a “neuroblastoma,” although many of them were historically similar. Hart and Earle [1] first introduced the term primitive neuroectodermal tumor in 1973 to describe predominantly undifferentiated tumors of the cerebrum (with 90-95% of the cells being undifferentiated) and did not fulfill the diagnostic criteria for neuroblastoma, ependymoblastoma, polar spongioblastoma, medulloepithelioma, or pineal parenchymal tumors. In 1983, Rorke [2] and Becker and Hinton [3] independently reviewed this concept in separate articles advocating that all central nervous system tumors predominantly composed by primitive neuroepithelial cells should be called PNETs. They then further subclassified these tumors based on differentiation. This concept has been widely accepted, although it is still controversial [4].

These tumors frequently disseminate or seed throughout the central nervous system via the cerebrospinal fluid and may rarely metastasize outside the CNS. Most cases of PNET involving the spinal cord are "drop" metastases from primary intracranial tumors.

Over twenty eight cases of primary intraspinal PNETs have been previously reported in the English literature and most of these cases, efforts were made to exclude primary intracranial lesions either by imaging (computed tomography, magnetic resonance of the brain mostly) and/or autopsy. In this case, MRI of the brain and spine failed to reveal any intracranial or other spinal tumor. This case therefore appeared to represent a primary intraspinal PNET of rare location which is in the sacral area [5].

A review of the English literatures show that primary intraspinal PNETs may arise at all levels of the spine and can be intramedullary, intra- and extramedullary, extramedullary, or extradural. The tumors seem to have a predilection for the cauda equina. It has been postulated that PNETs arise from neoplastic transformation of primitive neuroepithelial cells in subependymal zones [2]. The fact that the subependymal zone can persist in any part of the central nervous system may explain the presence of PNETs in locations other than the cerebellum.

In our case, the tumor showed strong homogeneous enhancement on contrast-enhanced T1WI MR images, which suggesting good vascularities. Mild hyperintense intensity on T2WI MR images indicating its compact cellularities as shown in pathological specimens.

Intraspinal PNETs appear to be more common in adults rather than children, in contrast to the predominance of intracranial PNETs in children. Frequent sites of metastases outside the CNS are lung, bones, and lymph nodes, a tendency shared by intracranial PNETs [6, 7]. Most patients were treated with a combination of surgery, radiotherapy, and chemotherapy, but despite treatment most did not response well. Most of these patients die within 2 years [8] and our case has been survived for at least 9 months. The aggressive nature of these tumors is evidenced by the rapid recurrence of the tumor in some reported cases. The cause of death in these patients included pneumonia, metastatic diseases, aggressive local spreading of the diseases, and progressive spinal cord involvement.
CONCLUSIONS

Primary intraspinal PNETs are rare lesions. They may be intramedullary, intradural extramedullary, combined intramedullary and extramedullary, or extradural. These tumors may metastasize to outside the central nervous system. The prognosis appears to be poor with most patients surviving for less than 2 years.

REFERENCES

一位 15 歲的男孩表現出漸進式的腰痛、小腿麻木和神經性膀胱功能異常。手術前的磁振造影檢查發現了一個薦椎腫瘤，並侵犯到硬腦膜外、薦骨和薦骨前區域，因此緊急開刀將腫瘤切除。病理檢查結果發現是薦椎原始神經外胚層細胞瘤。這類腫瘤對化療和放射線治療只有部份病例如有反應。英文期刊文獻至今只有少數病例被報告，而且此病例是非常少數的原始神經外胚層細胞瘤原發於薦椎被報告的病例。大多數原始神經外胚層細胞瘤患者存活期間少於 2 年。