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Case Report

Primitive neuroectodermal tumor of the lumbosacral nerve plexus: A case report ☆

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ABSTRACT

We report an uncommon case of primitive neuroectodermal tumor/ Ewing's sarcoma of the lumbar and sacral nerve plexus in a 17years old boy who presented with an intense pain in the lower back radiating to legs. Magnetic resonance imaging showed a soft tissue mass with thickening of lumbar and sacral spinal nerve roots (L5-S3 level), along with widening of the corresponding foramina. There was also posterior scalloping of L5/S1 vertebrae and invasion of the sacral bone. A Partial resection has been performed, and the ensuing histopathology confirmed the diagnosis of PNET/Ewing's sarcoma. MRI in conjunction with histopathology are the key to narrow down the differential diagnoses list. PNET of lumbosacral area remain scarce, and only few cases have been reported nowadays. Given to the aggressivity of these tumors, the prognosis is poor despite appropriate treatment.

Introduction

Primitive neuroectodermal tumors (PNET) are malignant, aggressive, and poorly differentiated neoplasms that occur predominantly in children and young adults [1,2]. It has been classified, along with Ewing sarcoma of the bone, as Ewing sarcoma family of tumors, due to their developmental, biological, and histological similarities [3,4]. Primitive intraspinal PNET, including intramedullary, intra dural-extra medullary or extradural, are scarce, and only few cases have been described in the literature nowadays [1]. Magnetic resonance imaging remains the imaging modality of choice, allowing initial diagnosis and showing extension of the tumor [4,9,10]. Imaging is also fundamental to rule out differential diagnoses and to determine the subsequent management of the patient. We report a case of an intradural lumbar and sacral PNET with sacral bone involvement.

Case report

A 17 years-old boy presented in our department with gradually increasing low back pain, radiating to the left leg and then to the right leg, on the path of L5 and S1 roots, lasting for

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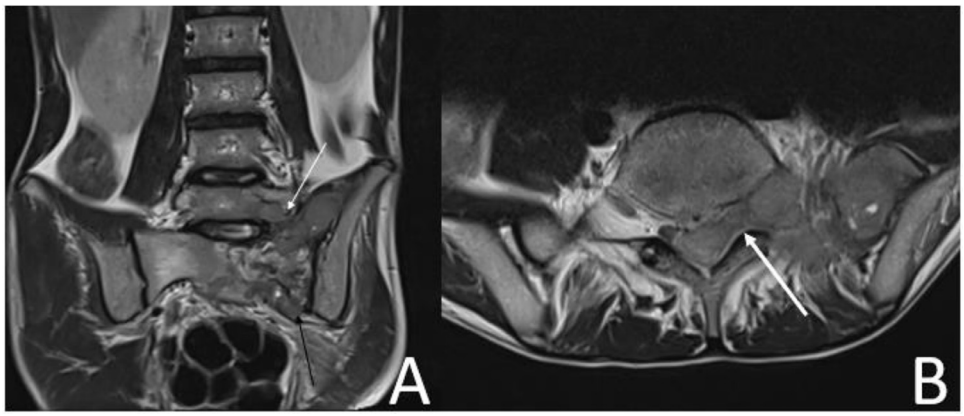

Fig. 1 – MRI images (A and B): T2W coronal (A) and T2W axial (B) show thickening with the ''hourglass sign'' in the level of fifth lumbar spinal nerve root on the left side (white arrow), and heterogenous signal at the level of S1 vertebrae (black arrow).

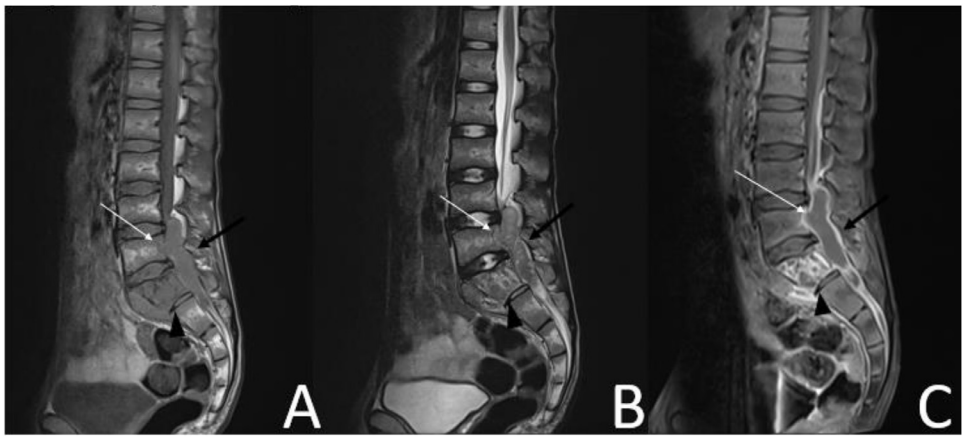

Fig. 2 – Sagittal MRI images (A-C): reveal extra medullary intra dural mass with respectively iso intense and heterogenous signal on T1 and T2W images (black arrows), extending from L4 to S2, with peripheral enhancement of the mass on post contrast sagittal T1W (C). Posterior scalloping of L5 vertebrae (white arrows) and abnormal signal within the S1 vertebrae (black head arrows) can also be noted.

10 months. The patient described the pain as severe, continuous, and resistant to analgesics. Lower extremity weakness and pain when walking have also been reported few days before. No history of trauma has been reported. The patient denied history of significant medical or surgical disease.

The neurological examination revealed weakness of proximal and distal left leg with strength testing estimated to 3/5. The Mingazzini sign was positive for the left leg. The patient had also difficulty standing up for a long time. The rest of physical examination and laboratory investigations were unremarkable.

Magnetic resonance imaging (MRI) of lumbar and sacral area has been performed, revealing a voluminous iso intense T1 and heterogenous T2 soft tissue mass, with peripheral enhancement, centered in the level of L5, where it makes the ''hourglass sign'' along a widened foramen (Fig. 1), and I. It has an intra dural extra medullary portion, extending from L4 to S2, pushing the remaining elements to the right side and causing posterior scalloping of lumbar and sacral vertebrae with cortical lysis (Fig. 2). This mass also extends though the first left sacral foramina, which is widened and lysed, and invade the left side of sacrum, more markedly at the level of S1 vertebrae. At this level, there are small area producing T2 hyper intensity, along with a slightly more heterogenous pattern of enhancement (Fig. 3). It extends through the second and third left sacral foramina with enhancement and enlargement of the corresponding sacral nerve, and infiltration of the left piriformis. There is a prevertebral extension pushing the abdominal content forward without invading it. Backward, bilateral longissimus thoracis and left psoas muscles are invaded. Based on this MRI features, the diagnosis of peripheral neuroectodermal tumor has been suspected. Few days later, a body scan has been performed, and showed significant S1 vertebrae destruction (Fig. 4).

The patient underwent emergent L4-L5-S1 laminectomy and partial resection of the mass has been done. The pathology report revealed proliferation of round cells, with scant cytoplasm and small rounded nuclei with finely nucleated chro-

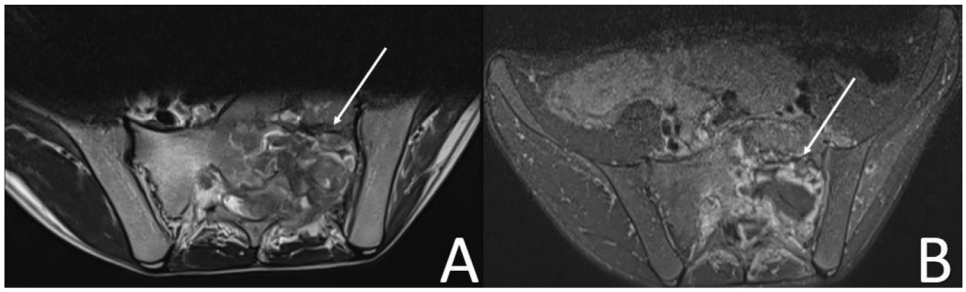

Fig. 3 – MRI images: Axial T2W (A) show heterogenous signal within the mass on the left side of sacral bone at the level of S1 vertebrae. Post contrast axial T1W (B) reveals heterogenous enhancement of the mass.

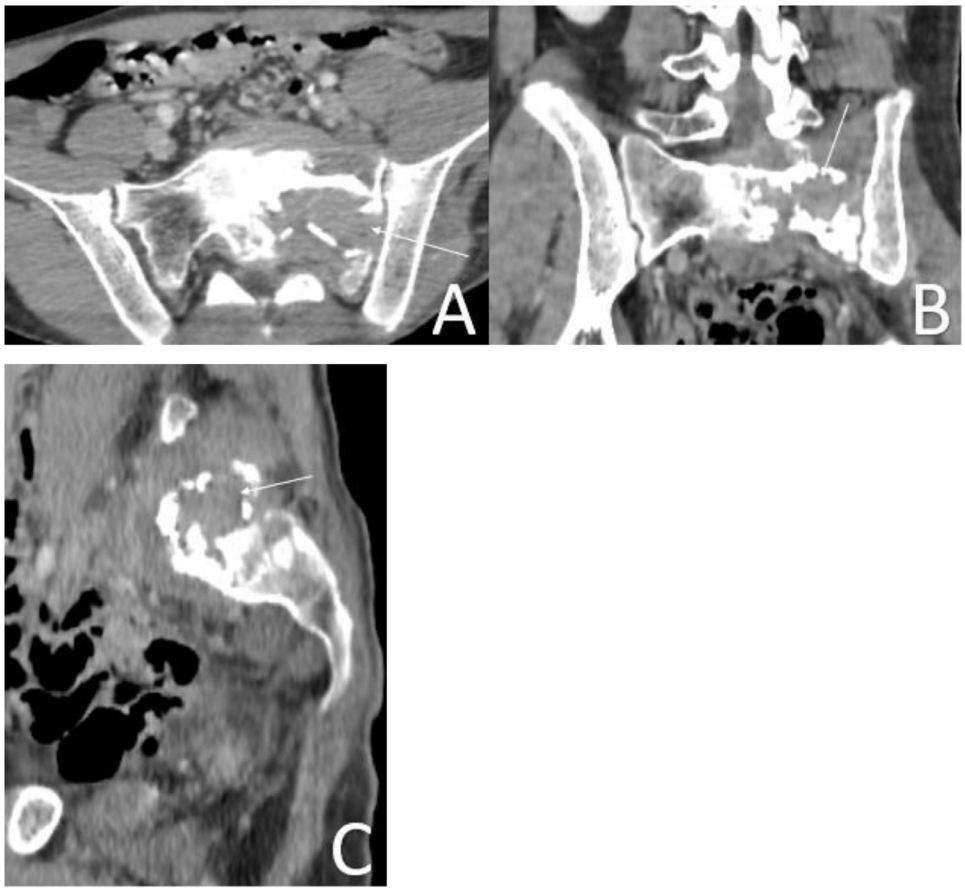

Fig. 4 – CT scan images: Axial (A), coronal (B) and sagittal (C) view showing destruction of S1.

matin, arranged in a fibrous background. Immunohistochemistry has been realized, showing the following positive markers: CD99, ERG, BCL2 and Cycline D1. The histopathological and immunohistochemical features were consistent with peripheral primitive neuroectodermal tumor (PNET/Ewing).

Discussion

PNET represent a group of malignant neoplasms arising from undifferentiated mesenchyme [4]. Theses tumors are mainly composed of undifferentiated sub ependymal neuro epithelial cells [1]. It was reported for the first time by Tefft et al in 1969 in patients presenting tumors of the paravertebral area with histopathological features similar to those of Ewing sarcoma [5]. The common histopathological aspect involves uniform round shaped cells with scanty cytoplasm and chromatic nuclei [4], which is compatible with our case. Patients in the second or third decade are the most concerned by this disease, with a very low incidence below age of 10 [6]. Contrary to Ewing's sarcoma of bone which present with a male predominance (ratio 2:1), PNET affects equally both male and female [7].

Commonly, the neoplasm seeding occurs through the central nervous system (CNS) via the cerebrospinal fluid. However, in some rare cases, it can metastasize outside the CNS. The majority of PNET invading the spinal cord represent drop metastasis that occurs in the setting of primary intracranial tumors [1,2]. In our case, the tumor arises from lumbar and sacral nerve roots, which is really uncommon. In the literature, it has been reported that theses tumors arise from subependymal primitive neuroepithelial cells. Considering that subependymal area can be present in any part of the CNS, PNET can arise from other locations than brain [2].

The common clinical features shared by lumbar/sacral PNET are related to the neurological compromise generated by theses tumors [8]. In the most of cases, the typical features of lumbosciatalgia are reported, consisting of low back pain with irradiation to lower limbs. In some cases, the cauda equina syndrome, including lower limb weakness and urinary/bowel problems, is also reported [1,2,4].

MRI is the most common initial diagnostic modality [9,10]. It is also considered as the imaging of choice, being more efficient than CT scan for showing the extension of the neoplasm and delimiting the soft tissues [4]. On MRI, the tumor is commonly iso to hypo intense on T1-weighted images, and heterogenous on T2-weighted images [1], which is consistent with our findings. Sometimes, necrotic or cystic area can be noticed within the tumor [1], which can explain the small area of T2 hyperintensity in our imaging. Bone destruction has also been reported in some other studies [11]. However, most of similar cases described in the literature report a homogenous enhancement after injection [1,2,4], contrasting with the peripheral/heterogenous enhancement in our case.

Given the fact that PNET arising from lumbar and sacral plexus are very uncommon, we have to eliminate other differential diagnosis that occur more frequently in this area. The benign tumors arising from nerve sheaths, as neurofibromas, are well circumscribed, with the classical and highly suggestive target sign related to hypo intense center and hyper intense rim on T2 weighted images. Collagen and dense Schwann cells that make up neurofibromas explain the enhancement on T1-weighted images [12]. However, the target sign is absent in our case. Benign tumors also include schwannoma which are well defined lesions with slow growing pattern, involving most commonly the lumbar area, and also exerting a mass effect on neighboring structures. Cystic component is also reported in more than 40% of cases [12]. Heterogenous enhancement can also be seen in large schwannoma [13]. Nevertheless, our MRI findings makes the diagnosis of malignancy more likely. Spinal ependymoma, especially the myxopapillary variant type, should be also considered as it involves the cauda equina region. It usually presents as an intra dural cavitation with symmetric cord expansion, showing hypo intense signal on T1-weighted images and hyper intense signal on T2-weighted images. Hypo intense rim on T2-weighted images, also known as ''cap sign'', is suggestive but not pathognomonic. However, the typical enhancement pattern is homogeneous (even if it can be variable depending on the importance of hemorrhage), and it is prone to hemorrhage [12]. Regarding spinal astrocytoma, they typically develop in an eccentric area within the spinal cord [14]. The thoracic and cervical region remain the most common location and the borders are not well defined. Given to these features, it makes the diagnosis of spinal astrocytoma less likely in our case [15]. Non-neuraxis tumors such as lymphomas (especially non-Hodgkin lymphoma) disseminate through the subarachnoid area and can result in extended thickening of the spinal nerve roots. Theses lesions show vivid enhancement after injection [12]. However, theses tumors generally occur in older people with a peak incidence between 40 and 60 years old [4]. Finally, metastasis, which show an extensive peri tumoral edema and which are more commonly found in the cervical and thoracic region, should be eliminated [15].

The treatment consists of surgery, radiotherapy and chemotherapy, which could be used separately, or in different combinations [7]. However, despite this treatment modalities, the majority of patients with intra spinal PNET die within 2 years [16]. The rapid recurrence of theses tumors explain why they are aggressive. The death is explained by several causes, including aggressive local dissemination of the disease, progressive spinal cord injury, and metastatic diseases [2].

Conclusion

Primitive neuroectodermal involving the lumbosacral region remain rare. Given to the number of potential differential diagnoses, MRI imaging features along with histopathological studies are the key to confirm the diagnosis. Despite appropriate treatment, the prognosis is poor, and the mortality rate remain high.

Patient consent

Informed consent for publication was obtained from patient.

REFERENCES

[1] Ozdemir N, Usta G, Minoglu M, Erbay AM, Bezircioglu H, Tunakan M. Primary primitive neuroectodermal tumor of the lumbar extradural space. J Neurosurg 2008;2(3):215–21. doi:10.3171/ped/2008/2/9/215.
[2] Hung J-J, Ho W-S. Sacral primitive neuroectodermal tumor: case report and review of the literature. Chin J Radiol 2008;33:281–5.
[3] Patnaik A, Mishra SS, Mishra S. Rare case of primary primitive neuroectodermal tumour of sacral region in a child and its follow-up. J Pediatr Neurosci 2013;8(1):75–7. doi:10.4103/1817-1745.111437.
[4] Narula M, Gupta N, Anand R, Kapoor S. ExtraosseousEwing's sarcoma/primitive neuroectodermal tumor of the sacral nerve plexus. Indian J Radiol Imaging 2009;19(2):151. doi:10.4103/0971-3026.50841.
[5] Tefft M, Vawter GF, Mitus A. Paravertebral "round cell" tumors in children. Radiology 1969;92:1501–9. doi:10.1148/92.7.1501.
[6] Gurney JG, Young JL, Roffers SD, Smith MA, Bunin GR. Soft tissue sarcomas. Cancer survival among children and adolescents: united states SEER program 1975-1995. Bethesda, MD: National Cancer Institute, SEER program; 1999. p. 111–23. NIH Pub No. 99-4649.

[7] Kaspers G-JJL, Veerman AJP, Kamphorst W, van de Graaff M, van Alphen HAM. Primary spinal epidural extraosseous ewing's sarcoma. Cancer 1991;68(3):648–54. doi:10.1002/1097-0142(19910801)68:3<648::aid-cncr2820680335>3.0.co;2-q.
[8] Musahl V, Rihn JA, Fumich FE, Kang JD. Sacral intraspinal extradural primitive neuroectodermal tumor. Spine J 2008;8(6):1024–9. doi:10.1016/j.spinee.2007.04.001.
[9] Kim YW, Jin BH, Kim TS, Cho YE. Primary intraspinal primitive neuroectodermal tumor at conus medullaris. Yonsei Med J 2004;45:533–8. doi:10.3349/ymj.2004.45.3.533.
[10] Virani MJ, Jain S. Primary intraspinal primitive neuroectodermal tumor (PNET): a rare occurence. Neurol India 2002;50:75–80.
[11] Zhang YW, Xia WH, Gao WC, Yan L, Xiao X, Xiao Y, et al. Giant primitive neuroectodermal pelvic tumour : a case report and literature review. J int Med Res 2020;48(6):030006052090674.
[12] Osborn Anne G. Illustrated by Julian Macck. Cysts, tumors, and tumorlike lesions of the spine and spinal cord. In: Diagnostic neuroradiology. Harcourt brace and company. Amsterdam, The Netherlands: Elsevier; 1994. p. 876–915.
[13] Skolnik AD, Loevner LA, Sampathu DM, Newman JG, Lee JY, Bagley LJ, Learned KO. Cranial nerve schwannomas: diagnostic imaging approach. Radiographics 2016;36:1463–77. doi:10.1148/rg.2016150199.
[14] Wein S, Sharma R, Rasuli B. Spinal astrocytoma. Reference article, Radiopaedia.org (Accessed June 2, 2024) doi:10.53347/rID-19274.
[15] Wein S, Rasuli B, Bickle I. Spinal primitive neuroectodermal tumor. Reference article, Radiopaedia.org (Accessed June 2, 2024) doi:10.53347/rID-19277.
[16] Hamed RK, Batool A. Primary intraspinal primitive neuroectodermal tumors: report of a case. Arch Iranian Med 2002;5:262–6.