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CASE REPORT

Malignant peripheral nerve sheath tumor in the paraspinal region mimicking a benign peripheral nerve sheath tumor: a case report

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Abstract
Purpose Malignant peripheral nerve sheath tumors are extremely rare in the general population and display a predilection for metastasis to the lungs. Here, we present a rare case of a malignant peripheral nerve sheath tumor located in the paraspinal region and highlight the importance of preoperative biopsy in diagnosis of spinal epidural peripheral nerve sheath tumors.
Methods We describe the clinical course of the patient as well as the radiological and pathological findings of the tumor.
Results A 14-year-old girl presented with a six-month history of sacral pain. Occasionally she experienced left leg pain and abnormal gait. General physical examination revealed sensorial loss in the L5–S1 regions. T1-weighted sagittal MRI showed a hypointense oval mass and the contrast-enhanced T1-weighted axial MRI image showed heterogeneous enhancement of the tumor. On CT imaging, this tumor characteristically appears as a dumbbell-like mass with punctate calcification and widening L5–S1 intervertebral foramen. Complete resection was performed using an anterior approach. Intraoperative pathological examination revealed evidence of malignancy and subsequent immunohistochemical analysis of the tumor confirmed the diagnosis of MPNST. The postoperative course was uneventful and the patient has had significant improvement in her symptoms 1 month postoperatively.
Conclusions Preoperative biopsy should be routinely performed for pathological differential diagnosis of spinal epidural PNSTs as well as surgical decision-making. Furthermore a combination of clinical manifestation, radiological findings and biopsy should also be pursued for diagnosing these tumors.

Keywords Malignant peripheral nerve sheath tumor (MPNST) · Benign peripheral nerve sheath tumor (BPNST) · Spinal epidural peripheral nerve sheath tumors (spinal epidural PNST) · Biopsy · Diagnosis

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are malignant tumors derived from a major or minor peripheral nerve branch or sheath of peripheral nerve fibers. MPNST usually occurs between 20 and 50 years of age, comprising about 5–10 % of soft tissue sarcomas [1, 2]. Although MPNST is uncommon in young patients, it also represents about 5–10 % of all soft tissue sarcomas of childhood [3, 4]. MPNSTs are most commonly located in the trunk and extremities [5]. Spinal MPNST is rare and primary spinal MPNST in children are rarer. Given its aggressive behavior, prompt and accurate diagnosis is critical for improving patient outcomes. Therefore, preoperative biopsy is necessary and helpful for surgical decision-making and preoperative preparation. In general, biopsies of spinal epidural peripheral nerve sheath tumors are performed in patients with presumed malignant tumors according to the clinical and imaging findings. However, preoperative biopsies of spinal epidural MPNSTs are often omitted due to their nonspecific and benign imaging findings.

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Here, we are describing a case of spinal epidural MPNST in the paraspinal region mimicking a BPNST to highlight the importance of preoperative biopsy in diagnosis of spinal epidural peripheral nerve sheath tumors (spinal epidural PNST).

Case report

A 14-year-old girl presented with a six-month history of sacral pain. Occasionally she experienced left leg pain and abnormal gait. During the last four months, her symptoms were aggravated and came to our hospital for treatment. General physical examination revealed sensorial loss in the L5–S1 regions. Bilateral deep tendon reflexes at both knees and ankles were normal. There were no signs of fasciculation, muscle atrophy, and upper extremity motor neuron dysfunction on her physical examination. There were no dermatologic signs of neurofibromatosis. She had no history of trauma. There were no defecation and urinary system complaints.

A plain radiograph demonstrated a mass lesion on the left sacrum and presacral space (Fig. 1). Lumbosacral CT showed a big and well-circumscribed mass extending from the L5–S1 intervertebral foramen into the left lateral aspect of the vertebra. A dumbbell-like mass with punctate calcification and widening L5–S1 intervertebral foramen was also revealed. Psoas muscle was pushed forward by the mass (Fig. 2a). After contrast enhancement, the mass presented a dumbbell-like shape and heterogeneous enhancement (Fig. 2b). The destructive and invasion of the spinal canal and erosion of the vertebral body was not revealed by CT. Moreover, CT of the chest showed no metastatic lesions in both lungs. MRI of the lumbar spine without contrast revealed a level. Preoperative MRI showed a well-circumscribed paraspinal lesion with its size 41 × 49 × 49 mm, no soft tissue involvement and complete tumor capsule. T1 weighted sagittal MRI showed a hypointense oval mass (Fig. 3a). After contrast enhancement, the mass presented a heterogeneous enhancement on T1-weighted axial MRI (Fig. 3b). On T2 weighted MRI without contrast, the mass showed a mixed signal of high and medium intensity (Fig. 3c).

On the basis of these imaging findings and clinical manifestation, we mostly suspected paraspinal benign peripheral nerve sheath tumors (BPNSTs) such as schwannoma and neurofibroma, although malignancy was not completely excluded. Then surgical resection was performed using an anterior approach. However, the intraoperative tumor capsule was incomplete and intraoperative pathological examination of a frozen section revealed evidence of malignancy. Thus, complete excision of the tumor was preferentially performed. Subsequent immunohistochemical reevaluation of the specimen showed that the tumor cells were positive for S-100, vimentin, CD31, CD34, CD56, CD117, Syn, Bcl-2, NSE, EMA, and negative for SMA, HMB45, melan-A,CD99, CgA, CK (AE1/AE3), CAM5.2, GFAP and desmin. The MIB-1 labeling index (Ki-67), which represents a marker of cell proliferation, was 30–40 %. Based on these findings, we finally diagnosed the tumor as a MPNST.

Postoperative MRI showed gross-total resection with some peri-resection site changes. The patient had a rapid recovery with no complications. Her gait is normal and her pain has completely resolved. She has no neurological deficits with no evidence of bowel/bladder dysfunction.

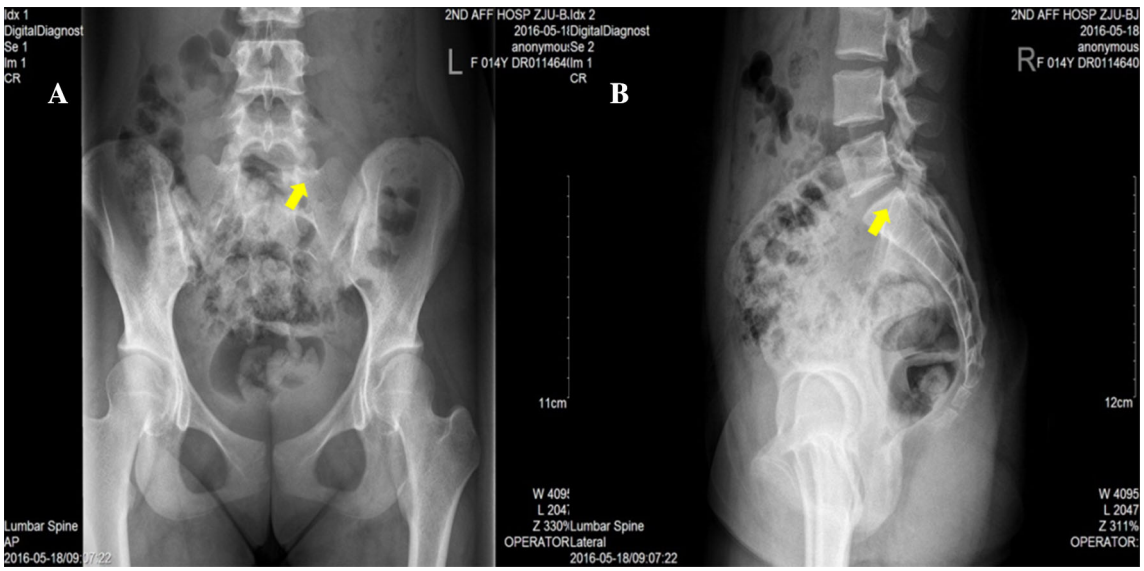

Fig. 1 Simple radiograph shows a mass lesion located in the left sacrum and presacral space (**a**, **b**)

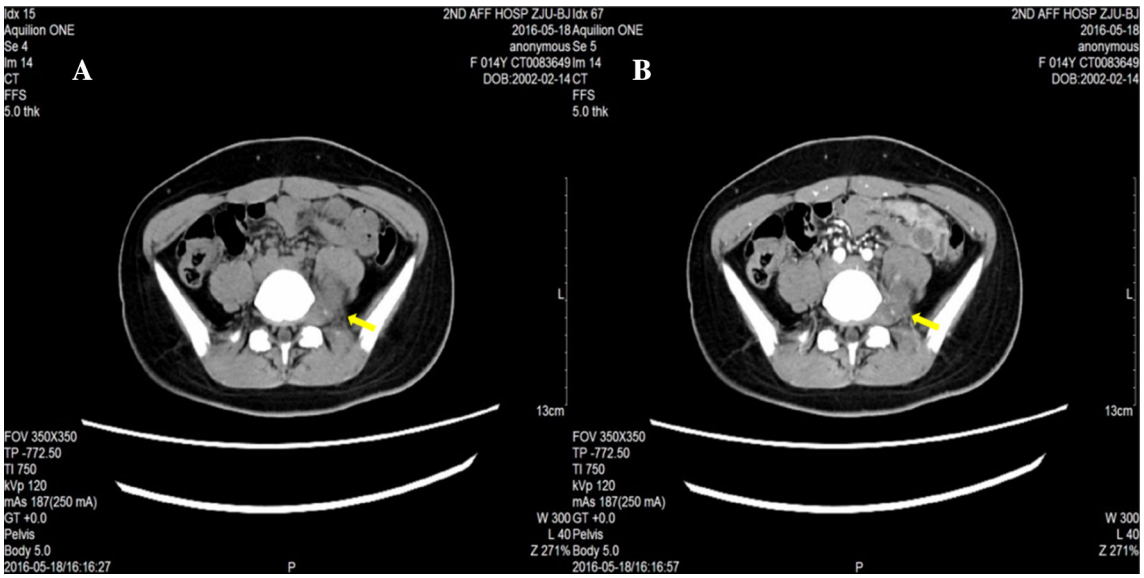

Fig. 2 Computed tomography showed a big mass extending from the L5–S1 intervertebral foramen into the left lateral aspect of the vertebra. A *dumbbell-like* and soft tissue mass with punctate calcification and widening L5–S1 intervertebral foramen was also revealed. Psoas muscle was pushed forward by the mass (**a**). After contrast enhancement, the mass presented a *dumbbell-like* shape and heterogeneous enhancement (**b**)

Follow-up MRI at 1 month showed the postoperative changes, with no evidence of residual tumor.

Discussion

MPNST has been previously known as neurofibrosarcoma, neurogenic sarcoma, malignant schwannoma, and malignant neurilemmoma [6]. The major differential diagnosis is BPNSTs. The mainstay treatment for MPNST is complete resection with the aim of achieving clear surgical margins [7]. While BPNSTs well respond to marginal excision with preservation of neurologic function [8]. Thus, the early and accurate diagnosis for spinal epidural PNSTs is particularly important for the choice of optimal treatment.

The preferred modality for imaging of MPNSTs is MRI for the primary lesion. However, benign imaging findings of our case with MPNST were actually shown on MRI and CT. T1 weighted sagittal MRI showed a hypointense oval mass and the contrast-enhanced T1-weighted axial MRI image showed heterogeneous enhancement of the tumor. CT chest was done to exclude pulmonary metastases. On CT imaging, this tumor characteristically appears as a dumbbell-like mass with punctate calcification and widening L5–S1 intervertebral foramen. As we all know, benign spinal dumbbell tumors (SDTs) such as schwannomas and neurofibromas occur frequently while malignant SDTs, including MPNSTs, are extremely rare [9]. So, paraspinal BPNSTs such as schwannoma and neurofibroma were mostly suspected. We agreed that preoperative diagnosis based on imaging findings would be helpful in selecting therapeutic methods for SDTs. Surprisingly, intraoperative pathological examination of a frozen section revealed evidence of malignancy and postoperative immunohistochemical reevaluation of the specimen showed MPNST. Additionally, the intraoperative tumor capsule is incomplete, which was contrary to preoperative MRI findings. Recently, evidence shows that MRI is unreliable to determine the diagnosis of MPNST and guide surgical decision-making [10]. Therefore, pre-operative biopsy is essential for precise diagnosis of spinal epidural PNSTs.

Matsumoto Y et al. has recently established a new scoring system, the dumbbell scoring system (DSS), for preoperative evaluation of the malignant potential of SDTs. This scoring system based on imaging findings includes four predictors namely size, boundary, irregularly shape and osteolytic bone destruction. If the DSS score is equal to or higher than 3, biopsies was recommended to confirm the histological diagnosis [11]. According to the rules of DSS, the DSS score of our case was 0, which meant preoperative biopsy was not definitely recommended. However, our patient without biopsy was finally diagnosed as a MPNST. Of course, the limitations of DSS included its retrospective design and small sample size, which was mentioned in the study [11]. Ozawa H et al. found that malignant dumbbell tumors were more common in children younger than 10 years of age than in older patients [9]. So other factors may also be considered in creating the new DSS in the future studies. Currently, however, we hold the

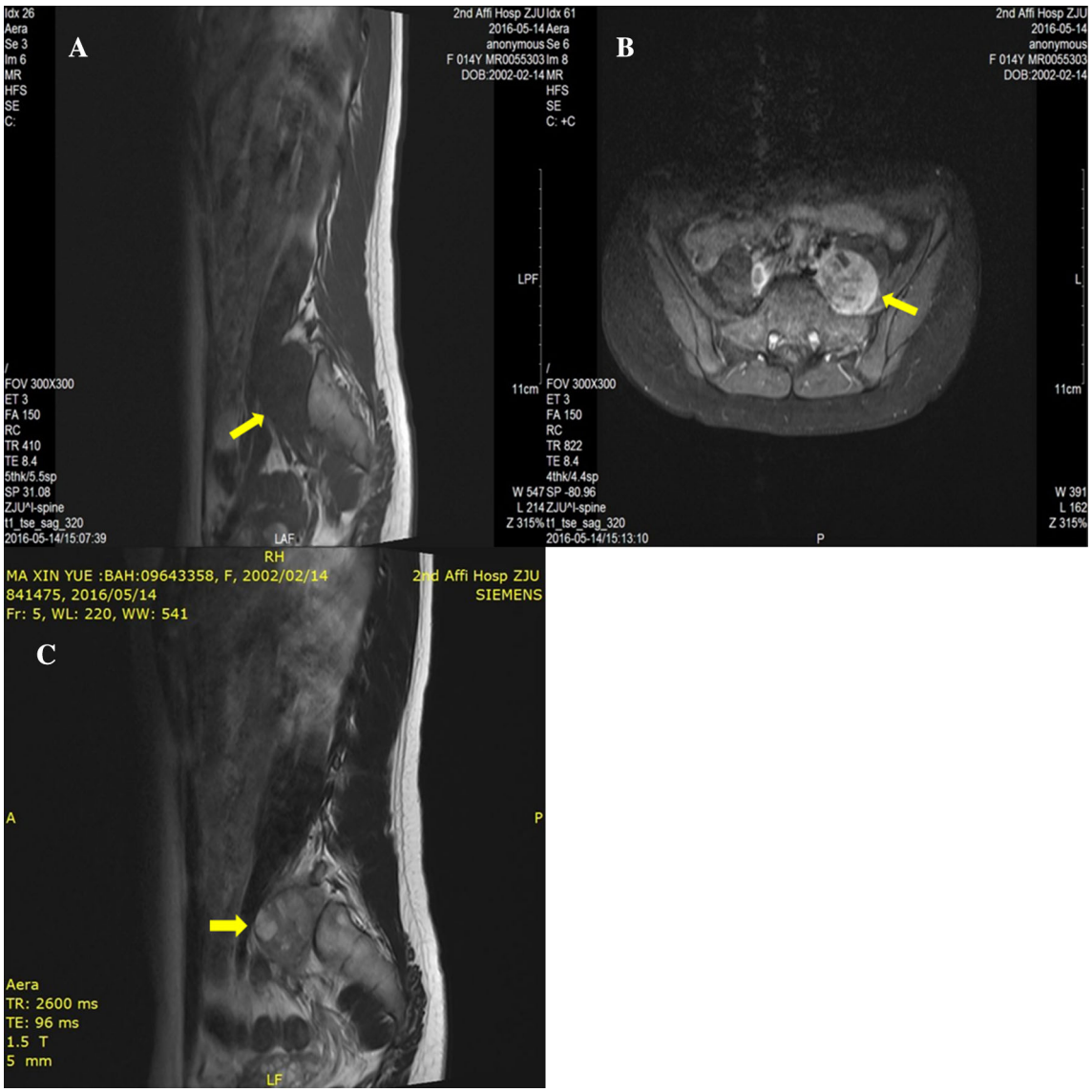

Fig. 3 T1-weighted sagittal MRI showed a hypointense oval mass (**a**). The contrast-enhanced T1-weighted axial MRI image showed heterogeneous enhancement of the tumor (**b**). The mass showed a mixed signal of high and medium intensity on T2-weighted MRI without contrast (**c**)

view that a combination of clinical manifestation, radiological findings and biopsy should be used for preoperative diagnosis of SDTs.

Biopsy of tumors located in the spine may be associated with pain, risk, expense, and needle tract seeding. But Pianta M et al. [12] reported that CT-guided core biopsy of PNSTs is a safe procedure with excellent diagnostic accuracy (100 %). The only occurring complication (12 %) was pain exacerbation but resolved within 1 week or after a longer duration. Needle tract seeding refers to implantation of tumor cells by contamination when biopsy needles are used to obtain tumor tissues. To our knowledge, while this iatrogenic phenomenon is reported in treating spinal metastases [13, 14], it has not been reported in spinal epidural PNSTs. Moreover, the incidence of the needle tract seeding may be avoided with precautionary measures and adopting the local treatment methods such as wide excision and radiotherapy which can remove or kill the implanted tumor cells along the needle tract [15]. In brief, biopsy of spinal epidural PNSTs is a safe and accurate procedure for diagnosing these tumors. Therefore, while weighing the pros and cons of preoperative biopsy, we believe it should be conducive to confirming the pathological nature of spinal epidural PNSTs, and developing a comprehensive treatment procedure.

Conclusions

On the whole, we recommend that preoperative biopsy should be routinely performed for pathological differential diagnosis of spinal epidural PNSTs as well as surgical decision-making. Furthermore a combination of clinical manifestation, radiological findings and biopsy should also be pursued for diagnosing these tumors.

Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest.

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Informed consent The patient and/or her family were informed that data from the case would be submitted for publication, and gave their consent.

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