

Case Report

Primary Intramedullary Spinal Sarcoma : A Case Report and Review of the Current Literatures

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Primary central nervous system (CNS) sarcomas are exceedingly rare, and, to the best of our knowledge, there has not yet been a report of intramedullary sarcoma. Here, we report a primary intradural intramedullary sarcoma of the spinal cord in a four-year-old boy who presented with low back pain and a radiculopathy involving both lower extremities. The tumor showed significant enhancement on magnetic resonance (MR) images due to its extreme vascularity. Gross total tumor removal was performed with microelectrical pulse recording, and the patient also received adjuvant radiotherapy and chemotherapy. After the operation, the patient's sensory deficits were improved. Because CNS dissemination is common, entire neuraxis evaluation is essential, although there was no evidence of dissemination in this case. The prognosis of primary CNS sarcoma is poor due to infiltrative nature and early CNS dissemination is common, and the treatment of choice is radical surgical resection. Adjuvant therapy is also beneficial with radiotherapy and chemotherapy.

KEY WORDS : Central nervous system · Intramedullary · Primary · Sarcoma.

INTRODUCTION

Primary sarcomas of the central nervous system are exceedingly rare. There have been reports of extramedullary spinal cord sarcomas; however, there has not yet been a report of intradural intramedullary primary sarcoma in humans. The only article we found was written by Vandeveldt et al.²⁰⁾ in 1976, and it reported an intramedullary sarcoma in a dog. Here, we describe an intramedullary sarcoma arising at the T11-T12 level in a four-year-old boy.

CASE REPORT

A four-year-old boy was admitted to our department with complaints of low back pain and a radiculopathy involving both lower extremities for the previous three weeks. He did not have a family history of cancer or other tumorous conditions. On neurologic examination, he did not have motor

weakness, although he did have sensory deficits at the anterolateral aspect of his thighs. Deep tendon reflexes were increased bilaterally at the knee and ankle, and pathologic reflexes, including ankle clonus and the Babinski sign, were also present. He had not experienced bowel or bladder incontinence.

Magnetic resonance imaging (MRI) revealed a 13 × 26 mm enhancing intramedullary mass with peritumoral edema and contrast enhancement of the pial layer at the T11-T12 level (Fig. 1). There were no other mass lesions along the neuraxis.

Two weeks after admission, he underwent an operation under general anesthesia for mass removal and histological confirmation. The T10, T11, and T12 laminae were removed by drilling, and there were no observed changes in the dura surface. The dural sac was then incised, and no tumor was found in the intradural extramedullary space. However, a tumor was found in the intradural intramedullary space at T11-T12 using a midline incision of the cord. The tumor was grayish, friable, and clearly demarcated from the spinal cord (Fig. 2). The tumor was removed with microscopic assistance using microelectrical pulse (NIM spine, Medtronic co. Minnesota, USA) recording. After hemostasis was achieved, the dura was closed with watertight fashion.

On histological examination, highly pleomorphic malig-

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nant cells including bizarre nuclei, frequent mitotic figures, and myxoid stroma were noted (Fig. 3A). According to the immunohistochemical report, the tumor was possibly of mesenchymal origin because of positive vimentin staining (Fig. 3B). The tumor was also positive for Ki-67 (20%) and p53 (10%). The other immunohistochemical stainings performed were negative including S-100, myoglobin, cytokeratin, MyoD1, synaptophysin, lens cullnaris agglutinin (LCA), ethidium monoazide (EMA), smooth muscle antibody (SMA), muscle specific actin (MSA), and CD99 and there was no morphologic evidence to suggest a neuroglial tumor. The pathologist concluded this tumor as highly malignant mesenchymal tumor - more like sarcoma - according to morphological and immunohistological pattern without the evi-

dence of neuroglial tumor. The patient did develop postoperative motor weakness, but this improved over a few weeks. The post-operative MRI on the 7 days showed intramedullary enhancement at the surgical site, which was possibly due to postoperative edema and hematoma (Fig. 4). After the operation, the patient received adjuvant conventional radiotherapy with 25 times fractionated 3,940 cGy and chemotherapy with ifofdomide and doxorubicin during 16 weeks. Follow-up MRI showed disappearance of the previous contrast-enhancing lesion and no tumor recurrence at one year postoperative (Fig. 5).

DISCUSSION

Primary sarcomas of the central nervous system are very rare. According to a paper by the Mayo clinic, only 66 primary CNS sarcomas have been identified (0.7%)¹⁹. While there have been several reports of extramedullary and epidural spinal sarcomas, a primary sarcoma of the spinal cord

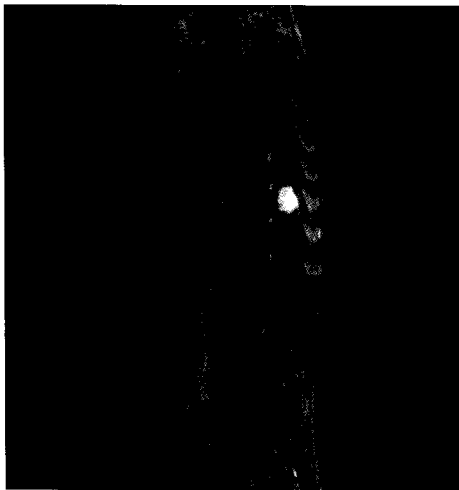


Fig. 1. Preoperative thoracic magnetic resonance images. A 13 × 26 mm bulging, contoured, strongly enhanced intramedullary mass is evident at T11-12. Peritumoral edema and contrast enhancement of the pial layer are also noted on the sagittal scan.



Fig. 2. Intraoperative microscopic view shows an intradural intramedullary mass. The mass is grayish, friable, and clearly demarcated from the spinal cord (blue arrow).

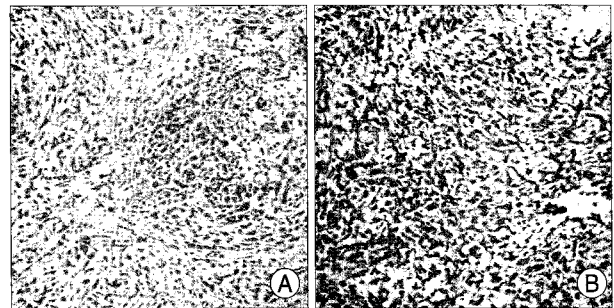


Fig. 3. Histological findings. A : H & E staining shows highly pleomorphic malignant tumor cells characterized by bizarre nuclei, frequent mitotic figures, and myxoid stromal background (× 200). B : Vimentin-positive cells are observed in immunohistochemical staining (× 200).

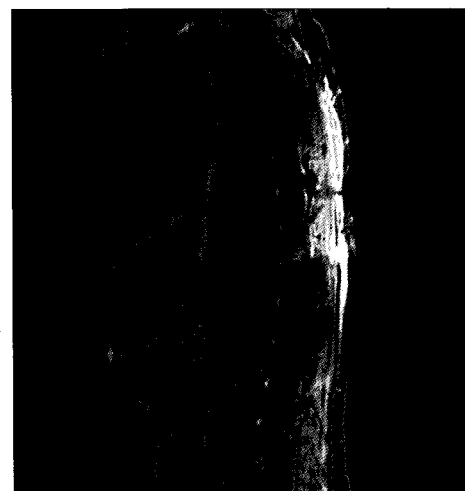


Fig. 4. Postoperative thoracic magnetic resonance images. There is a slight high signal change at T11-12, possibly due to postoperative hemorrhage, on the seventh postoperative day. Spinal cord edema is also noted at the operative site. The residual tumor is not identified.



Fig. 5. Thoracic magnetic resonance images reveal no tumor recurrence one year after the surgery.

had not yet been reported^{1,19)}.

The cellular origin of CNS sarcomas is controversial. The most widely accepted theory is that they originate from pluripotent primitive mesenchymal cells in the dura mater, the leptomeninges, or their pial extensions into the brain and the spinal cord along the periaxonal spaces, the tela choroidea, and the stroma of the choroid plexus^{2-4,12,13,16,18,20,25)}. However, intramedullary location has not been explained due to its rarity.

Recently, several risk factors including prior radiotherapy and chemotherapy for the development of primary sarcoma have been proposed^{9,11,14,22,24)}. However, of all the potential risk factors, only prior radiation therapy is well accepted as an inciting agent^{9,11,14,22,24)}, although other risk factors for primary central nervous system sarcoma have been suggested, such as trauma genetic factors and chemicals, the evidence is far from conclusive^{4,7,9,10,14-16)}.

The clinical presentation of patients with primary spinal cord sarcoma is non-specific and variable and is related to the level and location of the lesion. The same is true for other intrinsic and meningeal tumors.

The appearance of primary sarcoma on CT and/or MRI is also non-specific. There are no specific imaging criteria to differentiate primary sarcomas from other varieties of spinal tumors or inflammatory masses. Mubarak et al. reported that spinal sarcomas were heterogeneous in density, with cystic and solid components in seven of 17 cases¹⁾. According to other reports, enhancement is a typical feature of sarcomas because of their extreme vascularity^{1,20)}. Because of the high incidence of leptomeningeal spread of sarcomas, it is important to study the entire neuraxis at the time of diagnosis and during the follow-up period²⁰⁾. While CSF dissemination has been reported in about 40% of the prior reports, there were no malignant cells in the CSF of our patient. Addi-

tionally, no other masses involving the neuraxis were found¹⁾.

In reviewing the current literature, sarcoma in children tend to be more undifferentiated or show muscle differentiation, in contrary to fibrous histiocytomas and fibrosarcomas in adult^{21,23,28)}. The histology of our patient's tumor revealed that it was of the undifferentiated type which composed of highly pleomorphic small round cells by characterized with bizarre nuclei, frequent mitotic figures, and myxoid stromal background. Ki-67 immunolabeling has been found to be positive in over 80% of cases, and Andre et al. reported vimentin positivity in 12 of their 13 cases. For spinal sarcoma, immunohistochemical staining was only performed in three cases¹⁾, which showed positive staining for desmotin (one case), vimentin (two cases), and S100 (one case)¹⁹⁾. Our patient's tumor was positive for only vimentin, suggesting that it was a mesenchymal tumor, while there was no evidence to suggest the presence of neuroglial cells.

In general, primary CNS sarcomas carry a poor prognosis due to highly infiltrative nature and early CNS dissemination¹⁾. Most reports in the literature point toward radical surgical resection as the treatment of choice. There is no consensus on the utility of postoperative radiation or chemotherapy^{5-8,22,25,27)}. Merimsky et al.¹⁷⁾ reported that surgery without adjuvant radiation therapy yielded less favorable results comparing to only surgical excision, while the best results were achieved using high-energy electron or photon beams administered in a median dose of 50 Gy in 25 fractions. The value of chemotherapy is also controversial, but long-term survivors with mean 3.3 years who received chemotherapy have been reported¹⁾. Considering all those reports, the use of postoperative radiation and/or chemotherapy may be justified.

CONCLUSION

Primary sarcoma of the CNS is extremely rare, and a case of intraspinal intramedullary sarcoma has not previously been reported. As far as we know, this case was the first report of primary intramedullary sarcoma. The treatment of choice is radical surgical resection. However, due to high CNS dissemination rate adjuvant radiation and/or chemotherapy may improve control of the disease and may improve the overall prognosis.

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