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 Vandevelde et al.26) 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 laminas 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-
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 culinaris 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 evidence of neuroglial tumor. The patient did develop post-operative 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 ifosfamide 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%)\(^\text{19}\). While there have been several reports of extramedullary and epidural spinal sarcomas, a primary sarcoma of the spinal cord...
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 periadventitial spaces, the tela choroidea, and the stroma of the choroid plexus. 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. However, of all the potential risk factors, only prior radiation therapy is well accepted as an inciting agent; 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.

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 cases. According to other reports, enhancement is a typical feature of sarcomas because of their extreme vascularity.

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.

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.

References
1. Al-Gahtany M, Shroff M, Bouffet E, Dirks P, Drake J, Humphreys R, et al.: Primary central nervous system sarcomas in children: clinical, radiological, and pathological features. Childs Nerv Syst 19: 808-817, 2003
2. Arumugasamy N: Some neuropathologic aspects of intracranial sarcomas. Med J Malaya 23: 169-173, 1969
3. Bahr AL, Gayler BW: Cranial chondrosarcomas. Report of four cases.
and review of the literature. Radiology 124 : 151-156, 1977
4. Charman HP, Lowenstein DH, Cho KG, DeArmond SJ, Wilson CB : Primary cerebral angiosarcoma. Case report. J Neurosurg 68 : 806-810, 1988
5. Dropcho EJ, Allen JC : Primary intracranial rhabdomyosarcoma : case report and review of the literature. J Neurooncol 5 : 139-150, 1987
6. Gaspar LE, Mackenzie IR, Gilbert JJ, Kaufmann JC, Fisher BF, Macdonald DR, et al. : Primary cerebral fibrosarcomas. Clinicopathologic study and review of the literature. Cancer 72 : 3277-3281, 1993
7. Ho YS, Wei CH, Tsai MD, Wai YY : Intracerebral malignant fibrous histiocytoma : case report and review of the literature. Neurosurgery 31 : 567-571, 1992
8. Hockley AD, Hoffman HJ, Hendrick EB : Occipital mesenchymal tumors of infancy. Report of three cases. J Neurosurg 46 : 239-244, 1977
9. Ironside JW : Classification of primary intracranial sarcomas and other central nervous system neoplasms. Histopathology 18 : 483-486, 1991
10. Kishikawa T, Numaguchi Y, Fukui M, Komaki S, Ikeda J, Kitamura K, et al. : Primary intracranial sarcomas: radiological diagnosis with emphasis on arteriography. Neuroradiology 21 : 25-31, 1981
11. Lalitha VS, Rubinstein LJ : Reactive glioma in intracranial sarcoma : a form of mixed sarcoma and glioma (“sarcoglioma”) : report of eight cases. Cancer 43 : 246-257, 1979
12. Lam RM, Malik GM, Chason JL : Osteosarcoma of meninges: clinical, light, and ultrastructural observations of a case. Am J Surg Pathol 5 : 203-208, 1981
13. Legier JF, Wells HA Jr : Primary cerebellar rhabdomyosarcoma. Case report. J Neurosurg 26 : 436-438, 1967
14. Martinez-Salazar A, Supler M, Rojiani AM : Primary intracerebral malignant fibrous histiocytes tumor: immunohistochemical findings and etiopathogenetic considerations. Mod Pathol 10 : 149-154, 1997
15. Mayo CM, Barton KD : Concurrent glioma and primary intracranial sarcoma. A report of two cases and a review of the literature. Neurology 16 : 662-672, 1966
16. Mena H, Garcia JH : Primary brain sarcomas : light and electron microscopic features. Cancer 42 : 1298-1307, 1978
17. Merinosky O, Lepetichoux C, Terrier P, Vanel D, Delord JP, LeCene A : Primary sarcomas of the central nervous system. Oncology 58 : 210-214, 2000
18. Min K-W, Gyorkey F, Halbert B : Primary rhabdomyosarcoma of the cerebrum. Cancer 35 : 1405-1411, 1975
19. Oliveira AM, Scheithauer BW, Salomao DR, Parisi JE, Burger PC, Nasimiero AG : Primary sarcomas of the brain and spinal cord: a study of 18 cases. Am J Surg Pathol 26 : 1056-1063, 2002
20. Onofrio BM, Kernohan JW, Uihlein A : Primary meningeal sarcoma-tosis: A review of the literature and report of 12 cases. Cancer 15 : 1197-1208, 1962
21. Paulus W, Slowik F, Jellinger K : Primary intracranial sarcomas: histopathological features of 19 cases. Histopathology 18 : 395-402, 1991
22. Roosen N, Cras P, Paquier P, Martin JJ : Primary thalamic malignant fibrous histiocytoma of the dominant hemisphere causing severe neuropsychological symptoms. Clin Neuropathol 8 : 16-21, 1989
23. Shuangshoti S, Piyaratn P, Viriyapanich PL : Primary rhabdomyosarcoma of cerebellum—necropsy report. Cancer 22 : 367-371, 1968
24. Stone JL, Zavala G, Bailey OT : Mixed malignant mesenchymal tumor of the cerebellar vermis. Cancer 44 : 2165-2172, 1979
25. Tomita T, Gonzalez-Crussi F : Intracranial primary nonlymphomatous sarcomas in children: experience with eight cases and review of the literature. Neurosurgery 14 : 529-540, 1984
26. Vandevelde M, Higgins RJ, Greene CE : Neoplasms of mesenchymal origin in the spinal cord and nerve roots of three dogs. Vet Pathol 13 : 47-58, 1976
27. Younis GA, Sawaya R, DeMonte F, Hess KR, Albrecht S, Bruner JM : Aggressive meningeal tumors: review of a series. J Neurosurg 82 : 17-27, 1995
28. Zwartverwer FL, Kaplan AM, Hart MC, Hertel GA, Sphataro J : Meningeal sarcoma of the spinal cord in a newborn. Arch Neurol 35 : 844-846, 1978