Intradural Extramedullary Spinal Tumor Suspected Angiosarcoma Based on Clinical Course and Pathological Findings: A Case Report

Yasushi Iijima[1)], Takeo Furuya[2)], Toshiaki Kotani[1)], Tsuyoshi Sakuma[1)], Keita Nakayama[1)], Daisuke Sasai[3)], Satoshi Murakami[4)], Satoshi Maki[2)], Takuya Miyamoto[2)], Sho Okimatsu[2)], Shunji Kishida[1)], Yuta Muramatsu[1)], Yu Sasaki[1)], Keisuke Ueno[1)], Daisuke Kajiwara[1)], Seiji Ohtori[2)] and Shohei Minami[1)]

1) Department of Orthopedic Surgery, Seirei Sakura Citizen Hospital, Chiba, Japan
2) Department of Orthopedic Surgery, Graduate School of Medicine, Chiba University, Chiba, Japan
3) Department of Pathology, Seirei Sakura Citizen Hospital, Chiba, Japan
4) Department of Palliative Medicine, Seirei Sakura Citizen Hospital, Chiba, Japan

Keywords:
Angiosarcoma, Intradural extramedullary tumor, Radiation

Spine Surg Relat Res 2022; 6(5): 577-580
dx.doi.org/10.22603/ssrr.2021-0270

Intradural extramedullary spinal tumors are mostly benign and rarely malignant[1)]. Angiosarcoma is a rare malignant vascular tumor and occurs predominantly in soft tissues[2)]. Here we report an intradural extramedullary spinal tumor suspected as being an angiosarcoma.

A 47-year-old woman was referred to our hospital with a complaint of numbness and acute progression of paralysis of both lower limbs. Neurological examination of the patient at admission revealed analgesia in the lower limbs and anus bilaterally, and complete motor paralysis of the lower muscles bilaterally. Magnetic resonance imaging (MRI) showed an intradural mass located from T12 to L2 and compressing the spinal cord (Fig. 1). Based on these findings, an intradural hemorrhagic lesion due to an arteriovenous fistula or tumor was suspected. Emergency surgery consisting of a laminectomy and durotomy from T12 to L2 was conducted and a dark red mass was seen in the subarachnoid space (Fig. 2A). We performed a gross total resection (Fig. 2B).

After surgery, complete paralysis of lower limbs remained. Histopathological findings showed atypical nuclei with proliferation of blood vessels (Fig. 3A, B). Cluster of differentiation (CD) 31 and 34 antigen was positive (Fig. 3 C, D). S-100, epithelial membrane antigen, and glial fibrillary acidic protein staining were negative. Ki-67 was 10%-20%, but few mitotic cells were seen. Initial diagnosis was hemangioma. Two weeks postoperatively, MRI showed the mass was mostly resected, but a small mass remained below the conus medullaris (Fig. 4A, B). Imaging studies at this time showed no tumor on the cranial side above Th12. Hemangioma was diagnosed at that time, so conservation and follow-up were chosen. Five weeks postoperatively, MRI showed the mass had expanded and cerebrospinal fluid (CSF) dissemination was seen at C1 and L5 (Fig. 4C, D). A malignant tumor was suspected clinically and histopathology was reviewed. Spindle cells were seen in the stroma, but not in the vascular endothelium (Fig. 3A, B). CD31 and 34 antigen staining was limited to vascular endothelial cells, and not seen in stromal cells (Fig. 3C, D). Pathology found stromal tumors of unknown differentiation with a proliferation of blood vessels. Ultimately, an angiosarcoma was suspected by the clinical course, but a definitive diagnosis could not be made. The patient already had numerous disseminated lesions, and surgical resection was not an option. Radiation therapy to the whole brain and spinal cord was performed. However, MRI after radiotherapy showed the mass had expanded at the cervical spine (Fig. 4D). Chemotherapy was not chosen because durability was limited[3)]. The best supportive care was chosen, but 6 months postsurgery, the patient died of respiratory muscle failure.

To our knowledge, this is the first report of a primary intradural extramedullary spinal tumor suspected as an angiosarcoma, which usually occurs predominantly in the soft tissue of the head and neck[2)]. Primary spinal angiosarcomas have seldom been reported[4-7)]. Moreover, origins were all

Corresponding author: Yasushi Iijima, yi02008@yahoo.co.jp
Received: January 5, 2022, Accepted: February 17, 2022, Advance Publication: April 12, 2022

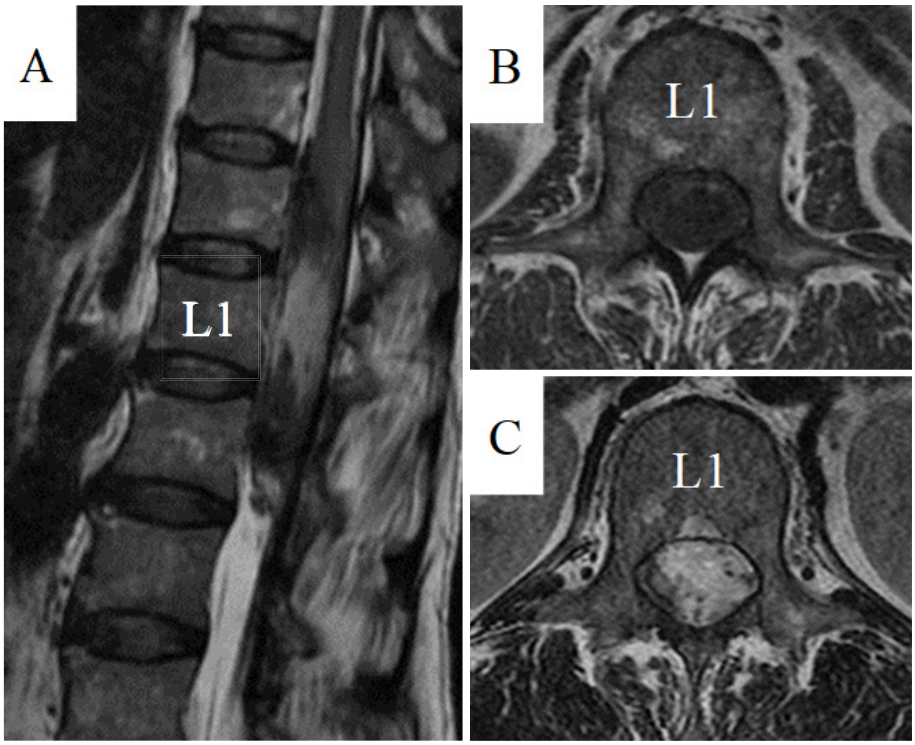

Figure 1. Preoperative MRIs.
Sagittal T2-weighted (A), axial T1-weighted (B), and axial T2-weighted (C) MRIs showing a poorly marginated mass with a T1-low and T2-mosaic pattern located intradurally from T12 to L2 and compressing the spinal cord.

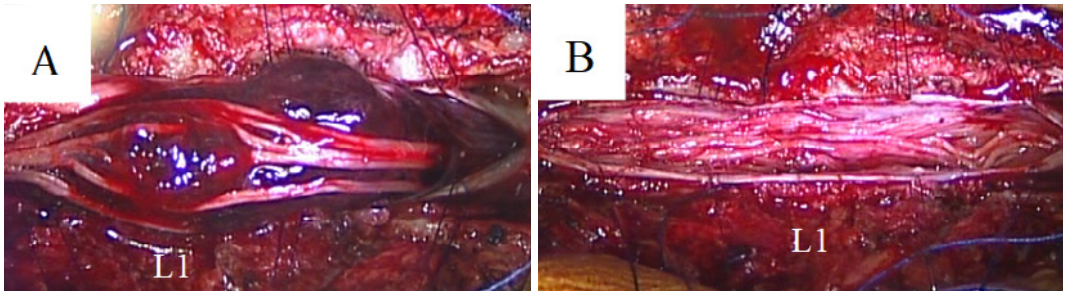

Figure 2. Intraoperative microscopic views.
Intraoperative photographs following laminectomy and durotomy at T12–L2 showing a dark red mass in the subarachnoid space (A). The mass was not connected to the dura matter or spinal cord or cauda equina. After resection, no bleeding lesion or arteriovenous fistula are seen on the pia mater (B).

vertebral, and a primary intradural angiosarcoma had not been reported to our knowledge. A histological feature of angiosarcomas is the presence of malignant endothelial cells[8], which are positive for endothelial markers such as CD31 and 34. By contrast, hemangiomas do not contain malignant cells[9]. In this case, it was unusual that malignant cells were seen in the stroma, but not in the vascular endothelium. Radical and complete resection with wide margins is recommended[8]. Radiotherapy with large doses (>50 Gy) and wide treatment fields is recommended in cases with high risk of local recurrence[8]. Overall, 5-year survival is about 35%[8]. Several targeted therapies are reported, but metastatic angiosarcoma remains incurable[3].

Although malignant intradural extramedullary spinal tumor is rare, we note that angiosarcomas are possible in tumors diagnosed with hemangiomas.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Sources of Funding: None

Author Contributions: Y. Iijima: Writing - original draft. T. Furuya: Conceptualization, Supervision, Writing - review & editing. T. Kotani: Writing - review & editing. T. Sakuma: Writing - review & editing. K. Nakayama: Writing - review & editing. D. Sasai: Data curation, Writing - review & editing. S. Murakami: Writing - review & editing. S.

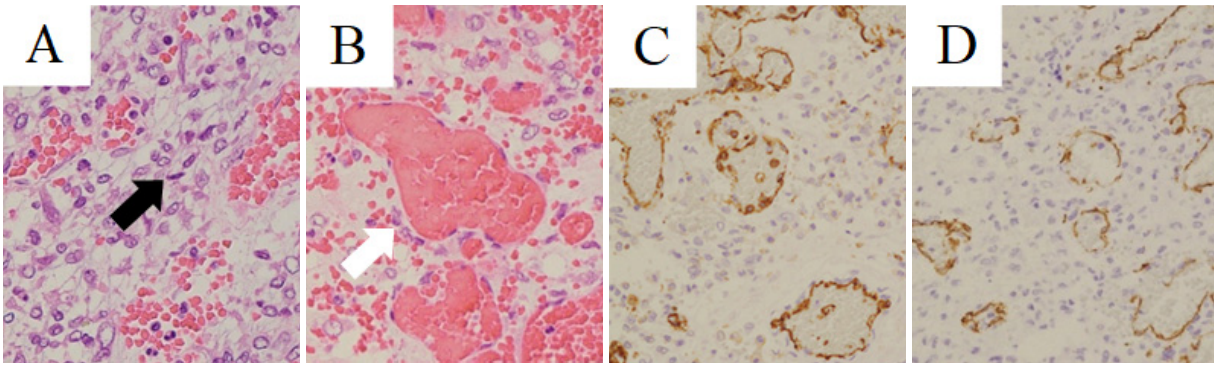

Figure 3. Histological findings.
Hematoxylin and eosin (H&E) staining ×400 (A, B). Cluster of differentiation (CD) 31 staining ×200 (C), and CD34 staining ×200 (D).
H&E staining showing atypical spindle cells in the stroma (black arrow). Proliferation of blood vessels is seen, but atypical cells are not seen in the vascular endothelium (white arrow). Cluster of differentiation (CD) 31 and 34 antigen staining is seen in vascular endothelial cells alone, but not stromal cells (C, D).

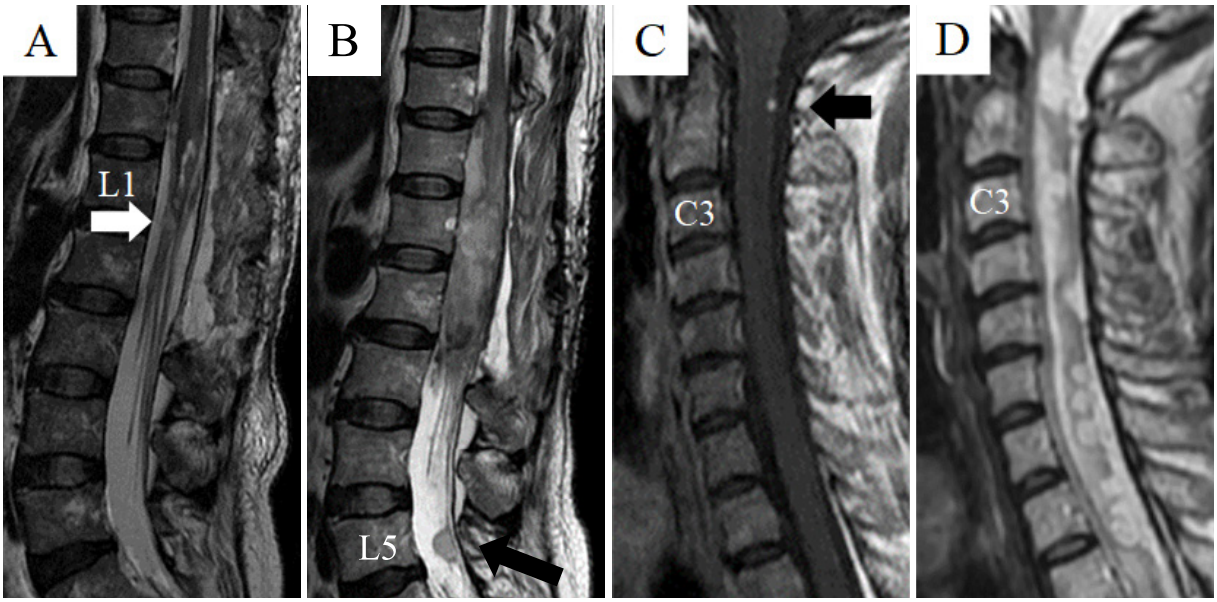

Figure 4. Postoperative MRIs.
Sagittal T2-weighted MRI (A) at 2 weeks after surgery showing the mass was mostly resected, but a small mass is seen below the conus medullaris (white arrow).
Sagittal T2-weighted MRI (B), and sagittal gadolinium-enhanced T1-weighted MRI (C) at 5 weeks after surgery showing an expanded mass and cerebrospinal fluid dissemination at C1 and L5 (black arrow).
Sagittal T2-weighted MRI (D) after radiation therapy showing an expanded mass at the cervical spine.

Maki: Writing - review & editing. T. Miyamoto: Writing - review & editing. S. Okimatsu: Writing - review & editing. S. Kishida: Writing - review & editing. Y. Muramatsu: Writing - review & editing. Y. Sasaki: Writing - review & editing. K. Ueno: Writing - review & editing. D. Kajiwara: Writing - review & editing. S. Ohtori: Writing - review & editing. S. Minami: Writing - review & editing.

Ethical Approval: Ethical approval was not necessary for clinical correspondence.

Informed Consent: The patient and her family consented to submitting data from the case for publication.

References

1. Koeller KK, Shih RY. Intradural extramedullary spinal neoplasms: Radiologic-pathologic correlation. Radiographics. 2019;39(2):468-90.
2. Gaballah AH, Jensen CT, Palmquist S, et al. Angiosarcoma: Clinical and imaging features from head to toe. Br J Radiol. 2017;90 (1075):20170039.
3. Florou V, Wilky BA. Current and future directions for angiosarcoma therapy. Curr Treat Options Oncol. 2018;19(3):14.
4. Liu ZH, Lee ST, Jung SM, et al. Primary spinal angiosarcoma. J Clin Neurosci. 2010;17(3):387-9.

5. Pülhorn H, Elliot T, Clark J, et al. Case report: Angiosarcoma of the cervical spine. J Clin Neurosci. 2017;45:129-31.
6. Huang W, Liu T, Duan R, et al. Primary multiple angiosarcoma of vertebra: A case report. Medicine (Baltimore). 2020;99(50): e23587.
7. Romero-Rojas AE, Diaz-Perez JA, Ariza-Serrano LM, et al. Vertebral bone primary angiosarcoma: A case report. Orthop surg. 2013;5(2):146-8.
8. Young RJ, Brown NJ, Reed MW, et al. Angiosarcoma. Lancet Oncol. 2010;11(10):983-91.
9. Panero I, Eiriz C, Lagares A, et al. Intradural-extramedullary capillary hemangioma with acute bleeding: Case report and literature review. World Neurosurg. 2017;108:988.e7-14.