

CASE REPORT

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Rapid Regrowth of a Capillary Hemangioma of the Thoracic Spinal Cord

—Case Report—

Yoichi KANEKO,¹ Kazutoshi YAMABE,² and Masamitsu ABE³

¹Department of Neurosurgery, Imamura Hospital, Tosu, Saga;

²Department of Medicine, Kitakyushu-Koga Hospital, Koga, Fukuoka;

³Department of Neurosurgery, Shin-Takeo Hospital, Takeo, Saga

Abstract

A 48-year-old man presented with a 2-week history of progressive gait disturbance. Neurological examinations showed mild weakness in his lower extremities and depreciation of deep sensation. Magnetic resonance (MR) imaging showed an intradural extramedullary enhanced lesion at the levels of the T10 and T11 vertebrae. Laminectomy of the T10 and T11 vertebrae was performed, and the vascular tumor on the spinal cord surface was completely resected. Histological analysis indicated that the lesion was a capillary hemangioma with an elevated proliferative index. Postoperatively, the patient showed rapid motor and sensory improvement. However, 6 months after the operation, MR imaging showed regrowth of the tumor although the clinical symptoms of the patient had not deteriorated. The patient has shown no tumor regrowth 9 years after the second operation. Capillary hemangiomas in the skin and soft tissues are often associated with high proliferative activity, and recurrence/regrowth is not infrequent. On the other hand, recurrence/regrowth of capillary hemangioma in the neuraxis after tumor resection has rarely been observed, even in cases of incomplete resection. The present case illustrates the treatment of recurrent capillary hemangioma of the spinal cord.

Key words: hemangioma, capillary hemangioma, spinal cord, central nervous system, recurrence

Introduction

Capillary hemangiomas are benign vascular lesions, most often found in the cutaneous, subcutaneous, or mucosal tissues during childhood.^{12,14,20,22} Histologically, these hemangiomas are characterized by a lobular architecture, separated from each other by fibrous septa that originate from the capsule. Each lobule consists of numerous capillaries lined by flattened endothelium. In spite of its benign property, capillary hemangioma shows increased proliferative activity, as indicated by an elevated MIB-1 index^{10,11};

moreover, recurrence/regrowth of capillary hemangioma in the skin or soft tissues is common.^{13,14,22} Capillary hemangioma rarely involves the central^{1,3,4,5,7-9,16,18,19,21,24-26,29} and peripheral^{17,32} nervous systems. Its proliferative activity along the neuraxis has been shown to be high^{2,5} similar to other lesions; however, recurrence/regrowth of the tumor in the central nervous system (CNS) has been extremely rare.²⁰ We report the case of an intradural capillary hemangioma of the thorax; this tumor showed a rapid regrowth 6 months after complete resection.

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Case Report

A 48-year-old man who experienced transient low back pain, followed by 2 weeks of progressive gait disturbances, was admitted to our clinic. Physical examination found no abnormalities. Neurological examinations showed mild motor weakness in both the left and right legs and depreciation of deep sensation associated with positive Romberg's sign. The patient's sensation to pain, touch, and temperature changes was intact and the bilateral Babinski's reflexes were negative. Sphincter function was normal. Routine laboratory investigations identified no abnormalities. Magnetic resonance (MR) imaging of the thoracic spine showed a round mass at the Th10-T11 levels; this mass was isointense and hyperintense on T₁- and T₂-weighted images, respectively (Fig. 1A, B). The lesion was markedly enhanced after gadolinium administration (Fig. 1C, D). The lesion was located in the intradural space on the dorsal surface of the spinal cord. A laminectomy was performed at the Th10-T11 levels, and a reddish-purple well demarcated polypoid lesion located on the dorsal surface of the spinal cord was observed through intradural exploration; this lesion was supplied by several feeding



Fig. 1 A: Sagittal T₁-weighted magnetic resonance (MR) image showing a well-defined, oval mass on the dorsal surface at the Th10-T11 levels (arrow) as isointense to the spinal cord. B: Sagittal T₂-weighted MR image showing the mass as hyperintense to the spinal cord. C: Sagittal T₁-weighted MR image with gadolinium showing homogeneous, intense enhancement of the mass. D: Axial T₁-weighted MR image with gadolinium showing the spinal cord markedly compressed by the dorsal mass.

vessels (Fig. 2A). The extramedullary mass was adhered to the nerve roots, especially on the right side. A larger part of the lesion was easily separated and resected from the spinal cord. However, the right lateral aspect of the mass firmly adhered to the spinal cord at the caudal end. Eventually, the tumor was completely resected (Fig. 2B). The patient's postoperative recovery was uneventful. The sensorimotor disturbance in the lower extremities improved rapidly, although slight numbness in the right thigh persisted.

The surgical specimen was fixed in 10% buffered formalin, routinely processed, and embedded in paraffin. Sections of 5- μ m thickness were prepared and stained with hematoxylin and eosin. Immunohistochemical analysis was performed using the avidin-biotin-peroxidase complex method and a monoclonal antibody against factor VIII, CD34, and Ki-67. Histological examination showed that the lesion had a distinctly lobular architecture, separated by thin fibrous septa arising from the capsule. Each lobule had numerous capillary-sized vessels lined by a single layer of cytologically benign endothelial cells (Fig. 3A, B). Slight stromal edema and occasional endothelial and stromal mitoses were observed. Scattered lymphocytes were also present. Immunostaining showed cells positive for factor VIII and CD34, in the lining of the blood vessels (Fig. 3C). Immunostaining also showed cells positive for Ki-67 among endothelial as well as stromal cells (Fig. 3D). The MIB-1 index (percentage of positive cells) was approximately 6%.

Postoperative MR imaging indicated no definite residual mass lesion (Fig. 4A). However, 6 months after the operation, MR imaging showed a regrowth of the intradural mass located on the right side of the thecal sac at the Th10-T11 levels (Fig. 4B). Despite the tumor regrowth, the patient's neurological findings did not worsen during the postoperative period. The patient was readmitted, and a second operation was performed. At the opening in the dural membrane, the lesion appeared as a non-exophytic mass located on the right lateral side of the spinal cord and covered by a thick regenerated arachnoid membrane (Fig. 5). The tumor was strongly adhered to the spinal cord as well as the posterior roots of the right 10th thoracic nerve. The tumor was dissected from the spinal cord and nerve roots and was thereafter completely excised. Postoperatively, the patient showed hypesthesia in the right T10 dermatome. Follow-up MR imaging has not shown any recurrent tumor for 9 years after the second operation.

Discussion

Capillary hemangiomas are common, benign, vascular lesions that most often involve the skin and soft tissues.^{12,14,20,22} Such lesions frequently develop during childhood and do not have a sex predilection, i.e., sex incidence is almost equal. Capillary hemangioma in the neuraxis, including the brain,²⁹ spinal cord,^{1,3,4,5,7-9,16,18,19,24,25} cauda equina,¹⁷ and nerve roots,³² have been rarely encountered. Twenty-four cases of capillary hemangioma that developed in the spinal cord have been reported, excluding tumors in the spinal nerve root or cauda equina. These 24

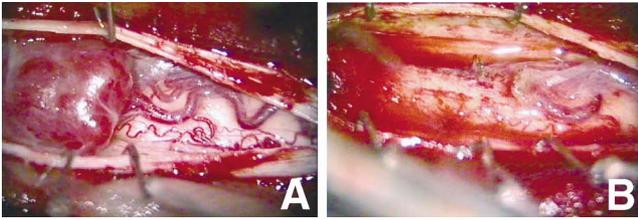


Fig. 2 Intraoperative photographs obtained at the first operation showing a red, polypoid mass lesion on the dorsal surface of the spinal cord (A), and the dorsal surface of the spinal cord after tumor resection (B).

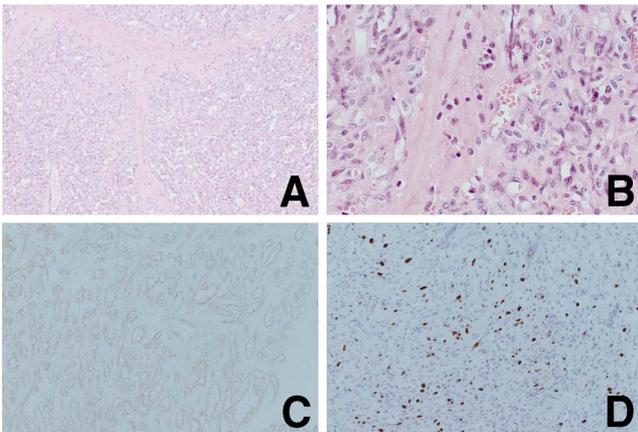


Fig. 3 A: Photomicrograph of the tumor showing the lobular architecture, separated by delicate fibrous septa, with each lobule consisting of numerous capillary channels of varying sizes. Hematoxylin and eosin stain, original magnification $\times 100$. B: Photomicrograph at a higher magnification showing blood vessels lined by flattened endothelium. Hematoxylin and eosin stain, original magnification $\times 400$. C, D: Immunostaining showing endothelial cells lining the capillary lumen positive for CD34 (C), and scattered nuclei positive for Ki-67 (D). Original magnification $\times 400$.

and the present patients included 21 men and 4 women aged from 3 months to 80 years, and 22 of these patients were over 40 years old (median age 52.6 years). The lesions in these patients were located between the T4 vertebra and the conus medullaris. Although capillary hemangioma of the spinal cord is usually present as an intradural extramedullary lesion, several cases of intramedullary lesions have been reported.^{18,19)} Capillary hemangioma of the spinal cord is likely to develop in middle- or older-aged men, and located at a level below the level of the mid-thoracic region.

Our patient presented with transient flank pain, followed by weakness in the legs and depreciation of deep sensation. Most capillary hemangiomas of the spinal cord, located at the thoracic level, cause progressive leg weakness with or without numbness in the lower extremities; this is frequently preceded by abdominal or low-back pain. Symptoms such as sensorimotor disturbance and pain are similar to all commonly occurring extra- or intramedulla-

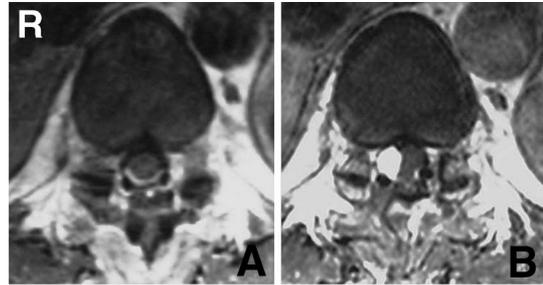


Fig. 4 A: Postoperative axial T₁-weighted magnetic resonance (MR) image showing no definite residual tumor. B: Axial T₁-weighted MR image with gadolinium at 6 months after the operation showing a dense enhanced mass in the right lateral half of the thecal sac at the Th10-T11 levels.

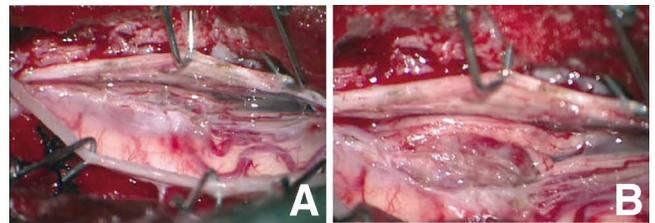


Fig. 5 Intraoperative photographs during the second operation showing a non-polypoid mass covered by a thick regenerated arachnoid membrane after incision and retraction of the dura mater (A), and a red mass between the spinal cord and posterior roots of the right 10th thoracic nerve after removal of the arachnoid membrane (B).

ry solid lesions. At the time of diagnosis, no severe neurological deficits are observed because of the non-infiltrative nature of such lesions. The neurological deficits usually reverse after total excision of the tumor.

Capillary hemangioma of the spinal cord is included in the differential diagnosis of spinal intradural extramedullary tumor. The findings of MR imaging in our patient are consistent with those of previous studies on capillary hemangioma of the spinal cord^{1,7,25)}; that is, the lesion is isointense and hyperintense relative to the spinal cord on T₁- and T₂-weighted images, respectively, and shows homogeneous, strong enhancement by gadolinium. Capillary hemangioma can be distinguished from meningioma or common intradural extramedullary tumor, because meningioma often appears as a mass of low intensity or isointensity on T₂-weighted images. The presence of enlarged abnormal vessels associated with the spinal capillary hemangioma should be considered as an indicator of a vascular mass.^{1,4,21)} Moreover, the absence of a peripheral T-rim owing to hemosiderin deposition can distinguish capillary hemangioma from cavernous hemangioma. However, spinal capillary hemangioma cannot be easily distinguished from schwannoma or other vascular tumors such as hemangiopericytoma and hemangioblastoma by MR imaging. The appearance of spinal capillary hemangioma on MR images is nonspecific, so histological exami-

nation is indispensable for correct diagnosis.

Although capillary hemangioma is a benign tumor, previous studies regarding the proliferative activity of capillary hemangioma involving the skin or soft tissues have indicated an elevated MIB-1 index of the tumor cells.^{10,11} A high proliferation index has also been described in cases of capillary hemangioma of the CNS.^{2,5} Because the recurrence/regrowth of capillary hemangioma in the skin or soft tissues is not infrequent,^{14,22} the surgical goal should be total excision of the lesion. However, pyogenic granuloma (capillary hemangioma) recurred despite surgical excision in 2 cases, which was initially thought to be sufficient.¹³ On the other hand, recurrence/regrowth of capillary hemangioma in the neuraxis is hardly known, even in cases of incomplete resection.^{1,3,7,21,25} An intracranial capillary hemangioma of the CNS showed rapid regrowth after tumor resection.²⁶ Capillary hemangioma has been known to develop rapidly and achieve a maximum size, ranging from several millimeters to a few centimeters, within a few weeks or months.¹⁴ Whereas capillary hemangiomas involving the skin or soft tissues are easily detected immediately after development, those involving the CNS can be hardly be detected before the presentation of symptoms. Therefore, the size of a capillary hemangioma of the CNS may be nearly its maximum at the time of operation, and the tumor may not recur after resection.

In our case, the capillary hemangioma of the spinal cord showed regrowth 6 months after the surgical resection. Underlying arteriovenous anastomosis may be associated with the pathogenesis of capillary hemangiomas,¹³ which may explain why a small remnant is possible even after gross total resection and is likely to show regrowth. The MIB-1 index of the current patient was approximately 6%, which is not as high as reported for previous cases in the CNS, where the MIB-1 index ranged from 2.2% to 12.0%.² A case of capillary hemangioma in the conus medullaris showed no recurrence/regrowth despite an MIB-1 index of more than 10%.⁵ Thus, the MIB-1 index is not a predictive factor for recurrence/regrowth in capillary hemangiomas. No other factors, including duration at diagnosis, age, sex, and location, were related to recurrence/regrowth. Interestingly, a well established lesion of capillary hemangioma in the skin or soft tissues is a polypoid, friable, purple-red mass; however, the recurrent tumor is often sessile.¹⁴ These findings are consistent with the findings of the present case: the first manifestation was a reddish-purple well-demarcated polypoid lesion, and the recurrent tumor was a non-exophytic and sessile mass.

The extent of tumor resection in cases of capillary hemangioma of the CNS depends on the location and properties of the tumor. If the tumor is hemorrhagic and strongly adhered to an eloquent area or the spinal cord, radical resection is not always performed. Furthermore, strong adherence of the tumor to the nerve roots renders tumor dissection difficult. On the other hand, surgical procedures may worsen the symptoms, especially after total excision,^{3,19,21,24} as in surgical excision of intramedullary tumors. Therefore, difficulties are occasionally encountered during radical resection of capillary hemangio-

mas of the CNS. Recently, several agents such as corticosteroids, interferon, and β -blockers have been reported to be effective for the treatment of capillary hemangioma in the skin or soft tissues.^{6,15,27,30,31} In addition to skin or soft-tissue lesions, residual tumor has been reported to show regression after medical treatment in patients with capillary hemangioma of the CNS.³ Furthermore, the effectiveness of radiation therapy for capillary hemangiomas has been established.^{23,28} If the presence of a residual tumor is obvious after the resection of a capillary hemangioma of the CNS, adjuvant therapies such as irradiation and drug administration should be considered. In the present case, the tumor showed rapid regrowth at 6 months after gross total resection. Whether the adjuvant therapies would immediately affect the rapidly regrowing tumor and prevent the development of irreversible neurological deficits remains unclear. Therefore, the regrown tumor was surgically resected instead of administering adjuvant therapies. However, radiation therapy may be a therapeutic option for recurrent/regrown capillary hemangiomas of the CNS.

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Address reprint requests to: Yoichi Kaneko, MD, Department of Neurosurgery, Imamura Hospital, 1523-6 Todoroki-machi, Tosu, Saga 841-0061, Japan.