Adjuvant multimodal treatment for spinal intradural extramedullary capillary hemangioma with subpial growth: illustrative case

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BACKGROUND Spinal intradural extramedullary capillary hemangiomas are rare vascular lesions. Although total resection is the first treatment, the efficacy of adjuvant therapies, such as steroid or radiation therapy, has not been investigated.

OBSERVATIONS A 74-year-old man presented with progressive back pain, gait disturbance, and left chest pain. Spinal magnetic resonance imaging (MRI) revealed an intradural extramedullary lesion at the middle thoracic level. Preoperatively, systemic steroid administration improved his chest pain and reduced the tumor size. The authors performed tumor extirpation. Because the tumor strongly adhered to the pia matter, in short, the tumor accompanied with partial subpial growth, subtotal resection was performed, leaving a thin layer of the tumor. A pathological examination revealed capillary hemangioma. His gait disturbance showed rapid improvement. Stereotactic cyber knife treatment was performed for the residual tumor at 1 month after the operation. Serial MRI showed a gradual decrease in the tumor size and no recurrence at 2 years after the operation.

LESSONS When a tumor adheres to the spinal cord or nerve root, immoderate total resection should not be performed to avoid exacerbating the clinical symptoms. Adjuvant therapies, such as a systemic steroid administration or radiation therapy, may provide satisfactory control of spinal capillary hemangiomas.

https://thejns.org/doi/abs/10.3171/CASE2314

KEYWORDS capillary hemangioma; intradural extramedullary tumor; radiation therapy; spine; steroid

Although capillary hemangiomas are benign vascular lesions most often found in the skin or mucosal soft tissues of the head and neck during childhood, they rarely involve the central and peripheral nervous systems.1–3 When they develop in the spine, they most frequently involve the vertebral body.4 Intradural extramedullary capillary hemangiomas are extremely rare. They are usually associated with the occurrence of slowly progressive myelopathy and radiculopathy. Total resection is the uncontroversial first treatment.4–6 However, when the tumor strongly adheres to the spinal cord or the nerve roots, total resection may not always be performed. Although some authors recommend the systematic administration of steroids, interferon-α, and β-blockers or radiation therapy for residual tumors, because such adjuvant therapies are effective for capillary hemangiomas in the skin or soft tissues,1,3,5,7–10 no report has proven the efficacy of adjuvant therapies for spinal intradural extramedullary capillary hemangiomas.

Herein, we describe a case of spinal intradural extramedullary capillary hemangioma treated with multimodal therapies, such as preoperative systemic steroid administration, subtotal resection, and postoperative radiation therapy because of strong adherence of the tumor to the spinal cord, in short, the tumor accompanied with partial subpial growth.

Illustrative Case

A 74-year-old man presented to our hospital with progressive back pain, gait disturbance, and left chest pain. He had a past history of hypertension. A neurological examination revealed gait disturbance due to posterior column impairment and radicular pain in the T7 area without motor paresis or bladder or rectal dysfunction. Spinal magnetic resonance imaging (MRI) revealed an intradural extramedullary mass at the middle thoracic level. The mass lesion showed isointensity on T1-weighted imaging and relative hyperintensity on T2-weighted imaging, along with spinal cord edema at...
the caudal side of the tumor (Fig. 1A and B). The mass lesion showed homogeneous contrast enhancement along with the dural tail sign on T1-weighted imaging (Fig. 1C). The lesion was located in the intradural space on the right posterolateral surface of the spinal cord (Fig. 1D).

Our initial diagnosis was meningioma, and tumor extirpation was planned. Because of the patient’s severe radicular pain and the existence of spinal edema, the preoperative systemic steroid administration (betamethasone valerate 4 mg/day) was performed for 3 days. After that, his severe radicular pain improved, and serial MRI showed a decrease in the tumor size and improvement in spinal cord edema (Fig. 1E and F). Since the tumor size decreased after the systemic administration of steroids, we also considered the presence of malignant lymphoma in the differential diagnosis. The patient underwent tumor extirpation with T6–7 osteoplastic laminotomy in the prone position under neurophysiological monitoring. After the dura mater was opened, a dark-reddish mass was seen to be compressing the spinal cord ventrally (Fig. 2A). The tumor showed neither adhesion to the dura nor bleeding when the dura was opened. A large part of the tumor was easily separated and resected from the spinal cord and nerve roots. However, the bottom of the tumor bed strongly adhered to the pia. Hence, subtotal resection was performed, leaving a thin layer of the tumor (Fig. 2B). His gait disturbance showed rapid improvement, and he could walk without assistance at 1 week after the operation. A histological examination revealed that the mass comprised a proliferation of capillary-sized vessels lined by flattened endothelial cells, and the mass was composed of nodules of small capillary-sized vessels supplied by a feeding vessel, indicating capillary hemangioma (Fig. 2C). Immunostaining showed CD31+ cells in the lining of the blood vessels (Fig. 2D).

Postoperative MRI performed 1 week after the operation showed residual tumor. To prevent regrowth of the tumor, stereotactic Cyber Knife treatment (marginal dose: 21 Gy; central dose: 25 Gy/3 fractions/week) was performed for the residual tumor at 1 month after the operation (Fig. 3A and B). Serial MRI showed a gradual decrease in the tumor size and no recurrence at 2 years after the operation (Fig. 3C–H).

Discussion

Observations

Spinal intradural extramedullary capillary hemangiomas are very rare. They are benign tumors or tumor-like vascular lesions and have been labeled as malformations or hamartomas. Based on recent literature reviews, they most frequently occur in middle-aged males, unlike capillary hemangiomas, which occur in the skin or mucosal soft tissue. The most frequent location is the lower thoracic spine, conus medullaris, and cauda equina. Spinal intradural extramedullary capillary hemangiomas involving the cervical or upper thoracic spine are extremely rare. In the intradural extramedullary space, capillary hemangiomas can arise from the blood vessels of the root nerves, the inner layer of the dura mater, or the pial surface of the spinal cord. They are slow-growing tumors and present symptoms like those of space-occupying lesions over a period of several months to years. Common symptoms

![Figure 1](https://example.com/fig1.png)

**FIG. 1.** Preoperative MRI. An intradural extramedullary lesion showing isointensity on a T1-weighted image (A) and relative hyperintensity accompanied by spinal cord edema on a T2-weighted image (B). The mass lesion showed homogeneous contrast enhancement on a T1-weighted image, accompanied by the dural tail sign (C) and spinal cord compression (D). The intradural extramedullary lesion showing a decrease in tumor size and improvement in spinal edema after systemic steroid administration (E and F). Th6 = 6th thoracic vertebrae.
include back pain and chronic progressive myelopathy or radiculopathy leading to motor and sensory deficits along with gait and sphincter disturbances. Although vascular tumors are associated with a risk of bleeding that may cause sudden-onset neurological deficits, the bleeding risk of capillary hemangioma is quite low in comparison to other vascular tumors, including caveromas. In fact, there is only 1 reported case of intradural capillary hemangioma with acute intratumoral hemorrhage followed by neurological deficits.

Most spinal intradural extramedullary capillary hemangiomas are radiologically misdiagnosed and/or mistaken for meningiomas or schwannomas. Capillary hemangiomas are typically described as well-defined lesions with a regular shape, isointense on T1-weighted MRI, and hyperintense on T2-weighted MRI, with homogeneous contrast enhancement. Moreover, the dural tail sign, considered a typical feature of meningiomas, is observed when the tumor arises from the inner surface of the dura matter. Hence, it is difficult to differentiate spinal intradural extramedullary capillary hemangiomas from other common intradural tumors by MRI. In our case, because the tumor showed the dural tail sign, our initial diagnosis was meningioma. Furthermore, the differential diagnosis included malignant lymphoma because systemic steroid administration resulted in a decrease in tumor size. After we observed the appearance of the tumor during the operation, we finally recognized that it was a vascular tumor.

In our case, we considered that the tumor might have arisen from a vessel of the pia mater because the tumor adhered to the pia mater rather than the inner surface of the dura mater. Interestingly, the tumor showed both the dural tail sign and spinal cord edema on MRI. These findings may contradict the tumor origin because it is believed that the dural tail sign in spinal capillary hemangiomas is detected when the tumor arises from the inner surface of the dura matter. On the other hand, it is believed that spinal cord edema is detected when the tumor arises from a vessel of the pia mater or accompanies an intramedullary component. We considered that focal dural inflammation caused by the tumor showed the dural tail sign because it is reported that vascular congestion or inflammation can cause the dural tail sign.

![FIG. 2.](image1) After the dura was opened, a dark-reddish mass lesion was seen on the dorsal surface of the spinal cord (A). A thin layer of the tumor was left on the dorsal surface of the spinal cord (B). The pathological diagnosis after surgery suggested capillary hemangioma (hematoxylin and eosin, C) with a positive reaction for CD31 (D). Original magnification ×200 (C and D).

![FIG. 3.](image2) Planning images for the Cyber Knife treatment based on computed tomography (A and B). Postoperative follow-up MRI 1 week after surgery showed a thin layer of residual tumor on the dorsal surface of the spinal cord (C and D). MRI 2 months after Cyber Knife treatment showed shrinkage of the residual tumor (E and F). MRI 2 years after Cyber Knife treatment showed gradual regression of the tumor (G and H). Th6 = 6th thoracic vertebrae.
Because spinal intradural extramedullary capillary hemangiomas mostly present as space-occupying lesions, total resection is the uncontroversial first treatment.\(^1\)–\(^4\) However, there are some matters that require particular attention in resection. First, piecemeal resection with the regrowth of mass or bleeding from residual tumors in the skin and soft tissue is not rare. Hence, subtotal resection. However, the recurrence of capillary hemangiomas after resection is rare, even in cases of incomplete resection.4,5,7,10,18–21 However, these therapies have not been investigated in the treatment of spinal intradural extramedullary capillary hemangiomas.2,5,7,22 Abe et al.\(^3\) reported that systemic administration of steroids and interferon-\(\alpha\) before surgery could ameliorate and improve outcomes because these drugs induced the regression of capillary hemangiomas in the skin, soft tissues, bone, or brain (Table 1).1,5–7,10,18–21 However, subtotal resection may lead to recurrence of neurological symptoms in association with the regrowth of mass or bleeding from residual tumor.2,17 In fact, Kaneko et al.\(^7\) reported that systemic administration of steroids and interferon-\(\alpha\) before surgery could ameliorate and improve outcomes because these drugs induced the regression of capillary hemangiomas in the brain. The recurrence or relapse of spinal intradural extramedullary capillary hemangiomas after resection is rare, even in cases of incomplete resection. However, the recurrence of capillary hemangiomas in the skin and soft tissue is not rare. Hence, subtotal resection should be avoided because of a high bleeding tendency.2,17 Second, immoderate total resection may exacerbate the symptoms because of a strong adhesion to the spinal cord or the nerve root.1,3–7 To resolve these problems, some adjunctive therapies are recommended: embolization of the feeding artery to avoid intraoperative bleeding; the systemic administration of steroids, \(\beta\)-blockers, or interferon-\(\alpha\); or radiation therapy are options because these therapies are effective in treating capillary hemangiomas in the skin, soft tissues, bone, or brain (Table 1).1,5–7,10,18–21 However, these therapies have not been investigated in the treatment of spinal intradural extramedullary capillary hemangiomas.2,5,7,22 Abe et al.\(^3\) reported that systemic administration of steroids and interferon-\(\alpha\) before surgery could ameliorate and improve outcomes because these drugs induced the regression of capillary hemangiomas in the brain. The recurrence or relapse of spinal intradural extramedullary capillary hemangiomas after resection is rare, even in cases of incomplete resection. However, the recurrence of capillary hemangiomas in the skin and soft tissue is not rare. Hence, subtotal resection may lead to recurrence of neurological symptoms in association with the regrowth of mass or bleeding from residual tumor.2,17 In fact, Kaneko et al.\(^7\) reported that systemic administration of steroids and interferon-\(\alpha\) before surgery could ameliorate and improve outcomes because these drugs induced the regression of capillary hemangiomas in the brain. The recurrence or relapse of spinal intradural extramedullary capillary hemangiomas after resection is rare, even in cases of incomplete resection. However, the recurrence of capillary hemangiomas in the skin and soft tissue is not rare. Hence, subtotal resection may lead to recurrence of neurological symptoms in association with the regrowth of mass or bleeding from residual tumor.2,17

**Lessons**

Spinal intradural extramedullary capillary hemangiomas are rare. Early total resection is the uncontroversial first treatment. Preoperative systemic steroid administration may be helpful for resection as it may reduce the tumor volume. When the tumor adheres to the spinal cord or nerve root, immoderate total resection should not be performed to avoid exacerbating the clinical symptoms. If partial resection is performed, postoperative radiation therapy may be effective for the residual tumor.

**References**


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**TABLE 1. Adjunctive nonoperative therapies for capillary hemangioma.**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Spinal Intradural Extramedullary</th>
<th>Brain</th>
<th>Skin, Mucosal Soft Tissue, or Bone</th>
</tr>
</thead>
<tbody>
<tr>
<td>Steroid</td>
<td>NR</td>
<td>Reported(^3)</td>
<td>Reported(^8)</td>
</tr>
<tr>
<td>Interferon-(\alpha)</td>
<td>NR</td>
<td>Reported(^2)</td>
<td>Reported(^9)</td>
</tr>
<tr>
<td>(\beta)-Blocker</td>
<td>NR</td>
<td>NR</td>
<td>Reported(^21)</td>
</tr>
<tr>
<td>Radiation therapy</td>
<td>Reported(^*)1</td>
<td>Reported(^16)</td>
<td>Reported(^10,20)</td>
</tr>
<tr>
<td>Feeder embolization w/ endovascular treatment</td>
<td>NR</td>
<td>NR</td>
<td>Reported(^75)</td>
</tr>
</tbody>
</table>

NR = not reported.
* Although there was a reported case with radiation therapy, the effect was not described.

Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Matsumoto. Acquisition of data: Matsumoto. Analysis and interpretation of data: Matsumoto. Drafting of the article: Matsumoto. Critically revising the article: Matsumoto, Shimokawa. Reviewed submitted version of the manuscript: Matsumoto, Sato. Approved the final version of the manuscript on behalf of all authors: Matsumoto. Administrative/technical/material support: Sato, Takami.

Study supervision: Matsumoto, Takami.

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