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Case Report

Intraspinal dissemination of intracranial hemangiopericytoma: Case report and literature review

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Abstract

Background: The authors report the case of a 53-year-old woman suffering from thoracic myelopathy caused by intraspinal dissemination of hemangiopericytoma. In literature, hemangiopericytoma is commonly found as an intracranial lesion, and often hematogenously metastasizes to the bone or liver; however, intradural spinal dissemination is extremely rare.

Case Description: The patient presented with gait disturbance due to thoracic myelopathy 6 years after surgical treatment for intracranial hemangiopericytoma. Magnetic resonance imaging demonstrated intradural disseminated lesions compressing the spinal cord. Although the patient underwent resection of the intradural spinal tumor, the lesion was tightly adherent to the dorsal surface of the spinal cord. Therefore, it resulted in subtotal removal. Immediately after the surgery, symptoms related to the thoracic myelopathy resolved. The patient was free from disease progression for 14 months after whole spine radiotherapy.

Conclusion: Recognition of this type of progression is important in the clinical management of intracranial hemangiopericytoma because intradural spinal dissemination dramatically degrades neurological functions.

Key Words: Hemangiopericytoma, myelopathy, spinal dissemination

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INTRODUCTION

Hemangiopericytoma is a neoplasm that commonly manifests as an intracranial mass lesion.[19] According to the 2007 World Health Organization classification, it is a "tumour of the meninges"[17] and constitutes approximately 2% of all such tumors.[8,9] Hemangiopericytoma is a clinically malignant tumor due to its high incidence of local recurrence and extraneural metastasis, which eventually leads to 5-year progression-free rates of 49–70%.[7,19,21]

Here, we report a case of intracranial hemangiopericytoma in which recurrence manifested as intraspinal dissemination. To our knowledge, subarachnoid spread via intraspinal dissemination has not been previously reported in English literature. It is important to recognize

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this rare form of disease progression to achieve better control in hemangiopericytoma.

CASE REPORT

Clinical history

In 2007, a 53-year-old woman was referred to our department with a chief complaint of double vision. She had an intracranial tumor originating from her right tentorium and underwent subtotal surgical resection [Figure 1]. After the tumor was pathologically diagnosed as hemangiopericytoma, the patient underwent local radiation therapy (60 Gy). After the radiotherapy was completed, she was referred to a local physician for serial brain magnetic resonance imaging (MRI).

In 2013, the patient was referred back to us with a recurrent intracranial lesion [Figure 2]. At that time, she underwent whole body positron emission tomography (PET) and spine MRI. PET revealed no extraneural metastasis. However, spine MRI demonstrated disseminated lesions along the spinal cord, which were compatible with intraspinal dissemination [Figure 2]. The spinal lesions were asymptomatic at that time, and the patient did not agree to proceed with spine irradiation. Therefore, intracranial recurrence was solely treated with gamma knife radiosurgery, which was effective in stabilizing the lesion.

Clinical course and examination

There was no further growth of the intracranial recurrent tumor. Six months after the radiosurgery, she acutely developed gait disturbance. Neurological examination showed left lower limb weakness with bilaterally increased patellar and Achilles tendon reflexes. Sensory examination revealed decreased superficial and deep sensations below the T12 dermatome. MRI demonstrated enlargement of multiple intraspinal lesions spanning the spinal cord [Figure 3]. They were isointense on T2-weighted images and homogeneously enhanced with gadolinium. Particularly, an enlarged lesion at T11/12 compressed the spinal cord. T2-weighted images detected abnormal hyperintensity in the thoracic spinal cord at T11/12.

Surgical interventions

The patient experienced aggravated symptoms related to thoracic myelopathy and agreed to undergo surgical resection of the T11 lesion. Under monitoring of motor and sensory evoked potentials, the patient was placed in the prone position. Following a left T11 hemilaminectomy, the tumor was exposed. Importantly, the tumor was located in the subarachnoid space and was tightly adherent to the spinal cord surface [Figure 4]. Although the tumor was resected without any deterioration in the motor and sensory evoked potentials, the tightly attached tumor did not allow total resection.

Pathological diagnosis

Pathological evaluations demonstrated a hypercellular tumor with numerous slit-like vascular channels called staghorn sinusoids [Figure 5]. Immunohistochemistry of the tumor cells was diffusely positive for vimentin, negative for epithelial membrane antigen, and intermediately positive for CD 34, which was compatible with the diagnosis of hemangiopericytoma. The degree of mitotic activity or nuclear atypia was not high enough to be the anaplastic counterpart of this clinical entity.[17]

Postoperative course

Immediately after the surgery, the symptoms due to the thoracic myelopathy improved. A month after the surgery, the patient did not have motor weakness. Her sensory

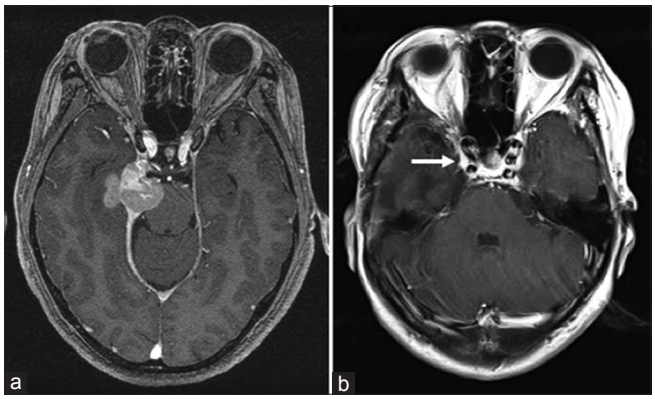

Figure 1: Magnetic resonance images from the original patient presentation in 2007. (a) Preoperatively, axial T1-weighted contrast images demonstrated a mass lesion located along the right tentorium, extending toward the temporal lobe and the pons. (b) Postoperative T1-weighted contrast image. A tumor invading into the cavernous sinus (arrow) remained. The extent of resection was judged as "subtotal"

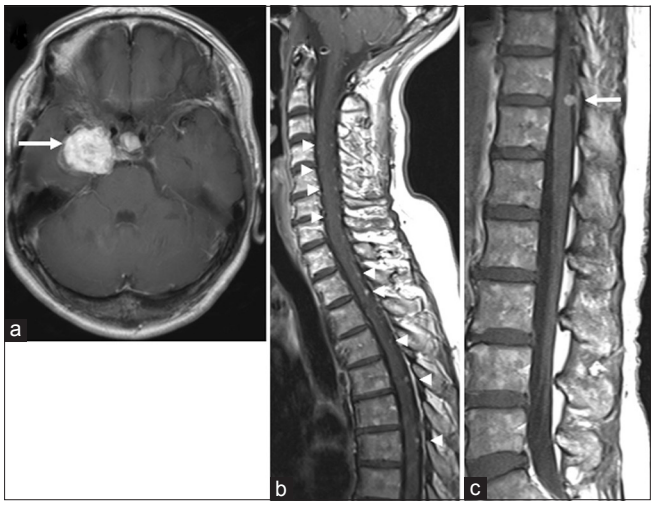

Figure 2: Magnetic resonance images from 2013 when the patient was referred back to us with a recurrent intracranial lesion. (a) Axial T1-weighted image of the brain, demonstrating a recurrent mass lesion (arrow). (b, c) Sagittal T1-weighted images demonstrating spinal disseminated lesions (arrowheads). The tumor at T11/12 (arrow) did not cause thoracic myelopathy at that time

symptoms were also resolved. To prevent other spinal lesions from becoming symptomatic, radiotherapy covering the whole spinal cord was performed (50.4 Gy). At the last follow-up, 16 and 14 months after spinal tumor removal and intraspinal radiotherapy, respectively, the patient was free from new symptoms or recurrences. MRI confirmed that the remnant lesions were stable in size [Figure 6]. She was able to independently perform activities of daily living.

DISCUSSION

Intracranial hemangiopericytoma is often complicated by extracranial metastasis. A large clinical series on intracranial hemangiopericytomas reported the rates of extracranial metastasis as 20–55%.[7,19,21] These studies agreed that the common sites for metastasis include the lung, bone, and liver. Spinal metastasis was less frequently encountered.[7,19,21]

We have summarized 10 previously reported cases of metastatic spinal hemangiopericytoma in Table 1.[1,2,4,6,12,13,16,20,22,23] We defined the types of spinal metastasis as "extradural," "intradural," and "intra-to-extradural and paravertebral," according to previous literature,[14] which are indicated in Table 1.

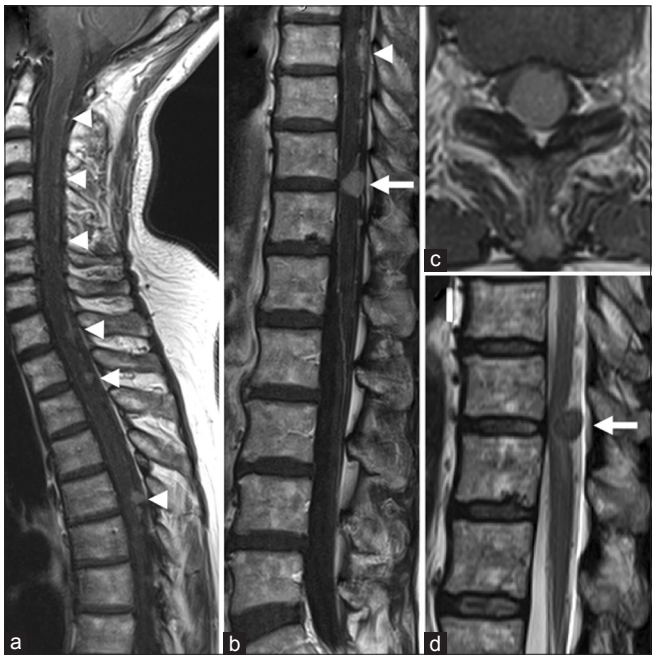

Figure 3: Preoperative magnetic resonance images of the spinal cord. (a, b) Sagittal T1-weighted images with gadolinium, demonstrating intraspinal disseminated lesions that were increased in size (arrowheads). Note the T11/12 lesion (arrow in B) compressing the spinal cord. (c) Axial T1-weighted contrast image demonstrating the T11/12 tumor now causing a deformity of the spinal cord. (d) Sagittal T2-weighted image demonstrating an abnormal hyperintense area in the dorsal spinal cord where the T11/12 lesion is located (arrow)

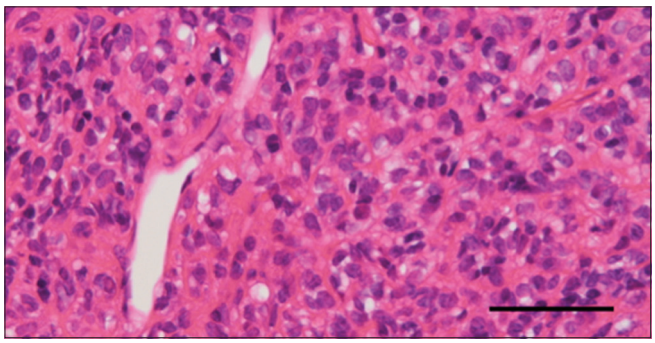

Figure 5: Histological examination of the tumor cells with hematoxylin and eosin staining, the findings of which were compatible with the diagnosis of hemangiopericytoma. Scale bar = 100 μm

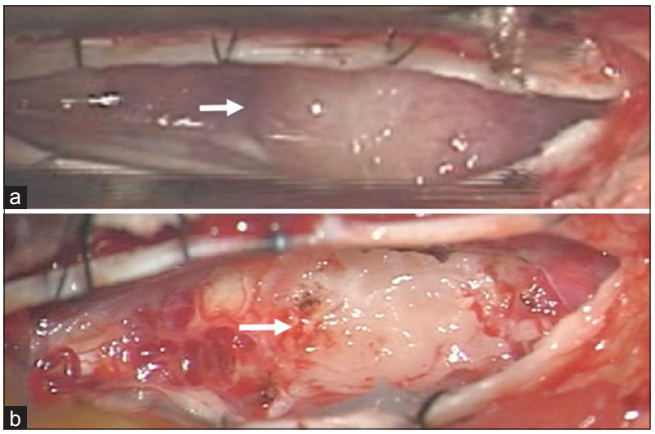

Figure 4: Intraoperative photographs following left T11 hemilaminectomy. (a) Following the dural opening, a mass lesion was visible underneath the arachnoid membrane (arrow). (b) Subtotal surgical resection was performed. A layer of the tumor was left attached to the dorsal spinal cord (arrow). Note that the left and right sides of the images were the rostral and caudal sides, respectively

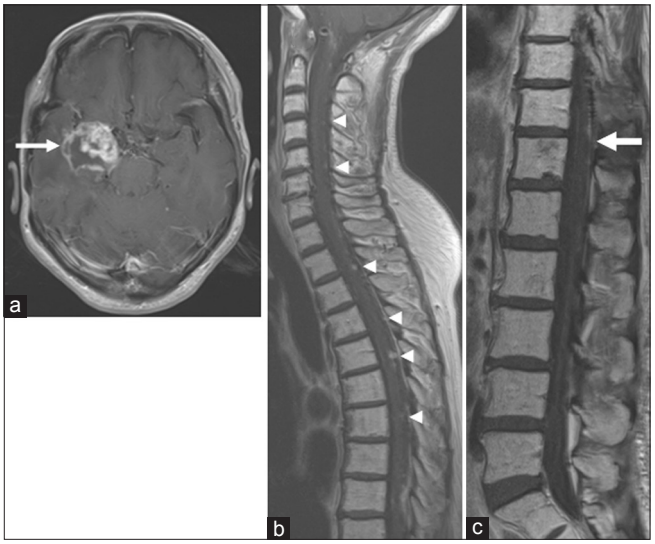

Figure 6: Follow-up magnetic resonance images 16 months after resection. (a) Axial T1-weighted image of the brain, demonstrating an intracranial mass lesion (arrow) that was stable in size. Note that it had an altered intensity following the gamma knife treatment. (b, c) Sagittal T1-weighted images demonstrating spinal disseminated lesions (arrowheads) that were stable in size. The resected spinal tumor (arrow) did not recur

Notably, the intradural type of spinal metastasis was extremely rare. To our knowledge, the current case is the first to illustrate multiple intraspinal disseminations from intracranial hemangiopericytoma, possibly through the subarachnoid space. A case of intradural metastasis to the cauda equina could be another example of intracranial hemangiopericytoma metastatically spreading through the cerebrospinal fluid (CSF), leading to drop metastases.[22]

Metastasis usually occurs late, well after the diagnosis of the primary lesion. Guthrie *et al*. reported 99 months as the average time to metastasis.[8] In the 10 cases that we summarized in Table 1, spinal metastases occurred at an average of 9.5 years after the diagnosis of the primary tumor. Having recognized that intracranial hemangiopericytoma could cause intradural drop metastases and disseminations, long-term follow-up, including MRI scans of the whole spine, is warranted in hemangiopericytoma patients.

To date, clear predisposing factors for metastatic hemangiopericytoma have not been established. However, the mechanisms or predisposing factors leading to drop metastasis in different intracranial pathologies besides hemangiopericytoma have been discussed. In glioblastoma, Shibahara *et al*. reported that the higher expression of CD133, a stem cell marker, correlated with an increased incidence of distant metastasis. In oligodendroglioma, the proximity to cisterns and ventricles facilitated the spread through the CSF.[5]

In hemangiopericytoma, a size of ≥6 cm and a non-skull base location were found to be predictive of local recurrences.[18] The extent of resection, especially total resection, was associated with better local control and prolonged recurrence-free survival.[7,11] However, these factors could not be correlated with extracranial metastases.[7]

Comparing the grades of hemangiopericytoma,[3] metastases were encountered in 36% and 25% of grade II and grade III cases, respectively. This difference did not reach statistical significance.[3] Grade II tumors have better survival than grade III. However, a higher pathological grade did not necessarily correlate with metastatic potential. This was also true in cases harboring primary spinal lesions.[14] Among five spinal metastatic hemangiopericytoma cases for which pathological information was available [Table 1], four were grade II and one was grade III.[1,4,6,23]

There is no consensus on the best treatment for metastatic hemangiopericytoma. When considering surgical resection, the high vascularity of this disease should be taken into account.[1,4,22] In the current case, although we were able to control the bleeding from the tumor, the tight adherence between the

Table 1: Previously reported cases of metastatic spinal hemangiopericytoma

Case	Authors, Year	Age, Sex	Vertebral Level	Classification	Duration	Pathology	Treatment	Radiation	Clinical outcome
1	El Hindy *et al*., 2013[4]	46, M	T12	Intra-to extradural and paravertebral	15 years	Initially Grade II Grade III at recurrence	Embolization and Complete Excision	Yes	No recurrence for 6 months
2	Fukuda *et al*., 2015[6]	36, M	T10	Extradural	17 years	Grade II	Complete	No	No recurrence for 2 years
3	Cole *et al*., 2009[2]	36, F	C3	Extradural	6 years	NA	Complete	Yes	No recurrence for 4 years
4	Nonaka *et al*., 1998[16]	40, F	T8	Intra-to extradural and paravertebral	9.5 years	NA	Subtotal	Yes	No recurrence for 2 years
5	Taniura *et al*., 2007[22]	30, F	L4-S1	*Intradural*	4 years	NA	Partial	Yes	No recurrence for 1 year
6	Woitzik *et al*., 2003[23]	40, F	C6-T2	Intra-to extradural and paravertebral	8 years	Grade III	Complete	Yes	Recurrence in L2 in 1 year
7	Lee *et al*., 2006[13]	48, F	C6-C7	Intra-to extradural and paravertebral	6.5 years	NA	Partial	Yes	Recurrence in 8 months
8	Brass *et al*., 2004[1]	53, M	C6-T5	Extradural	5 years	Grade II	Embolization and Partial	Yes	Progression to paraparesis in 1 year, alive for 2 years.
9	Kruse 1961[12]	22, F	Lumbar	Extradural	8 years	NA	Surgery	No	Death in 5 years
10	Scott *et al*., 1974[20]	38, M	T12/L1	NA	16 years	NA	Surgery	Yes	Recurrence in 3 years
	Current case	53, F	T11/12	*Intradural*	6 years	Grade II	Subtotal	Yes	No recurrence for 1.5 years

F: Female, M: Male, C: Cervical, T: Thoracic, L: Lumbar, NA: Not Addressed

tumor and the spinal cord made complete removal difficult [Figure 4]. In a clinical series on primary spinal hemangiopericytomas, the infiltrative nature of hemangiopericytomas was also pointed out.[14] In the current case, we did not pursue complete resection since there were other metastatic lesions along the spinal cord. Rather, postoperative radiotherapy covering the whole spinal cord provided good control of all the spinal lesions [Figure 6]. For intracranial hemangiopericytoma, postoperative radiotherapy is recommended for better local control.[7,19,21] Distinctive roles and indications for the radiotherapy of spinal lesions remain to be established by large clinical series. Nonetheless, 8 out of 10 reported cases harboring spinal metastases received postoperative radiotherapy for disease control [Table 1]. To date, there is no standardized chemotherapeutic protocol for hemangiopericytoma. Although some chemotherapeutic agents are promising,[10,15] postoperative radiotherapy still plays an important role in this clinical entity.[18]

CONCLUSIONS

Intracranial hemangiopericytoma can cause intraspinal dissemination. This rare form of disease progression should be recognized for better therapeutic management.

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

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Conflicts of interest
There are no conflicts of interest.

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