Intraspinal dissemination of intracranial hemangiopericytoma: Case report and literature review

Hosam Shata Mohammed Ali, Toshiki Endo, Hidenori Endo, Kensuke Murakami, Teiji Tominaga

1Department of Neurosurgery, Tohoku University, Graduate School of Medicine, Sendai, Japan, 2Department of Neurosurgery, Mansoura University, Mansoura, Egypt, 3Department of Neurosurgery, Kohnan Hospital, 4Department of Neurosurgery, Sendai Medical Center, Sendai, Japan

E-mail: Hosam Shata Mohammed Ali - hosamshata@gmail.com; Toshiki Endo - endo@nsg.med.tohoku.ac.jp; Hidenori Endo - hideendo@gmail.com; Kensuke Murakami - murakami@nsg.med.tohoku.ac.jp; Teiji Tominaga - tomi@nsg.med.tohoku.ac.jp

Received: 05 July 16 Accepted: 04 October 16 Published: 12 December 16

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

INTRODUCTION

Hemangiopericytoma is a neoplasm that commonly manifests as an intracranial mass lesion. According to the 2007 World Health Organization classification, it is a “tumour of the meninges” and constitutes approximately 2% of all such tumors. 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%. 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...
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.

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.
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 \(\text{et al.}\) 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 \(\text{et al.}\) reported that the higher expression of CD133, a stem cell marker, correlated with an increased incidence of distant metastasis.\(^{[14]}\)

In oligodendroglioma, the proximity to cisterns and ventricles facilitated the spread through the CSF.\(^{[5]}\)

In hemangiopericytoma, a size of \(\geq 6\) cm and a non-skull base location were found to be predictive of local recurrences.\(^{[9]}\) 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.\(^{[1,4,6,22]}\)

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

| Case | Authors, Year | Age, Sex | Vertebral Level | Classification | Duration | Pathology | Treatment | Radiation | Clinical outcome |
|------|---------------|---------|----------------|----------------|----------|-----------|-----------|-----------|-----------------|
| 1    | El Hindy \(\text{et al.}, 2013\)\(^{[6]}\) | 46, M   | T12            | Intra-to extradural and paravertebral | 15 years | Initially Grade II | Embolization and Complete Excision | Yes       | No recurrence for 6 months |
| 2    | Fukuda \(\text{et al.}, 2015\)\(^{[15]}\) | 36, M   | T10            | Extradural              | 17 years | Grade II at recurrence | Complete | No         | No recurrence for 2 years |
| 3    | Cole \(\text{et al.}, 2009\)\(^{[7]}\)  | 36, F   | C3             | Extradural             | 6 years  | NA        | Complete      | Yes       | No recurrence for 4 years |
| 4    | Nonaka \(\text{et al.}, 1998\)\(^{[16]}\) | 40, F   | T8             | Intra-to extradural and paravertebral | 9.5 years | NA        | Subtotal | Yes       | No recurrence for 2 years |
| 5    | Taniura \(\text{et al.}, 2007\)\(^{[22]}\) | 30, F   | L4-S1          | Intradural              | 4 years  | NA        | Partial       | Yes       | No recurrence for 1 year  |
| 6    | Woitzik \(\text{et al.}, 2003\)\(^{[23]}\) | 40, F   | C6-T2          | Intra-to extradural and paravertebral | 8 years  | Grade III | Complete | Yes       | Recurrence in L2 in 1 year |
| 7    | Lee \(\text{et al.}, 2006\)\(^{[13]}\)  | 48, F   | C6-C7          | Intra-to extradural and paravertebral | 6.5 years | NA        | Partial | Yes       | Recurrence in 8 months |
| 8    | Brass \(\text{et al.}, 2004\)\(^{[11]}\) | 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 \(\text{et al.}, 1974\)\(^{[90]}\) | 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.

Acknowledgements
We thank Enago for English review.

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

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Brass SD, Guiot MC, Albrecht S, Glikstein R, Mohr G. Metastatic hemangiopericytoma presenting as an epidural spinal cord lesion. Can J Neurol Sci 2004;31:550-3.
2. Cole CD, Schmidt MH. Hemangiopericytomas of the spine: Case report and review of the literature. Rare Tumors 2009;1:e43.
3. Damodaran O, Robbins P, Knuckey N, Bynevelt M, Wong G, Lee G. Primary intracranial haemangiopericytoma: Comparison of survival outcomes and metastatic potential in WHO grade II and III variants. J Clin Neurosci 2014;21:1310-4.
4. El Hindy N, Ringelstein A, Forsting M, Sure U, Mueller O. Spinal metastasis from malignant meningeal intracranial hemangiopericytoma: One-staged percutaneous Onyx embolization and resection—a technical innovation. World J Surg Oncol 2013;11:152.
5. Elefante A, Peca C, Del Basso De Caro ML, Russo C, Formica F, Marinelli G, et al. Symptomatic spinal cord metastasis from cerebral oligodendroglioma. Neurol Sci 2012;33:609-13.
6. Fukuda Y, Watanabe K, Toyama Y, Mikami S, Matsumoto M. Metastasis of intracranial meningeal hemangiopericytoma to thoracic spine 17 years after surgical excision: A case report. J Orthop Sci 2015;20:425-9.
7. Ghia AJ, Chang EL, Allen PK, Mahajan A, Penas-Prado M, McCutcheon IE, et al. Intracranial hemangiopericytoma: Patterns of failure and the role of radiotherapy. Neurosurgery 2013;73:624-31.
8. Guthrie BL, Ebersold MJ, Scheithauer BW, Shaw EG. Meningeal hemangiopericytoma: Histopathological features, treatment, and long-term follow-up of 44 cases. Neurosurgery 1989;25:514-22.
9. Jaakkeleinen J, Servo A, Haltia M, Wahlstrom T, Valtonen S. Intracranial hemangiopericytoma: Radiology, surgery, radiotherapy, and outcome in 21 patients. Surg Neurol 1985;23:227-36.
10. Johnson DR, Kimmel DW, Burch PA, Cascino TL, Giannini C, Wu W, et al. Phase II study of subcutaneous octreotide in adults with recurrent or progressive meningioma and meningeal hemangiopericytoma. Neuro Oncol 2011;13:530-5.
11. Kim JH, Jung HW, Kim YS, Kim CJ, Hwang SK, Paek SH, et al. Meningeal hemangiopericytomas: Long-term outcome and biological behavior. Surg Neurol 2003;59:47-54.
12. Kruse F, Jr. Hemangiopericytoma of the meninges (angioblastic meningioma of Cushing and Eisenhardt). Clinico-pathologic aspects and follow-up studies in 8 cases. Neurology 1961;11:771-7.
13. Lee JY, Kim SH, Joo SP, Kim TS, Jung S, Kim JH, et al. Spinal metastasis from cranial meningeal hemangiopericytomas. Acta Neurochir 2006;148:787-90.
14. Liu HG, Yang AC, Chen N, Yang J, Qiu XG, Zhang JG. Hemangiopericytoma in the spine: Clinical features, classification, treatment, and long-term follow-up in 26 patients. Neurosurgery 2013;72:16-24.
15. Lorigan P, Verweij J, Papai Z, Rondhuis S, Le Cesne A, Leahy MG, et al. Phase III trial of two investigational schedules of ifosfamide compared with standard-dose doxorubicin in advanced or metastatic soft tissue sarcoma: A European Organisation for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group Study. J Clin Oncol 2007;25:3144-50.
16. Nonaka M, Kohmura E, Hirata M, Hayakawa T. Meningeal metastasis of hemangiopericytoma of thoracic spine. Clin Neurol Neurosurg 1998;100:228-30.
17. Perry A, Louis DN, Scheithauer BW, Budka H, von Deimling A. Meningeal tumours. In: Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, editors. WHO classification of tumours of the central nervous system. Lyon: IARC; 1998;100:63-8.
18. Scott M, Kellett G, Peale A. Angioblastic meningioma (hemangiopericytoma) of the spine. Surg Neurol 1974;2:35-8.
19. Rutkowski MJ, Bloch O, Jian BJ, Chen C, Sughrue ME, Tihan T, et al. Management of recurrent intracranial hemangiopericytoma. J Clin Neurosci 2011;18:1500-4.
20. Rutkowski MJ, Jian BJ, Bloch O, Chen C, Sughrue ME, Tihan T, et al. Intracranial hemangiopericytoma: Clinical experience and treatment considerations in a modern series of 40 adult patients. Cancer 2012;118:1628-36.
21. Scott M, Kellett G, Peale A. Angioblastic meningioma (hemangiopericytoma) of the cerebellar fossa with metastases to the temporal bone and the lumbar spine. Surg Neurol 1974;2:35-8.