SNI
SURGICAL NEUROLOGY INTERNATIONAL
SNI: Neuro-Oncology
OPEN ACCESS
For entire Editorial Board visit : http://www.surgicalneurologyint.com
Editor: Ekkehard Kasper, M.D., Harvard Medical School, Boston, MA, USA

Case Report

Intramedullary and intratumoral hemorrhage in spinal hemangioblastoma: Case report and review of literature

Satoshi Kiyofuji[1,2], Christopher S. Graffeo[1], Munehiro Yokoyama[3], Shigeo Sora[2]

[1]Department of Neurologic Surgery, Mayo Clinic, Rochester, MN, USA, [2]Departments of Neurosurgery and [3]Pathology, Tokyo Metropolitan Police Hospital, Tokyo, Japan

E-mail: *Satoshi Kiyofuji - skiyofu1@gmail.com; Christopher S. Graffeo - Graffeo.Christopher@mayo.edu; Munehiro Yokoyama - muneyoko@mrh.biglobe.ne.jp; Shigeo Sora - mastersurgeon@gmail.com
*Corresponding author

Received: 09 October 18 Accepted: 31 October 18 Published: 04 December 18

Abstract

Background: Intramedullary hemorrhages involving spinal hemangioblastomas are rare. They are frequently associated with devastating neurologic outcomes, despite with emergent surgical intervention. Here, we presented an example of an intramedullary hemorrhage occurring in a spinal hemangioblastoma, where the patient markedly improved with surgery. Additionally, the appropriate literature was reviewed (including intraoperative video).

Case Description: A 49-year-old female with a 4-year history of tingling in the left lower extremity presented with vomiting, stepwise worsening of bilateral scapular pain, new upper motor neuron signs, and severe sensory loss bilaterally below C4 on the left and T4 on the right. The magnetic resonance imaging demonstrated a well-circumscribed, uniformly enhancing intramedullary tumor at the C2 level with hyperintensity on the T2 study consistent with acute hemorrhage and cord edema. An urgent C2 laminectomy was performed for gross total tumor resection. Intraoperatively, intramedullary hemorrhage was identified anterior to the tumor mass and was confirmed histopathologically. Postoperatively, the patient had no new sensorimotor deficits and fully recovered within two postoperative months.

Conclusions: Patients presenting with acute intramedullary hemorrhage within hemangioblastomas of the spinal cord may demonstrate significant postoperative neurological recovery.

Key Words: Hemangioblastoma, intramedullary hemorrhage, intramedullary spine tumor, intratumor hemorrhage, isolated spine hemangioblastoma

Video available on: www.surgicalneurologyint.com

Access this article online
Website: www.surgicalneurologyint.com
DOI: 10.4103/sni.sni_344_18
Quick Response Code:

INTRODUCTION

Hemangioblastomas are the third most frequent spinal cord tumors after ependymomas and astrocytomas and account for 2–15% of all intramedullary tumors.[7] Although they occur at all spinal levels, they most commonly involved the cervical spine.[2] A considerable number of these patients are diagnosed with von Hippel–Lindau disease (e.g. 20–30%).[8] Hemangioblastomas typically follow an indolent course, with occasional

How to cite this article: Kiyofuji S, Graffeo CS, Yokoyama M, Sora S. Intramedullary and intratumoral hemorrhage in spinal hemangioblastoma: Case report and review of literature. Surg Neurol Int 2018;9:250.
http://surgicalneurologyint.com/Intramedullary-and-intratumoral-hemorrhage-in-spinal-hemangioblastoma:-Case-report-and-review-of-literature/

tumors demonstrating acute intramedullary hemorrhage resulting in profound neurologic deficits.[2] Here, we report an acute intramedullary hemorrhage into a hemangioblastoma occurring at the C2 spinal level in a 49-year-old female who presented with acute cervical spastic quadriparesis.

CASE PRESENTATION

Patient history

A 49-year-old female presented with 1 week of bilateral scapular pain, and 1 day of worsening of the scapular pain accompanied by vomiting. Her neurological examination revealed no gross weakness but moderate dysesthesias (5/10) below the C4 spinal level on the left, and T4 on the right. Vibratory sensation was decreased in both lower extremities, left more than right, and she exhibited diffuse hyperreflexia throughout the upper and lower extremities.

MR and computed tomography imaging

The cervical magnetic resonance imaging (MRI) demonstrated a uniformly contrast-enhancing mixed -density intramedullary spinal tumor, with confluent T2 hyperintensity noted from the upper cervical spine to the T1 level. A region of low signal intensity on T2* was identified within and extending from the anterior limit of the tumor, suggestive of acute hemorrhage into an intramedullary hemangioblastoma [Figures 1a, b and 2a, b]. The contrast-enhanced computed tomography identified a well-circumscribed hypervascular mass measuring 13 mm × 9 mm × 11 mm, with a prominent, tortuous vessel emerging from the posterior aspect of the tumor [Figure 3a and b].

Surgery

The patient underwent an immediate C1–C3 laminectomy performed under microscope visualization and with intraoperative monitoring (e.g., somatosensory-evoked potential and motor-evoked potential monitoring).

Upon opening of the arachnoid, the tumor was immediately apparent on the dorsal surface of the spinal cord, associated with multiple neoplastic vessels. Indocyanine green (ICG) angiography was performed (15 mg IV) and identified five major feeding arteries and two draining veins. Temporary clips were applied to the feeding arteries, and the operation was paused for 10 min to confirm stability of the electrical potentials. Vessels were then cut and cauterized, followed by circumferential resection of tumor. Frank intramedullary hemorrhage was noted anterior to the tumor itself and removed. Repeat ICG injection was negative for residual circulation to the tumor and demonstrated only subtle distal drainage through the remaining draining vein, which was cauterized. Ultimately, a complete en-bloc resection was achieved [Video 1].

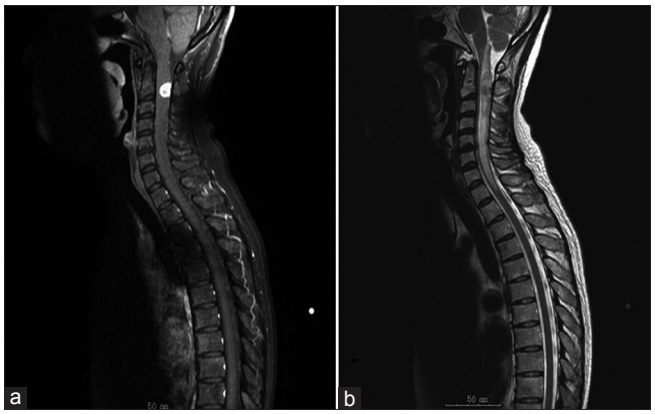

Figure 1: T1-weighted images (T1WI) with contrast of magnetic resonance imaging (MRI) show a uniformly enhanced intramedullary tumor with contrast at C2 level (a). In T2-weighted images (T2WI), significant edema was noted down to T1 level (b)

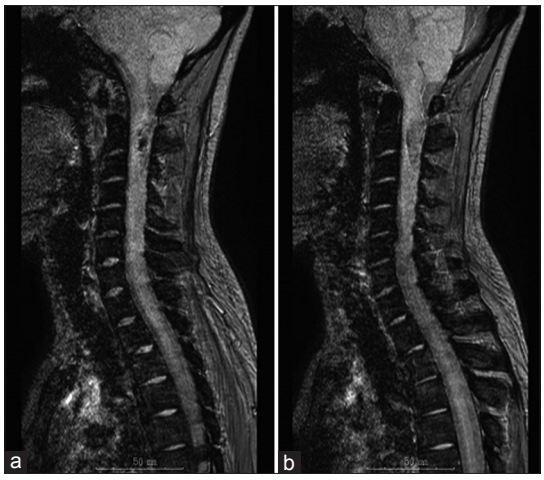

Figure 2: In T2* sequence of MRI, low intensity area was noted within (a) and anterior to the tumor (b), which suggested intratumoral/intramedullary hemorrhages from the tumor

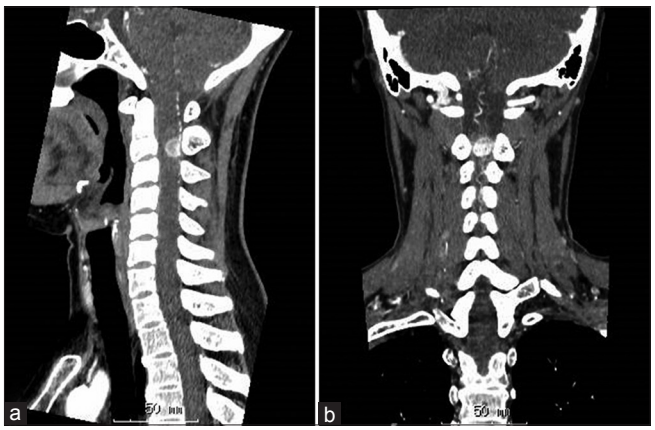

Figure 3: Computed tomography (CT) images of sagittal (a) and coronal (b) section demonstrate a clearly circumscribed hypervascular tumor sized in 13 × 9 × 11 mm with a tortuous vessel posteriorly to the tumor

Postoperative course

Postoperatively, the patient demonstrated no motor deficit, but an increased severity of the left upper and lower extremity sensory disturbance. Successive postoperative MRI studies demonstrated complete resection of the tumor [Figure 4a] and progressive diminution of the T2 cord hyperintensity [Figure 4b-d]. Two months postoperatively, the patient had no residual motor deficit, but a residual severe upper and lower sensory deficit bilaterally below T5. Her neurological status remained unchanged at 6 postoperative months.

Histology

The final histopathology was consistent with a hemangioblastoma with intratumor hemorrhage (WHO grade I; Figure 5a and b).

DISCUSSION

A patient with a cervical spinal hemangioblastoma accompanied by an acute intramedullary hemorrhage had preserved motor function but a severe residual sensory deficit below the T5 level following surgical decompression. Roonprapunt *et al*. described 19 patients with intramedullary spine hemangioblastomas, accompanied by edema, syrinx formation, and/or tumor cysts presenting in an indolent fashion over 29.8 months; the patients did well following surgical resection/decompression.[6] Lonser *et al*. also described resolution of symptoms/signs for symptomatic hemangioblastomas contributing to cord compression.[4]

When these lesions hemorrhage, it typically occurs into the subarachnoid space rather than within the cord itself (e.g., intramedullary). Five prior reports describe six patients with intramedullary hemangioblastomas presenting with intratumoral hemorrhages [Table 1].[1-3,8,10] Typically, patients presented with severe acute motor deficits (e.g., paraplegia, quadriplegia), none of which recovered postoperatively. In our case, the patient uniquely had no motor deficit but showed profound sensory changes; postoperatively,

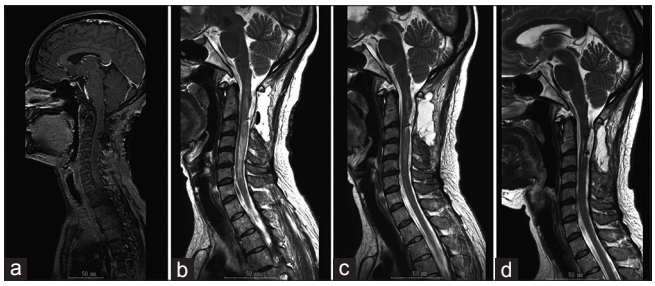

Figure 4:T1WI with contrast in MRI demonstrated complete resection of the tumor (a). In T2WI, high-intensity area significantly improved over time [(b) postoperative day (POD) 6; (c) POD 11; (d) POD 18)

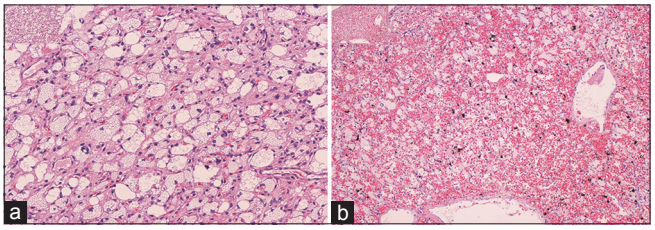

Figure 5: Pathology specimen pictures of the tumor. In high power view of hematoxylin and eosin (HE) staining, tumor cells had an abundant clear vacuolated cytoplasm and small uniform nuclei, which is characteristic to hemangioblastoma (a). Intratumor hemorrhage was identified (b)

Table 1: Literature review of intramedullary hemorrhage of spine hemangioblastoma

Patient number	Studies (authors and year)	Age (years), sex	Location	Size (cm)	VHL disease	Symptoms (McCormick Scale)	Surgery	Surgery timing	Follow-up	Recovery (McCormick Scale)
1	Yu *et al*., 1994[10]	31, F	T6	5	–	Paraplegia, preceding dysesthesia and neck pain (V)	T3-6 LAM, GTR	Immediate	7 YR	Complete thoracic myelopathy at T2 (V)
2		28 M	T7	Large	+	Paraplegia, preceding pain in lower back, groin, buttocks, and feet (V)	T7-8 LAM, GTR	Immediate	6 MO	Spastic paraplegia, spastic bladder (V)
3	Glasker *et al*., 2005[1]	34 F	T9-11	2.2	+	Sudden acute paraparesis, conus cauda syndrome (V)	N/A	N/A	N/A	N/A
4	Sharma *et al*., 2009[8]	39 M	T3-6	2	–	Paraplegia and bilateral upper extremity weakness (V)	T3-8 LP, GTR	Emergent	7 MO	Paraplegic, some improvement in bilateral upper extremity strength (V)
5	Gluf *et al*., 2014[2]	22 F	C7	N/A	–	Acute-onset quadriparesis (V)	C5-T2 LP, resection	Urgent	3 YR	Full upper extremity function, no use of lower extremities, no bladder function (V)
6	Koda *et al*., 2014[3]	40 M	T8	N/A	–	Acute paraplegia (V)	T7-9 LAM, GTR	Elected: one MO after onset	Unknown	No apparent neurologic recovery: paraplegia (V)

LAM=Laminectomies; LP=Laminoplasty; GTR=Gross total resection; MO=Months; YR=Years

the motor function remained intact, but the sensory deficit was more severe.

Surgical indications for intramedullary hemangiobastomas with acute intratumoral hemorrhages

It is unclear whether patients with spinal intramedullary hemorrhages within hemangioblastomas benefit from surgery. Excepting one report from Koda *et al*.,[3] all operations were performed urgently. Although limited recovery may be expected in patients with mild-to-moderate injuries, those with profound plegias may only partially benefit from early surgery.

Steiger *et al*. conclude in their review of 20 patients that the benefits of early surgery for patients with moderate-to-severe deficits were effectively zero.[9] However, others have separately demonstrated good preoperative neurologic function, which predicts optimal postoperative functional outcome with early surgery.[5,9]

CONCLUSIONS

Intramedullary hemorrhages within spine hemangioblastomas are rare. Surgery is optimized by utilizing intraoperative monitoring and vascular visualization techniques (e.g., intraoperative ICG angiography). This case and literature review demonstrated that surgery may result in favorable postoperative neurological outcomes.

DISCLOSURE

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

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Glasker S, Van Velthoven V. Risk of hemorrhage in hemangioblastomas of the central nervous system. Neurosurgery 2005;57:71-6; discussion 71-76.
2. Gluf WM, Dailey AT. Hemorrhagic intramedullary hemangioblastoma of the cervical spinal cord presenting with acute-onset quadriparesis: Case report and review of the literature. J Spinal Cord Med 2014;37:791-4.
3. Koda M, Mannoji C, Itabashi T, Kita T, Murakami M, Yamazaki M, *et al*. Intramedullary hemorrhage caused by spinal cord hemangioblastoma:A case report. BMC Res Notes 2014;7:823.
4. Lonser RR, Oldfield EH. Spinal cord hemangioblastomas. Neurosurg Clin N Am 2006;17:37-44.
5. Montano N, Papacci F, Trevisi G, Fernandez E. Factors affecting functional outcome in patients with intramedullary spinal cord tumors: Results from a literature analysis. Acta Neurol Belg 2017;117:277-82.
6. Roonprapunt C, Silvera VM, Setton A, Freed D, Epstein FJ, Jallo GI. Surgical management of isolated hemangioblastomas of the spinal cord. Neurosurgery 2001;49:321-7; discussion 327-328.
7. Samartzis D, Gillis CC, Shih P, O'Toole JE, Fessler RG. Intramedullary spinal cord tumors: Part I-epidemiology, pathophysiology, and diagnosis. Global Spine J 2015;5:425-35.
8. Sharma GK, Kucia EJ, Spetzler RF. Spontaneous intramedullary hemorrhage of spinal hemangioblastoma: Case report. Neurosurgery 2009;65:E627-8; discussion E628.
9. Steiger HJ, Turowski B, Hanggi D. Prognostic factors for the outcome of surgical and conservative treatment of symptomatic spinal cord cavernous malformations: A review of a series of 20 patients. Neurosurg Focus 2010;29:E13.
10. Yu JS, Short MP, Schumacher J, Chapman PH, Harsh GRt. Intramedullary hemorrhage in spinal cord hemangioblastoma. Report of two cases. J Neurosurg 1994;81:937-40.