Case Report

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

Satoshi Kiyofuji¹,², Christopher S. Graffeo¹, Munehiro Yokoyama³, Shigeo Sora²

¹Department of Neurologic Surgery, Mayo Clinic, Rochester, MN, USA, ²Departments of Neurosurgery and ³Pathology, 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

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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 neurologic recovery.

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

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.⁶ 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

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tumors demonstrating acute intramedullary hemorrhage resulting in profound neurologic deficits. 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].
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. Lonser et al. also described resolution of symptoms/signs for symptomatic hemangioblastomas contributing to cord compression.

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]. 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, the patient demonstrated a severe residual sensory deficit below the T5 level following surgical decompression.
the motor function remained intact, but the sensory deficit was more severe.

**Surgical indications for intramedullary hemangioblastomas 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., 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. However, others have separately demonstrated good preoperative neurologic function, which predicts optimal postoperative functional outcome with early surgery.

**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/

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