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Surgical management of a cervical intramedullary hemangioblastoma presenting with intracystic hemorrhage by hemi-semi-laminectomy via a posterior approach

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Abstract

Background and Importance: Cervical spinal cord hemangioblastoma with intracystic hemorrhage is a rare entity and presents a challenge for clinicians with regard to its timely diagnosis and appropriate treatment.

Case Presentation: A 35-year-old man presented with sudden-onset acute and progressive neck pain and severe radicular pain in his left upper limb. Motor weakness and numbness in the left upper and lower limbs with gait ataxia followed 2 days later. His initial diagnosis was acute myelitis, and he was treated with glucocorticoids for 2 weeks. Follow-up contrast-enhanced magnetic resonance imaging (MRI) suggested a spinal cystic hemangioblastoma with intracystic hemorrhage at the C3-4 level. The tumor was totally removed by minimally invasive unilateral hemi-semi-laminectomy via a posterior transcystic approach. The postoperative course was uneventful, and postoperative MRI revealed no residual tumor. The patient showed full

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neurological recovery at the 1.5-year follow-up, and computed tomography with a volume-rendering technique showed regrowth of the left C3 lamina.
Conclusion: Close MRI follow-up and thin-section imaging are invaluable for the timely diagnosis of spinal hemangioblastoma with intracystic hemorrhage, which was safely removed via minimally invasive microsurgery in the present case.

Keywords
Cervical intramedullary hemangioblastoma, intracystic hemorrhage, minimally invasive surgery, magnetic resonance imaging, hemi-semi-laminectomy, posterior transcystic approach

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Introduction

Spinal cord hemangioblastomas are highly vascularized benign tumors that represent 2% to 15% of intramedullary tumors. They can either be solitary or develop as presentations of von Hippel–Lindau disease.[1] Despite microsurgical resection being the gold standard treatment,[2] total resection of spinal cord hemangioblastomas, especially intramedullary ones, without causing neurological deficits remains a challenging task for neurosurgeons.[3] Despite their highly vascularized nature, an intramedullary cystic hemangioblastoma accompanied by intracystic hemorrhage is quite rare. We herein report such a case in the cervical spine and suggest the importance of its differential diagnosis from acute myelitis because both share similar symptoms and radiological features. Close monitoring by magnetic resonance imaging (MRI) may help to establish an accurate diagnosis and lead to timely surgical treatment.

Clinical presentation

A 35-year-old man presented with sudden-onset acute neck pain that became progressively aggravated and radiated to his left upper limb. Two days later, he further developed weakness and numbness in the left upper and lower limbs with gait ataxia, sensory deficits in the right limbs, and fecal and urine retention. He also had a low-grade fever at that time and was initially diagnosed with acute myelitis at the outpatient clinic according to his history and the first MRI findings (Figure 1(a), (b)). On admission to the Department of Neurology, physical examination revealed motor strength of 4/5 in the left limbs with superficial hypoesthesia. After intravenous administration of glucocorticoids for 2 weeks, his limb weakness and numbness were slightly improved. A second MRI scan showed syringomyelia/cystic change at the C3-4 level, indicating myelomalacia and possible cystic hemangioblastoma (Figure 1 (c), (d)). Considering his relieved symptoms, a contrast-enhanced follow-up MRI scan was recommended. Approximately 1.5 months later, a third contrast-enhanced MRI scan revealed a small intramedullary contrast-enhanced cystic nodule at the C3-4 level with a non-enhancing rim (Figure 1(e), (f)). A final diagnosis of cervical spinal cord cystic hemangioblastoma was established. The patient was then referred to the Department of Neurosurgery for surgical management, and postoperative MRI

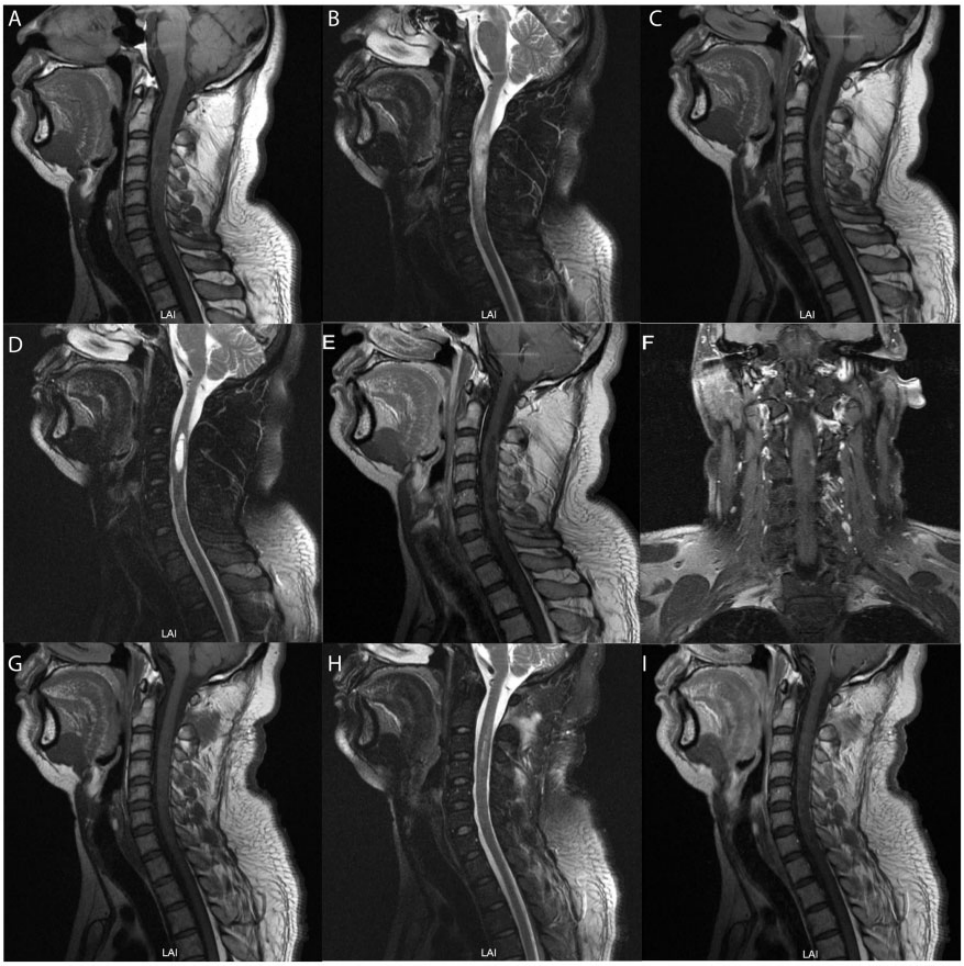

Figure 1. Sagittal magnetic resonance imaging (MRI) demonstrated an intramedullary patchy lesion that was (a) hypointense on T1-weighted imaging (T1WI) and (b) hyperintense on T2-weighted imaging (T2WI) from C2 to C6 and a markedly expanded spinal cord from C3 to C5. Two weeks later, follow-up MRI revealed an intramedullary cyst with (c) hypointense signals on T1WI and (d) hyperintense signals from the cyst on T2WI from C3 to C4. The previous spinal cord enlargement and peritumoral edema were markedly reduced on T2WI. (e, f) Approximately 1.5 months later, contrast-enhanced MRI indicated an intramedullary enhancing nodule on T1WI, about 2 mm in diameter, and a non-enhancing cyst wall at the left dorsal aspect of the spinal cord. MRI performed 1 week postoperatively showed a marked decrease in the size of the syrinx on sagittal (g) T1WI and (h) T2WI, and (i) sagittal T1-weighted contrast-enhanced MRI confirmed complete tumor removal.

suggested total resection of the tumor (Figure 1(g)–(i)).

The patient underwent general anesthesia in the prone position. Intraoperative electrophysiological monitoring was routinely used. The spinal level was determined using fluoroscopy. A 4-cm midline skin incision was centered over the C3 lamina, and the left paraspinal muscles were stripped subperiosteally. The left

upper part of the C3 lamina was opened with a unilateral hemi-semi-laminectomy. The dura was opened longitudinally along the midline under microscopic vision. After sharp dissection of the arachnoid matter, a reddish tumor nodule measuring $1.0 \times 0.5 \times 0.5\,cm^3$ was identified beneath the left dorsolateral nerve root, together with a long segment of swollen spinal cord. The major arterial feeder and draining vein were identified at the pial surface, coagulated with great care, and cut with microscissors (Figure 2(a)). The tension of the spinal cord was significantly decreased after puncturing the cyst with a fine needle caudal to the tumor nodule and draining about 5 mL of yellowish-brown liquid from the pseudocyst. The plane between the tumor and neural tissue was gradually developed by gentle traction of the tumor with microscissors, and the tumor nodule was removed en bloc (Figure 2(b)). The cyst wall was left untouched. After careful hemostasis, the dura was closed with

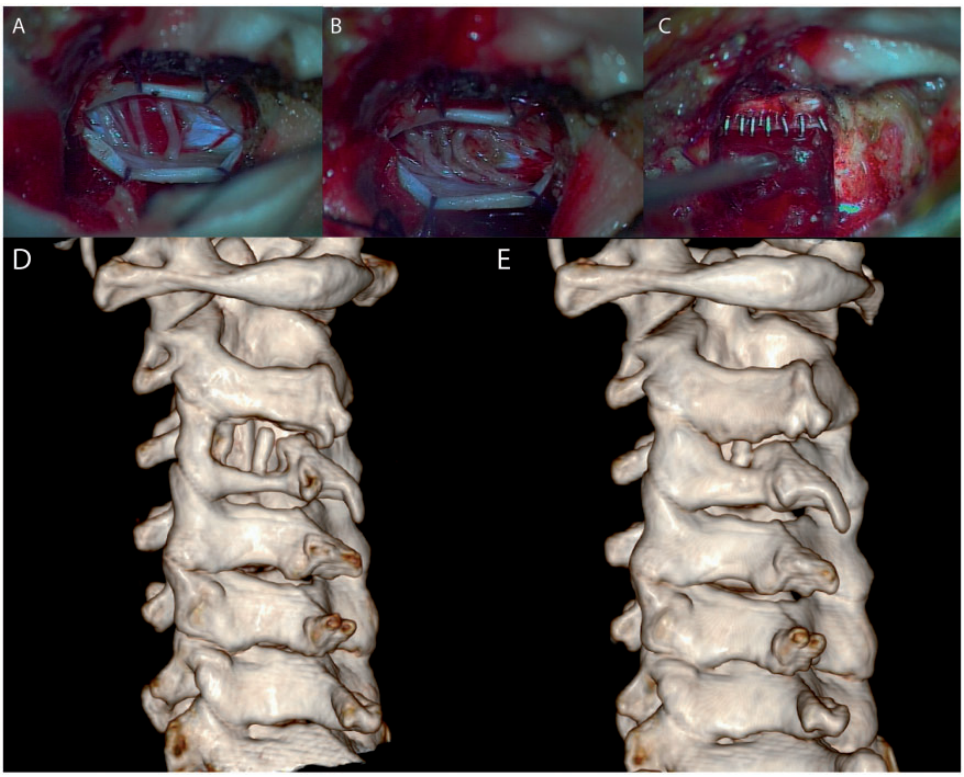

Figure 2. (a) Intraoperative image showed the characteristic bright red appearance of a spinal cord hemangioblastoma arising from the left dorsolateral pia at the C3 level with a long segment of spinal cord swelling. Note that the C3 dorsal root fascicles tightly coursed over the dorsal surface of the tumor and partly obscured the left posterolaterally located tumor. The arterial feeders and draining veins of the tumor were located at the pial surface. (b) Intraoperative image after removal of the tumor showed that the dorsolateral nerve rootlets were left intact. (c) After careful hemostasis, the dura was closed with titanium dural clips. (d) Postoperative computed tomography reconstruction of the spinal column with a volume-rendering technique (CT-VRT) suggested minimal removal of the bony structure. (e) Follow-up CT-VRT 1.5 years later and showed regrowth of the left C3 lamina.

titanium dural clips (Figure 2(c)). The paraspinal muscle, fascial, subcutaneous, and cutaneous layers were closed in separate layers.

The postoperative course was uneventful; neither complications nor neurological deterioration occurred. Postoperative computed tomography (CT) reconstruction of the spinal column with a volume-rendering technique (VRT) showed minimal removal of bony structures by this hemi-semi-laminectomy and preservation of the left C3 lamina as a bony "bridge" (Figure 2 (d)). Histopathological examination confirmed the diagnosis of hemangioblastoma (World Health Organization grade I). One week after surgery, the patient was discharged with no neurological deficits. He was symptom-free at follow-up in our clinic 1.5 years later, and CT-VRT showed regrowth of left C3 lamina (Figure 2(e)).

The patient provided informed consent for reporting of his individual data and participation in the study. This study was approved by the Medical Ethics Committee of the authors' hospital.

Discussion

Histologically, hemangioblastoma is a microvascular structure that is identical to a normal capillary. Therefore, it is resistant to high-pressure blood flow and is less likely to be a hemorrhagic lesion.[4] Only 0.24% of hemangioblastomas develop spontaneous hemorrhage,[5] and even fewer are associated with subarachnoid hemorrhage (0.06%).[6] To the best of our knowledge, only one case report has described intracystic hemorrhage in a patient with spinal hemangioblastoma, which was located in the epiconus.[5] Additionally, only two case reports have described the surgical management of cervical intramedullary hemangioblastoma with hemorrhage (C2 and C7, respectively), and both showed uniform contrast enhancement.[6,7] These radiological features allowed them to be much more easily differentiated from other intramedullary lesions than in our current case, which mainly showed cystic change and intracystic hemorrhage. However, once intramedullary bleeding occurs, especially in the cervical cord, it usually leads to catastrophic consequences such as quadriparesis. In contrast, patients with only subarachnoid hemorrhage usually recover from mild neurological deficits after surgery. Although our patient had intramedullary hemorrhage as evidenced by follow-up MRI, his neurological deficits did not deteriorate after the acute phase. This may be partially explained by the hemorrhage having been confined within the cyst, which may also have contributed to his benign clinical course and quick recovery after surgery. Routine laminoplasty was performed in the two above-mentioned cases of cervical intramedullary hemangioblastoma,[6,7] while we further minimized surgery-related trauma in the present case by performing hemi-semi-laminectomy.

Because the patient's initial cervical MRI findings resembled those of longitudinally extensive transverse myelitis, thin-section MRI follow-ups were conducted for differential diagnosis. The second MRI scan only showed an intramedullary cystic lesion (syringomyelia or myelomalacia). However, the third contrast-enhanced MRI scan revealed a small, well-demarcated intramedullary nodule with intense contrast enhancement and development of a pseudocyst, which strongly indicated a spinal hemangioblastoma. Thin-section MRI was invaluable because it is quite easy to miss a tumor with a diameter of only 2 mm on a thick-section scan. Finally, a posterior translaminar approach was used for total removal of the tumor, which was confirmed by postoperative MRI. Because the cyst capsule was composed of neuroglial tissues, it was left

untouched to avoid potential neural impairment.

In our case, the tumor nodule was located superficially on the dorsal aspect of the spinal cord. This location made it extremely easy to access via a posterior approach. Additionally, puncture of the tumor cyst and release of the cyst contents provided more working space for the surgical maneuvers. These features made the operation safe and simple. A recent article also described a posterior approach to manage anterior intramedullary cystic hemangioblastomas because drawbacks associated with the anterior approach can be avoided.[8] We agreed with these authors that a large pseudocyst not only provides a safe working corridor to the tumor but also offers extra working space after releasing the cyst contents. In the present case, these features made it possible for en bloc removal of the tumor via a small bony window through the cervical lamina and preservation of a bone "bridge" after hemi-semi-laminectomy, facilitating regrowth of the lamina and maintenance of spinal stability as evidenced by our follow-up CT. Improvements in imaging modalities and surgical equipment like endoscopes have made it possible to access spinal intradural–extramedullary lesions via a less-invasive interlaminar approach for common intraspinal tumors.[9] We also recently reported the use of percutaneous spinal endoscopy for resection of a cervical foraminal schwannoma,[10] sparing the bony structures and greatly maintaining the postoperative spinal stability.

Conclusion

We have herein reported a rare case of cervical spinal cord hemangioblastoma with intracystic hemorrhage. The patient's clinical course and initial MRI findings made the differential diagnosis from acute myelitis difficult. Thin-section MRI follow-up examinations were indispensable for the correct diagnosis, and the tumor was successfully removed by hemi-semi-laminectomy.

Abbreviations

MRI, magnetic resonance imaging; CT, computed tomography; VRT, volume-rendering technique.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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