Case report

Hemorrhagic intramedullary hemangioblastoma of the cervical spinal cord presenting with acute-onset quadriparesis: Case report and review of the literature

Wayne M. Gluf¹, Andrew T. Dailey²

¹Department of Neurological Surgery, University of Texas Southwestern Medical Center, Dallas, TX, USA,

²Department of Neurosurgery, University of Utah Health Sciences Center, Salt Lake City, UT, USA

Context: Hemangioblastomas of the spinal cord are uncommon vascular tumors. Patients commonly present with subtle neurologic findings that are thought to represent growth of the lesion over time. Hemorrhage of an intramedullary hemangioblastoma presenting as acute neurologic deficit is an extremely rare occurrence. Although the cervical spine is the most common location for hemangioblastoma of the spinal cord, there have been no previously published cases in the literature of intramedullary hemorrhage from such a lesion.

Findings: A 22-year-old woman with a previously undiagnosed spinal cord hemangioblastoma presented with sudden-onset dense quadriparesis due to intramedullary hemorrhage in the cervical spinal cord. The patient did not have any clinical findings of von-Hippel Lindau disease. Laminoplasty from C5 to T2 and posterior midline myelotomy for resection of the intramedullary tumor with hematoma evacuation were completed without complication.

Conclusion: Intramedullary hemangioblastoma of the spinal cord is uncommon, and hemorrhage from a cervical spinal cord lesion has not previously been reported. Symptoms from these usually indolent lesions are commonly associated with tumor growth, edema, or associated syrinx, whereas devastating acute neurologic deficit from hemorrhage is exceedingly rare. Microsurgical resection should be done in cases of symptomatic lesions and considered in isolated symptomatic lesions without the known diagnosis of von Hippel-Lindau disease.

Keywords: Cervical vertebrae, Hemangioblastoma, Hemorrhage, Spinal cord neoplasms

Hemangioblastomas are well-described vascular tumors of the central nervous system that may occur as part of the presentation of von Hippel-Lindau disease (VHL) or as isolated lesions not associated with VHL. These lesions frequently follow an indolent course, and neurologic symptoms are commonly attributed to mass effect from increasing tumor size or associated syringomyelia and only very rarely are the result of an acute and significant intramedullary hemorrhage. Hemangioblastomas are found at all levels of the spinal cord, with the cervical spinal cord the most frequent location; however, there are no documented reports of significant intramedullary hemorrhage from a cervical cord hemangioblastoma

presenting with devastating neurologic manifestations. The authors present their experience with this condition and review the published literature describing similar cases.

Case report

History and examination

A 22-year-old woman without significant medical history presented to the emergency room with sudden-onset quadriparesis. The patient reported experiencing neck pain that began 2 days before the development of her acute neurologic deficit. She managed her neck pain with an alternating regimen of heat and cold packs, which provided symptomatic relief. On the day of presentation, while lying on a heating pad at home, the patient became acutely unable to move her upper or lower

Correspondence to: Wayne M. Gluf, Department of Neurosurgery, University of Texas Southwestern Medical Center, 5323 Harry Hines Blvd., Dallas, TX 75390-8855 USA. Email: Wayne.Gluf@UTSouthwestern.edu

Figure 1 (A) Sagittal T2-weighted magnetic resonance image demonstrating large intramedullary hemorrhage and markedly expanded spinal cord diameter. (B) Sagittal T1-weighted magnetic resonance image with gadolinium enhancement showing the hemangioblastoma located in the ventral spinal cord at C7. (C) Axial T2-weighted magnetic resonance image demonstrating large hemorrhage within the spinal cord.

extremities secondary to painless quadriplegia. After evaluation at the community hospital, she was transported via air ambulance to our facility for neurosurgical evaluation and management. On admission, she was fully awake, alert, oriented, and cooperative. Her respiratory rate and oxygen saturation were normal. Her motor examination demonstrated motor strength of 4/5 in bilateral trapezius, 1/5 in her right biceps, 3/5 in the left biceps, and flaccid paralysis of bilateral triceps, wrist flexors/extensors, grip, and all lower extremity muscle groups (McCormick Grade V). The patient was sensitive to light touch and pinprick above the level of T2; deep tendon reflexes were absent in the bilateral upper and lower extremities. Joint position sense of the great toe was absent bilaterally, and digital rectal examination revealed absent sphincter tone. We identified no abnormal skin lesions, and her funduscopic examination results were normal bilaterally.

The findings of all hematologic studies (complete blood count, serum chemistry analysis, coagulation panel) were normal. Magnetic resonance imaging of the brain and spinal axis showed an intramedullary enhancing lesion at C7 and a large intramedullary hemorrhage extending from C5 to T2 (Fig. 1). The spinal cord diameter was markedly enlarged from C5 to T2, corresponding to the lesion and its associated hemorrhagic component. No other abnormal lesions were identified throughout the neuraxis. VHL was ruled out based on the findings on the physical and radiographic examinations.

Operation

Because the patient was admitted more than 8 hours after onset of quadriplegia methylprednisolone was

not administered and dexamethasone was given before surgery. Laminoplasty from C5 to T2 and microsurgical resection of the intramedullary tumor with hematoma evacuation were completed without complication. A posterior, midline myelotomy was done for evacuation of the large intramedullary hematoma and resection of the tumor, which was identified at the ventral aspect of the spinal cord. Intraoperative somatosensory-evoked potentials were monitored during the surgical procedure. Histopathologic analysis of the resected lesion confirmed the diagnosis of hemangioblastoma.

Postoperative course

The patient awoke after surgery with no new neurologic deficits. On postoperative Day 4, she was transferred to the hospital's inpatient physical rehabilitation ward. Throughout her treatment the patient made progressive neurologic recovery, and she was discharged home on postoperative Day 46. She had regained full bilateral arm and hand function and could perform all personal hygiene and bladder catheterization with minimal assistance (McCormick Grade III). At 6 months, she had reliable sensation throughout her right lower extremity, although her left leg remained insensate even 3 years after surgery. At 3-year follow-up, she had regained neither the use of her lower extremities nor bowel/bladder function.

Discussion

Hemangioblastoma of the spinal cord, whether associated with VHL disease or presenting as an isolated lesion, is an uncommon pathological condition with a slight male predominance¹ that accounts for less than

3% of intramedullary tumors.^{2,3} More than 70% of spinal cord hemangioblastomas occur as sporadic, single lesions, unassociated with VHL.² The patient described in this report did not have any clinical findings of VHL. Specifically, she had a normal retinal examination, no additional central nervous system lesions on magnetic resonance imaging, no tumors visible on abdominal/pelvic computed tomography scan, and negative genetic testing for VHL. Approximately 60% of intramedullary hemangioblastomas are located in the cervical spine, with the thoracic cord the next most common site at nearly 40%, followed by a small number originating in the lumbar spinal cord.^{2,4} Most spinal cord hemangioblastomas are located in the dorsum of the spinal cord, but the patient described herein had a ventrally originating lesion, just deep to the anterior spinal artery within the spinal cord parenchyma.^{3,5-7} Nearly 33% of patients identified with an intraspinal hemangioblastoma have been shown to possess a tandem lesion elsewhere in the central nervous system.^{3,8}

As is the case with many intramedullary tumors, pain is the most common presenting complaint among patients with spinal hemangioblastoma and frequently precedes motor or sensory dysfunction.³ Development of neurologic symptoms from spinal cord hemangioblastoma is thought to be associated with subtle tumor progression, mass effect, presence of associated syrinx, or syrinx enlargement over time.^{5,6,9,10} Associated spinal cord syrinx in patients with intramedullary hemangioblastoma has been shown to be a function of tumor size.¹⁰ Hemorrhage with devastating neurologic sequelae from these lesions remains exceptionally rare.

In their series of 160 consecutive patients, Lonser and Oldfield^{6,11} reported that symptoms were referable to either tumor size as a result of more rapid growth or mass effect of an associated syrinx. In this large patient series, there was no report of neurologic signs or symptoms as a result of intramedullary hemorrhage. Roonprapunt *et al.*¹ presented 19 patients with intramedullary hemangioblastoma with significant variance in neurologic deficits at presentation. The mean duration of symptoms in their study was 20.8 months (range 2–84 months), with no acute hemorrhage reported. These findings led to their conclusion that hemangioblastoma is an indolent process with symptoms resulting primarily from edema or syrinx/cyst formation. The patient presented herein had symptoms of isolated neck pain without neurologic findings for only 2 days preceding her intramedullary hemorrhage and reported

no previous neck pain or neurologic symptoms that could be attributed to her cervical hemangioblastoma. McCormick and Stein¹² reported that hemangioblastomas commonly presented with an insidious onset in their series of intramedullary spinal cord tumors, and only rarely is there an acute event due to subarachnoid hemorrhage. No mention of acute intramedullary hemangioblastoma was noted in their extensive clinical series. Yu *et al.*¹³ reported two patients with acute intramedullary hemorrhage who were found to have hemangioblastomas involving the thoracic spinal cord; both caused acute paraplegia. One of these patients had a known diagnosis of VHL prior to presenting with hemorrhage, while the other did not. Both Rothstein¹⁴ and Sharma *et al.*³ documented cases of a single patient who presented with acute paraplegia from ruptured thoracic intramedullary hemangioblastoma. Despite the fact that the majority of spinal cord hemangioblastomas occur in the cervical cord, there have been no prior reports of intramedullary hemorrhage in this location in neurosurgical literature.

Subarachnoid hemorrhage has a spinal location in approximately 0.06% of presenting cases.¹⁵ In very rare instances, hemangioblastoma has been identified as the cause of the subarachnoid hemorrhage, but an intramedullary hemorrhage is the exception.¹⁶⁻²⁰

Treatment for spinal cord hemangioblastoma is reserved for patients who present with a neurologic deficit in a previously undiagnosed tumor or with development of a neurologic deficit in a previously diagnosed, but asymptomatic tumor.^{6,21} The patient in this report had neither prior neurologic symptoms referable to the cervical spinal cord nor a known diagnosis of intramedullary tumor. Patients with VHL frequently harbor numerous lesions, and surgery is limited to symptomatic lesions, whether directly from the hemangioblastoma or from its associated syrinx or spinal cord edema. A patient with an isolated lesion without VHL is more likely to undergo surgery in an attempt to establish tissue diagnosis. Microsurgery remains the definitive treatment for hemangioblastoma.^{1,5,6,9,14,21-24} It has been suggested that patients with isolated, sporadic intramedullary hemangioblastomas have better surgical outcomes than those with similar lesions with the diagnosis of VHL.⁸ Although gamma knife radiosurgery has been used to treat some patients, especially in cases of multiple hemangioblastomas, the long-term results for this treatment method are not yet available.^{1,4,9,14} Accordingly, symptomatic lesions should be completely resected with microsurgery. Fenestration of an associated syrinx is not required, as it is

common for the syrinx to regress after resection of the hemangioblastoma.^{14,21}

Conclusions

Intramedullary hemangioblastoma of the spinal cord is uncommon, and although the cervical spinal cord is the most common location for this lesion, no prior report of a hemorrhage in this area of the cord has been reported. Symptoms referable to such lesions are commonly associated with tumor growth, edema, or associated syrinx. An indolent course is usually seen, whereas devastating acute neurologic deficit from hemorrhage is exceedingly rare. Microsurgical resection should be done in cases of symptomatic lesions and considered in isolated symptomatic lesions without the known diagnosis of VHL.

Acknowledgment

The authors thank Kristin Kraus for her efforts in the preparation of this paper.

Disclaimer statements

Contributors Both authors contributed substantially to the conception, analysis, and writing of this paper.

Conflicts of interest There are no conflicts of interest.

Ethics approval Ethical approval was not necessary for a single-case report.

Funding None.

References

- Roonprapant C, Silvera VM, Setton A, Freed D, Epstein FJ, Jallo GI. Surgical management of isolated hemangioblastomas of the spinal cord. *Neurosurgery* 2001;49(2):321–7; discussion 327–8.
- Neumann HP, Eggert HR, Weigel K, Friedburg H, Wiestler OD, Schollmeyer P. Hemangioblastomas of the central nervous system. A 10-year study with special reference to von Hippel-Lindau syndrome. *J Neurosurg* 1989;70(1):24–30.
- Sharma GK, Kucia EJ, Spetzler RF. Spontaneous intramedullary hemorrhage of spinal hemangioblastoma: case report. *Neurosurgery* 2009;65(3):E627–8.
- Ammerman JM, Lonser RR, Dambrosia J, Butman JA, Oldfield EH. Long-term natural history of hemangioblastomas in patients with von Hippel-Lindau disease: implications for treatment. *J Neurosurg* 2006;105(2):248–55.
- Cohen-Gadol AA, Spencer DD, Krauss WE. The development of techniques for resection of spinal cord tumors by Harvey W. Cushing. *J Neurosurg Spine* 2005;2(1):92–7.
- Lonser RR, Oldfield EH. Spinal cord hemangioblastomas. *Neurosurg Clin N Am* 2006;17(1):37–44.
- Tihan T, Chi JH, McCormick PC, Ames CP, Parsa AT. Pathologic and epidemiologic findings of intramedullary spinal cord tumors. *Neurosurg Clin N Am* 2006;17(1):7–11.
- Conway JE, Chou D, Clatterbuck RE, Brem H, Long DM, Rigamonti D. Hemangioblastomas of the central nervous system in von Hippel-Lindau syndrome and sporadic disease. *Neurosurgery* 2001;48(1):55–63.
- Parsa AT, Chi JH, Jr. Acosta FL, Ames CP, McCormick PC. Intramedullary spinal cord tumors: molecular insights and surgical innovation. *Clin Neurosurg* 2005;52:76–84.
- Wanebo JE, Lonser RR, Glenn GM, Oldfield EH. The natural history of hemangioblastomas of the central nervous system in patients with von Hippel-Lindau disease. *J Neurosurg* 2003;98(1):82–94.
- Lonser RR, Butman JA, Oldfield EH. Pathogenesis of tumor-associated syringomyelia demonstrated by peritumoral contrast material leakage. Case illustration. *J Neurosurg Spine* 2006;4(5):426.
- McCormick PC, Stein BM. Intramedullary tumors in adults. *Neurosurg Clin N Am* 1990;1(3):609–30.
- Yu JS, Short MP, Schumacher J, Chapman PH, Harsh GR 4th. Intramedullary hemorrhage in spinal cord hemangioblastoma. Report of two cases. *J Neurosurg* 1994;81(6):937–40.
- Rothstein TL. Paraplegia resulting from rupture of previously asymptomatic intramedullary hemangioblastoma during coitus. *Ann Neurol* 1985;17(5):519.
- Hawkins CP, Heron JR. Subarachnoid hemorrhage from spinal tumor (in the absence of spinal symptoms or signs). *J Neurol Neurosurg Psychiatry* 1988;51(2):305–7.
- Cerejo A, Vaz R, Feyer PB, Cruz C. Spinal cord hemangioblastoma with subarachnoid hemorrhage. *Neurosurgery* 1990;27(6):991–3.
- Dijindjian M, Djindjian R, Houdart R, Hurth M. Subarachnoid hemorrhage due to intraspinal tumors. *Surg Neurol* 1978;9(4):223–9.
- Iwasaki Y, Koyanagi I, Hida K, Abe H. Anterior approach to intramedullary hemangioblastoma: case report. *Neurosurgery* 1999;44(3):655–7.
- Jallo GI, Kothbauer KF, Epstein FJ. Intrinsic spinal cord tumor resection. *Neurosurgery* 2001;49(5):1124–8.
- Van Hille PT, Abbott RJ, Cameron MM, Holland IM, Loizou LA. Recurrent spontaneous subarachnoid haemorrhage due to spinal haemangioblastoma. *J Neurol Neurosurg Psychiatry* 1987;50(5):639–40.
- Lonser RR, Oldfield EH. Microsurgical resection of spinal cord hemangioblastomas. *Neurosurgery* 2005;57(4 Suppl):372–6.
- Guidetti B, Fortuna A. Surgical treatment of intramedullary hemangioblastoma of the spinal cord. Report of six cases. *J Neurosurg* 1967;27(6):530–40.
- Parsa AT, Lee J, Parney IF, Weinstein P, McCormick PC, Ames C. Spinal cord and intradural-extramedullary spinal tumors: current best care practices and strategies. *J Neurooncol* 2004;69(1–3):291–318.
- Yasargil MG, Antic J, Laciga R, de Preux J, Fidelier RW, Boone SC. The microsurgical removal of intramedullary spinal hemangioblastomas. Report of twelve cases and a review of the literature. *Surg Neurol* 1976;3:141–8.