www.surgicalneurologyint.com

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Spine

Editor
Nancy E. Epstein, MD
Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook

ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

SNI
Open Access

Case Report

Intramedullary hemangioblastoma of the thoracic cord with a microsurgical approach: A case report and literature review

Eduardo Cattapan Piovesan[1], Werner Petry Silva[2], Adroaldo Baseggio Mallmann[2], Felipe Severo Lanzini[1], Bruna Zanatta de Freitas[1], Francisco Costa Beber Lemanski[1], Charles André Carazzo[1,2]

[1]Department of Neurosurgery, University of Passo Fundo, [2]Department of Neurosurgery, São Vicente de Paulo Hospital, Passo Fundo, Rio Grande do Sul, Brazil.

E-mail: *Eduardo Cattapan Piovesan - eduardocattapanpiovesan@gmail.com; Werner Petry Silva - wernerpetry@hotmail.com; Adroaldo Baseggio Mallmann - abmallmann@hotmail.com; Felipe Severo Lanzini - felanzini@hotmail.com; Bruna Zanatta de Freitas - bruunamx@gmail.com; Francisco Costa Beber Lemanski - franlemanski@hotmail.com; Charles André Carazzo - cacarazzo@hotmail.com

***Corresponding author:**
Eduardo Cattapan Piovesan, Department of Neurosurgery, University of Passo Fundo, Passo Fundo, Rio Grande do Sul, Brazil.

eduardocattapanpiovesan@gmail.com

Received : 22 March 2023
Accepted : 30 March 2023
Published : 14 April 2023

DOI
10.25259/SNI_252_2023

Quick Response Code:

ABSTRACT

Background: Spinal cord hemangioblastomas (HBs) account for 2–15% of all spinal cord neoplasms. They are the third most common primary intramedullary tumor (1–5%). Here, 72-year-old female presented with a thoracic intramedullary spinal HB that responded well to surgery.

Case Description: A 72-year-old female presented with a 3–4 years of progressive paresthesias and paraparesis. On examination, she exhibited diffuse distal weakness of the lower extremities. The magnetic resonance scan showed an intramedullary expansive lesion at the T1–T2 level that markedly enhanced with contrast with both proximal and distal hydromyelia. Surgery included a C7 partial and T1–T2 total laminectomies performed under microscope visualization with intraoperative monitoring. At surgery, there was a well-documented cleavage plane between the tumor and the cord; excision was facilitated using the cavitron ultrasonic surgical aspirator device.

Conclusion: Surgery is the gold standard treatment for treating/resecting HBs and should include utilization of an operating microscope and intraoperative monitoring.

Keywords: Hemangioblastoma, Neurosurgery, Spine surgery, Spine, Spine tumor

INTRODUCTION

Spinal hemangioblastomas (HBs) are benign and highly vascular tumors that can occur anywhere throughout the central nervous system. They account for 2–15% of all spinal cord neoplasm and are the third most common primary intramedullary tumors (1–5%). Notably, they most commonly involve the cervical spine and are often associated with perilesional edema, and/or peritumoral cysts.[4,8,9] Clinically, they contribute to varying degrees of myelopathy that corresponds with their size/location.[2] Here, a 72-year-old female presented with an expansive intramedullary HB at the T1–T2 spinal level resulting in a significant paraparesis that resolved following tumor resection.

CASE REPORT

Clinical presentation

A 72-year-old female presented with a 6 month history of progressive paresthesias and paraparesis. On examination, she exhibited distal lower extremity weakness (4/5 level dorsiflexion along with diffuse lower extremity hyperreflexia and bilateral Babinski signs). The magnetic resonance imaging (MRI) demonstrated an intramedullary expansive lesion at the T1–T2 level that markedly enhanced with contrast; it was also accompanied by cephalad and caudad hydromyelia [Figures 1 and 2].

Surgery

The patient underwent for a C7 partial and T1–T2 total laminectomies. Using microscope visualization and intraoperative monitoring, the dura was opened revealing an expansive and very vascular intramedullary cord lesion [Figure 3]. A cleavage plane between the tumor and cord facilitated gross-total operative resection of the lesion using the cavitron ultrasonic surgical aspirator device [Figure 4].

Pathology

The histopathological examination confirmed the diagnosis of an HB. The tumor included a rich vascular network and vacuolated cells with mild nuclear enlargement and clear cytoplasm. Immunohistochemical staining revealed stromal cells positive for inhibin A, S100, and CD31, and was immunonegative for CD56, and AE1-3 [Figure 5].

Postoperatively

Postoperatively, the patient's paraparesis markedly improved. The postoperative MRI revealed that the tumor had been totally removed, and the syrinxes/hydromyelia cephalad/caudad were diminished. One-year later, the patient was fully neurologically intact, and the magnetic resonance (MR) continued to confirm the lack of tumor recurrence along with further reduction of the cephalad/caudad syrinxes/hydromyelia [Figure 6].

DISCUSSION

Patients typically present with symptoms/signs of spinal HB during the fourth decade of life.[9] Approximately 70% of spinal hemangioblastoma (HB) are sporadic single lesions, with the remaining 30% representing von Hippel-Lindau (VHL)-associated familial cases.[10] The posterior fossa is the most common location for HB (45–50%), followed by the spinal cord (40–45%, where they typically extend over 1–2 vertebral levels) and the brain stem (5–10%). Over 75% of spinal HB are intramedullary (75%) and comprise the third most common intramedullary spinal cord neoplasm, following ependymomas and astrocytomas.[4,7] Approximately 60% of intramedullary HBs are located in the cervical spine, 40% involve the thoracic cord, followed by the lumbar spine (5–10%) and cauda equina (<1%) [Table 1].[3,5]

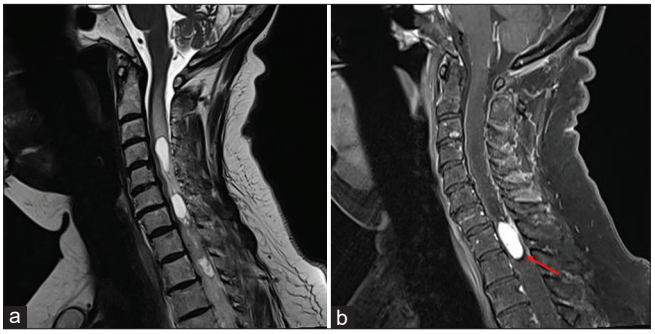

Figure 1: Magnetic resonance imaging of the cervical spine. (a) Sagittal T2-weighted revealed a syrinx with an intramedullary expansive lesion. (b) Sagittal T1-weighted dixon Gadolinium (Gd) enhancement image revealing an intramedullary intradural hyperintense mass (red arrow) at the T1-T2 level.

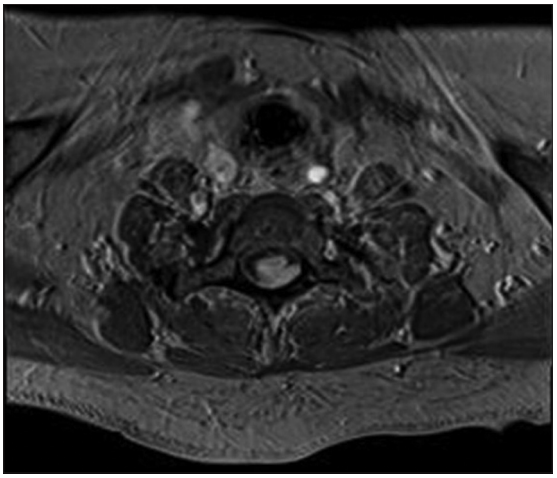

Figure 2: Axial T1-weighted image with Gd enhancement demonstrates the posterolateral hyperintense lesion infiltrating the spinal cord located in the posterior arch of T1.

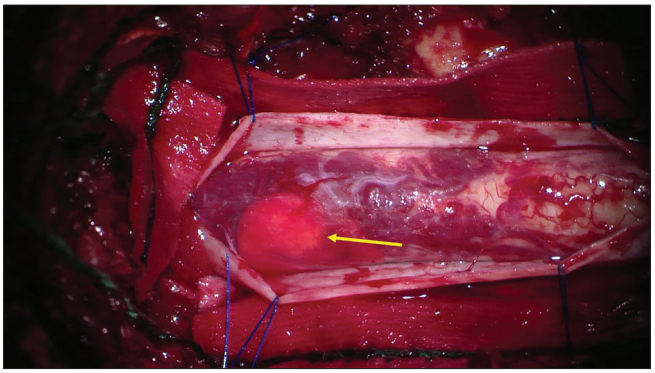

Figure 3: Posterolateral sulcus approach for the thoracic spinal cord intramedullary tumor. Tumor (yellow arrow).

Table 1: Summary of the cited cases, including the present case report.

Article	Patient's age	Sex	Level of the lesion	Symptoms	Surgical approach
Colamaria *et al.*[2]	32	Female	C2. Intramedullary – dorsal area	Numbness in the upper extremities with gait instability	Suboccipital craniectomy and total C1 laminectomy
Gluf and Dailey.[3]	22	Female	C7. Intramedullary – ventral area	Quadriparesis	Laminoplasty from C5 to T2. Hematoma evacuation
Mandigo *et al.*[6]	42	Male	T2. Intramedullary – dorsal area	Paresthesias	Laminectomy
	54	Male	C7–T1. Intramedullary – dorsal area	Numbness, spasticity	Laminectomy
	53	Female	T3. Intramedullary – dorsal area	Numbness	Laminectomy
	29	Female	T3–T4. Intramedullary – dorsal area	Weakness, numbness	Laminectomy
	45	Male	T3–T4. Intramedullary – dorsal area	Pain, weakness	Laminectomy
	38	Female	T11. Intramedullary – dorsal area	Numbness	Laminectomy
	42	Male	T7. Intramedullary – dorsal area	Pain, numbness	Laminectomy
	58	Male	C5–C6. Intramedullary – dorsal area	Pain	Laminectomy
	43	Female	T11–T12. Intramedullary – dorsal area the spinal cord	Headache, numbness	Laminectomy
	28	Female	C2. Intramedullary – dorsal area	Pain, numbness	Laminectomy
	55	Male	T2. Intramedullary – dorsal area	Pain, numbness	Laminectomy
	49	Female	T8–T9. Intramedullary – dorsal area	Numbness, weakness	Laminectomy
	45	Female	T3–T4. Intramedullary – dorsal area	Numbness	Laminectomy
	29	Male	T10–T11. Intramedullary – dorsal area the spinal cord	Paresthesias, numbness	Laminectomy
	28	Female	T10–T11, T12–L1. Intramedullary – dorsal area	Pain	Laminectomy
Mariniello *et al.*[7]	80	Female	C4–C5. Extradural	Pain in her left shoulder and weakness of her left arm	C4–C5 laminectomy
	25	Female	C6–C7. Extradural	Left brachial pain	C6–C7 laminectomy
Piovesan *et al.* (Current study)	72	Female	T1–T2. Intramedullary – dorsal area the spinal cord	Paresthesias and paraparesis	C7 partial and T1–T2 total laminectomies

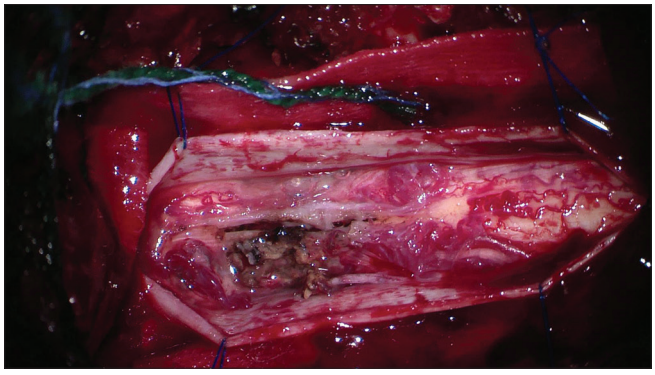

Figure 4: Macroscopic complete resection of the spinal cord lesion.

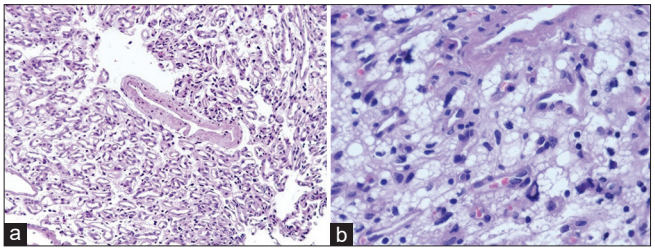

Figure 5: (a and b) Histopathological examination with hematoxylin and eosin staining revealed a large and vacuolated stromal cells, and intertwined with thin-walled capillaries, and clear cytoplasm. Immunohistochemical staining with inhibin A showing marked positivity of the neoplastic cells.

MRI gold standard examination for HB

MRI remains the gold standard for diagnosing HB; lesions are typically iso- to hypointense on T1-weighted sequences, and iso- to hyperintense on T2-weighted studies; further, they markedly enhance with contrast.[1] For patients with HB, it is critical to obtain MRI studies of the entire spine and brain before proceeding with definitive spinal surgery.[8]

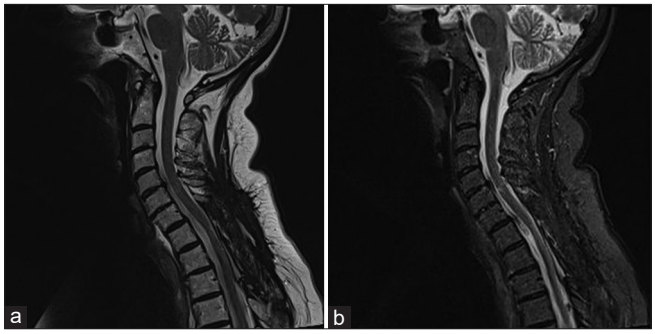

Figure 6: (a and b) Follow-up magnetic resonance imaging showing gross total removal of the hemangioblastoma (HBL) and reduction of the syrinx.

Indications for surgery

For patients with HB, the clinical and radiographic findings both determine whether surgical intervention is warranted. The optimal surgical management includes gross-total resection that is typically achieved in over 90% of the cases; this usually results in a 96% incidence of full functional recovery of the cases. Postoperatively, patients' symptoms often regress within 1–2 postoperative weeks, while follow-up contrast MR studies help confirm complete tumor resection, along with documenting regression of cephalad/caudad cysts (i.e., syrinxes/hydromyelia over 3–6 months).[2,6]

CONCLUSION

A 72-year-old female presented with myelopathy/paraparesis attributed to a T1–T2 HB. Following complete tumor resection, the patient regained normal neurological function, and the follow-up MR up to 6 months lateral confirmed no tumor recurrence and regression of the cephalad/caudad syrinxes/hydromyelia.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Baker KB, Moran CJ, Wippold FJ, Smirniotopoulos JG, Rodriguez FJ, Meyers SP, *et al*. MR imaging of spinal hemangioblastoma. AJR Am J Roentgenol 2012; 174:377-82.
2. Colamaria A, Fochi NP, Laguado YA, Blagia M, Leone A, Carbone F. Cervical intra and extramedullary hemangioblastoma with associated syringomyelia: A case report and review of the literature. Surg Neurol Int 2022;13:448.
3. 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.
4. Intramedullary Spinal Cord Tumors-StatPearls-NCBI Bookshelf. Available from: https://www.ncbi.nlm.nih.gov/books/NBK442031 [Last accessed on 2023 Jan 06].
5. Lonser RR, Oldfield EH. Spinal cord hemangioblastomas. Neurosurg Clin N Am 2006;17:37-44.
6. Mandigo CE, Ogden AT, Angevine PD, McCormick PC. Operative management of spinal hemangioblastoma. Neurosurgery 2009;65:1166-77.
7. Mariniello G, Corvino S, Corazzelli G, Franca RA, de Caro MD, Maiuri F. Spinal cervical extradural hemangioblastoma. J Craniovertebr Junction Spine 2022;13:192-7.
8. Messerer M, Cossu G, Pralong E, Daniel RT. Intramedullary hemangioblastoma: Microsurgical resection technique. Neurochirurgie 2017;63:376-80.
9. Yasuda T, Hasegawa T, Yamato Y, Kobayashi S, Togawa D, Banno T, *et al*. Relationship between spinal hemangioblastoma location and age. Asian Spine J 2016;10:309-13.
10. Yoda RA, Cimino PJ. Neuropathologic features of central nervous system hemangioblastoma. J Pathol Transl Med 2022;56:115-25.

How to cite this article: Piovesan EC, Petry Silva W, Baseggio Mallmann A, Lanzini FS, Zanatta de Freitas B, Lemanski FC, *et al*. Intramedullary hemangioblastoma of the thoracic cord with a microsurgical approach: A case report and literature review. Surg Neurol Int 2023;14:137.