CASE REPORT

Dorsal hemangioblastoma with holocord syringomyelia: case report

Alejandra Arévalo Sáenz, MD[1], Manuel Pedrosa Sánchez, MD[2]

1. Department of Neurosurgery, Hospital Clinic San Carlos, Madrid, Spain
2. Department of Neurosurgery, University Hospital La Princesa, Madrid, Spain

Abstract

Introduction: Intramedullary hemangioblastomas are usually accompanied by syringomyelia. However, a holocord syringomyelia is rare. The most common cause of syringomyelia continues to be Chiari disease, and only 10 cases of hemangioblastomas with holocord syringomyelia reported so far.

Case report: We present a case of a 35-year-old patient with a two-month history of cervicobrachialgia at the C7-C8 root level, previously preceded by pain at the D1-D2 level. Cervico-dorso-lumbar MRI revealed a medullar tumor with hyper-uptake mural nodule at the conus medullaris level accompanied by an extensive syringomyelic cavity from C5 to L1 compatible with medullary hemangioblastoma. The patient underwent surgery for tumor resection with subsequent resolution of her painful symptoms.

Conclusion: It is important to note that the surgery is aimed at treating the origin of this syringomyelia and not the syringomyelia itself. Although the majority of patients with holocord syringomyelia have Chiari as its cause, the possibility of focal spinal intramedullary tumors as being responsible for syringomyelia should not be forgotten.

Keywords: hemangioblastoma; holocord syringomyelia; spinal neoplasm; Chiari malformation; microsurgery

DOI: https://doi.org/10.55005/v2i1.4

Introduction

Spinal hemangioblastomas comprise 1.6-2.1% of all primary spinal cord tumors. They are the third most common primary spinal cord tumor, following astrocytoma and ependymoma, and represent 2%–6% of all intramedullary tumors. They are preferably located in the cervical or thoracic region and typically present as a highly vascularized solid lesion with an associated cystic component. Although between 50-70% of these tumors are associated with syringomyelia, they very rarely present a holocord syringomyelic cyst[1-3] Hemangioblastomas and ependymomas are the most common tumors associated with syringomyelia, while astrocytomas are usually more diffuse and with less tendency to present cysts[4].

Most hemangioblastomas occur sporadically, but up to 32% are part of the von Hippel-Lindau syndrome. 80% of these tumors are solitary[5].

Case report

A 35-year-old woman, with no history of interest, who debuted with acute symptoms of intermittent right cervicobrachialgia in C7-C8 dermatome of two months´evolution. The pain was preceded by right dorsal pain at the level of the T1-T2 territory. On examination, there is no neurological focality.

Whole spine magnetic resonance imaging (MRI) (***Figure 1***) diagnosed an intramedullary tumor with a hyperenhanced mural nodule accompanied by an extensive syringomyelic cavity from C5 to L1 compatible with spinal hemangioblastoma. The rest of the pre-operative tests were normal.

The patient underwent scheduled surgery for tumor resection under neurophysiological control (somatosensory and motor potentials and root control). A T10 laminectomy with complete microsurgical excision of a hypervascularized subpial spinal cord lesion with multiple pial vascular microafferents was performed (***Figure 2***). The surgery was performed in the anti-Trendelenburg position to avoid the massive leakage of cerebrospinal fluid from the extensive syringomyelic cavity.

In the postoperative period, the patient had dysesthesia at the T10 level and symptoms of fluid hypotension that remitted with acetazolamide. The pathology showed a result of hemangioblastoma.

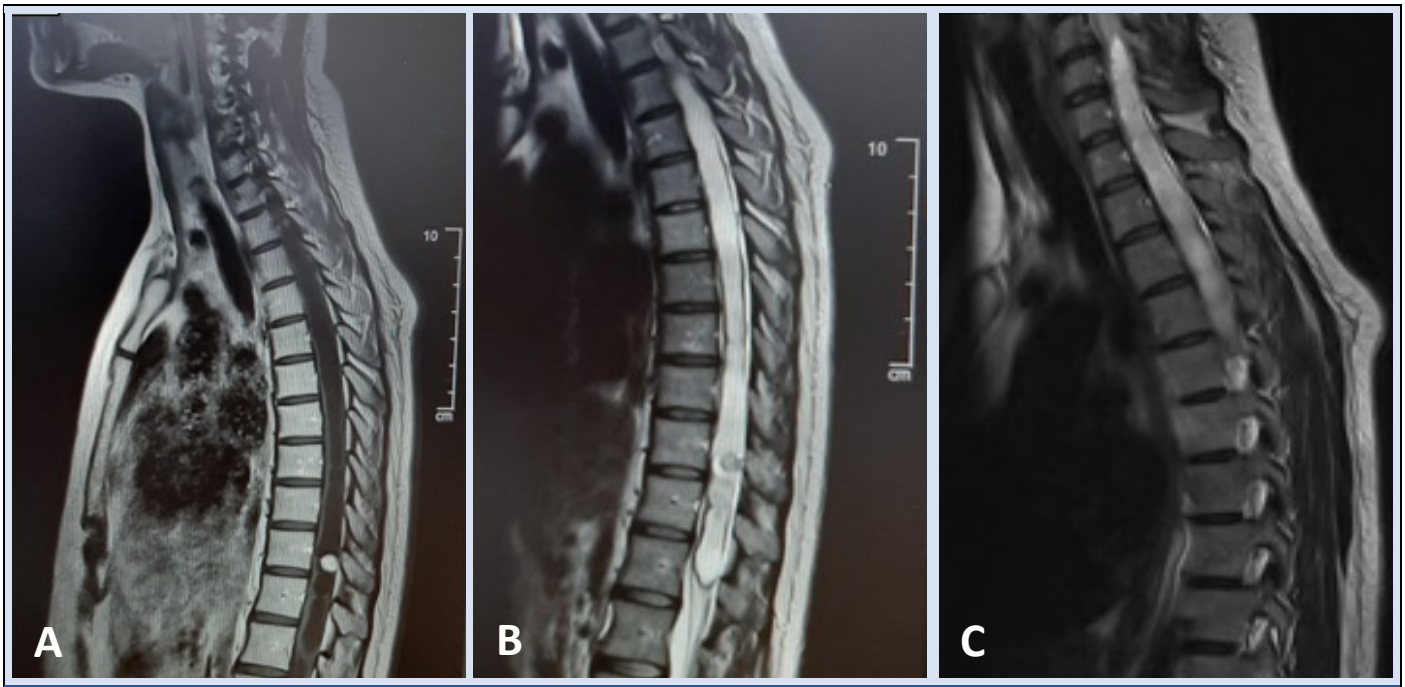

Figure 1. 3D Preoperative whole spine MRI. Sagittal slices on T1 (image A) and sagittal slices on T2 (images B and C). They demonstrate the existence of a well-defined intramedullary nodule at the level of T10, hypointense on T2, hyperintense on T1 with intense contrast enhancement surrounding it. It associates a prominent syringomyelic cavities from C5-C6 above to D12 below, occupying almost the entire cord.

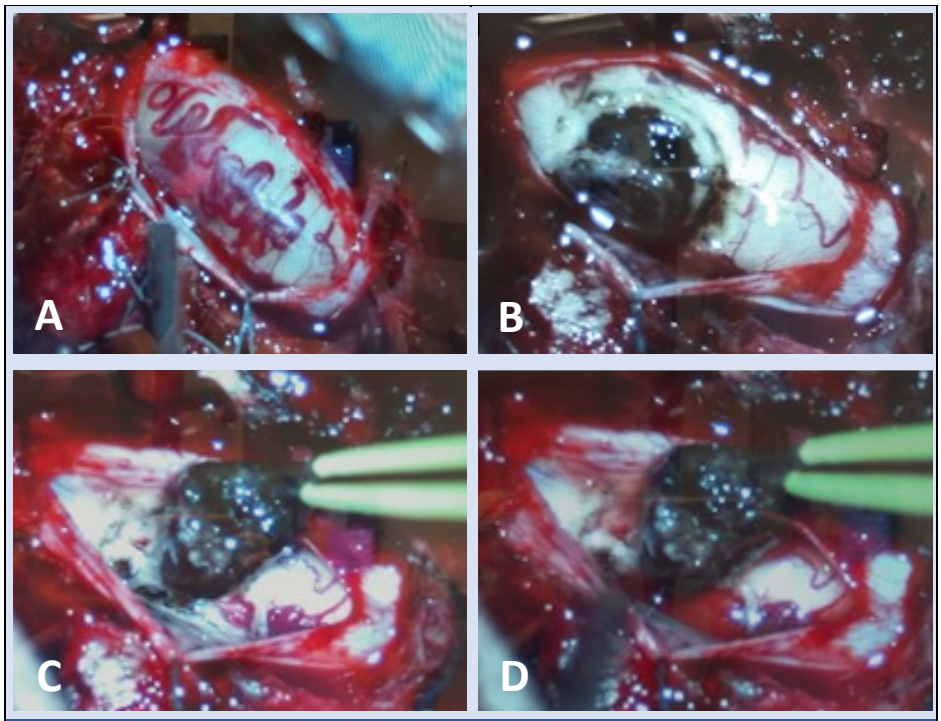

Figure 2. Intraoperative images after T10 dorsal laminectomy and dural opening showing: **A.** the intramedullary nodule with pial vascular afferents. **B.** lesion once disconnected from the pial vascular supply. **C.** the moment of total resection; **D.** removal of the lesion.

Discussion

The appearance of a cyst or syrinx was reported in 53.94% of spinal hemangioblastomas, which most often appeared in the cervical and thoracic spine[3].

The theory behind the formation of syringomyelic cysts is that the high interstitial pressure of an intramedullary tumor leads to extravasation of plasma into the central canal of the spinal cord; that is, the fluid that expands the central canal comes directly from the tumor or its associated vascularization[4]. Studies about high protein concentrations in the cerebrospinal fluid of tumoral intramedullary cysts support this theory[6]. Other theories also combine the obstruction of the cerebrospinal fluid and/or the obstruction of the flow of perimedullary extracellular fluid[1,7].

Varying degrees of asymptomatic central spinal medullary stenosis, common in healthy adults based on autopsy studies[8], may limit further expansion of the central canal and explain the multiseptated appearance often seen in cysts associated with intramedullary tumors. It should be noted that the higher the tumor level, the higher the incidence of an accompanying cyst[6]. However, there is no association between the levels of syringomyelia and severity of symptoms[3].

Syringomyelia is mainly associated with Chiari malformation and also with inflammatory pathologies, spinal attacks, trauma and intramedullary tumors of the spinal cord. Holocord syringomyelia is more frequently associated with Chiari malformation[9]. In a patient with this diagnosis, an exhaustive evaluation of the entire spine should be performed to rule out other pathologies.

In the literature there are only 11 cases[1,6,10-15] of hemangioblastomas with holocord syringomyelia (***Table 1***). The main difference in this group of patients is the surgical treatment, since the massive leakage of cerebrospinal fluid from the cavity can lead to permanent neurological symptoms. The resolution of the cavity is carried out naturally once the cause of the cavity has been treated, either by tumor resection or Chiari treatment. Based on our experience and previous literature, we believe that intraoperative the opening of the syrinx is not necessary during surgery, as associated edema and syringomyelia usually disappear over time after complete surgery resection[3].

Table 1. General characteristics of hemangioblastomas with holocord syringomyelia previously reported in the literature

Reference	Year	Number of patients	Age	Gender	Clinical presentation	Duration	Tumor location	Extension syringomyelia
Pai SB et al.	2003	2	17	M	Progressive quadriparesis and bladder involvement	1 month	Medullary conus	C2 - medullary conus
			35	M	Dysesthetic pain in both lower limbs with progressive paraparesis	6 months	T8	Cervicomedullary -T10
Wu et al.	2005	1	20	M	Intermittent right upper extremity numbness	3 months	T10-T11	Cervicomedullary -T11
Borkar SA et al.	2009	1	38	F	Spastic quadriparesis and hesitancy with urge incontinence	6 months	C7/T1	Cervicomedullary -conus
Cosgrove et al.	2015	1	50	M	Muscle wasting and weakness of both legs	6 years	T6/T7	Cervicomedullary -D12
Dutta et al.	2018	1	21	F	Progressive numbness/tingling of the face, hands and feet, gait instability, and hesitancy/ urge incontinence	6 months	T4	Cervicomedullaris - T12
Pojskic et al.	2018	1	30	M	Sudden onset urinary incontinence	Days	T11	C1 - T11
Maejima et al.	2019	3	42	M	Abnormal sensation in left upper extremity	2 months	T6	-
			43	F	Left hand numbness	3 years	T4	-
			42	M	Back pain	years	T7-T8	-
Knoop et al.	2019	1	37	M	Screening for von Hippel Lindau disease	-	T1-T2 and T11-T12	Cervicomedullary -T10

Conclusions

Although most patients with holocord syringomyelia are caused by Chiari malformation, others may have focal spinal intramedullary tumors. Therefore, it´s essential to assess the entire spine to avoid missing these injuries and missing the opportunity to properly treat these patients.

Disclosures

Conflict of Interest: All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent: Informed consent was obtained from the reported individual.

Funding: No funding was received for this research.

Correspondence

Alejandra Arévalo Sáenz

✉ Department of Neurosurgery, Hospital Clinic San Carlos, Calle Profesor Martín Lagos, s/n, 28040 Madrid, Spain

☏ +34913303000

@ praimale@hotmail.com

References

1. Pai SB, Krishna KN. Secondary holocord syringomyelia with spinal hemangioblastoma: a report of two cases. Neurol India. 2003 Mar;51(1):67-8.
2. Sarikaya S, Acikgöz B, Tekkök IH, Güngen YY. Conus ependymoma with holocord syringohydromyelia and syringobulbia. J Clin Neurosci. 2007 Sep;14(9):901-4. doi: 10.1016/j.jocn.2006.06.005.
3. Jankovic D, Hanissian A, Rotim K, Splavski B, Arnautovic KI. Novel Clinical Insights into Spinal Hemangioblastoma in Adults: A Systematic Review. World Neurosurg. 2022 Feb;158:1-10. doi: 10.1016/j.wneu.2021.10.105.
4. Stoodley M. The filling mechanism. In: Flint G, Rusbridge C, eds. Syringomyelia—a disorder of CSF circulation. Berlin Heidelberg: Springer-Verlag, 2014:87–101.
5. Chu BC, Terae S, Hida K, Furukawa M, Abe S, Miyasaka K. MR findings in spinal hemangioblastoma: correlation with symptoms and with angiographic and surgical findings. AJNR Am J Neuroradiol. 2001 Jan;22(1):206-17.
6. Wu TC, Guo WY, Lirng JF, Wong TT, Chang FC, Luo CB, et al. Spinal cord hemangioblastoma with extensive syringomyelia. J Chin Med Assoc. 2005 Jan;68(1):40-4. doi: 10.1016/S1726-4901(09)70131-5.
7. Samii M, Klekamp J. Surgical results of 100 intramedullary tumors in relation to accompanying syringomyelia. Neurosurgery. 1994 Nov;35(5):865-73; discussion 873. doi: 10.1227/00006123-199411000-00010.
8. Milhorat TH, Kotzen RM, Anzil AP. Stenosis of central canal of spinal cord in man: incidence and pathological findings in 232 autopsy cases. J Neurosurg. 1994 Apr;80(4):716-22. doi: 10.3171/jns.1994.80.4.0716.
9. Kumar J, Kumar A, Gupta S. Neurological pictures. Chiari I malformation with holocord syrinx. J Neurol Neurosurg Psychiatry. 2007 Feb;78(2):146. doi: 10.1136/jnnp.2006.099440.
10. Pojskic M, Arnautovic KI. Microsurgical Resection of Spinal Cord Hemangioblastoma: 2-Dimensional Operative Video. Oper Neurosurg (Hagerstown). 2018 Dec 1;15(6): E88-E89. doi: 10.1093/ons/opy123.
11. Dutta G, Singh D, Singh H, Srivastava AK, Jagetia A, Agrawal A. Dorsal hemangioblastoma manifesting as holocord syringomyelia. Surg Neurol Int. 2018 Apr 5; 9:73. doi: 10.4103/sni.sni_47_18.
12. Maejima R, Kawaguchi R, Aoyama M, Takeuchi M, Joko M, Takayasu M. [Three Cases of Small Hemangioblastoma with a Large Syringomyelia]. No Shinkei Geka. 2019 May;47(5):573-576. Japanese. doi: 10.11477/mf.1436203984.
13. Cosgrove J, Warren D, Derham C, Jamieson S. Holocord syrinx associated with haemangioblastoma. Pract Neurol. 2015 Dec;15(6):485-7. doi: 10.1136/practneurol-2015-001251.
14. Borkar SA, Kasliwal MK, Suri A, Sharma BS. Cervical Hemangioblastoma with holocord syrinx. Surg Neurol. 2009 Oct;72(4):437-8. doi: 10.1016/j.surneu.2008.11.014.
15. Knoop N, Seidel C, Frydrychowicz C, Meixensberger J. Combined Microsurgery and Radiotherapy for Multiple Spinal Cord Hemangioblastomas with Holocord Syrinx in von Hippel-Lindau Disease: A Case Report. J Neurol Surg Rep. 2019 Oct;80(4):e46-e50. doi: 10.1055/s-0039-3401808.