Case Report

Cervical intra and extramedullary hemangioblastoma with associated syringomyelia: A case report and review of the literature

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ABSTRACT

Background: Spinal hemangioblastoma (HB) is a highly vascularized tumor commonly presenting in the lower thoracic and lumbar segments. It typically causes spinal compression, extensive bleeding, and/or syringomyelia.

Case Description: A 32-year-old female presented with persistent headaches with a cervical MRI showing an intradural and extradural mass extending from the obex to C2. Following surgical tumor resection, the patient's symptoms resolved.

Conclusion: Resection of spinal HB requires direct removal of the tumor mass as the accompanying cystic components typically spontaneously regress.

Keywords: Cervical hemangioblastoma, Fenestration, Hemangioblastoma surgery, Intra and extradural, Syringomyelia

INTRODUCTION

Spinal hemangioblastomas (HBs) are slow-growing and highly vascularized tumors, accounting for 2–15% of primary spinal cord malignancies. They can arise as an isolated lesion or as multiple tumors spread throughout the central nervous system in association with syringomyelia, intramedullary hemorrhages, or Von Hippel-Lindau (VHL) syndrome (in 20–30% of the cases). Clinically, they contribute to varying degrees of myelopathy that correlate with the location of the tumor mass and accompanying abnormalities. Here, we describe a 32-year-old female with a cervical, intra- and extra-medullary HB extending from the obex to C2 resulting in headaches and numbness in the upper extremities.

CASE PRESENTATION

A 32-year-old female presented with occipital headaches and sudden onset of numbness in the upper extremities with gait instability without focal neurological deficits (Mc Cormick
grade II). The cervical MRI with gadolinium showed an oval mass in the intradural space at the obex extending to the superior margin of C2 markedly enhancing following the administration of contrast and associated with syringomyelia extending from the obex to C6 [Figure 1].

Surgery

The patient underwent suboccipital craniectomy and total C1 laminectomy for complete removal of the intramedullary mass under continuous spinal cord monitoring. Intraoperatively, the lesion was red and elastic in consistency, richly vascularized, and extended to the central canal of the spinal cord [Figure 2].

Postoperative course

Postoperatively, the gait was slightly impaired, while the headaches subsided, but the hypoesthesia in the upper extremities remained unchanged. The postoperative MRI demonstrated total tumor removal with a reduction of the syrinx. The patient was discharged 7 days later without focal deficits (Mc Cormick Grade I). At 6 months of follow-up, the patient had no residual complaints or deficits. Further, the MRI confirmed no tumor recurrence and a significant reduction of the accompanying syringomyelia [Figure 3].

Pathology

The pathological examination confirmed the diagnosis of a capillary HBL. It showed groups of large polygonal, lipid-laden stromal cells, interspersed with thin-walled, and closely packed blood-filled channels or vessels. Immunohistochemical staining revealed the presence of stromal cells positive for inhibin A and NSE while immunonegative for CD10 and EMA [Figure 4].

DISCUSSION

Sporadic spinal HBs usually present as a single lesion, mostly occupying the cervical area, followed by the thoracic and lumbosacral segment, with a relative incidence of 50.4%, 36.4%, and 13.4%, respectively. Most of the reported spinal

Figure 1: (a) Sagittal and (b) coronal T1-weighted image demonstrating the posterolateral hyperintense lesion infiltrating the spinal cord located between obex and the posterior arch of C2 (red arrow) with associated syringomyelia extending from the tumor to C6 (white arrow).

Figure 2: (a and b) Intraoperative images showing the subdural, intra/extra-axial lesion localization, which was completely removed after spinal cord exposure, following a complete C1 laminectomy and suboccipital craniotomy.

Figure 3: Follow-up MRI showing gross total removal of the HBL and reduction of the syrinx.
HBs are intramedullary\textsuperscript{6,10} and located in the dorsal area of the spinal cord [Table 1].\textsuperscript{3,7,9} Specifically in the posterior region of the denticulate ligament. MRI remains the gold standard, with T1-weighted imaging showing homogeneous, hyperintense signal, and T2-weighted imaging (T2WI) reflecting the highly vascularized nature that commonly characterizes these lesions. Additional information obtained with T2WI includes the demarcation of myelocoele in adjacent segments and the presence and extension of associated syringomyelia. Regardless of the location of the tumor in relation to the dura mater, these lesions almost always exhibit a marked enhancement after gadolinium administration.\textsuperscript{2}

### Management options

The most effective and definitive treatment for spinal HB are represented by microsurgical resection, which should only be considered if neurological deficits are present. In VHL patients with multiple small lesions, gamma-knife radiosurgery has shown promising results although further evidence is foreseen to recommend this treatment.\textsuperscript{3,7} Other factors that need to be acknowledged when considering preoperative embolization are the location of the tumor, its gross vascular anatomy, and the embolization material.\textsuperscript{3,8}

### Prognosis

Gross-total resection (i.e., minimally invasive cytoreductive surgery under neurophysiological monitoring) can be achieved in over 90\% of the cases and can result in full functional recovery in 96\% of cases.\textsuperscript{3,5,10} In general, symptoms

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**Table 1: Summary of the cited cases.**

| Article       | Patient's age | Sex | Level of the lesion                                      | Associated syrinx | Surgical approach                                                                 | Outcome                                                                 |
|---------------|---------------|-----|---------------------------------------------------------|-------------------|----------------------------------------------------------------------------------|------------------------------------------------------------------------|
| Barrey et al.\textsuperscript{2} | 31            | Female | Right C5-C6 intervertebral foramen. Intra-extraspinal | Absent            | Lateral approach. Limited bone drilling to enlarge the foramen. Dura cut around the tumor. Dural defect closed by packing fat with fibrin glue | Radiologic GTR. Partial neurologic recovery, no new neurologic deficits. Patient retained some deficits. |
| Chang et al.\textsuperscript{3}  | 32            | Male  | C3-C4                                                   | NA                | Laminoplasty from C5 to T2 and posterior midline myelotomy. Hematoma evacuation  | Radiologic GTR. Partial neurologic recovery, no new neurologic deficits. Patient retained some deficits. |
| Gluf et al.\textsuperscript{6}   | 22            | Female | C7, intramedullary, ventral aspect of the spinal cord. Hemorrhage extending from C5 to T2 | Absent            | Preoperative embolization. Piecemeal resection. Anterior and posterior approach. | Substantial debulking. Neurologic recovery, no new neurologic deficits. Patient retained deficits from the first operation. GTR. Total neurologic recovery, no new neurologic deficits. |
| Kim et al.\textsuperscript{7}    | 59            | Male  | C6-C7, recurrent (1° surgery 12 years before) left spinal nerve root. | Absent            | Laminectomy from C4 to C6. En bloc removal.                                      | Radiologic GTR. Neurologic recovery, no new neurologic deficits.         |
| Li et al.\textsuperscript{8}     | 72            | Male  | C5, left intradural extramedullary                     | Absent            | En bloc resection                                                                | GTR: Gross-total resection                                              |
| D’Oria et al.\textsuperscript{9} | 49            | Female | L1-L2, intradural extramedullary                        | Absent            |                                                                                   |                                                                        |

GTR: Gross-total resection

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**Figure 4:** (a) (× 40) and (b) (× 100) pathological examination with hematoxylin and eosin staining revealed large, stromal cells rich in lipids, and intertwined with thin-walled capillaries. C Immunohistochemical staining with inhibit A showing marked positivity of the neoplastic cells.
tend to spontaneously regress within 1–2 weeks after surgical tumor resection, whereas improvement of associated conditions such as cord enlargement, cyst, or syrinx requires between 3 and 6 months.[10] The use of radiosurgery or presurgical embolization remains highly controversial, as they provide no clear therapeutic benefit but can result in hemorrhage, medullary infarction, and radionecrosis.[1,3,10]

Recurrence of HB

Moreover, recurrence is frequently seen after partial resections[10] accounting for 6.25–7.7% of the cases.[10] Survival rates for sporadic HBs at 10 years are >90% and causes of death are mostly associated with other factors, that is, other cancerous manifestations of VHL.[5]

CONCLUSION

Spinal cervical HBs are rare malignancies and are best managed with gross total resection without the need for complete syrinx excision.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ampie L, Choy W, Khanna R, Smith ZA, Dahdaleh NS, Parsa AT, et al. Role of preoperative embolization for intradural spinal hemangioblastomas. J Clin Neurosci 2016;24:83-7.
2. Barrey C, Kalamadrides M, Polivka M, George B. Cervical dumbbell intra-extradural hemangioblastoma: Total removal through the lateral approach: Technical case report. Neurosurgery 2005;56:E625.
3. Chang H, Li J, Wang P, Lu X, Li B. Microsurgical treatment of cervical spinal hemangioblastoma. Neurochirurgie 2020;66:56-60.
4. Colamaria A, Sacco M, Iodice S, D’Oria, Parbonetti G, Carbone F, et al. Intradural extramedullary cavernous hemangioma of the cervicothoracic junction: A case report and review of the literature. Surg Neurol Int 2022;13:53.
5. D’Oria S, Giraldi D, Fanelli V, D’Angelo V. Sporadic hemangioblastoma of cauda equina: A case-report and brief literature review. Neurocirugía 2022.
6. Gluf WM, Dailey AT. Hemorrhagic intramedullary hemangioblastoma of the cervical spinal cord presenting with acute-onset quadripareisis: Case report and review of the literature. J Spinal Cord Med 2014;37:791-4.
7. Kim JH, Joo SM, Cho YE, Ha SW, Suh SH. Percutaneous onyx embolization of recurrent cervical nerve root hemangioblastoma: A case report and review of the literature. Clin Neuroradiol 2021;31:1209-13.
8. Li D, Choe S, Borys E, Serrone JC, Germanwala AV. Primary intradural extramedullary sporadic spinal hemangioblastomas: Case report and systematic review. World Neurosurg 2021;152:84-94.
9. Sun HI, Özduman K, Usseli MI, Özgen S, Pamir MN. Sporadic spinal hemangioblastomas can be effectively treated by microsurgery alone. World Neurosurg 2014;82:836-47.
10. Takai K, Taniguchi M, Takahashi H, Usui M, Saito N. Comparative analysis of spinal hemangioblastomas in sporadic disease and Von Hippel-Lindau syndrome. Neurol Med Chir (Tokyo) 2010;50:560-7.