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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).[2,4] 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 intra and extramedullary 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].

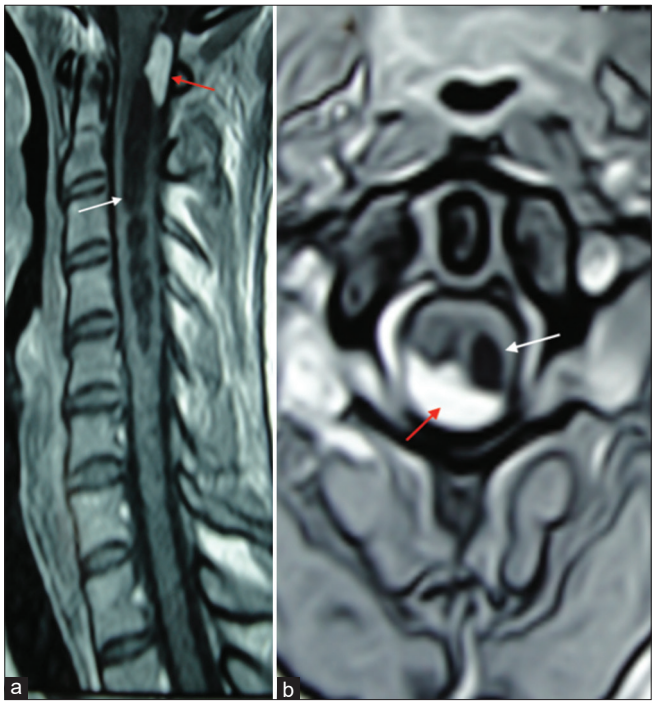

Figure 1: (a) Sagittal and (b) coronal T1-weighted image demonstrating the posterolateral hyperintense lesion infiltrating the spinal cord located between obex and the posterior arch of C2 (red arrow) with associated syringomyelia extending from the tumor to C6 (white arrow).

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,[7] 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.[7,10] Most of the reported spinal

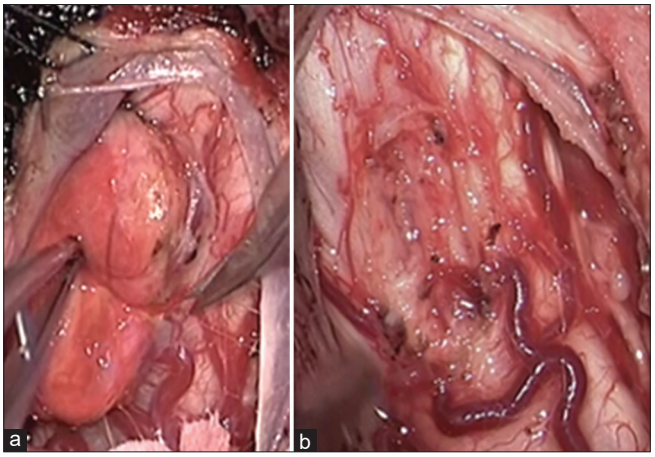

Figure 2: (a and b) Intraoperative images showing the subdural, intra/extra-axial lesion localization, which was completely removed after spinal cord exposure, following a complete C1 laminectomy and suboccipital craniotomy.

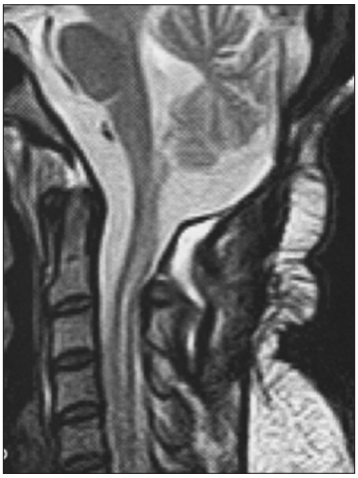

Figure 3: Follow-up MRI showing gross total removal of the HBL and reduction of the syrinx.

Table 1: Summary of the cited cases.

Article	Patient's age	Sex	Level of the lesion	Associated syrinx	Surgical approach	Outcome
Barrey *et al*.[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*.[3]	32	Male	C3-C4	NA	NA	Radiologic GTR. Partial neurologic recovery, no new neurologic deficits.
Gluf *et al*.[6]	22	Female	C7, intramedullary, ventral aspect of the spinal cord. Hemorrhage extending from C5 to T2	Absent	Laminoplasty from C5 to T2 and posterior midline myelotomy. Hematoma evacuation	Partial neurologic recovery, no new neurologic deficits. Patient retained some deficits.
Kim *et al*.[7]	59	Male	C6-C7, recurrent (1° surgery 12 years before) left spinal nerve root.	Absent	Preoperative embolization. Piecemeal resection. Anterior and posterior approach.	Substantial debulking. Neurologic recovery, no new neurologic deficits. Patient retained deficits from the first operation.
Li *et al*.[8]	72	Male	C5, left intradural extramedullary	Absent	Laminectomy from C4 to C6. *En bloc* removal.	GTR. Total neurologic recovery, no new neurologic deficits.
D'Oria *et al*.[5]	49	Female	L1-L2, intradural extramedullary	Absent	En bloc resection	Radiologic GTR. Neurologic recovery, no new neurologic deficits.

GTR: Gross-total resection

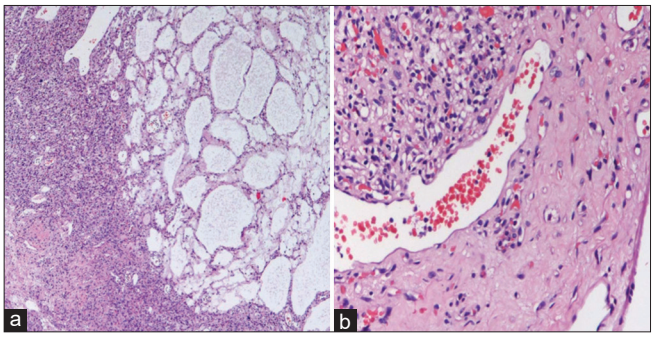

Figure 4: (a) (× 40) and (b) (× 100) pathological examination with hematoxylin and eosin staining revealed large, stromal cells rich in lipids, and intertwined with thin-walled capillaries. C Immunohistochemical staining with inhibin A showing marked positivity of the neoplastic cells.

HBs are intramedullary[6,10] and located in the dorsal area of the spinal cord [Table 1],[5,7,9] 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.[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.[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.[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.[3,5,10] In general, symptoms

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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