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Case Report

Intramedullary cervical spinal cord and cerebellar hemangioblastoma: A case report

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Received : 07 June 2022
Accepted : 22 June 2022
Published : 08 July 2022

DOI
10.25259/SNI_525_2022

Quick Response Code:

ABSTRACT

Background: Hemangioblastomas are benign tumors that develop in the central nervous system. They represent 1.5–2.5% of all intracranial tumors, and about 2–15% of all spinal cord tumors. They are highly associated with von Hippel–Lindau disease.

Case Description: A 36-year-old female presented with a 4-year history of progressive right upper extremity distal weakness and cervical pain. The magnetic resonance imaging demonstrated a homogeneously, contrast enhancing intradural/intramedullary tumor at C6–C7 with perilesional edema and a syrinx accompanied by a cerebellar cyst with a mural nodule. Surgery included excision of the spinal lesion and decompression and excision of the cerebellar cyst and mural nodule (i.e., median suboccipital craniectomy and cervical C5–C7 laminectomy).

Conclusion: Surgery is the gold standard treatment for symptomatic hemangioblastomas, and surgical approaches should minimize risk.

Keywords: Hemangioblastoma, Neurosurgery, Spine surgery, Spine, von Hippel-Lindau

INTRODUCTION

Hemangioblastomas are histologically benign tumors that develop in the central nervous system. They represent 1.5–2.5% of all intracranial tumors, where they are most often found is the posterior fossa (7–12%).[1,3,6] Spinal cord hemangioblastomas constitute 2–15% of all spinal cord tumors and are most often encountered in the thoracic followed by the cervical regions.[2,4] Hemangioblastomas occurs as a sporadic tumor or as a component of the hereditary von Hippel-Lindau disease. In case of absence of family history, patient must have two or more hemangioblastomas, to meet the diagnostic criteria.[5] Here, we report a patient with two hemangioblastomas, respectively, located in the cerebellum and spinal cord in a patient ultimately diagnosed with von Hippel–Lindau disease.

CASE REPORT

Clinical presentation

A 36-year-old female presented with a 4-year history of progressive right upper extremity distal weakness and cervical pain. On examination, she exhibited hypoesthesia in both upper

extremities accompanied by a hemispheric cerebellar syndrome and hyperreflexia. A family history of von Hippel–Lindau disease was ruled out.

Magnetic resonance imaging (MRI)

The MRI demonstrated a cerebellar cyst with a mural nodule [Figure 1], accompanied by a homogeneously, contrast enhancing intradural/intramedullary tumor at the C6–C7 level with perilesional edema and syrinx formation [Figure 2].

Surgery

The patient underwent tumor resection through a median suboccipital craniectomy for the cerebellar lesion [Figure 3] and C5–C7 laminectomy with a posterolateral sulcus approach to resect the C6–C7 spinal cord tumor [Figures 4 and 5]. The surgery was completed without complications. The histopathological examination of the specimen confirmed the diagnosis of a hemangioblastoma. The patient was extubated the next day. Postoperatively, the patient recovered strength from the right upper extremity, and the cervical pain disappeared. In the immediate postoperative, the patient developed a neurogenic bladder which resolved 1 month following the surgery. The postoperative brain MRI revealed that the tumor had been removed with no obvious residual tumor, while the cervical MR findings indicated only a mild inflammatory process and no obvious residual tumor [Figure 6].

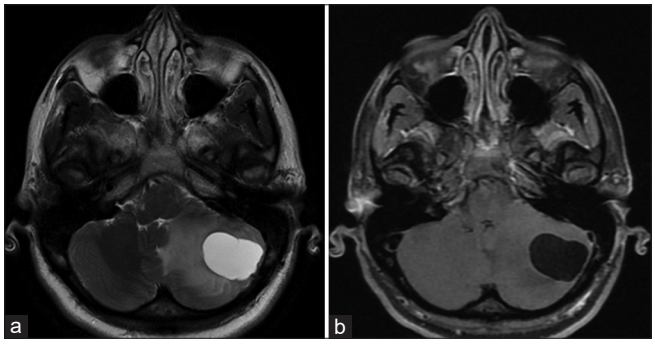

Figure 1: MRI of the cerebellum. (a) T2-weighted MR images revealed a cyst lesion with edema. (b) T1-weighted Gd-enhanced axial image revealing an hypointense mass at the cerebellar left hemisphere.

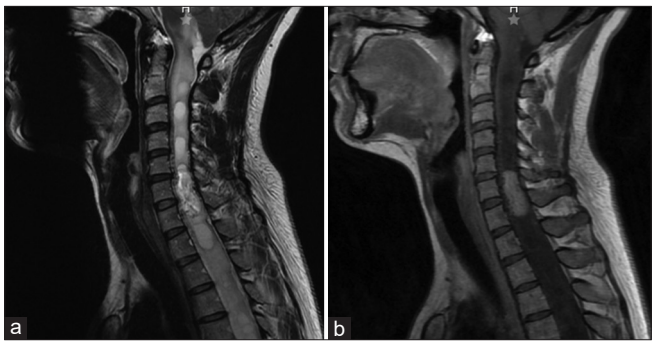

Figure 2: MRI of the cervical spine. (a) T2-weighted MR images revealed a syrinx with edema. (b) T1-weighted Gd-enhanced sagittal image revealing an intramedullary intradural isointense mass (arrow) at the C6-C7 level.

DISCUSSION

Surgical resection offers definitive therapy for sporadic, isolated hemangioblastomas, particularly those arising in the cerebellum. The role of surgery in patients with von Hippel–Lindau disease is less well-defined due to the frequent occurrence of multiple lesions. The optimal timing of surgery for patients these patients is uncertain. Most neurosurgeons do not operate these patients until they demonstrate progressive neurological deficits, tumor or cyst growth, or renewed hemorrhage. The preoperative neurologic deficit noted in this patient warranted surgical intervention that resulted in symptomatic improvement.

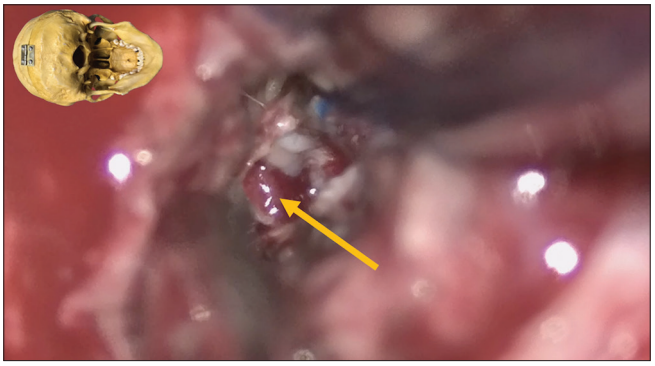

Figure 3: Median suboccipital craniectomy, transcortical approach showing mural nodule (yellow arrow).

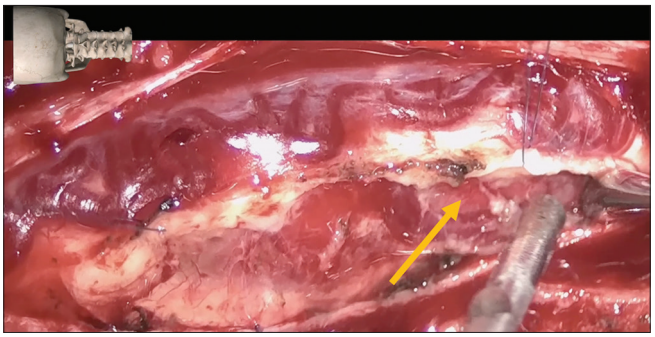

Figure 4: Posterolateral sulcus approach for the cervical spinal cord intramedullary tumor. Tumor (yellow arrow).

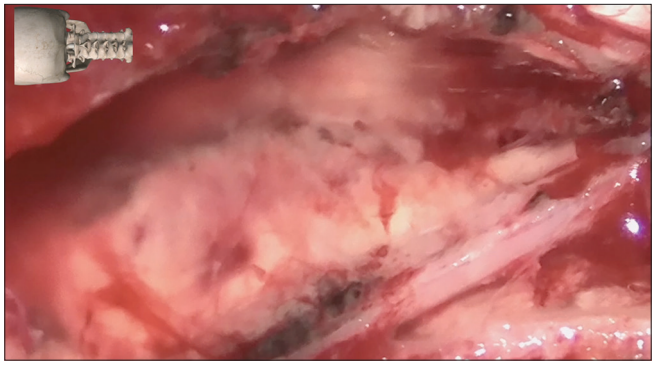

Figure 5: Macroscopic complete resection of the spinal cord lesion.

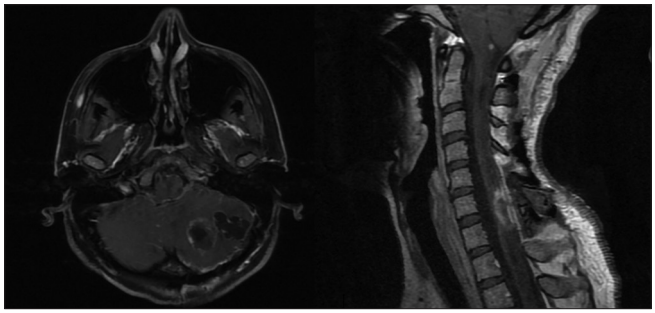

Figure 6: Postoperative MRI.

CONCLUSION

We reported a case of symptomatic multiple hemangioblastomas, in cerebellum and spinal cord. Complete resection of hemangioblastomas is very important to reduce the risks of lesion recurrence. This patient underwent complete resection of two symptomatic hemangioblastomas involving the cerebellum and spinal cord. In this case, the diagnosis of von Hippel–Lindau disease was confirmed by the presence of two or more central nervous system hemangioblastomas even without a positive genetic test.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Tirado-Ornelas HA, Olivares-Peña JL, Olivares-Camacho JL, Santos-Franco JA, Ochoa-González MV. Intramedullary cervical spinal cord and cerebellar hemangioblastoma: A case report. Surg Neurol Int 2022;13:294.