

## Case Report

# A case report of surgical management of hemangiopericytoma at the foramen magnum

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## Abstract

**Background:** Hemangiopericytoma (HPC) is a highly vascularized mesenchymal tumor known for its high rates of recurrence and metastasis. The extent of tumor removal is known to be the most trustful prognostic factor. Skull base HPCs are challenging to treat because of the difficulty of the surgical approach and proximity to vital vascular and neuronal structures. We successfully treated a case of HPC at the ventral foramen magnum through surgical gross tumor removal via a far-lateral transcondylar approach.

**Case Description:** A 38-year-old male complained of neck pain and bilateral paresthesia of his shoulders for 2 months, for which he was referred to our hospital. A magnetic resonance image (MRI) showed a 20 mm diameter mass at the ventral foramen magnum, which compressed his medulla oblongata. The tumor was gross totally removed via a far-lateral transcondylar approach. During the surgery, marked bleeding disturbed the surgical field until the main feeding artery from the direction of the dura mater was coagulated and cut. A relatively wide surgical field and a transcondylar approach were helpful to control the bleeding. The pathological examination revealed the tumor to be a HPC. After an uneventful recovery period of 9 days, the patient was discharged without neurological sequelae.

**Conclusion:** We successfully and completely removed an HPC near the foramen magnum, employing a wide surgical field and a transcondylar approach to help control bleeding. When the tumor is suspected preoperatively to be a hemangiopericytoma or vascular-rich tumor, a surgical approach that can secure a wide surgical field should be selected.

**Key Words:** Far-lateral approach, foramen magnum, hemangiopericytoma, transcondylar approach

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## INTRODUCTION

Hemangiopericytomas (HPCs) are highly vascularized mesenchymal tumors that derive from the pericytes forming the walls of capillaries and postcapillary venules. They are relatively rare intracranial tumors with a reported incidence of 0.4% of all primary central nervous system (CNS) tumors.<sup>[7,12]</sup> HPCs are well known

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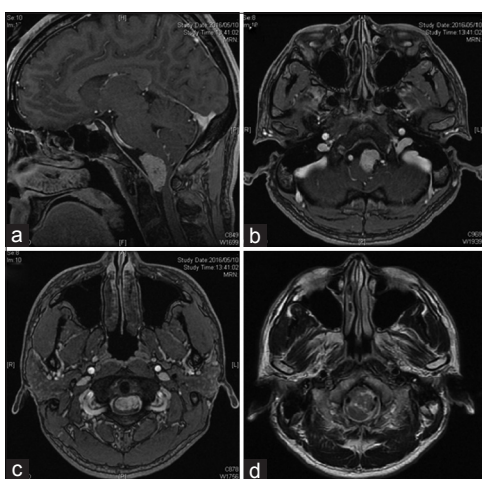
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for being both invasive and highly vascularized. They recur frequently and often metastasize, even outside the CNS, which significantly shortens a patient's overall survival time. The tremendous vascularity makes the radical resection of the tumor difficult. When gross total removal is achieved, a better prognosis can be anticipated.<sup>[8,14]</sup> However, when found in the foramen magnum, which is surrounded by many vital neuronal and vascular structures, removal is more difficult.<sup>[11,18]</sup> For such tumors, several approaches including the lateral suboccipital approach, transoral approach, transcervical, and far-lateral approach have been described previously. Of the useful approaches, some are less invasive, and some, especially the far-lateral approach, can offer a wider surgical corridor.<sup>[5]</sup> In the present report, we report on such a case that we treated successfully through surgery.

## CASE REPORT

A 38-year-old man was referred to our institution with chief complaints of bilateral sensory disturbance of the hands, intermittent headache, and nausea that persisted for 2 months. He had no apparent past medical history nor familial medical history.

On admission, neurological examination revealed sensory disturbance of the bilateral upper extremities. T1-weighted magnetic resonance imaging (MRI) with gadolinium enhancement revealed a well-demarcated, solid mass at the ventral foramen magnum that was compressing the patient's medulla oblongata [Figure 1a-c]. The tumor size was 12 × 13 × 22 mm and extended from the height of the C2 cervical vertebra to that of the medulla oblongata.

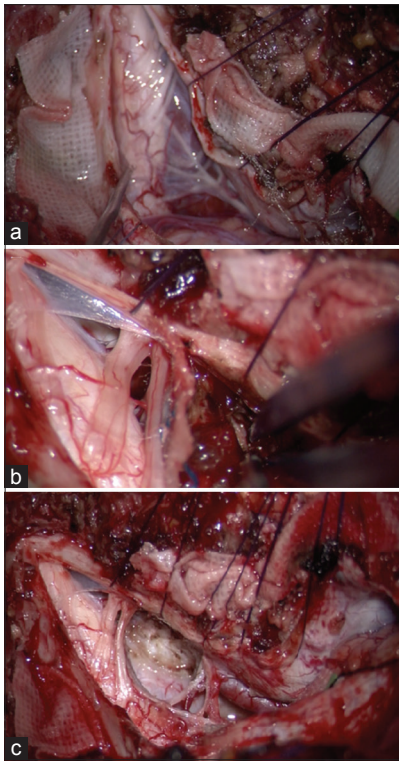


**Figure 1:** T1-weighted, gadolinium-enhanced MR images; a sagittal image shows a homogeneously enhanced mass at the ventral foramen magnum, whose size is around 20 mm in diameter (a). On axial image, the tumor seemed to be attached to dura mater around C2. The tumor severely compressed the ventral medulla oblongata. (b and c) The axial, T2-weighted MR image showed no medullary edema (d)

A T2-weighted MRI showed no apparent edema inside the medulla [Figure 1d]. Computed tomography angiography (CTA) revealed no obvious feeding arteries; however, the tumor was strongly enhanced. At first, cerebral angiography was considered as it is possible to embolize the feeders if detected. However, we thought surgical resection of feeders with appropriately wide surgical corridor can control the hemorrhage. It can result in almost the same outcome compared to TAE procedure, which carries some risks. Moreover, the patient was reluctant to undergo angiography. Consequently, we did not perform preoperative angiography. A preoperative diagnosis of meningioma was made, and the patient was recommended surgical removal of the tumor. Because the CTA image suggested high vascularity of the tumor, we determined that a far-lateral approach to secure wide surgical corridor was needed for safely managing the hemorrhage, as described above. The patient underwent surgical resection of the tumor under motor evoked potential (MEP) and sensory evoked potential (SEP) monitoring, utilizing a transcondylar approach in conjunction with a lateral suboccipital craniotomy. With this approach, the dorsomedial part of the left occipital condyle and the laminae of C1 and part of C2 were drilled. After cutting the dura, the reddish tumor was found ventral to the C1/C2 root and the dentate ligament, without retraction of the cervical spine. The dentate ligament was cut, and the tumor was detached from the surrounding neurovascular structures. After detailed inspection, the tumor was found to be attached to the dorsal dura at the odontoid process. The tumor bled easily and did not stop bleeding until multiple feeding arteries from the direction of the dura were coagulated and cut. After cutting the feeding arteries, the tumor became pale, and was easily removed with no further hemorrhage. The tumor was gross totally removed (Simpson's grade 2) [Figure 2].

Histologically, the tumor cells had high, dense nuclear, and branching vessels inside the tumors producing a staghorn appearance [Figure 3a and b]. Immunohistochemical labeling was positive for cluster of differentiation (CD) 34, CD 31, and vimentin; it was negative for epithelial membrane antigen (EMA) and S-100. The Ki-67 proliferation labelling index was less than 3% [Figure 3c and d]. The above findings were compatible with HPC.

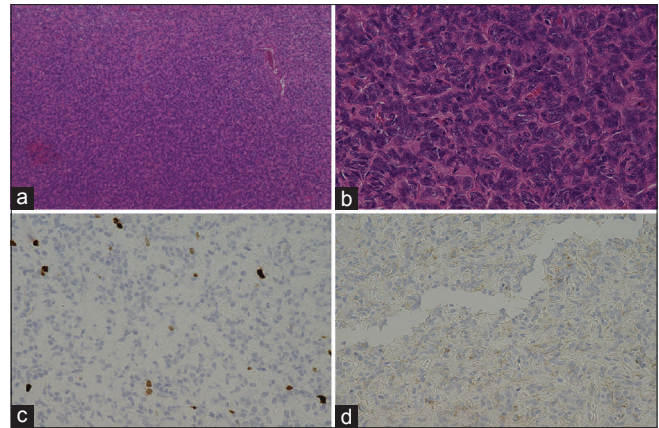
After surgery, no additional neurological sequelae appeared, and his initial symptoms of bilateral sensory disturbance of the hands completely disappeared. He was discharged on the 9<sup>th</sup> day after surgery. Follow-up MRIs were conducted periodically, and no tumor recurrence was detected at the 10-month follow-up. Radiation therapy has not been offered to the patient till now. We saved the choice of treatment in case of tumor recurrence. Postoperative MRI showed no residual or occurrence of the tumor [Figure 4].



**Figure 2:** After opening the dura, the tumor, covered by the arachnoid, was identified. The tumor compressed the medulla (a). It had no severe adhesion to the brain stem or any cranial nerves. When approaching the tumor to detach it from the dura, massive hemorrhaging occurred that was difficult to control (b). After cutting the main feeder from the direction of the dura, the tumor became white and easy to manage. The whole dural attachment of the tumor was detached. Plate c shows the final view, after gross total removal of the tumor was achieved (Simpson Grade 2)

## DISCUSSION

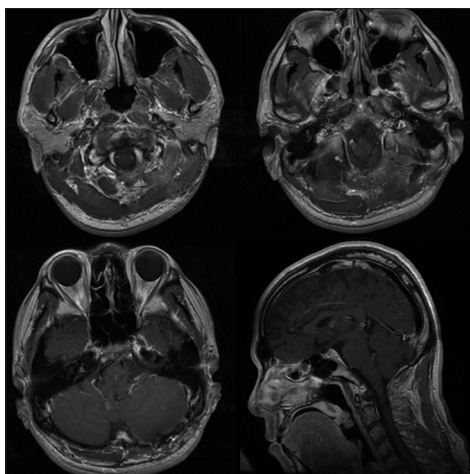
Tumors at the foramen magnum are challenging to treat because of their proximity to important vascular and nervous structures. Many critical neurovascular structures in this area are sensitive to injury.<sup>[11,18]</sup> Furthermore, surgical access to tumors at the foramen magnum, especially tumors attached to the anterior rim, is difficult to manage.<sup>[9]</sup> Thus far, several distinctive surgical approaches for the region have been investigated and discussed in the literature.<sup>[1,5,9-11,16]</sup> The transoral or transcervical approaches are widely recognized for the resection of extradural or ventral tumors in the foramen magnum,<sup>[10,16]</sup> however, some reports indicate that there are many more complications, such as pharyngeal fistula and cerebrospinal fluid (CSF) leakage, than there are benefits.<sup>[4,15]</sup> The posterior suboccipital midline approach is an easy and straightforward method and VAs and cranial nerves are easily manageable. Regarding meningiomas at the foramen magnum, Samii *et al.* reported that in most cases this approach is enough to achieve adequate tumor exposure.<sup>[13]</sup> However, especially in ventral or ventrolateral lesions, surgery would require unavoidable brainstem



**Figure 3:** (a) Hematoxylin-eosin staining showing hypercellular tumor with staghorn appearance (arrow) (original magnification  $\times 10$ ). (b) The arrow showed mitosis. (original magnification  $\times 40$ ). (c) Ki-67 expressed in the HPC, nucleus positive, positive cells were less than 3% (original magnification  $\times 40$ ). (d) The tumor was negative for EMA staining (original magnification  $\times 40$ )

retraction, as well as higher rates of subtotal resection and postoperative morbidity due to inadequate exposure.<sup>[2,6]</sup> Moreover, HPCs are highly vascularized, which makes complete removal difficult. A previously reported foramen magnum HPC during a pregnancy was resected with a suboccipital approach, which resulted in only partial resection to avoid surgical complications.<sup>[3]</sup> Hypervascular tumors such as HPCs require a strategy of first managing the feeders or dural attachment. From this viewpoint, the far-lateral approaches have a significant advantage. Far-lateral approaches were invented and developed by Bertalanffy,<sup>[1]</sup> and can provide lateral exposure of the brainstem with minimal retraction; however, it is more time-consuming and includes higher risks of CSF leakage and craniocervical instability. Suhardja *et al.* showed that the far-lateral approach afforded a significantly larger surgical field.<sup>[17]</sup> The entire longitudinal and lateral portions of the tumor, as well as the dural attachment and feeders, can be visualized, which makes it considerably easier to seek the attachment and achieve radical dissection of the tumor. Previously, our group reported that the far-lateral transcondylar approach is especially beneficial in managing foramen magnum meningiomas located on the anterior rim of the foramen magnum.<sup>[5]</sup>

Recently, less invasive surgery with narrow surgical view such as keyhole surgery or endoscopic surgery has been admired all around the world. In contrast, the hemorrhage in this case could not have been controlled with such a narrow corridor. Moreover, we could not have achieved gross total removal due to its poor view. This case teaches us a lesson that sufficiently wide corridor would be safe and reliable if preoperative neuroimaging showed high vascularity of tumors assumed to be challenging to remove. It is natural that to curtail the superfluous invasiveness and narrow the range of bone



**Figure 4: No residual tumor in the images**

drilling as much as possible is very important. However, we think this inclination should not always be applied to all cases. From our experience, we aimed to create a stir in this trend and share this experience to cope with tumors case by case flexibly.

In the present case, there was massive hemorrhage during the operation while cutting the tumor. Because of the bleeding, the tumor itself and the surrounding anatomy was not fully visible. Thus, performing fine manipulation was nearly impossible until the bleeding was managed by coagulating the feeding artery. In this case, the far-lateral approach provided a large enough surgical field to find the attachment easily. After managing this, we controlled the hemorrhage and attained gross total removal.

## CONCLUSION

We experienced a challenging case of HPC at the ventral foramen magnum, which was gross totally removed via a far-lateral transcondylar approach. This approach provided a significantly large surgical corridor compared to the lateral suboccipital approach, and thus allowed us to manage the tumor attachment with minimal retraction of the brainstem and relatively easy control of the bleeding, which is necessary for cases of highly vascularized tumors.

## Informed consent

The patient has consented to submission of this case report to the journal.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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