

A Large Solitary Hemangioblastoma of the Lateral Ventricles: A Case Report and Literature Review

Alireza Tabibkhouei, MD;¹
Arash Fattahi, MD;¹
Ali Rostami, MD

Department of Neurosurgery, School of Medicine, Iran University of Medical Sciences, Tehran, Iran

Correspondence:

Arash, Fattahi, MD;
2nd Floor, No. 15, Bahar St., Marzadaran St., P.O. Box: 14618-86461, Tehran, Iran
Tel: +98 21 66503890
Fax: +98 21 66509120
Email: fatahi.a@iums.ac.ir
Received: 05 February 2019
Revised: 13 May 2019
Accepted: 09 June 2019

What's Known

- Supratentorial hemangioblastoma (HB) is rare and only a few cases are reported on the intraventricular location of a tumor. HB of the lateral ventricle is even rarer.
- Perioperative complications are associated with the surgical treatment of a large and highly vascular intraventricular tumor.

What's New

- Since 1935, the present study describes the fourth reported case and the largest solitary intraventricular HB in the literature.
- Because of intrasessional hemorrhage, the tumor mass showed a heterogeneous enhancement, which is unusual for HB and poses a preoperative diagnostic challenge.

Abstract

Hemangioblastoma (HB) in the supratentorial region of the brain is rare and only a few cases are reported on intraventricular HB. HB of the lateral ventricles is even rarer. We present a case of a 30-year-old man with generalized tonic clonic seizures. The brain computed tomography showed a 5.5 cm heterogeneous mass extending into both lateral ventricles with partial enhancement. Based on the size and imaging features, we present the fourth documented case of a large solitary intraventricular HB. Our approach to this unique case and some treatment complexities are also described.

Considering the rarity of the case and the patient's imaging features, the present study provides a better understanding of HB and recommends HB to be considered in the differential diagnosis of masses in the lateral ventricles. In addition, some preventable pitfalls in the treatment of such complex cases are described.

Please cite this article as: Tabibkhouei AR, Fattahi A, Rostami A. A Large Solitary Hemangioblastoma of the Lateral Ventricles: A Case Report and Literature Review. *Iran J Med Sci.* 2020;45(1):67-72. doi: 10.30476/ijms.2019.81095.

Keywords • Hemangioblastoma • Solitary • Lateral ventricle • Magnetic resonance imaging • Hydrocephalus • Seizure

Introduction

Hemangioblastoma (HB) is a benign vascular tumor of the central nervous system (CNS) and responsible for 2% of all intracranial tumors.¹ This tumor is associated with von Hippel-Lindau disease (VHL) in one-third of cases and the remaining are sporadic cases. The infratentorial region of the brain is the most common site of these tumors.² Supratentorial HB is rare and there are only a few reports on the intraventricular location of a tumor, especially in the lateral ventricles.^{3, 4} To the best of our knowledge, there are only three reported cases of solitary intraventricular HB of the lateral ventricles. Based on the size and imaging features, we present the fourth and the largest reported case with partial central enhancement. Furthermore, our approach to this case and some treatment complications are also described.

Case Presentation

A 30-year-old man was presented to our medical center with a 12-month history of progressive blurred vision and headache. He had a generalized tonic-clonic seizure four days prior and was referred to us after an initial diagnostic workup. On examination, he had bilateral papilledema and positive bilateral Hoffmann's

reflex. We did not observe any neurologic deficit and his medical history was not significant. The brain computed tomography (CT) showed a 5.5 cm heterogeneous mass located in the midline position extending into both lateral ventricles with a small high-density area; most probably due to small intralesional bleeding (figure 1a). The magnetic resonance imaging (MRI) revealed a large heterogeneous mass extended to both lateral ventricles from the splenium of corpus callosum toward the foramen of Monro. The mass was hypo-isointense on T1-weighted and hyper-intense on T2-weighted images. Its enhancement was greater in the central portion of the mass with weak and irregular patterns (figures 1b and 1c).

Based on imaging features, our primary preoperative differential diagnosis was an intraventricular ependymoma. Other possible preoperative diagnoses were glioma and central neurocytoma. We decided to perform a right-sided trans-parietal trans-ventricular approach for tumor resection via the right parieto-occipital craniotomy. In the operating room, the patient was placed in a semi-lateral position. During surgery, when we reached the tumor bulk, a large

mass with the signs of previous bleeding was observed. Intraoperatively, the tumor was found to be highly vascular and hemorrhagic. Finally, after removing all visible parts of the tumor and meticulous intraventricular hemostasis, a ventricular drain was placed into the right lateral ventricle. The postoperative brain CT scan showed a small remnant of the tumor in the left lateral ventricle (figure 1d). Histopathological report of the mass indicated HB, which was not among our preoperative diagnoses (figure 2). In search of other signs for the possible presence of VHL disease, we performed abdominal, renal, and ophthalmological examinations, as well as laboratory tests (e.g. 24-hour urine vanillylmandelic acid); the results were negative.

After the surgery, the patient was in good health without neurological deficits and did not show any of the signs and symptoms reported preoperatively. Four months later, the postoperative MRI revealed a near-total resection of the tumor with a small remnant in the left lateral ventricle. It also showed a right-sided fronto-parietal chronic subdural hematoma (SDH) due to the reduction of the mono-ventricular hydrocephalus of the right

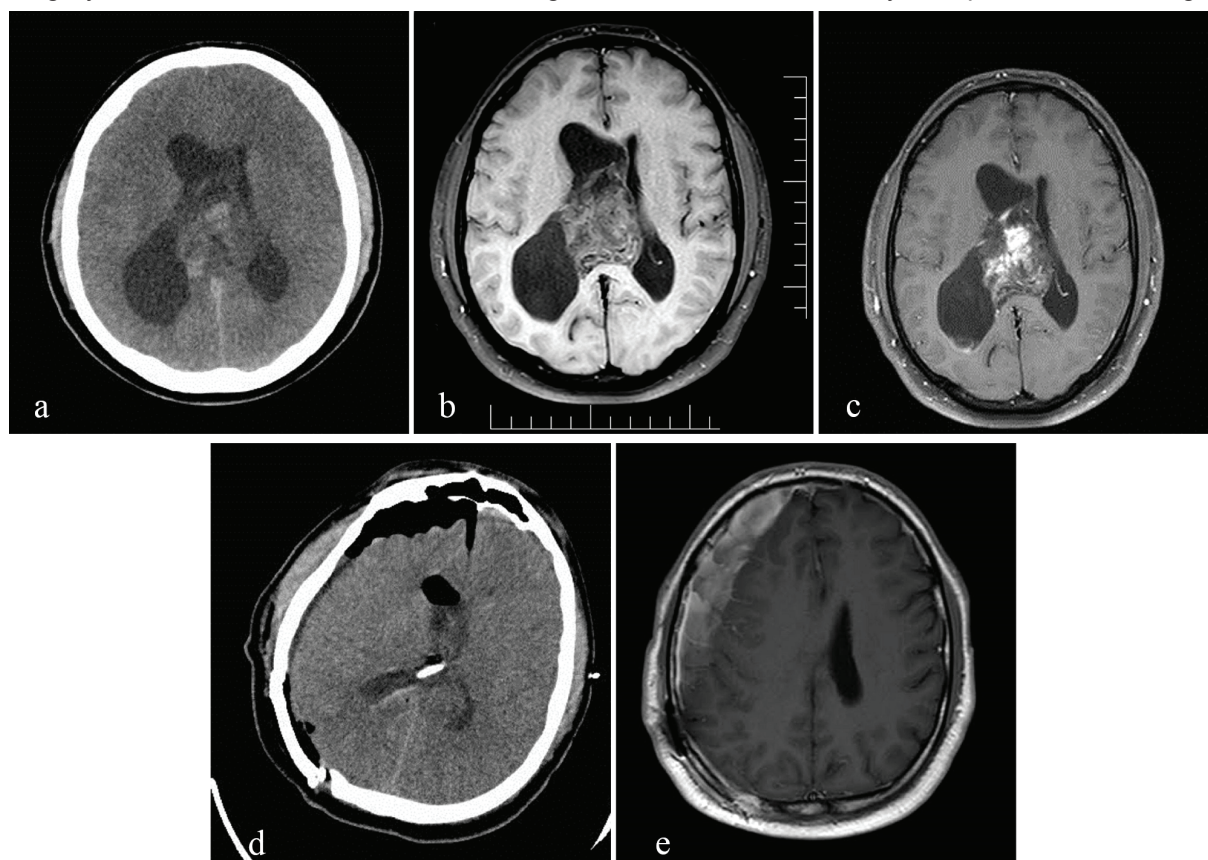


Figure 1: Preoperative and postoperative images of a 30-year-old man with intraventricular hemangioblastoma. A 5.5 cm heterogeneous mass located in midline position extending into both lateral ventricles (a). An irregular enhancement, greater in the central portion of the mass, on axial without (b) and axial with gadolinium injection (c) views. The postoperative brain computed tomography scan revealed a small remnant of the tumor in the left lateral ventricle (d). The right-sided fronto-parietal chronic epidural hematoma on the last magnetic resonance imaging (axial view) was drained via one burr hole (e).

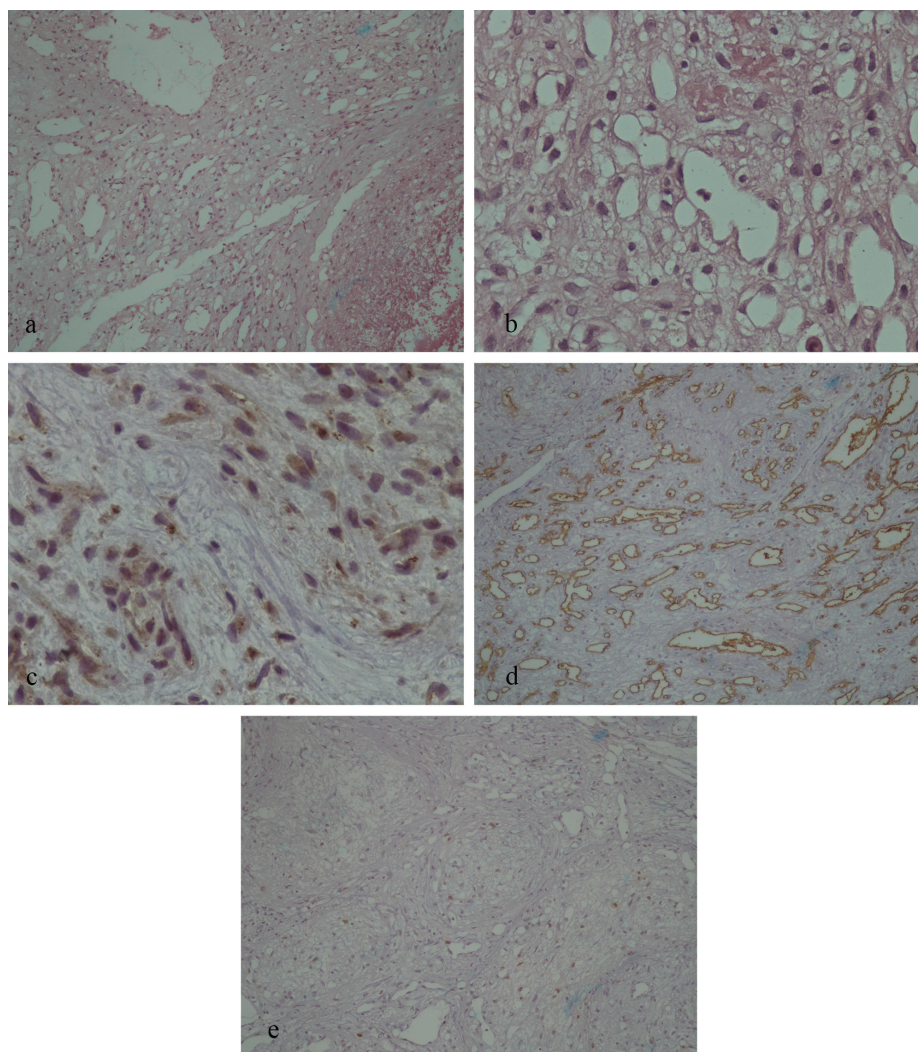


Figure 2: Histopathologic features of intraventricular hemangioblastoma (HB) in the patient. These revealed a highly vascular mass with a predominant capillary background in concomitant with an expansion into the venous caverns due to previous recurrent of asymptomatic intralesional bleedings (a: H&E $\times 40$; 2b: H&E $\times 200$). Immunohistochemistry study of samples revealed positive reactivity for neuron-specific enolase (c). CD34 highlighted vascular channels (d). Ki67 was positive in 2-3% of tumoral cells (e). Histopathologic appearances and the results of immunohistochemical staining matched with HB diagnosis.

lateral ventricle (figure 1e). Subsequently, the SDH was drained via burr hole and we decided to monitor the remnant of the tumor through brain MRI. Two years later, a follow-up showed a small increase in the size of tumor remnants. However, considering the satisfactory neurological examination, we decided on patient follow-up only. Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Discussion

HBs are highly vascular tumors of the CNS with benign behavior and two-thirds of the cases are of the sporadic type. The other one-third of the cases are associated with VHL disease, an inherited disease with multi-organ involvement (e.g., pheochromocytoma, endolymphatic sac

tumors, renal and pancreatic tumors and cysts, ependymal cystadenoma, retinal HB, broad ligament cystadenoma).⁵ HB is more common in the infratentorial area, mostly in the cerebellum, though the supratentorial region of the brain is a rare site for HB; accounting for 4% of the sporadic and 13% of the VHL-related types.⁶

Supratentorial HB is more prevalent in men.⁷ In the supratentorial region of the brain, HB of the lateral ventricles is very rare.⁸ As presented in table 1, there were only a few reports on lateral ventricular HB. Only the case report by Diehl and Symon was similar to our case in terms of the tumor size, but was involved the temporal horn of the right lateral ventricle.¹⁰ However, because of the midline position, our patient was bi-ventricular in both lateral ventricles resulting in mono-ventricular hydrocephalus due to the foramen of Monro obstruction. Both cases

Table 1: A brief review of literature presenting lateral ventricle HB

Author	Sex, age*	Signs and symptoms	VHL-related	Site	Preoperative imaging features	Diameter (cm)
Vecchi et al. (1935) ⁹	Female, 80	Incidental postmortem	N/A [†]	N/A	N/A	N/A
Diehl et al. (1981) ¹⁰	Male, 20	Progressive visual loss, headache, bilateral papilledema	Yes	Right temporal horn	CT scan, homogenous	5
Murakami et al. (1985) ¹¹	Female, 31	Headache, gait disturbance, bilateral papilledema	Positive Family history	Trigone of right lateral ventricle	CT scan, homogenous	N/A
Ho et al. (1990) ¹²	Female, 44	Headache	Positive Family history	Trigone of right lateral ventricle	MRI, homogenous	3
Lyo et al. (1992) ¹³	Male, 36	Headache, diplopia, right abducens nerve palsy, confusion	No	Left lateral ventricle wall	N/A	N/A
Prieto et al. (2005) ⁶	Male, 37	Confusion, cognitive deficit, dysphasia	Yes	Trigone of left lateral ventricle	MRI, homogenous	3
Jaggi et al. (2009) ¹⁴	Male, 30	Headache	No	Left lateral ventricle	MRI, homogenous	3
Takeuchi et al. (2011) ¹⁵	Male, 33	Headache	Yes	Right lateral ventricle	MRI, solid-cystic homogenous	N/A
Al-Najar et al. (2013) ¹⁶	Male, 70	Headache, vomiting, dizziness, left facial palsy, left hemiparesis, intraventricular hemorrhage	No	Right occipital horn	MRI, heterogeneous	3.3
Anderson et al. (2014) ¹⁷	Male, 24	Vertigo, right arm weakness, transient right facial paresis and slurring of speech, headache	N/A	Midline and greater in left lateral ventricle	MRI, solid-cystic with diffuse enhancement	N/A
Present case (2017)	Male, 30	Seizure, blurred vision, headache, bilateral papilledema	No	Midline in both lateral ventricle	MRI, heterogeneous	5.5

*Years; [†]Not available; HB: Hemangioblastoma; VHL: Von Hippel-Lindau disease; CT: Computed tomography; MRI: Magnetic resonance imaging

described solitary HB, but our case was of the sporadic type, whereas the case reported by Diehl and Symon concerned the VHL-related type.

Imaging modalities can be used to diagnose any brain mass and facilitate accurate planning of surgeries or other adjuvant therapies. Considering the availability of diverse imaging modalities, accurate preoperative diagnosis of brain tumors is within reach; at least it can assist in reducing the number of differential diagnoses. The MRI of HB has a diffuse enhancement, which indicates high-flow vessels in the mass; termed as flow void. Usually, HB is hypo-isointense on T1-weighted and hyper-intense on T2-weighted images. If the tumor is HB, it is unlikely to see calcification or peritumoral edema. In the MRI (with gadolinium-based contrast agent) of a patient with HB, homogeneous enhancement is the most prevalent characteristic of the imaging. However, as in our case and other studies,^{16, 18, 19} partial enhancement is a highly unusual finding. Heterogeneity of the mass may indicate previous asymptomatic intralesional bleeding resulting in the expansion into the venous caverns; as found

in our histopathological evaluation.

In patients with HB, angiographic study shows a highly vascular mass. If HB diagnosis is the objective, angiography should be part of the preoperative assessment or even pre-surgical treatment (e.g., embolization). Due to the unusual findings on the preoperative MRI images, we did not perform angiography and consequently, our patient had severe intraoperative bleeding. Therefore, it is recommended to perform an angiographic evaluation in the case of a huge intraventricular mass presented with hemorrhage and hydrocephalus.

In the post-surgical period, our patient developed a right-sided fronto-parietal chronic SDH complication, which required surgical treatment. We believe that the inevitable extensive decompression of the tumor and overtreatment resulting from ventricular drain insertion were the cause of this complication. In any ventricular procedure with preoperative hydrocephalus, placement of an external ventricular drain (EVD) followed by short-time tapering and subsequent EVD removal could be an option. The ventricular surgery by itself could

be a cause of postoperative SDH, however, we believe that EVD placement may increase the possibility of its occurrence.¹⁸ We recommend to place the EVD drainage bag at least at 15 cm height for slow and continuous drainage and then remove it as soon as possible. On a final note, choosing the right surgical trajectory to an intraventricular hemorrhagic mass is pivotal; narrower view channels through the interhemispheric transcallosal approach may be harmful due to the limited access to mass borders for controlling hemorrhage.

Conclusion

We present a patient with solitary HB in both lateral ventricles. The MRI only showed an irregularly enhanced imaging in the central part. The main complication in our treatment was the occurrence of a chronic SDH. Therefore, we recommend an accurate use of postoperative EVD to reduce such undesirable complications and to perform a preoperative angiography for the detection of large intraventricular hemorrhagic masses.

Conflict of Interest: None declared.

References

- 1 Ammerman JM, Lonser RR, Dambrosia J, Butman JA, Oldfield EH. Long-term natural history of hemangioblastomas in patients with von Hippel-Lindau disease: implications for treatment. *J Neurosurg.* 2006;105:248-55. doi: 10.3171/jns.2006.105.2.248. PubMed PMID: 17219830.
- 2 Richard S, Graff J, Lindau J, Resche F. Von Hippel-Lindau disease. *Lancet.* 2004;363:1231-4. doi: 10.1016/S0140-6736(04)15957-6. PubMed PMID: 15081659.
- 3 Conway JE, Chou D, Clatterbuck RE, Brem H, Long DM, Rigamonti D. Hemangioblastomas of the central nervous system in von Hippel-Lindau syndrome and sporadic disease. *Neurosurgery.* 2001;48:55-62. doi: 10.1097/00006123-200101000-00009. PubMed PMID: 11152361.
- 4 Mills SA, Oh MC, Rutkowski MJ, Sughrue ME, Barani IJ, Parsa AT. Supratentorial hemangioblastoma: clinical features, prognosis, and predictive value of location for von Hippel-Lindau disease. *Neuro Oncol.* 2012;14:1097-104. doi: 10.1093/neuonc/nos133. PubMed PMID: 22723428; PubMed Central PMCID: PMC3408260.
- 5 Matsumoto K, Kannuki S. [Hemangioblastoma and von Hippel-Lindau disease]. *Nihon Rinsho.* 1995;53:2672-7. PubMed PMID: 8538024.
- 6 Prieto R, Roda JM. Hemangioblastoma of the lateral ventricle: case report and review of the literature. *Neurocirugia (Astur).* 2005;16:58-62. doi: 10.1016/s1130-1473(05)70436-x. PubMed PMID: 15756413.
- 7 Ferrante L, Acqui M, Mastronardi L, Fortuna A. Supratentorial hemangioblastoma. Report of a case and review of the literature. *Zentralbl Neurochir.* 1988;49:151-61. PubMed PMID: 3071932.
- 8 Black ML, Tien RD, Hesselink JR. Third ventricular hemangioblastoma: MR appearance. *AJNR Am J Neuroradiol.* 1991;12:553. PubMed PMID: 2058513.
- 9 Vecchi B, Patrassi G. Angioreticuloma del plessi corioidei, con" aree di Gamna. *Schweiz Med Wochenschr.* 1935;65:242-6.
- 10 Diehl PR, Symon L. Supratentorial intraventricular hemangioblastoma: case report and review of literature. *Surg Neurol.* 1981;15:435-43. doi: 10.1016/s0090-3019(81)80033-x. PubMed PMID: 7197059.
- 11 Murakami H, Toya S, Otani M, Sato S, Ohira T, Takenaka N. A case of concomitant posterior fossa and supratentorial hemangioblastomas. *No Shinkei Geka.* 1985;13:175-9. PubMed PMID: 4039417.
- 12 Ho YS, Plets C, Goffin J, Dom R. Hemangioblastoma of the lateral ventricle. *Surg Neurol.* 1990;33:407-12. doi: 10.1016/0090-3019(90)90154-h. PubMed PMID: 2190345.
- 13 Lyo IU, Kim CJ, Kwon Y, Rhim SC, Lee JK, Kwun BD, et al. A Case of Hemangioblastoma in the Lateral Ventricle. *J Korean Neurosurg Soc.* 1992;21:195-200.
- 14 Jaggi RS, Premeasagar IC, Abhishek. Hemangioblastoma of the lateral ventricle. *Neurol India.* 2009;57:677-9. doi: 10.4103/0028-3886.57797. PubMed PMID: 19934579.
- 15 Takeuchi S, Takasato Y. Supratentorial intraventricular hemangioblastomas. *Acta Neurol Belg.* 2011;111:353-6. PubMed PMID: 22368981.
- 16 Al-Najar M, Al-Hadidy A, Saleh A, Al-Tamimi A, Al-Darawish A, Obeidat F. Sporadic Lateral Ventricular Hemangioblastoma presenting with Intraventricular and Subarachnoid Haemorrhage. *Sultan Qaboos Univ Med J.* 2013;13:597-600. doi: 10.12816/0003324. PubMed PMID: 24273675; PubMed Central PMCID: PMC3836654.
- 17 Anderson I, Kumar R, Patankar T, Tyagi A. Lateral ventricle haemangioblastoma: the role of perfusion scanning and embolisation in diagnosis and management. *BMJ Case Rep.* 2014;2014. doi: 10.1136/bcr-2014-205736.

- PubMed PMID: 25257888; PubMed Central PMCID: PMC4180557.
- 18 John JK, Robin AM, Pabaney AH, Rammo RA, Schultz LR, Sadry NS, et al. Complications of ventricular entry during craniotomy for brain tumor resection. *J Neurosurg.* 2017;127:426-32. doi: 10.3171/2016.7.JNS16340. PubMed PMID: 27813467.
- 19 Iplikcioglu AC, Yaradanakul V, Trakya U. Supratentorial haemangioblastoma: appearances on MR imaging. *Br J Neurosurg.* 1997;11:576-8. doi: 10.1080/02688699745772. PubMed PMID: 11013633.