A Solitary Hemangioblastoma in the Lateral Ventricle: Case Report and Review of the Literature

Wenjing Yu

The First Clinical Medical College of Zhejiang Chinese Medical University

Yifan Guo

The First Affiliated Hospital of Zhejiang Chinese Medical University

Zhijian Cao (✉ 20001010@zcmu.edu.cn)

The First Affiliated Hospital of Zhejiang Chinese Medical University

Case Report

Keywords: Hemangioblastoma, Lateral ventricle, Magnetic resonance imaging, Solitary

Posted Date: March 1st, 2022

DOI: <https://doi.org/10.21203/rs.3.rs-1374907/v2>

License: © ⓘ This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

Abstract

Background: Hemangioblastoma is a benign tumor occurring in the central nervous system. As known, the most common sites are the cerebellum and spinal cord with the typical appearance which is a nodule with a cyst and contrast-enhanced scan demonstrates the obvious enhancement of the nodule and the local cyst wall. However, it's difficult to diagnose the hemangioblastoma when it occurs in the lateral ventricle, due to the rare incidence of hemangioblastoma in the lateral ventricle.

Case presentation: A 28-year-old man with a 1-month history of recurrent headache accompanied by slight dizziness, then the computed tomography (CT) and magnetic resonance imaging (MRI) of brain showed the cystic solid tumor with obvious enhancement in the parenchymal part and local cystic wall, also, there was abnormal dilated blood vessel in the parenchymal part which was considered as the hemangioblastoma in the lateral ventricle.

Conclusions: Total resection of the tumor was performed under neuronavigation, then the tumor was pathologically confirmed to be hemangioblastoma which was the Grade WHOI. Based on this study, we have increased the understanding of hemangioblastoma in the differential diagnosis of tumors located in the lateral ventricle and provide a new method of resection of the tumor.

Background:

Hemangioblastoma is a benign tumor, accounting for 2% of intracranial tumors.¹ Radiographically, the typical appearance is a nodule with a cyst, Contrast-enhanced scan demonstrates the obvious enhancement of the nodule and the local cyst wall.² About two-thirds of the hemangioblastoma happened alone, and the remaining one-third was associated with Hippel - Lindau syndrome (VHL).³ As known, the most common sites are the cerebellum and spinal cord.⁴ In this paper, we report a case of hemangioblastoma in the lateral ventricle.

Case Presentation:

A 28-year-old man was presented to our medical center with a 1-month history of recurrent headache accompanied by slight dizziness, other than this, there were no nausea, vomiting, memory loss or other discomfort. His parents were healthy and had no similar family history or medical history. We gave him the CT and MRI of the brain, then, we found a mass in the left lateral ventricle and we considered the mass was benign tumor with rich blood supply. After the whole lesion was resected under neuronavigation, the tumor was pathologically confirmed to be hemangioblastoma which was the Grade WHOI.

Plain CT of the brain showed a heterogeneous mass located in the left lateral ventricle with cystic components, causing the midline structure to shift to the right. The total size of the tumor was about

35mm*32 mm, Contrast-enhanced CT revealed heterogeneous enhancement of the solid components and thickened vascular shadow was seen in the solid components (Fig. 1).

MRI revealed a large heterogeneous mass with slight edema in the left lateral ventricle. Cyst on T1WI presented low signal and nodule with equal signal; Cyst on T2WI presented high signal and nodule with slightly high signal and strip-like low signal shadow, for us, the strip-like low signal shadow was considered as the flow-empty actions. In addition to that, nodule showed hypo-isointense on DWI and slightly high signal on ADC. The enhancements of the solid nodule and cyst wall were significant (Fig. 2).

Then, the whole lesion was resected under neuronavigation. The intraoperative findings were as follows: the tumor was located in the left lateral ventricle, the supplying arteries were dural vessels. The tumor was round and slightly tough in texture. Microscopically, capillaries and mesenchymal cells of different maturation stages were observed after postoperative histopathological staining, among them, some of the cells showed mild atypia, these signs suggested the tumor was hemangioblastoma (WHO grade I) (Fig. 3).

one month after surgery, the patient recovered well without any discomfort such as dizziness and headache.

Discussion And Conclusions:

The age of onset of the hemangioblastoma is between 20 and 50 years old,⁵ it mainly occurs in the cerebellum, brainstem and spinal cord,⁶ the supratentorial area is relatively rare.⁷

To date, a little of hemangioblastoma in the lateral ventricle have been reported. The main clinical manifestations include progressive blurred vision, headache, cognitive impairment and generalized tonic-clonus in severe cases.^{3,8-9} As presented in **table1**,¹⁰⁻¹⁹ there were only a few reports on lateral ventricular HB. The case reports by Ho et al, Prieto et al, Jaggi et al and Al-Najar et al were similar to our case in terms of the tumor size, but only the case report by Jaggi et al was very similar to our case, which had no typical clinical symptoms and no VHL-relation. Among these hemangioblastomas of lateral ventricular, only ours had both a solid and cystic component.

Table 1
Comparison of previously reported cases of lateral ventricle haemangioblastoma.

Author/year	Sex/age	Clinical symptoms	Von Hippel-Lindau disease(VHL)-related	Solid/cystic	Diameter(cm)
Vecchi et al, 1935 ¹⁰	Female/80	Incidental finding	Not available	Not available	Not available
Diehl et al. 1981 ¹¹	Male/20	Progressive visual loss, headache and bilateral papilledema	Yes	Cystic	5
Murakami et al. 1985 ¹²	Female/31	Headache and gait disturbance	Family history	Cystic	Not available
Ho et al. 1990 ¹³	Female/44	Depressive and headache	Family history	Not available	3
Prieto et al. 2005 ¹⁴	Male/37	Confusion, cognitive deficit, dysphasia	Yes	Solid	3
Jaggi et al. 2009 ¹⁵	Male/30	Headache	No	Not available	3
Takeuchi et al. 2011 ¹⁶	Male/33	Headache	Yes	Cystic	Not available
Al-Najar et al. 2013 ¹⁷	Male/70	Headache, vomiting, dizziness, left facial palsy, left hemiparesis	No	Solid	3.3
Anderson et al. 2014 ¹⁸	Male/24	Vertigo, right arm weakness, headache	Not available	Solid	Not available
John et al. 2017 ¹⁹	Male/30	Seizure, blurred vision, headache and bilateral papilledema	No	Not available	5.5
Present case (2021)	Male/28	recurrent headache, slight dizziness	No	Solid, Cystic	3.5

So, according to the imaging findings of this patient, the ependyma, central neurocytoma and choroidal papilloma were mainly considered in the differential diagnosis of tumors in the lateral ventricle.²⁰⁻²²

To diagnose the intracranial hemangioblastoma, MRI is superior to CT.²³ On MRI, there are several types of hemangioblastoma²⁴⁻²⁵: (1) Large unenhanced cyst with enhanced small nodule; (2) Simple cyst without enhancement; (3) Simple cyst with enhanced wall; (4) The cyst with enhanced wall is accompanied by enhanced small nodule. In this case, the tumor was consistent with the last type. However, it is very rare for hemangioblastoma to occur in the lateral ventricle, Therefore, differential diagnosis should be made with common tumors in the lateral ventricle before definite diagnosis, mainly including ependymoma, central neurocytoma and choroid plexus papilloma. Firstly, the age of onset of ependyma and central neuroblastoma overlays with the age of onset of hemangioblastoma,²⁶⁻²⁷ and the solid components of all the three tumors are heterogeneous enhancement,²⁸⁻²⁹ These have brought some difficulties to the diagnosis. However, the dispersion of the solid components of the first two is limited on DWI and ADC: the DWI is mostly characterized by high signal and lower ADC value than that of hemangioblastoma,³⁰ which has become the key point of differential diagnosis. Moreover, central neurocytomas showed foam-like changes in cystic lesions on T2WI.³¹ Typical choroidal papilloma is relatively easy to distinguish from hemangioblastoma, but when choroidal plexus papilloma presents as rare cysts with nodules, it is difficult to distinguish, under the circumstances, the main distinguishing point is that choroidal plexus papilloma often presents hydrocephalus.³²

Due to the common tumor types in the lateral ventricle, The MRI and DWI should be combined for comprehensive consideration in the differential diagnosis of tumors in the lateral ventricle. Hemangioblastoma is a benign tumor, it presents hypo-isointense on DWI and has a high ADC value. Meanwhile, the tumor is rich in vascular components, and the strip-like low signal shadow on T2WI is helpful to the diagnosis, suggesting the possibility of vascular empty flow. The imaging findings of this case are roughly consistent with those of hemangioblastoma, which can be diagnosed as hemangioblastoma after differential diagnosis.

In conclusion, we presented a case of hemangioblastoma occurring in the lateral ventricle and made a differential diagnosis from common tumors in the lateral ventricle. Although hemangioblastoma occurring in the lateral ventricle is relatively rare, the possibility of hemangioblastoma should be considered in combination with the imaging findings.

Declarations:

Ethics approval and consent to participate

The studies involving human participants were reviewed and approved by the Ethics Committee of the First Affiliated Hospital of Zhejiang Chinese Medical University.

Consent for publication

The images and data involved in this case report have been informed consent of the patients.

Availability of data and materials

The datasets used during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

Funding

This work was supported by the Medicine and Healthcare Scientific Research General Program of Zhejiang Province of China(2021KY829) and The Scientific Research Project of Zhejiang Provincial Health and Family Planning Commission(2014KYA155).

Authors' contributions

Wenjing Yu and Yifan Guo took the lead in drafting the manuscript and provided magnetic resonance images. Zhijian Cao provided supervision and participated in the literature review and in drafting the manuscript. All authors read and approved the final manuscript.

Acknowledgements

Not applicable

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(2):248-255.
2. Wang Q, Zhang S, Cheng J, Liu W, Hui X. Radiologic Features and Surgical Strategy of Hemangioblastomas with Enhanced Cyst Wall. *World Neurosurg.* 2017;108:143-150.
3. Tabibkhooei A, 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.
4. de San Pedro JR, Rodríguez FA, Níguez BF, et al. Massive hemorrhage in hemangioblastomas Literature review. *Neurosurg Rev.* 2010;33(1):11-26.
5. Nguyen TH, Pham T, Strickland T, Brewer D, Belirgen M, Al-Rahawan MM. Von Hippel-Lindau with early onset of hemangioblastoma and multiple drop-metastases like spinal lesions: A case report. *Medicine (Baltimore).* 2018;97(39):e12477.
6. Kosty J, Staarman B, Zimmer LA, Zuccarello M. Infundibular Hemangioblastoma in a Patient with Neurofibromatosis Type 1: Case Report and Review of the Literature. *World Neurosurg.*

- 2016;88:693.e7-693.e12.
7. 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(8):1097-1104.
 8. Diehl PR, Symon L. Supratentorial intraventricular hemangioblastoma: case report and review of literature. *Surg Neurol.* 1981;15(6):435-443.
 9. Prieto R, Roda JM. Hemangioblastoma of the lateral ventricle: case report and review of the literature. *Neurocirugia (Astur).* 2005;16(1):58-62.
 10. Vecchi B, Patrassi G. Angioreticuloma del plessi corioidei, con" aree di Gamna. *Schweiz Med Wochenschr.* 1935;65:242–6
 11. Diehl PR, Symon L. Supratentorial intraventricular hemangioblastoma: case report and review of literature. *Surg Neurol.* 1981;15(6):435-443.
 12. Murakami H, Toya S, Otani M, Sato S, Ohira T, Takenaka N. No Shinkei Geka. 1985;13(2):175-179.
 13. Ho YS, Plets C, Goffin J, Dom R. Hemangioblastoma of the lateral ventricle. *Surg Neurol.* 1990;33(6):407-412.
 14. Prieto R, Roda JM. Hemangioblastoma of the lateral ventricle: case report and review of the literature. *Neurocirugia (Astur).* 2005;16(1):58-62.
 15. Jaggi RS, Premsagar IC; Abhishek. Hemangioblastoma of the lateral ventricle. *Neurol India.* 2009;57(5):677-679.
 16. Takeuchi S, Takasato Y. Supratentorial intraventricular hemangioblastomas. *Acta Neurol Belg.* 2011;111(4):353-356.
 17. 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(4):597-600.
 18. 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:bcr2014205736. Published 2014 Sep 25.
 19. John JK, Robin AM, Pabaney AH, et al. Complications of ventricular entry during craniotomy for brain tumor resection. *J Neurosurg.* 2017;127(2):426-432.
 20. Zhang XP, Liu Y, Zhang D, et al. Cerebellar ependymoma with overlapping features of clear-cell and tancytic variants mimicking hemangioblastoma: a case report and literature review. *Diagn Pathol.* 2017;12(1):28. Published 2017 Mar 20.
 21. Scheithauer BW, Bruner JM. Central nervous system tumors. *Clin Lab Med.* 1987 Mar;7(1):157-79. PMID: 3030612.
 22. García-Valtuille R, Abascal F, García-Valtuille AI, et al. Adult choroid plexus papilloma of the posterior fossa mimicking a hemangioblastoma. Case report. *J Neurosurg.* 2000;92(5):870-872.

23. Elster AD, Arthur DW. Intracranial hemangioblastomas: CT and MR findings. *J Comput Assist Tomogr.* 1988;12(5):736-739.
24. Lee GJ, Jung TY, Kim IY, et al. The clinical experience of recurrent central nervous system hemangioblastomas. *Clin Neurol Neurosurg.* 2014;123:90-95.
25. Richard S, Campello C, Taillandier L, Parker F, Resche F. Haemangioblastoma of the central nervous system in von Hippel-Lindau disease. French VHL Study Group. *J Intern Med.* 1998;243(6):547-553.
26. Laviv Y, Germano IM, Mahadevan A, Kasper EM. Surgery for posterior fossa ependymomas in adults. *J Neurosurg Sci.* 2018;62(1):63-70.
27. Dutta SW, Kaleem TA, Muller DA, et al. Central neurocytoma: Clinical characteristics, patterns of care, and survival. *J Clin Neurosci.* 2018;53:106-111.
28. Kuai XP, Wang SY, Lu YP, Xiong J, Geng DY, Yin B. MRI Features of Intracranial Anaplastic Ependymomas: A Comparison of Supratentorial and Infratentorial Lesions. *Front Oncol.* 2020;10:1063. Published 2020 Jul 21.
29. Zhang D, Wen L, Henning TD, et al. Central neurocytoma: clinical, pathological and neuroradiological findings. *Clin Radiol.* 2006;61(4):348-357.
30. Sun PF, Ma L, Ye BQ, Pei YY. Application of diffusion-weighted imaging combined with apparent diffusion coefficient in differential diagnosis between central neurocytoma and ependymoma. *Neuroradiology.* 2020;62(4):439-445.
31. Donoho D, Zada G. Imaging of central neurocytomas. *Neurosurg Clin N Am.* 2015;26(1):11-19.
32. Muly S, Liu S, Lee R, Nicolaou S, Rojas R, Khosa F. MRI of intracranial intraventricular lesions. *Clin Imaging.* 2018;52:226-239.

Figures

Figure 1

CT appearance of the tumor. (A1) Contrast-enhanced CT revealed heterogeneous enhancement of the solid components and (A2) thickened vascular shadow (yellow arrow).

Figure 2

MRI appearance of the tumor. (A) T1WI and (B1) T2WI revealed a large heterogeneous mass with slight edema in the left lateral ventricle, strip-like low signal shadow was seen on the (B2) T2WI (yellow arrow); (C) Contrast-enhanced MRI revealed the nodule and cyst wall were significantly enhanced; (D) DWI showed the nodule with hypo-isointense.

Figure 3

The pathological result of tumor. Microscopically, capillaries and mesenchymal cells of different maturation stages were observed.