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Supratentorial intraventricular hemangioblastomas

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Abstract

A 33-year-old male presented with a headache. He had a history of a previous surgical procedure for excisions of cervical spine hemangioblastomas 13 years prior. He had a family history of von-Hippel Lindau (VHL) disease, and a VHL mutation was identified. Brain magnetic resonance imaging showed enhanced mass lesions in both the third and right lateral ventricles as well as in the cerebellum. The lesion in the right lateral ventricle also had a cystic component. Two-staged resections of the tumors in the third and right lateral ventricles and ventriculo-peritoneal shunt were performed. A histopathological specimen was compatible with a hemangioblastoma. Supratentorial intraventricular hemangioblastomas are extremely rare. We reviewed the literature and discussed the features.

Key words: Hemangioblastoma; von-Hippel Lindau disease; lateral ventricle; third ventricle; supratentorial.

Introduction

Hemangioblastomas of the central nervous system (CNS) are the most frequent vascular tumors (1). Although they are usually isolated tumors, sometimes they are associated with von Hippel-Lindau (VHL) disease in 3% to 38% of cases as a major manifestation (2). They are predominantly found in the cerebellum, spinal cord, and brainstem (2). Supratentorial location accounts for 4% to 13% of the cases (2). However, supratentorial intraventricular hemangioblastomas are extremely rare and only 13 cases have been reported in the literatrue (3-15). We report a case of hemangioblastomas in both the third and lateral ventricles and review the literature.

Case report

A 33-year-old male presented with a headache. He had a history of a previous surgical procedure for excisions of cervical spine hemangioblastomas 13 years ago. A VHL mutation was identified and the patient was diagnosed with VHL disease. The patient's family history is of particular relevance. His mother underwent excisions of multiple cerebellar hemangioblastomas 13 years ago and 3 years ago. His brother underwent an excision of a cerebellar hemangioblastoma 1 year ago. On admission, neurological examination revealed normal findings. Computed tomographic scan of the abdomen showed no abnormality in the kidneys. Brain magnetic resonance imaging with gadolinium showed enhanced mass lesions in both the third and right lateral ventricles as well as the cerebellum, associated

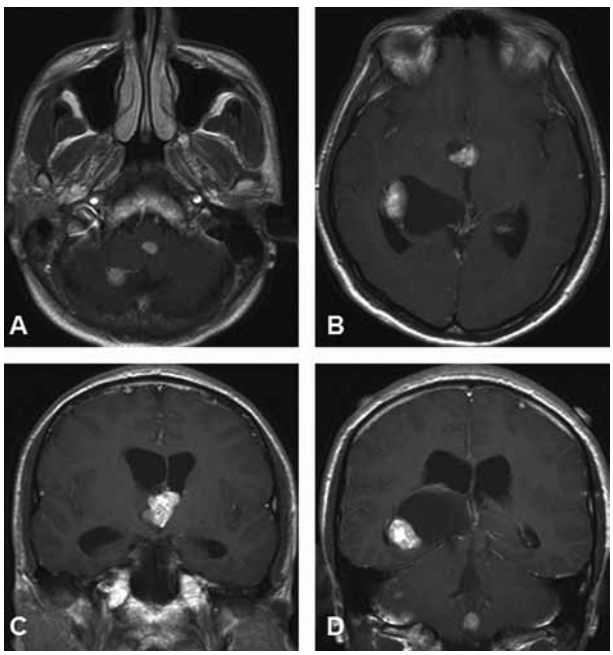

FIG. 1. — Brain magnetic resonance imaging with gadolinium showing enhanced mass lesions in the third and right lateral ventricles and cerebellum, associated with ventriculomegaly. Note that the lesion in the right lateral ventricle also has a cystic component (A, B: axial view; C, D: coronal view).

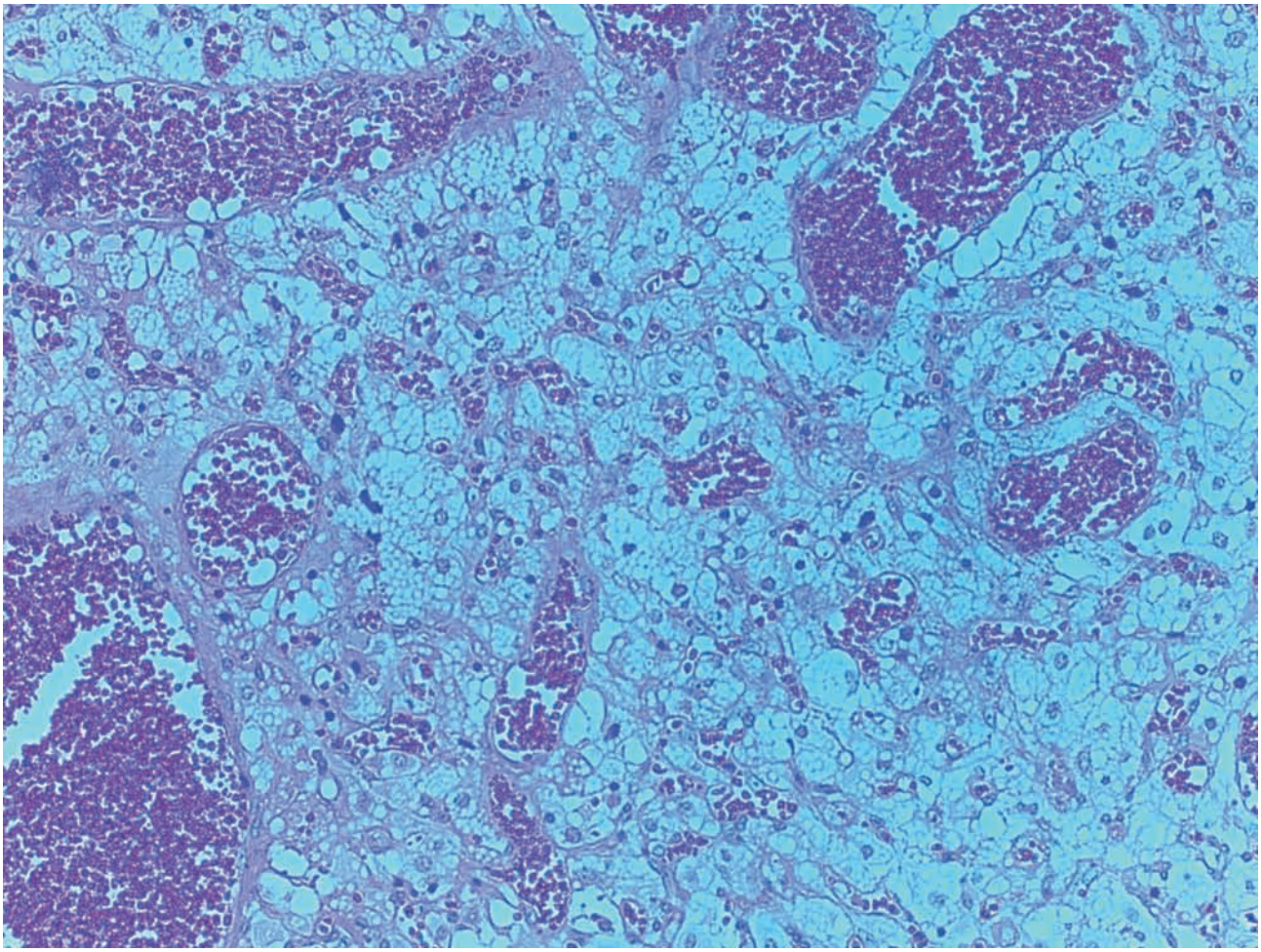

FIG. 2. — Histopathology of the resected specimen from the lesion in the right lateral ventricle showing tumors that are composed of a fine network of blood spaces separated by numerous polygonal stromal cells, with a lightly stained cytoplasm, which is compatible with a hemangioblastoma (hematoxylin and eosin; original magnification: ×200).

with ventriculomegaly (Fig. 1). The lesion in the right lateral ventricle also had a cystic component (Fig. 1). Two-staged resections of the tumors in the third and right lateral ventricles and ventriculoperitoneal shunt were performed. A histopathological examination showed that tissue of the tumors was composed of a fine network of blood spaces separated by numerous polygonal stromal cells, with a lightly stained cytoplasm (Fig. 2). Immunostaining showed a strong immunopositivity in several stromal cells for vimentin and neuron specific enolase – typical of a hemangioblastoma. The patient was discharged with a slight memory disturbance.

Discussion

To the best of our knowledge, only 15 tumors in 14 cases with supratentorial intraventricular hemangioblastomas, including our case, have been reported (3-15). The clinicoradiological features of all 14 cases are summarized in Table 1.

Interesting features of patients with supratentorial intraventricular hemangioblastomas include a high frequency of VHL disease association (9 of 14 patients), solitary tumors (9 of 14 patients), and solid tumors (13 of 15 tumors). The frequency of hemangioblastomas of the third ventricle was similar in patients with or without VHL disease, whereas hemangioblastomas of the lateral ventricle, except for one case, were only found in patients with VHL disease. There is also a similar frequency of the sites (the third ventricle in seven patients, lateral ventricle in six, and both in one). Furthermore, Ho *et al.* (9) reported that hemangioblastomas of the lateral ventricle are associated with a better prognosis than in the third ventricle; however, we found that prognosis

Table 1

Summary of patients with supratentorial intraventricular hemangioblastomas

Author	Age/Sex	VHL	Characteristics related to VHL disease	Site	Other hemangioblastomas	Nature	Treatment	Outcome
Vecchi *et al.* (3)	80/F	Yes	NA	LV	NA	Solid	None	Dead
Rho *et al.* (4)	58/M	Yes	Pheochromocytoma, paraganglioma, hypernephroma, positive FH	TV	Cerebellum, spinal cord	Solid	None	Dead
Diehl *et al.* (5)	20/M	Yes	Retinal angiomatosis , epididymal cyst, positive FH	LV	Cerebellum, medulla oblongata	Solid	TR	Transient partial homonymous field defect
Loftus *et al.* (6)	63/M	No	None	TV	None	Solid	TR	Dead
Murakami *et al.* (7)	31/F	Yes	Positive FH	LV	Cerebellum	Solid	TR	Transient hemiparesis
Katayama *et al.* (8)	30/M	No	None	TV	None	Solid	TR	Panhypopituitarism
Ho *et al.* (9)	44/F	Yes	Positive FH	LV	None	Solid	TR	Homonymous lower quadrantanopia
Black *et al.* (10)	15/M	No	None	TV	None	Solid	TR	NA
Isaka *et al.* (11)	47/F	Yes	Polycythemia	TV	Cerebellum	Solid	TR and radiation	Transient SIADH
Kouri *et al.* (12)	20/F	Yes	NA	TV	None	Solid	TR	Panhypopituitarism, DI
Prieto *et al.* (13)	73/M	Yes	Renal cell carcinoma	LV	None	Solid	TR	Transient mutism
Miyata *et al.* (14)	59/F	No	None	TV	None	Cystic	TR	Panhypopituitarism
Jaggi *et al.* (15)	30/M	No	None	LV	None	Solid	TR	Transient sensory aphasia
Present case	33/M	Yes	Positive FH, VHL mutation	TV, LV	Cerebellum Spinal cord	Solid (TV), cystic (LV)	TR	Memory disturbance

CNS, central nervous system; DI, diabetes insipidus; F, female; FA, family history; LV, lateral ventricle; M, male; NA, not available; SIADH, syndrome of inappropriate antidiuretic hormone secretion; TR, tumor removal; TV, third ventricle; VHL, von Hippel-Lindau disease.

was similar in both. Our patient was exceptionally unique because tumors were located in both the third and the lateral ventricles, and the tumor in the lateral ventricle was cystic but not solid.

In conclusion, supratentorial intraventricular hemangioblastomas are rare and their discovery should raise a high degree of suspicion for VHL disease.

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REFERENCES

1. Strugar J, Criscuolo G. Primary intracranial vascular tumors. In: Vecht C, ed. Handbook of clinical neurology. Neuro-Oncology. 1997;Part II:269-286.
2. Conway J, Chou D, Clatterbuck R, Brem H, Long DM. *et al.* Hemangioblastomas of the central nervous system in von Hippel-Lindau syndrome and sporadic disease. Neurosurgery. 2001;48:55-62.
3. Vecchi B, Patrassi G. Angioretriculoma del plessi corioidei, con "aree di Gamna". Schweiz Med Wochenschr. 1935;65:242-246.
4. Rho YM. Von Hippel-Lindau's disease: a report of five cases. Can Med Assoc J. 1969;101:135-142.
5. Diehl PR, Symon L. Supratentorial intraventricular hemangioblastoma: case report and review of literature. Surg Neurol. 1981;15:435-443.
6. Loftus CM, Marquardt MD, Stein BM. Hemangioblastoma of the third ventricle. Neurosurgery. 1984;15:67-72.
7. Murakami H, Toya S, Otani M, Sato S, Ohira T. *et al.* A case of concomitant posterior fossa and supratentorial hemangioblastomas. No Shinkei Geka. 1985;13:175-179.
8. Katayama Y, Tsubokawa T, Miyagi A, Goto T, Miyagami M. *et al.* Solitary hemangioblastoma within the third ventricle. Surg Neurol. 1987;27:157-162.
9. Ho YS, Plets C, Goffin J, Dom R. Hemangioblastoma of the lateral ventricle. Surg Neurol. 1990;33:407-412.
10. Black ML, Tien RD, Hesselink JR. Third ventricular hemangioblastoma: MR appearance. AJNR Am J Neuroradiol. 1991;12:553.
11. Isaka T, Horibe K, Nakatani S, Maruno M, Yoshimine T. Hemangioblastoma of the third ventricle. Neurosurg Rev. 1999;22:140-144.
12. Kouri JG, Chen MY, Watson JC, Oldfield EH. Resection of suprasellar tumors by using a modified transsphenoidal approach. Report of four cases. J Neurosurg. 2000;92:1028-1035.
13. Prieto R, Roda JM. Hemangioblastoma of the lateral ventricle: case report and review of the literature. Neurocirugia (Astur). 2005;16:58-62.
14. Miyata S, Mikami T, Minamida Y, Akiyama Y, Houkin K. Suprasellar hemangioblastoma. J Neuroophthalmol. 2008;28:325-326.
15. Jaggi RS, Premsagar IC, Abhishek. Hemangioblastoma of the lateral ventricle. Neurol India. 2009; 57:677-9.

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