

Hemangioblastoma of the Fourth Ventricle: Case Report¹

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Tumors arising from ventricular wall are usually ependymomas, subependymomas, and subependymal giant cell astrocytomas. Intraventricular hemangioblastomas are very unusual, especially hemangioblastomas in the fourth ventricle. Here, we described a hemangioblastoma of the fourth ventricle which was completely excised.

Index words : Hemangioblastoma
Fourth ventricle

The ventricles are surrounded by a lining of ependymal cells and a subependymal plate composed of glial cells. Accordingly, these layers give rise to ependymomas, subependymomas, and subependymal giant cell astrocytomas (1). Hemangioblastomas are benign vascular tumors with an uncertain origin. They account for 1-2% of all intracranial tumors, and are a component of the von Hippel-Lindau syndrome in 30% of cases (2, 3). Hemangioblastomas usually occur in the infratentorial region, especially the cerebellum, the pons, medulla oblongata, spinal cord, and rarely the supratentorial locations (2, 3). Moreover, intraventricular hemangioblastomas are also very unusual, as are hemangioblastomas of the fourth ventricle. Here, we described a hemangioblastoma of the fourth ventricle which was completely excised.

Case Report

A 55-year-old man admitted to our hospital with a several day history of dizziness. Prior to the dizziness, the patient was in good health. A neurologic examination performed on admission was unremarkable. A laboratory investigation revealed a hemoglobin level of 16 g/dL and a hematocrit of 44.3%. None of his relatives had any relevant illness or any lesion of the VHL disease.

A Magnetic Resonance Imaging (MRI) was performed and revealed hypo-isointense on T1-weighted images compared to the normal white matter and hyperintense T2-weighted images with vascular signal voids. A gadolinium-enhanced T1-weighted MRI demonstrated a solitary, lobulated intraventricular mass measuring 2 cm in diameter, located at the fourth ventricle (Fig. 1A-D).

These findings are consistent with a hemangioblastoma, but diagnostic discrimination is still required with other hypervascular masses such as ependymoma, meningioma, and hemangiopericytoma.

The patient underwent a craniotomy and a C1 partial laminectomy. A grayish yellow-colored mass, 2 cm in diameter, was seen on the floor of the fourth ventricle with slight adhesion to the ventricular wall. However, it

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was clearly separated from ventricular wall.

A histologic examination of the tumor specimens revealed numerous vacuolated stromal cells consistent with the characteristics of a hemangioblastoma (Fig. 2A, B).

After three months, there is no remaining residual tumor mass in the fourth ventricle on a follow-up MRI (Fig. 3).

Discussion

Hemangioblastomas are benign vascular tumors of

with an uncertain origin and consist of three types of cells: endothelial cells, pericytes, and stromal cells (4). Further, hemangioblastomas account for 1–2% of all intracranial tumors, and are a component of the von Hippel-Lindau syndrome in 30% of cases (2, 3). The site most frequently affected by a hemangioblastoma is the posterior fossa with the cerebellar hemispheres (80–85%), followed by the spinal cord and brainstem. Supratentorial hemangioblastomas are rare, accounting for only 4% of cases in patients with sporadic disease and 13% for VHL disease (5).

Hemangioblastomas are usually found in young

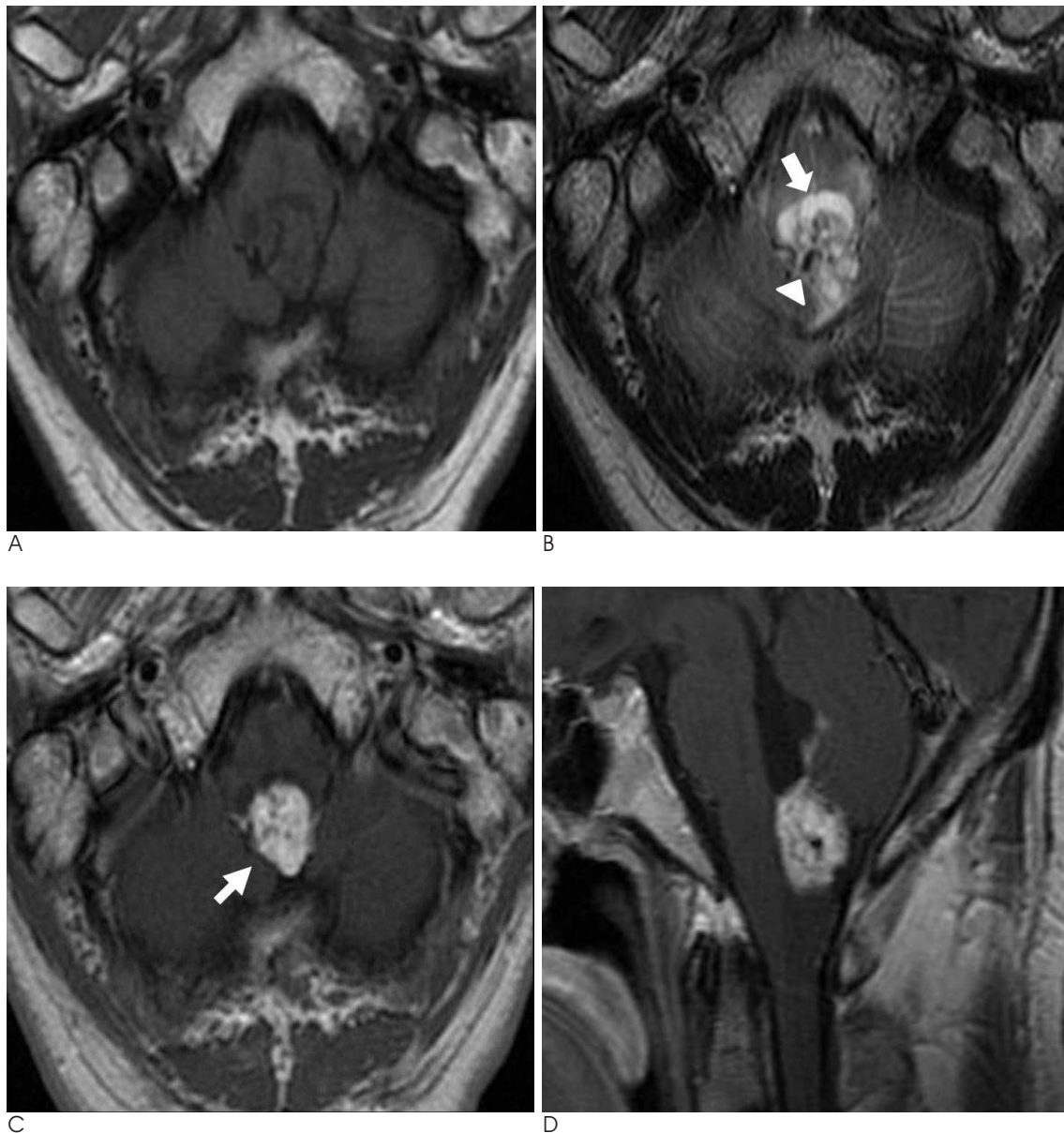


Fig. 1. Axial T1-weighted image reveals a hypo-intense mass at fourth ventricle (A). An axial T2-weighted image reveals a heterogeneous mass with a focal cyst (arrow) and multiple signal voids (arrow head) (B). Contrast-enhanced axial (C) and sagittal (D) images reveals strong enhancement (arrow, Fig. 1C).

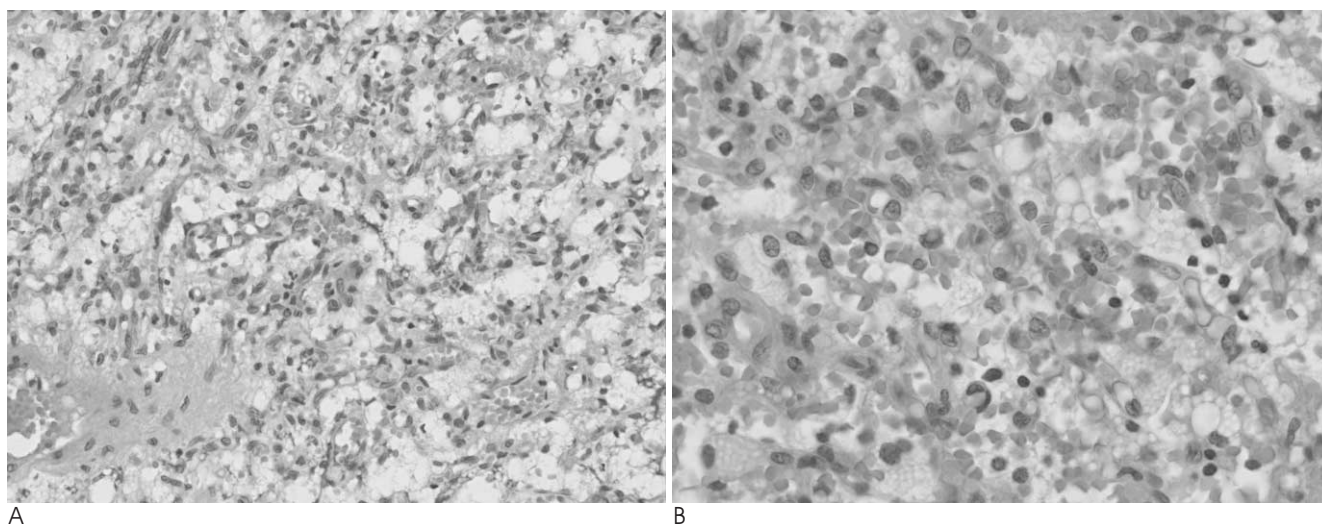


Fig. 2. Histologic findings of the brain tumor. Numerous vacuolated stromal cells are present between several small blood vessels (hematoxylin-eosin, original magnification $\times 200$, $\times 400$) (A, B).

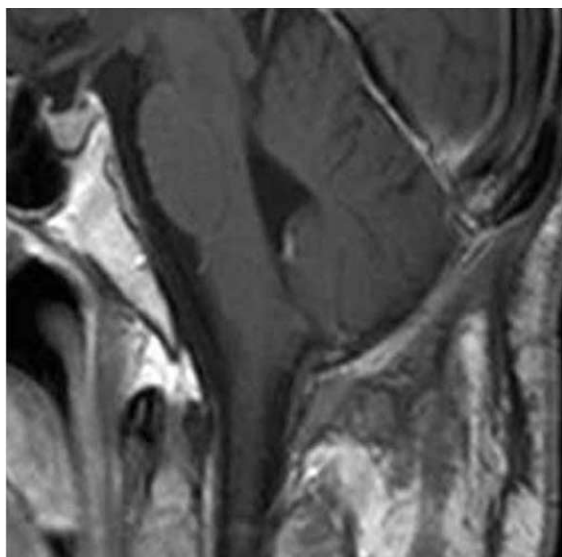


Fig. 3. Post-operative follow-up MRI. No residual tumor mass is observed on the T1-weighted sagittal image with contrast enhancement.

adults, with the mean patient age at presentation in most series being the third or fourth decade of life (3). Approximately 80% of these tumors arise sporadically. Moreover, 70% of infratentorial hemangioblastomas and 49% of supratentorial hemangioblastomas are associated with cysts (2, 6).

Hemangioblastomas can be divided into four groups according to the cystic component: simple cystic, macrocystic, solid, and microcystic solid types (6). Polycythemia can be detected in 10–50% of patients with infratentorial hemangioblastoma, but has been rarely reported in patients with supratentorial heman-

gioblastoma (7).

The neuroradiological features of infratentorial hemangioblastomas have been well documented. On CT, the cyst fluid has a higher density than that of CSF. Moreover, solid tumors and peripheral pial-based mural nodules of cystic tumors are isodense with brain tissue and show marked homogenous contrast enhancement. Calcification and surrounding edema are an uncommon finding.

The solid part of the tumor is usually hypointense or isointense on a T1-weighted MRI. The solid part shows diffuse enhancement with contrast medium and the cystic part is hyperintense on T2-weighted images.

Total extirpation of the tumor prevents recurrence and postoperative hemorrhage. Radiotherapy is an alternative or adjuvant therapy for multiple, subtotally resected, and recurrent cases.

Tumors arising from the ventricular wall are usually ependymomas, subependymomas, and subependymal giant cell astrocytomas. To our knowledge, a heman-gioblastoma in fourth ventricle is very rare. But it should be considered, when found the intraventricular mass.

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제 4 뇌실의 혈관모세포종: 증례 보고¹

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뇌실 벽으로부터 기원한 종양들은 대개 상의세포종, 상의하세포종, 그리고 상의하 거대세포 성상세포종 등이다. 뇌실 내의 혈관모세포종은 매우 드물며 제 4 뇌실은 더욱 그러하다. 수술적 제거 후 혈관모세포종으로 확진된 증례에 대해서 보고하고자 한다.