Tentorial Vascularization in Solid Hemangioblastoma

—Case Report—

M. Faik OZVEREN, Cahide TOPSAKAL, Fatih S. EROL, Metin KAPLAN,
Koichi UCHIDA*, and Canan TANIK**

*Department of Neurosurgery, Firat University Medical School, Elazig, Turkey; *Department of Neurosurgery, Keio University Medical School, Tokyo; **Department of Pathology, Sisli Etfal State Hospital, Istanbul, Turkey*

Abstract

A 40-year-old female was admitted to the hospital with complaints of headache worsening gradually over a 1-month duration. Her past history included surgery to treat a left cerebellar cystic lesion 3 years before, and an untreated small solid right supracerebellar lesion of 1 cm diameter. On admission, magnetic resonance imaging showed that the right cerebellar lesion had grown to approximately 4 cm diameter abutting the tentorium and causing obstructive hydrocephalus. She also had two more small lesions, a right supratentorial solid lesion with cystic component near the splenium and an intramedullary cystic lesion at the C-2 level. Right suboccipital craniectomy was done. The vascular attachments between the superior aspect of the tumor and the tentorium were coagulated and the tumor was totally removed. C1-2 laminectomy was also performed to drain the intramedullary cyst. The patient deteriorated and lost consciousness with respiratory arrest 6 hours postoperatively and was reoperated for intracerebellar hematoma due to oozing from the tentorial vessels. Histological investigation revealed hemangioblastoma. Dural tentorial vascular attachments in solid hemangioblastomas located subjacent to the tentorium may cause early postoperative complications of hematoma at the site of vascular attachment following the resection. Computed tomography study in the early postoperative period is helpful to identify this problem.

Key words: postoperative hemorrhage, solid hemangioblastoma, tentorial vascularization

Introduction

Hemangioblastomas are benign vascular lesions and account for 1-2.5% of all intracranial tumors. Hemangioblastomas originating from the pia-arachnoid junction form 7.3-8.8% of lesions in the posterior fossa.^{5,15)} Basically, hemangioblastoma occurs in infratentorial locations but can occur in supratentorial or spinal sites as well.^{10,11,19)} The presentation is usually sporadic, but 10-20% manifest as a component of autosomal dominant-familial transmitted von Hippel-Lindau disease characterized by retinal angiomas, pheochromocytoma, as well as renal, pancreatic, and bone cysts.^{1,8)}

Hemangioblastomas can be subdivided into four groups: simple cystic (4.1%), macrocystic (60%), solid (26%), and microcystic solid types (9%).^{3,18)}

Solid hemangioblastomas are relatively less common and require different treatment because of the high vascularization. We report the significance of tentorial vascularization of a solid cerebellar hemangioblastoma which occurred as a component of multiple hemangioblastomas without the stigmata of von Hippel-Lindau disease.

Case Report

A 40-year-old female was admitted to the Neurosurgery Department of Firat University Medical School with complaints of headache, nausea, and vomiting worsening gradually for a month. The patient had papilla edema, but otherwise no neurological abnormalities were found. Her past history included surgery to treat a left cerebellar cystic lesion 3 years previously, and an untreated small solid right cerebellar lesion of 1 cm diameter (Fig. 1A). On admission, magnetic resonance (MR) imaging showed the right cerebellar lesion was approximately 4 cm di-

Fig. 1 A: Computed tomography scans taken 3 years before admission, showing the cystic mass lesion on the left cerebellar hemisphere (arrowhead) as well as a discrete infratentorial small lesion attached to the right tentorium (arrow). B: Magnetic resonance image on admission, showing contrast enhancement of the right infratentorial lesion, which had grown to 4 cm diameter and caused obstructive hydrocephalus as well as brainstem distortion.

ameter, abutting the tentorium and causing obstructive hydrocephalus (Fig. 1B). In addition, a right supratentorial solid lesion with cystic component was found near the splenium, and another intramedullary cystic lesion at the C-2 level (Fig. 2). MR imaging demonstrated the right cerebellar lesion as an isointense area on T_1 -weighted images and a hyperintense area on T_2 -weighted images with contrast enhancement (Fig. 1B). Angiography revealed arterial feeding maintained by the right superior cerebellar, dural arteries and venous drainage of the tumor into the sigmoid sinus (Fig. 3). There was also a discrete arterial blush consistent with the small supratentorial lesion.

The patient was operated on in the sitting position. Right suboccipital craniectomy was performed to treat the first lesion. The vascular attachments between the uppermost aspect of the lesion and tentorium were cauterized. The cleavage plane between the arachnoid membrane and cerebellum was followed completely around the lesion and total removal was achieved (Fig. 4). C1-2 laminectomy was performed to treat the second lesion. The intradural cyst with mural nodule was drained. The patient was awakened at the 2nd postoperative hour without neurological deficit. However, she suddenly deteriorated and became unconscious, followed by respiratory arrest at the end of the 6th hour. Computed tomography (CT) was not available, so she was reoperated and a hematoma was discovered due to oozing from the vascular elements at the inferior aspect of tentorium. After drainage, meticulous hemostasis was performed.

Histological examination of the tumor specimens revealed cellular elements with large vacuolized foamy cytoplasm, and tiny vesicular nuclei consis-

Fig. 2 Magnetic resonance images, showing a right supratentorial solid mass with a cystic component near the splenium (A), and an intramedullary cystic lesion at the C-2 level (B).

Fig. 3 Right lateral vertebral angiogram, indicating that the infratentorial lesion is fed mainly by the superior cerebellar artery (arrow) (A). Right lateral carotid angiogram, showing rich dural vessels located posterior to the petrous bone, and the solid part of the supratentorial lesion enhanced densely by contrast medium (arrow) (B).

Fig. 4 Photograph, showing the solid lesion removed en bloc.

Fig. 5 Photomicrograph, showing the tumor tissue characterized by rich vascular elements encircled by foamy translucent cytoplasmic cells. Factor-VIII-positive tumor cells contain intracytoplasmic brown granules (arrows). HE stain, $\times 100$.

tent with hemangioblastoma (Fig. 5). Immunostaining showed positive reaction for factor-VIII in the vascular elements of the hemangioblastoma, which confirmed the diagnosis. No neurological deficit, and no tumor recurrence on CT was noted at the 2-year follow-up examination (Fig. 6).

Discussion

Intracranial hemangioblastomas are usually unifocal, but are multifocal in 3.8–11.9% of cases.^{3,18)} Patients with von Hippel-Lindau disease show central nervous system involvement in 42–63% of cases.^{5,8)} Cranial multifocal lesions are more common in familial cases.^{9,11)} Hemangioblastomas are usually located in the posterior fossa, and rarely in the spinal cord and supratentorial region.^{5,11)} Supratentorial hemangioblastoma resembles a cerebellar lesion, but unifocal lesions can present a problem in the differential diagnosis.¹¹⁾ In our case, the pre-operative diagnosis of the supratentorial lesion was hemangioblastoma due to the solid and cystic components delineated on MR imaging and hypervascularity on angiography. The vascular origin of the tumor was confirmed by positive factor-VIII immunostaining. The patient had no familial predisposition and von Hippel-Lindau disease was excluded despite the concomitant supratentorial, infratentorial, and spinal lesions.

Hemangioblastomas are benign lesions, so the symptoms and signs usually manifest in the late

Fig. 6 Magnetic resonance image, showing no recurrence at the 2-year follow-up examination.

period.¹⁸⁾ Latency can vary from 5 to 33 years.¹⁶⁾ The mean presentation age is 29–30 years in cases of von Hippel-Lindau disease, but 42–47 years in sporadic cases.^{5,11)} In our case, the cerebellar cystic lesion manifested at the age of 37 years, and developed within 4 years to the extent of causing mass lesion symptoms. Most presenting symptoms are due to direct compression of neural structures, hydro-

cephalus, cerebellar syndrome, brain stem signs, and increased intracranial pressure.^{5,16,18)} Ventricular hemorrhage has also been reported.¹⁶⁾ During surgery, attention must be paid to the fourth cranial nerve which has an intimate relationship with solid tumors located near the tentorium.¹²⁾ In our patient, increased intracranial pressure symptoms were noted due to obstructive hydrocephalus.

Hemangioblastomas consist mainly of a solid component even in the cystic forms, which is 83% of the lesion on CT and 95% on angiography.⁵⁾ This solid part consists of the mural nodule composed of feeding and drainage vessels in cystic lesions and the entire solid mass in pure solid tumors. Therefore, mural nodule should be excised after cyst removal, whereas the mass should be removed "en bloc" in solid forms similar to arteriovenous malformation.^{12,18)} The significance of total extirpation is well known because of the possibility of postoperative hemorrhage complications.⁷⁾ Preoperative embolization can be considered to avoid excessive perioperative bleeding.¹⁷⁾ Intraparenchymal cerebellar lesions are fed mainly by the posterior inferior cerebellar artery.^{12,16,18)} However, lesions at the superior cerebellar location are fed mainly by the superior cerebellar artery¹²⁾ and appear similar to tentorial meningiomas on neuroimaging.¹⁷⁾ This similarity is due to both the rich vascularized nature of the tumor and the tentorial dural vascular attachments. The tentorial artery supply is significant in hemangioblastoma.^{2,13)} In our case, the tumor was nourished mainly by the superior cerebellar artery, but also by the dural arteries from the tentorium, causing the lesion to resemble a meningioma. The presence of the transverse, superior petrosal, and straight sinuses, as well as venous complexes draining into these sinuses and the sinus variations, require careful attention to hemostasis.^{4,16)} Careful measures must be taken to prevent postoperative hematoma formation resulting from improper cauterization or gradual venous oozing facilitated by the increased intracranial pressure. In our case, total extirpation and meticulous cauterization was performed, but intracerebellar hematoma developed due to indolent venous oozing through the tentorium.

The arachnoid membrane circumscribing the tumor along the cleavage plane prevents neural tissue damage at the time of surgery,¹⁰⁾ and allows total removal of the tumor.¹⁸⁾ This feature is important in preventing recurrences.¹⁶⁾ Subtotal resection for brain stem extension is recommended because of the increased mortality resulting from cauterization of tumor bed in that area.^{7,12,16)} The effects of neural damage most likely due to cauterization are notori-

ous, particularly in the field of the hypoglossal trigone.¹⁸⁾ The lesion in our patient had no arachnoid cleavage plane at the site of the dural attachment, as the contact was maintained directly by vessels.

Radiotherapy can hinder or regress tumor enlargement in cases of recurrent multifocal posterior fossa hemangioblastoma that was not totally removed.^{9,14,15,19)} Recently, gamma knife radiotherapy has been found efficacious in cases of small or medium solid hemangioblastomas, but cystic lesions usually require surgical intervention.⁶⁾ In our case, we did not perform radiotherapy as total extirpation had been achieved. No recurrence was noted at the follow-up examination 2 years later.

Solid hemangioblastoma located subjacent to the tentorium cerebelli can cause postoperative hematoma complications because of dural vascular attachments. Therefore, meticulous hemostasis is needed at the time of tumor resection from the tentorium, and CT should be performed in the early postoperative period. Total extirpation prevents recurrence, but radiotherapy can be implemented as an adjunct therapy in cases of subtotal resection or multifocal lesions.

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Address reprint requests to: M. F. Ozveren, M.D., Firat Universitesi, Tip Fakultesi, Norosirurji Anabilim Dalı, Elazig, Turkey.