Cerebellar Hemangioblastoma with Intracystic Hemorrhage

—Case Report—

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Abstract

A 45-year-old male presented with sudden onset of severe headache, mild disorientation, and gait disturbance due to intracystic hemorrhage from a cerebellar hemangioblastoma. He was successfully treated with ventricular drainage followed by total removal of the tumor. Reported cases of intracranial hemangioblastoma with massive hemorrhage show male predominance, tend to be solid rather than cystic, and occur in supratentorial more than infratentorial locations. Thin-walled and dilated vessels in the present and other cases were a possible cause of hemorrhage. Massive hemorrhage from hemangioblastoma is rare but needs prompt treatment because of the relatively high mortality.

Key words: hemangioblastoma, cerebellum, intratumoral hemorrhage, computed tomography, magnetic resonance imaging

Introduction

Hemangioblastoma constitutes 1–3% of intracranial neoplasms, occurs chiefly in the posterior fossa,^{4,13)} and tends to be associated with cyst formation. Hemangioblastoma is composed of a large number of capillary vessels separated by stroma cells, and sometimes contains hemosiderin deposits which imply intratumoral bleeding. However, massive hemorrhage from this tumor is rare, with only 13 reported cases of intracranial hemangioblastoma with clinically significant hemorrhage.^{1,3,5,9–12,17)}

We describe a rare case of hemangioblastoma with hemorrhage into the cyst.

Case Report

A 45-year-old male had suffered from mild occipitalgia for 6 months. He was admitted to our hospital on April 5, 1994 with sudden onset of headache in the occipital region, nausea, and vomiting, followed by gait disturbance. He had no

history of hypertension or specific family history.

On admission, he was slightly drowsy and disoriented with left cerebellar ataxia. Precontrast computed tomography (CT) showed a crescent-shaped high density area in the vermis (Fig. 1 *left*). The fourth ventricle had collapsed and the lateral ven-

Fig. 1 Precontrast CT scan showing a crescent-shaped intracystic hemorrhage and an isodense mass in the cerebellar vermis (*left*). Postcontrast CT scan revealing the well-enhanced tumor mass (*right*).

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Fig. 2 T₁-weighted MR images without (*left*) and with (*right*) gadolinium enhancement demonstrating a well-demarcated tumor mass with many flow-void areas in the cerebellar vermis.

Fig. 3 Left vertebral angiograms (*left*: anteroposterior view, *right*: lateral view) demonstrating a tumor stain fed by vermian branches of the left posterior inferior cerebellar artery.

tricles and third ventricle were dilated. Postcontrast CT revealed a well-enhanced mass (4 × 4 × 3 cm) in the vermis just behind the crescent-shaped high density area (Fig. 1 *right*). Magnetic resonance (MR) imaging also revealed a well-enhanced mass containing flow-void areas in the vermis (Fig. 2). Vertebral angiography revealed a tumor stain fed mainly by vermian branches of the left posterior inferior cerebellar artery without apparent venous drainage (Fig. 3). Based on these findings, the lesion was diagnosed as cerebellar hemangioblastoma with intracystic hemorrhage. Laboratory examinations revealed normal levels of urine vanillylmandelic acid and serum erythropoietin, and no erythrocytosis. Optic funduscopy revealed no vascular anomaly.

On April 5, right ventricular drainage was im-

Fig. 4 Photomicrographs of the tumor specimen demonstrating small capillary vessels separated by stroma cells (*upper*: HE stain, ×300), and some dilated vessels with thin walls surrounded by massive hemorrhage (*lower*: HE stain, ×80).

placed to control the intracranial pressure with careful management to avoid overdrainage. On April 20, total removal of the reddish nodule in the cerebellar vermis was performed through a bilateral suboccipital craniotomy. The associated cyst containing xanthochromic fluid was exposed on the ventral surface of the nodule.

Histological examination of the tumor specimen using HE staining revealed prominent vascular capillary networks with endothelial linings separated by so-called stroma cells, findings typical of hemangioblastoma (Fig. 4 *upper*). Some vessels with thin walls were abnormally dilated and may have been the source of the hemorrhage (Fig. 4 *lower*).

Postoperative angiography showed no residual tumor, and CT showed a gradual normalization of the size of the ventricles. His postoperative course was uneventful and he resumed his normal activities 1 month after the operation. Abdominal echography revealed no apparent mass or cystic lesion.

Discussion

Spontaneous massive hemorrhage occurs in 1–5% of brain tumors, especially malignant tumors such as glioblastoma multiforme and metastatic tumors, and benign tumors such as meningioma and pituitary adenoma.^{2,8,14,18} Hemangioblastoma is rarely associated with massive hemorrhage despite its rich vascular capillary networks. The 14 reported cases of intracranial hemangioblastoma with massive hemorrhage are listed in Table 1.

Hemangioblastoma with massive hemorrhage has a male predominance (male:female = 11:3) compared with the ratio for all hemangioblastoma (male:female = 1–2:1).^{4,13} Familial hemangioblastoma was confirmed in one case⁹ and suspected in another.¹ There is also a supratentorial predominance (supratentorial:cerebellum:brainstem = 6:7:1), considering that supratentorial hemangioblastoma is quite rare. Obvious cyst formation occurred in only two cerebellar cases including ours. Both had intracystic hemorrhage, implying that the solid type of hemangioblastoma bleeds more easily than the cystic type.

Hemangioblastoma with hemorrhage was diagnosed preoperatively by angiography in the most recent four cerebellar cases, including ours,^{9,10,12} but all six supratentorial cases^{1,3,17} and four other infratentorial cases^{5,11,17} were diagnosed only after operation or at autopsy by histological examination. Angiography may not visualize vascular abnormalities because the tumor is destroyed by hemorrhage or the hematoma compresses the feeding

artery and tumor. Careful radiological studies including postcontrast CT and angiography are mandatory. MR imaging in two recent cases including ours⁹ visualized the tumor clearly. MR imaging is particularly useful for the diagnosis of small supratentorial and brainstem hemangioblastoma.

Proposed mechanisms for hemorrhage from brain tumors are as follows^{6,7,15,16}: hemorrhagic infarction due to stenosis or occlusion of vessels by endothelial proliferation or tumor emboli, rupture of thin and extended fragile vessels due to compression by the tumor, rupture of vessels due to the direct invasion of the tumor cells, rupture of vessels due to loss of perivascular supporting tissue, rupture of fragile neovasculature, vascular degeneration due to radiation or chemotherapy, and sudden structural changes due to reduction of intracranial pressure by surgical procedures. In familial hemangioblastoma, genetic factors and associated hypertension due to visceral tumors such as pheochromocytoma may also contribute to the fragility of vascular walls.

At least nine of the 14 previous cases, including ours,^{9,12,17} had some thin-walled and dilated vessels which might be the cause of hemorrhage. One patient¹² received radiation therapy (Linac 49.5 Gy) before the second hemorrhage. Four of the eight patients with infratentorial hemangioblastoma including ours received ventricular drainage¹¹ or ventriculoperitoneal shunting,^{9,12} two of whom suffered from subsequent hemorrhage.^{9,12} Our patient underwent ventricular drainage for acute hydrocephalus, with the draining pressure set above 25 cmH₂O for the first 3 days to avoid sudden changes of in-

Table 1 Reported cases of hemangioblastoma with hemorrhage

Author (Year)	Age/ Sex	Site	Size of nodule (cm)	Cyst	CE on CT	Angiography	Treatment	Outcome
Fukuma <i>et al.</i> (1977) ⁹	60/M	vermis	3	+	—	tumor stain	total (twice)	alive
Bachmann <i>et al.</i> (1978) ³	26/M	rt parietal	3?	?	+	small tangle	total	good
Nishio <i>et al.</i> (1981) ¹¹	27/M	medulla oblongata	1	—	?	not performed	drainage	died
Adegbite <i>et al.</i> (1983) ¹¹	29/M	lt frontotemporal	3	—	?	not performed	total	died
Wakai <i>et al.</i> (1984) ¹⁷	2/M	superior vermis	1	—	—	tumor blush	total	fair
	49/M	lt cerebellum	1	—	—	mass effect	total	good
	32/F	lt parieto-occipital	?	—	—	mass effect	subtotal	good
	35/M	lt parieto-occipital	1	—	—	tumor blush	total	good
	74/M	lt parieto-occipital	5	—	—	mass effect	total	good
	72/F	lt parieto-occipital	3	—	—	mass effect	total	good
Matsumura <i>et al.</i> (1985) ¹⁰	54/M	rt cerebellum	2	?	+	tumor stain	total	good
Orita <i>et al.</i> (1985) ¹²	52/F	lt cerebellum	2	—	+	tumor stain	drainage, VP shunt, radiation, total (three times)	good
Kikuchi <i>et al.</i> (1994) ⁹	69/M	vermis-brainstem	5	—	+	tumor stain	VP shunt	died
Present case	45/M	vermis	4	+	+	tumor stain	drainage, total	good

CE: contrast enhancement, VP: ventriculoperitoneal.

tracranial pressure, rebleeding, and upward herniation.

Massive intratumoral hemorrhage from hemangioblastoma should be considered in the diagnosis of intracranial hemorrhage. Prompt diagnosis and appropriate surgical treatment are required because of the relatively high mortality.

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