

Case Report

Rapid Progression of Cerebral Infarction after Intraventricular Hemorrhage in Adult Moyamoya Disease

Hiroaki Matsumoto, M.D., Ph.D., Yasuhisa Yoshida, M.D.

Department of Neurosurgery, Eisyokai Yoshida Hospital, Kobe, Japan

The authors present a rare case of adult moyamoya disease in which a patient experienced rapid progression of cerebral infarction after intraventricular hemorrhage (IVH). A healthy 39-year-old woman was admitted to our hospital with sudden headache, a decreased level of consciousness and mild tetraparesis. Initial magnetic resonance imaging revealed small cerebral infarction and IVH. Although the patient underwent conservative therapy including hypervolemia, hemodilution, keeping moderate hypertension and administration of a free radical scavenger, she showed a fulminant clinical course of cerebral infarction. The authors discuss the possible pathophysiology and suggest the treatment for such cases.

Key Words : Moyamoya disease · Cerebral infarction · Intraventricular hemorrhage.

INTRODUCTION

Moyamoya disease is characterized by progressive bilateral occlusion of the internal carotid artery (ICA)¹⁾. Although moyamoya disease is usually classified as either ischemic or hemorrhagic type, unusual clinical cases of moyamoya disease such as rapid progression from unilateral to bilateral involvement, infarction following the acute presentation of hemorrhagic-type moyamoya, or concomitant cerebral infarction and hemorrhage have rarely been reported²⁻¹³⁾. Herein, we report one adult case of moyamoya disease in which a patient experienced rapid progression of cerebral infarction after intraventricular hemorrhage (IVH).

CASE REPORT

A healthy 39-year-old woman visited our hospital with sudden headache and a decreased level of consciousness. On admission, she was drowsy with mild tetraparesis [manual muscle testing (MMT) score 4/5] and dysarthria. Her blood pressure was 230/100 mm Hg. Magnetic resonance imaging (MRI) revealed IVH on diffusion-weighted imaging in addition to small

cerebral infarction in the bilateral frontal lobe (Fig. 1A-C), and MR angiography revealed bilateral occlusion at the terminal of the ICA (Fig. 1D). There was asymptomatic, old cerebral infarction in the left frontal lobe on fluid-attenuated inversion recovery imaging (Fig. 1E). No old hemorrhage was detected on T2-star-weighted imaging (Fig. 1F). Routine laboratory tests were normal. We diagnosed that ischemic-type moyamoya disease had changed into hemorrhagic-type. After admission, the patient underwent conservative treatment with edaravone, a free radical scavenger. Anti-platelet therapy was not performed because of the possibility of progression of IVH. Because there was old cerebral infarction, dehydration might occur ischemia. Hence, hyperosmotic agent such as glycerol was not given. To maintain adequate cerebral hemodynamics, her systolic blood pressure was kept at 180-200 mm Hg, and she received hypervolemia and hemodilution. Her body temperature was around 37.0°C, and her urine output was adequate. Nevertheless, on the second hospital day, the right hemiparesis deteriorated (MMT score 1/5) with motor aphasia, and the infarction area continued to expand without progression of IVH (Fig. 2). On the 7th day, the left hemiparesis deteriorated (MMT score 1/5) and she became apallic. On the 14th day, repeated MRI re-

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• Address for reprints : Hiroaki Matsumoto, M.D., Ph.D.

Department of Neurosurgery, Eisyokai Yoshida Hospital, Daikai-dori9-2-6, Hyogo-ku, Kobe 652-0803, Japan

Tel : +81-78-576-2773, Fax : +81-78-577-2792, E-mail : hiroaki-matsu@umin.ac.jp

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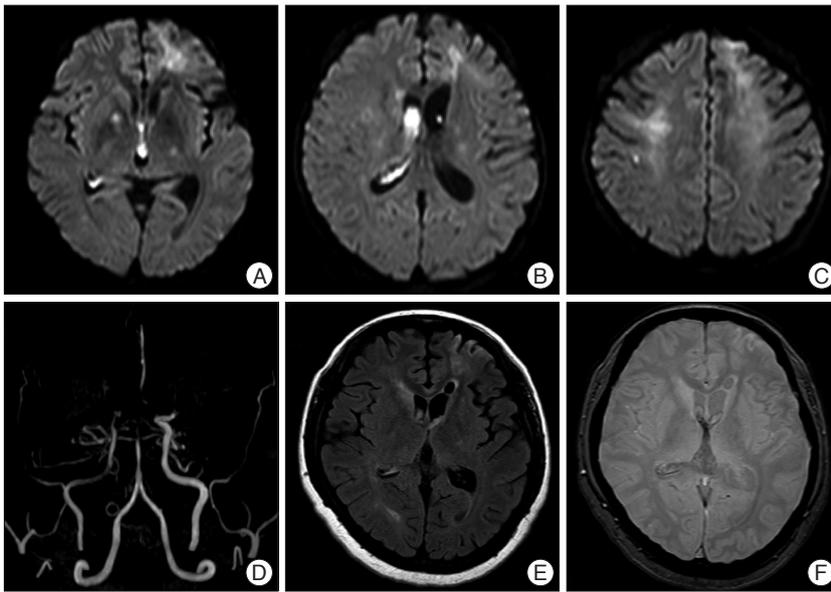


Fig. 1. Magnetic resonance (MR) imaging obtained on admission. Diffusion weighted imaging reveals intraventricular hemorrhage and acute cerebral infarction in bilateral frontal lobes (A, B and C). MR angiography reveals bilateral occlusion at the terminal of the internal carotid artery, indicating moyamoya disease (D). Fluid-attenuated inversion recovery imaging reveals asymptomatic, old cerebral infarction in the left frontal lobe (E). T2-star-weighted imaging does not reveal low intensity (F).

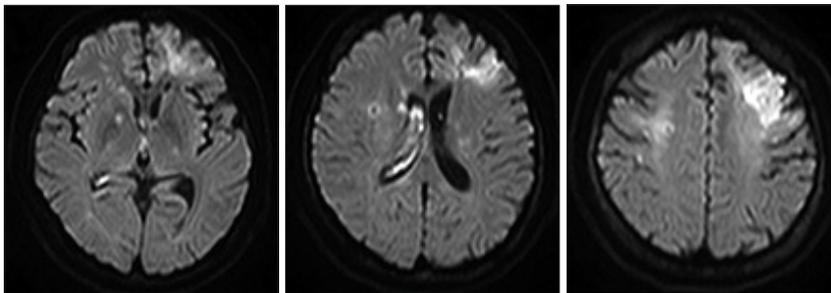


Fig. 2. Diffusion weighted imaging obtained on the second hospital day showing expansion of cerebral infarction in the left cerebral hemisphere without progression of IVH.

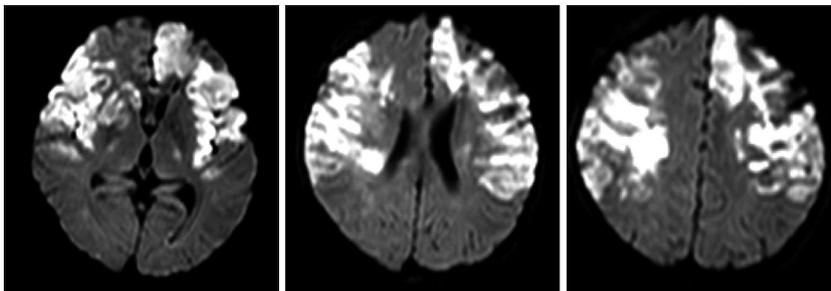


Fig. 3. Diffusion weighted imaging obtained on the 14th hospital day showing expansion of cerebral infarction in the bilateral cerebral hemispheres without ventriculomegaly.

vealed massive cerebral infarction in the bilateral frontal lobes and expansion of the cerebral infarction stopped at last (Fig. 3).

DISCUSSION

Moyamoya disease is classified as either the ischemic or hemorrhagic type. In general, cerebral ischemia predominates in chil-

dren, and intracranial hemorrhage is more common in adults^{1,5}. The symptoms and course of moyamoya disease vary, ranging from no symptoms to a transient disorder, to fixed neurological deficits of slight or severe degree¹.

In our case, the patient experienced rapid progression of cerebral infarction after IVH. Including the present case, 20 cases of moyamoya disease with ischemic stroke after intracranial hemorrhage have been reported, and the Table 1 summarized the clinical features. The patients were predominantly female (14 female patients and six male patients) and adult (13 adult patients and seven pediatric patients). As to hemorrhage type, IVH was common. Of the 20 patients, 18 patients had IVH with or without ICH. Infarction occurred in these patients mainly during the subacute stage between postical days 7 and 20, with the exception of four patients in whom infarction occurred during the acute stage. Of the four patients, three patients including our case had infarction within 24 hours after hemorrhage. Although the overall outcome tended to be moderate, the patients in whom infarction occurred during the acute stage had tendency to be poorer than the patients during the subacute stage.

Increased intracranial pressure (ICP), dehydration, vasospasm, and shrinkage of the ruptured vessels have been proposed as the pathophysiology²⁻¹³. In our case, this injury is not due to rupture or shrinkage of moyamoya vessels because ischemia involves bilateral regions. We consider the pathophysiology as described below. The patient presented with the ischemic type of moyamoya disease possibly because of an asymptomatic, old cerebral infarction revealed by MRI. As cerebral hemodynamics decrease with the progression of moyamoya disease, blood pressure increases to maintain cerebral hemodynamics secondary. Such secondary hypertension or essential hypertension may cause IVH due to rupture of fragile moyamoya vessels. And then, vasospasm due to IVH may have promoted the progression of cerebral infarction. In general, vasospasm is unlikely to be the etiology of the infarction in the absence of subarachnoid hemorrhage (SAH)^{2,6,7}. However, Yanaka

Table 1. Summary of recent reported cases of cerebral infarction after hemorrhage in moyamoya disease

Author (years)	Age (years)	Sex (M/F)	Day after hemorrhage	Hemorrhage type	Treatment in the acute phase	Revascularization	Outcome
Iwama et al. (2001) ²⁾	10	F	14	IVH	Conservative	Direct procedure	MD
	18	M	16	IVH	Conservative	Direct procedure	MD
	46	F	11	IVH+ICH	Conservative	-	Dead
Nakai et al. (2002) ⁷⁾	29	F	11	IVH	Conservative	-	Dead
Rafay et al. (2006) ⁹⁾	10	F	15	IVH	Conservative	Indirect procedure	MD
	11	M	7	IVH+ICH	Conservative	Indirect procedure	MD
Nagasaka et al. (2007) ⁶⁾	9	F	0	IVH+ICH	Endoscopic removal of IVH+EVD	-	MD
Suyama et al. (2008) ¹²⁾	7	F	16	IVH+ICH	Conservative	Direct+indirect procedure	MD
	9	F	7	IVH+ICH	Conservative	Direct+indirect procedure	MD
Su et al. (2008) ¹¹⁾	29	F	15	IVH	Conservative	Direct procedure	MD
Kim et al. (2001) ³⁾	41	M	13	IVH+ICH	ND	ND	ND
	52	M	6	IVH+ICH	Conservative	-	GR
	42	F	15	IVH+ICH	ND	ND	ND
	22	F	11	IVH+ICH	Stereotactic ICH aspiration+EVD	Indirect procedure	MD
	47	F	9	IVH	Conservative	Indirect procedure	SD
	56	F	8	SAH	ND	ND	ND
Tomii et al. (2011) ¹³⁾	39	F	1	IVH+ICH	Hematoma evacuation	-	Dead
Patra et al. (2012) ⁸⁾	12	M	20	IVH	Conservative	Stenting	GR10
Shibamoto et al. (2012) ¹⁰⁾	31	M	15	ICH	Hematoma evacuation	-	MD
Our case	39	F	1	IVH	Conservative	-	PVS

EVD : external ventricular drainage, GR : good recovery, ICH : intracerebral hemorrhage, IVH : intraventricular hemorrhage, HA : hyperosmotic agent, MD : moderate disability, ND : not described, PVS : persistent vegetative state, SD : severe disability

et al.¹⁴⁾ have reported on vasospasm after IVH without SAH in patients with ruptured arteriovenous malformation and speculated that factors derived from a ventricular clot might play an important role in the pathogenesis of vasospasm. Hence, it is not surprising that IVH in moyamoya disease resulted in symptomatic vasospasms in subacute stages, because moyamoya vessels are hypertensive to some physiological conditions²⁾.

What is the best treatment? In the majority of reported cases, conservative therapy was chosen in the acute stage, and then revascularization was performed in the chronic stage. In our case, prevention of both rebleeding and progression of cerebral infarction are essential. To prevent progression of IVH, anti-platelet therapy and excessive hypertension should be avoided because of the high risk of rebleeding. Hyperosmotic agent must be used cautiously because administration may result in dehydration²⁾. The patient received intensive treatment including hypervolemia, hemodilution, keeping moderate hypertension and administration of a free radical scavenger without hyperosmotic agent because hypotension and dehydration are lethal in patients with moyamoya disease. Despite such measures, the patient showed a fulminant clinical course of cerebral infarction.

What should we have done for the patient to prevent progression of cerebral infarction? Adequate control of ICP and systemic circulation is essential for management of the hemorrhagic type of moyamoya disease because rapidly increased ICP

may decrease blood flow from collateral pathways including moyamoya vessels, resulting in cerebral infarction²⁾. Hence, Iwama et al.²⁾ have proposed that central venous pressure (CVP) and ICP should be monitored to maintain adequate cerebral perfusion and hydration. We should have used hyperosmotic agents under the CVP monitoring not only to control ICP but also to avoid dehydration. On the other hand, surgical procedures have been proposed. Iwama et al.²⁾ have proposed that ventricular drainage should be performed when increased ICP is suspected. Moreover, Nagasaka et al.⁶⁾ have proposed endoscopic removal of the intraventricular hematoma besides ventricular drainage for the prevention of vasospasm. If ICP is suspected to be elevated, we should have performed removal of the intraventricular hematoma and ventricular drainage on admission in spite of administration of hyperosmotic agents. Although there is a possibility that surgical intervention may accelerate cerebral infarction because moyamoya vessels are hypertensive to some physiological conditions.

CONCLUSION

Rapid progression of cerebral infarction after hemorrhage in moyamoya disease is rare and the best strategy for acute treatment has not yet been determined. Adequate control of ICP and prevention of vasospasm in addition to control of systemic cir-

ulation is essential for the management.

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