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

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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 fulminating 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.
dren, and intracranial hemorrhage is more common in adults\(^1\). The symptoms and course of moyamoya disease vary, ranging from no symptoms to a transient disorder, to fixed neurological deficits of slight or severe degree\(^1\).

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 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 pathophysiology\(^2\)-\(^13\). 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)\(^2\)-\(^7\). However, Yanaka

**DISCUSSION**

Moyamoya disease is classified as either the ischemic or hemorrhagic type. In general, cerebral ischemia predominates in children, and intracranial hemorrhage is more common in adults\(^1\). The symptoms and course of moyamoya disease vary, ranging from no symptoms to a transient disorder, to fixed neurological deficits of slight or severe degree\(^1\).

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 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.

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et al.\(^6\) 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 conditions\(^6\).

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 dehydration\(^1\). 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 infarction\(^7\). 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-
culation is essential for the management.

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