

Acute cerebral ischemia following intraventricular hemorrhage in moyamoya disease: early perfusion computed tomography findings

Case report

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The authors present a rare case of an infarction complication 15 days following acute intraventricular bleeding due to moyamoya disease. Before the infarction occurred, perfusion CT imaging disclosed early but reversible ischemic injury on the day of hemorrhage. Dehydration and hypotension are both possibly contributing factors of progressive injury from reversible ischemia due to infarction. Although the patient underwent successful bypass surgery, 1 month after the ictus the neurobehavior evaluation still showed marked executive dysfunction. The authors address that, in hemorrhagic-type moyamoya disease, early perfusion CT scanning is not only a powerful tool to identify the high-risk group of patients who could experience subacute infarction, but also alarms neurosurgeons to eliminate any predisposing factors when it shows reversible ischemic injuries. (DOI: 10.3171.JNS.2008.109.12.1049)

KEY WORDS • cerebral infarction • intraventricular hemorrhage • moyamoya disease • perfusion computed tomography

MOYAMOYA disease is characterized by idiopathic stenosis of bilateral supraclinoid ICAs and formation of collateral arteries, known as moyamoya vessels. The disease is classified as 1 of 2 types: infarction or hemorrhagic. The former has been reported to be frequently encountered in the pediatric or juvenile population, whereas the latter seems to be more prevalent in adults.^{4,11} Generally speaking, the prognosis of hemorrhagic-type is poorer than infarction-type moyamoya disease.

Infarction following the acute presentation of hemorrhagic-type moyamoya disease has been rarely reported; we identified 7 cases after reviewing the literature. Various mechanisms have been proposed, but they remain controversial.^{5–7,9} Herein, we report on a patient with moyamoya disease who developed cerebral infarction 15 days after suffering a hemorrhage. Before this irreversible injury, ev-

idence of reversible cerebral ischemia had already been noted on perfusion CT scanning on the day of bleeding. Given that early radiological detection of this ischemic injury following IVH in moyamoya disease has never been described, the usefulness of perfusion CT is especially addressed.

Case Report

History and Examination. This 29-year-old woman presented to our hospital with sudden headache and decreased level of consciousness. There was no motor weakness. Routine laboratory tests revealed normal findings. An initial CT scan revealed primary IVH, mainly in the left lateral ventricle (Fig. 1A). No evidence of cerebral infarction was observed. Perfusion CT scanning revealed increased CBV in bilateral frontal cortices. The CBF was markedly reduced, and the MTT was abnormally prolonged in these regions (Fig. 1B–D). Cerebral angiography demonstrated occlusion of the bilateral supraclinoid ICA and the presence of a rich but tortuous

Abbreviations used in this paper: CBF = cerebral blood flow; CBV = cerebral blood volume; ICA = internal carotid artery; ICP = intracranial pressure; IVH = intraventricular hemorrhage; MTT = mean transit time.

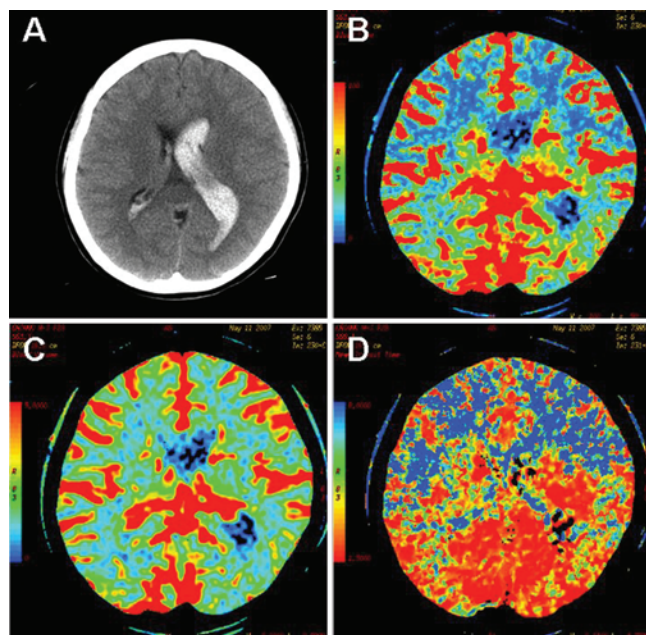


FIG. 1. A: Admission CT scan showing primary IVH. B–D: Perfusion CT scans obtained on the day of bleeding, revealing decreased CBF (B), increased CBV (C), and prolonged MTT (D) in the bifrontal areas.

vascular network in the lenticulostratial regions, a typical pattern of moyamoya disease. The anterior and middle cerebral arteries were not demonstrated bilaterally.

Treatment. After admission, the patient underwent conservative treatment with osmotic diuretic and isotonic solutions. Her blood pressure was $\sim 100/70$ mm Hg, and her urine output was adequate. However, her oral intake was inadequate due to poor appetite and nausea. On postictal Day 12, the patient fainted suddenly while walking around. Repeated head CT scanning revealed neither a new hemorrhage nor an ischemic lesion. On Day 15, the patient became obtunded. An emergency head CT scan disclosed bilateral frontal infarction (Fig. 2C and D). The patient then received hypervolemia and induced hypertension in the intensive care unit. One month later, she underwent successful superficial temporal artery–middle cerebral artery anastomosis on the left side. Aspirin and Persantin were prescribed after the operation.

Posttreatment Course. One week after the bypass surgery, the patient's higher cortical functions were evaluated by a neuropsychological test battery.^{2,3,8,12} The test results showed a moderate level of spontaneity and logopenia, whereas it revealed a remarkable executive dysfunction and impairments of verbal episodic memory.

Discussion

Infarction Complications Following Bleeding From Moyamoya Disease

Infarction complications secondary to hemorrhagic-type moyamoya are rarely reported, and their pathogenesis are largely undetermined. Only 7 cases have been described

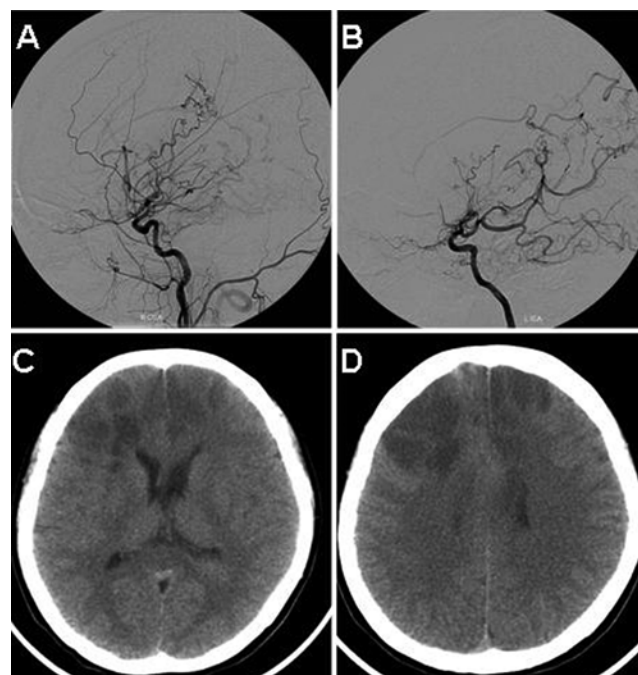


FIG. 2. Left (A) and right (B) carotid artery angiograms showing occlusion of supraclinoid segments of ICA and their bifurcations. Moyamoya vessels were prominent around the base of the brain. Computed tomography scans (C and D) obtained 15 days after acute bleeding, showing cerebral infarction at the bifrontal regions.

in the English-language literature.^{5–7,9} Summarizing all these cases, we found that infarction mainly occurs during the subacute stage after bleeding, typically between postictal Days 7 and 16. Dehydration,⁵ unrelieved increased ICP,⁹ shrinkage of the ruptured vessel,⁶ and vasospasm⁷ are all proposed contributing factors of infarction complications. In our patient, bilateral frontal infarction developed 15 days after IVH, and the proposed precipitating factors included dehydration and borderline hypotension. Therefore, a powerful tool that helps define the high-risk group of infarction complication becomes essential.

Usefulness of Perfusion CT Scanning in Predicting the High-Risk Group for Infarction Complications

Perfusion CT scanning provides a rapid and noninvasive survey of cerebral hemodynamics. It produces a color mapping of CBV, CBF, and MTT, and it is applied in the evaluation of various disease entities. For instance, this tool is commonly used to evaluate the evolution of acute ischemic stroke.^{10,13} Three distinct patterns have been described. The first consists of a cerebral hypoperfusion without true ischemia. In response to autoregulatory effects, perfusion CT scanning usually shows a prolonged MTT, but CBV and CBF remain undisturbed. When autoregulation is impaired, the second pattern, so-called reversible ischemia, occurs. At this stage, prolonged MTT is associated with a reduction in CBF. The CBV remains normal or slightly increased. As autoregulation is no longer maintained, irreversible ischemia develops, which is now characterized by an increased MTT in association with decreased CBF and CBV.

Early perfusion CT findings in hemorrhagic-type moyamoya disease

Some authors have suggested that, in hemorrhagic-type moyamoya, the only hemodynamic change before bleeding is cerebral hypoperfusion without ischemic injury. It corresponds to the first pattern of acute ischemic stroke. For patients with preexisting cerebral hypoperfusion, increased ICP resulting from acute bleeding may lead to development of reversible ischemia, the second pattern of ischemic stroke. Perfusion CT scanning in our patient demonstrated this pattern during the acute stage of IVH. This injury is due neither to vasospasm because of the acute onset,^{1,10} nor to shrinkage of the ruptured moyamoya vessel because ischemia involves bifrontal regions. Instead, this likely reflects the effects of generalized edema and early impairment of oxidative metabolism after IVH.^{1,10}

Without paying attention to this early ischemic insult, further dehydration, unrelieved increased ICP, or even vasospasm during the subacute stage will possibly lead to irreversible ischemia and subsequent infarction, the third pattern of ischemic stroke. In the 8 reported cases, including ours, the authors of 7 reported infarction complications during the subacute stage of bleeding. We highly suspect that, before infarction develops, all these patients may already have early but reversible ischemia immediately after acute IVH.

Herein, we have proposed that the susceptibility to further ischemic injury in hemorrhagic-type moyamoya can be determined by perfusion CT scanning in the early stage of bleeding. When perfusion CT scanning shows reversible ischemia, it alarms neurosurgeons to eliminate any predisposing factors. Aggressive control of increased ICP and adequate hydration are mandatory to prevent the infarction complications following hemorrhage in patients with moyamoya disease, especially when early but reversible ischemic pattern is documented by perfusion CT scanning at the acute stage of bleeding.

Conclusions

Ischemic injury following IVH is rare, but patients at high risk for subacute infarction can be predicted using perfusion CT scanning in the acute stage of bleeding. These patients may further suffer from neurobehavioral disturbances, which probably depend on lesion locations. Elimination of any possible unfavorable factors, such as increased ICP and dehydration, is essential to prevent infarction complications following hemorrhage in moyamoya disease, especially for those with radiological evidence of acute but reversible ischemic injury.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Manuscript submitted August 22, 2007.

Accepted December 17, 2007.

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