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Case Report

Extremely Rare Breakdown of the Moyamoya Vessels Resulting in Intraventricular Hemorrhage after Direct Bypass Surgery in a Pediatric Patient with Moyamoya Disease

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Revascularization surgery is established for both ischemic and hemorrhagic moyamoya disease (MMD), although hemorrhagic complication is a serious problem especially in adult MMD patients showing postoperative hyperperfusion. Herein, we present an extremely rare case with pediatric MMD showing intraventricular hemorrhage the day following direct bypass surgery, possibly due to a breakdown of the terminal branch of well-developed perforators working as moyamoya vessels. Clinicians should consider this rare complication after bypass surgery for MMD regardless of preoperative hemodynamics or patient age.

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Key words: moyamoya disease; pediatric; direct bypass; intracranial hemorrhage

Introduction

The majority of cases of pediatric moyamoya disease (MMD) present with ischemic symptoms, while approximately 50% of adult MMD cases present with intracranial hemorrhage due to long-term hemodynamic stress to the collateral moyamoya vessels. The Japan Adult Moyamoya Trial has provided recent evidence that direct bypass surgery has a preventive effect against rebleeding in adult MMD patients showing intracranial hemorrhage over a 5-year follow-up [1]. Moreover, direct bypass surgery has been reported to obliterate peripheral artery aneurysms by decreasing hemodynamic stress [2]. Herein, we present an extremely rare case of pediatric MMD showing postoperative hemorrhage due to a breakdown of the moyamoya vessels on the day following bypass surgery.

Case description

A 9-year-old healthy female presented with bilateral transient hemiparesis. Head magnetic resonance (MR) imaging and digital subtraction angiography (DSA) revealed stenosis of the terminal portion of the bilateral internal carotid arteries, proximal portion of the anterior cerebral arteries and middle cerebral arteries with well-developed moyamoya vessels, fulfilling the diagnostic criteria of MMD [3] (Fig. 1a, b). No cerebral microbleeds were detected in T2* weighted MR imaging, and no microaneurysms were detected with DSA. Hemodynamic assessment using ^{123}I-IMP single photon emission computed tomography (SPECT) revealed a decrease in cerebral blood flow (CBF) in the right anterior cerebral artery and middle cerebral artery territory (Fig. 1c), although

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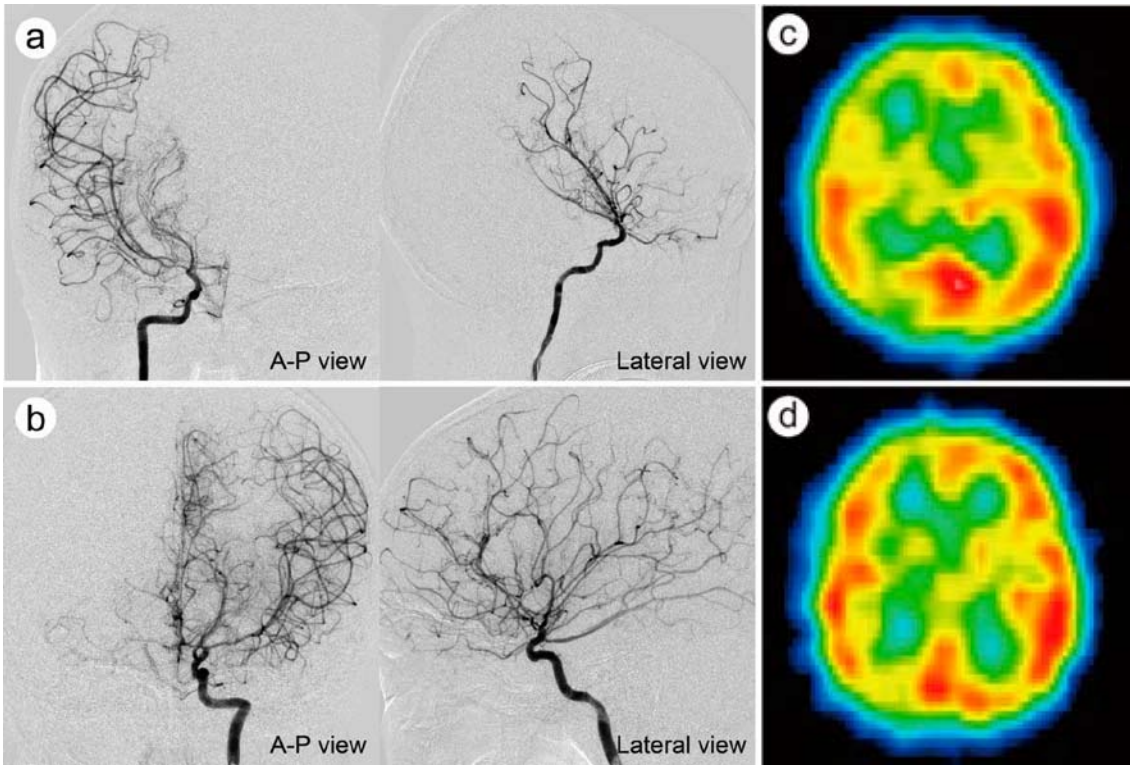

Figure 1. Digital subtraction angiography demonstrates stenosis of the terminal position of the internal carotid artery, proximal position of the anterior cerebral artery and middle cerebral artery with well-developed moyamoya vessels (a: right, b: left). Initial hemodynamic assessment using single photon emission computed tomography shows a decrease in cerebral blood flow in the right anterior middle cerebral artery territory (c), which improved at 4 months after right bypass surgery (d). A-P: anterior-posterior.

acetazolamide challenge was not performed because of her unstable symptoms. The patient underwent volume loading without antithrombotic agent and her symptoms improved. One month later, direct/indirect combined bypass surgery (superior temporal artery-middle cerebral artery anastomosis with encephaloduromyosynangiosis) was initially performed in the right hemisphere, and improvement of CBF was confirmed at one month after surgery (Fig. 1d). The same surgery was performed for the left hemisphere at 4 months after the first surgery. In direct bypass, one of M4 segment of middle cerebral artery in frontal cortex was selected as the recipient. Her postoperative course was uneventful and patency of direct anastomosis was verified by MR angiography, and she showed no hyperperfusion on SPECT. Nevertheless, computed tomography and T2* MR imaging surprisingly detected asymptomatic intraventricular hemorrhage in the ipsilateral hemisphere the day following surgery (Fig. 2a, b). Blood pressure lowering (a systolic blood pressure between 80 and 120 mm Hg) was performed by continuous intravenous infusion of nicardipine hydrochloride, if necessary. She was discharged without any symptoms at 9 days after the surgery. Postoperative DSA was not performed because of patient refusal, although no aneurysmal formation was detected on follow-up MR angiography.

Discussion

Bypass surgery is an established treatment for both ischemic and hemorrhagic MMD with compensating cerebral blood flow, resulting in reduced hemodynamic stress to the moyamoya vessels in the long term [1, 4]. However, perioperative complications have been reported including hyperperfusion and watershed shift infarction, which can affect morbidity and mortality [5, 6]. MMD specific vasculature can contribute to these complications with a rapid increase in blood flow until cerebral hemodynamics reach stabilization. As such, careful perioperative management is mandatory [7].

Herein, we present a pediatric MMD case showing postoperative breakdown of the moyamoya vessels causing intraventricular hemorrhage. Epidemiologically, it is difficult to explain this hemorrhagic event as a natural course of pediatric MMD [8]. Microangiographical analyses demonstrated a clear anastomosis between the peripheral branches

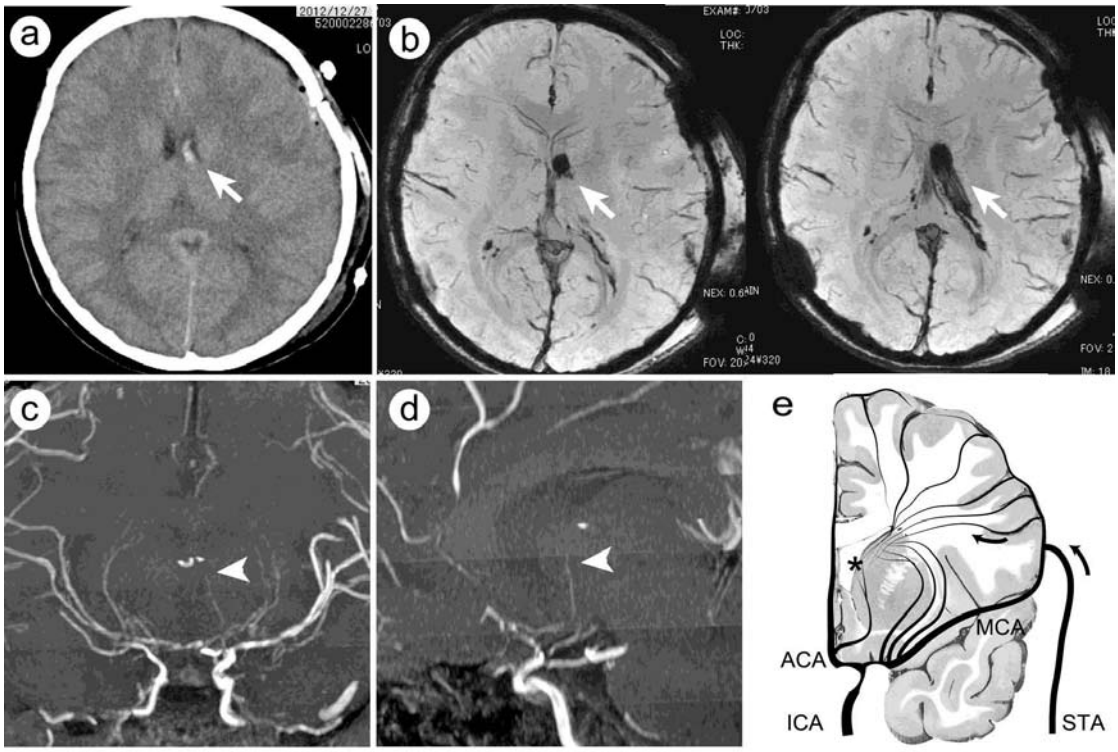

Figure 2. Postoperative evaluations. Postoperative computed tomography (a) and T2* magnetic resonance (MR) images (b) showing intraventricular hemorrhage (arrow) in the left lateral ventricle. Multiplanar reconstruction images of the time-of-flight MR angiography reveal a well-developed left Heubner artery extending to the anterior horn of the lateral ventricle, where the hemorrhage occurred (arrow head, c: coronal view, d: sagittal view). The schema shows the mechanism of intraventricular hemorrhage after bypass surgery (e). Asterisk indicates the estimated point of hemorrhage. ACA: anterior cerebral artery, ICA: internal carotid artery, MCA: middle cerebral artery, STA: superficial temporal artery.

of the perforating arteries and medullary branches from the cortical arteries, which may explain the compensatory mechanism of cerebral blood flow in pediatric MMD [9]. Retrospectively, we confirmed a well-developed left Heubner artery extending to the anterior horn of the lateral ventricle, where the hemorrhage occurred (Fig. 2c, d). We believe that a rapid but tiny increase in local blood flow from the direct bypass that couldn't be detected by hemodynamic assessment contributed to the peripheral moyamoya vessel breakdown through the medullary branches originating from the cortical arteries in this pediatric MMD case (Fig. 2e). In adult MMD, hemorrhagic presentation as a natural course results from persistent hemodynamic stress of the moyamoya vessels and occurs in the basal ganglia, thalamus or periventricular region [4]. Moreover, peripheral aneurysms in the collateral vessels or moyamoya vessels can be identified on cerebral angiography in some adult MMD patients [10, 11]. In an extensive review of adult MMD cases showing intraventricular hemorrhage, Irikura et al. reported more frequent occurrence of marked enlargement of the choroidal arteries and the derived medullary arteries [12]. These findings suggest that the hemodynamic load in the vessels supplying the walls of the ventricles and the periventricular region can affect hemorrhagic presentation in adult MMD. A similar mechanism may contribute to the intraventricular hemorrhage following bypass surgery in our pediatric MMD. In a review of postoperative complications in pediatric MMD patients after revascularization surgery, hemorrhagic complications occurred in 1.5% of cases after indirect bypass surgery, while no cases were reported after direct bypass surgery [13]. We found only one pediatric patient who developed hemorrhage in ipsilateral basal ganglia after direct revascularization surgery in the report [14]. Therefore, to our knowledge this is the second reported case showing postoperative hemorrhage due to breakdown of the moyamoya vessels, potentially due to direct surgery, in pediatric MMD.

Cerebral microbleeds are occasionally detected in MMD, indicating minor rupture of the moyamoya vessels, which were suggested to be a potential risk factor for hemorrhage following direct bypass surgery in patients with MMD [15]. Although, cerebral microbleeds were not detected in the present case, our findings provide support for this hypothesis that direct bypass can affect the fragile moyamoya vessels even in pediatric patients. Therefore, clinicians should consider

this complication, and provide mandatory strict control of the blood pressure after bypass surgery for MMD regardless of preoperative hemodynamics or patient age.

References

1. Miyamoto S, Yoshimoto T, Hashimoto N et al. Effects of extracranial-intracranial bypass for patients with hemorrhagic moyamoya disease: results of the Japan Adult Moyamoya Trial. *Stroke* 45:1415-1421, 2014
2. Kuroda S, Houkin K, Kamiyama H, Abe H. Effects of surgical revascularization on peripheral artery aneurysms in moyamoya disease: report of three cases. *Neurosurgery* 49:463-467; discussion 467-468, 2001
3. Research Committee on the P, Treatment of Spontaneous Occlusion of the Circle of W, Health Labour Sciences Research Grant for Research on Measures for Infractable D. Guidelines for diagnosis and treatment of moyamoya disease (spontaneous occlusion of the circle of Willis). *Neurol Med Chir (Tokyo)* 52:245-266, 2012
4. Kuroda S, Houkin K. Moyamoya disease: current concepts and future perspectives. *Lancet Neurol* 7:1056-1066, 2008
5. Fujimura M, Shimizu H, Inoue T, Mugikura S, Saito A, Tominaga T. Significance of focal cerebral hyperperfusion as a cause of transient neurologic deterioration after extracranial-intracranial bypass for moyamoya disease: comparative study with non-moyamoya patients using N-isopropyl-p-[(123)I]iodoamphetamine single-photon emission computed tomography. *Neurosurgery* 68:957-964; discussion 964-965, 2011
6. Hayashi T, Shirane R, Fujimura M, Tominaga T. Postoperative neurological deterioration in pediatric moyamoya disease: watershed shift and hyperperfusion. *J Neurosurg Pediatrics* 6:73-81, 2010
7. Fujimura M, Inoue T, Shimizu H, Saito A, Mugikura S, Tominaga T. Efficacy of prophylactic blood pressure lowering according to a standardized postoperative management protocol to prevent symptomatic cerebral hyperperfusion after direct revascularization surgery for moyamoya disease. *Cerebrovasc Dis* 33:436-445, 2012
8. Suyama K, Yoshida K, Takahata H et al. Pediatric moyamoya disease presenting with intracerebral hemorrhage--report of three cases and review of the literature. *Clin Neurol Neurosurg* 110:270-275, 2008
9. Kodama N, Suzuki J. Cerebrovascular Moyamoya disease. IIIrd report--the study on the aging of the perforating branches and the possibility of collateral pathway. *Neurol Med Chir (Tokyo)* 14 pt 1:55-67, 1974
10. Iwama T, Morimoto M, Hashimoto N, Goto Y, Todaka T, Sawada M. Mechanism of intracranial rebleeding in moyamoya disease. *Clin Neurol Neurosurg* 99 Suppl 2:S187-190, 1997
11. Sadatoh A, Yonekawa Y, Morooka Y, Imakita T. [A case of moyamoya disease with repeated intraventricular hemorrhage due to a ruptured pseudoaneurysm]. *No Shinkei Geka* 17:755-758, 1989
12. Irikura K, Miyasaka Y, Kurata A et al. A source of haemorrhage in adult patients with moyamoya disease: the significance of tributaries from the choroidal artery. *Acta Neurochir* 138:1282-1286, 1996
13. Fung LW, Thompson D, Ganesan V. Revascularisation surgery for paediatric moyamoya: a review of the literature. *Childs Nerv Syst* 21:358-364, 2005
14. Guzman R, Lee M, Achrol A et al. Clinical outcome after 450 revascularization prosedures for moyamoya disease. *J Neurosurg* 111: 927-935, 2009
15. Kikuta K, Takagi Y, Nozaki K et al. Asymptomatic microbleeds in moyamoya disease: T2*-weighted gradient-echo magnetic resonance imaging study. *J Neurosurg* 102:470-475, 2005