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Case Report

Moyamoya Disease Emerged with Corpus Callosum Hemorrhage: A 3D Computer Graphic Analysis

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The authors present a rare case of moyamoya disease emerged with corpus callosum hemorrhage. A 31-year-old woman suddenly complained of severe headache followed by consciousness disturbance. Radiological examinations revealed the bleeding in the splenium of corpus callosum, which was associated with intraventricular hemorrhage. On cerebral angiography, the carotid fork was severely stenotic on both sides, and a marked dilatation was observed in the anterior/posterior choroidal arteries and posterior pericallosal artery as well as the lenticulostriate arteries. Therefore, she was diagnosed as moyamoya disease. She successfully underwent superficial temporal artery to middle cerebral artery (STA–MCA) anastomosis and indirect bypass on both sides. Postoperative course was uneventful. Follow-up cerebral angiography performed 4 months after surgery showed well-developed surgical collaterals via the external carotid system and a marked decrease of the dilated moyamoya vessels. She has been free from any cerebrovascular events for 36 months after surgery. Radiological findings strongly suggest that splenial bleeding occurred due to the rupture of the dilated abnormal collateral vessels that originate from the medial posterior choroidal artery and penetrate the corpus callosum in this case. Three-dimensional computer graphic analysis was useful to determine the complex collateral circulation in moyamoya disease.

Keywords: moyamoya disease, corpus callosum, hemorrhage, bypass surgery

Introduction

Moyamoya disease is characterized by progressive occlusion of the supraclinoid internal carotid artery (ICA) and its main branches within the circle of Willis. This occlusion results in the formation of a fine vascular network (the moyamoya disease) which functions as collateral pathways.[1,2] Although most of pediatric patients with moyamoya disease develop transient ischemic attack or ischemic stroke, about half of adult patients emerge with intracranial hemorrhage. Susceptible age for hemorrhagic stroke is thirties to forties. Intracranial bleeding is known to occur from the fragile, dilated moyamoya vessels or from the saccular aneurysm within the circle of Willis. Hemodynamic stress in the collateral routes is playing a key role in both types of hemorrhagic stroke in moyamoya disease. The dilated moyamoya vessels readily cause the bleeding in the basal ganglia or subventricular white matter.[3,4] In this report, we present a case of adult moyamoya disease that presented with hemorrhage in the splenium of corpus callosum. Callosal bleeding is quite rare in moyamoya disease. More importantly, there are almost no studies that denote the origin of bleeding in the corpus callosum, probably because of the complicated anatomical course of moyamoya vessels around the splenium of corpus callosum as well as a small number of cases with moyamoya disease-related callosal bleeding.

Case Report

A 31-year-old woman suddenly complained of severe headache followed by consciousness disturbance, and was admitted to an emergent hospital. She had frequently complained of headache and weakness of the extremities in her childhood, but she was not referred to any hospitals. She had no family history of moyamoya disease. Neurological examinations on admission showed severe consciousness disturbance (III-100 on Japan Coma Scale). Plain CT scan revealed the bleeding in the splenium of corpus callosum, which was associated with intraventricular hemorrhage (Fig. 1A). She underwent emergent ventricular drainage on the right sides. Two months later, she completely recovered, and she was transferred to our hospital for further examinations and treatments. On admission, her neurological status was intact. T_2- and T_2*-weighted MRI clearly identified the site of hemorrhage in the splenium of corpus callosum (Fig. 1B, C). Cerebral angiography demonstrated severe stenosis of the carotid fork on both sides. Basal moyamoya vessels were well developed and anterior choroidal arteries were markedly dilated on both sides. Right external carotid angiography demonstrated spontaneously developed collaterals from the superficial temporal artery (STA) and middle meningeal artery (MMA) through previous burr hole (Fig. 2). Left vertebral angiography revealed well-developed pial anastomosis via the posterior cerebral arteries (PCAs) on both sides. In addition, the posterior medial choroidal arteries and posterior pericallosal arteries were notably developed and functioned as the collateral to the anterior cerebral arteries. Especially, extensive collateral circulation was noted in the choroid plexus of the third ventricle near the foramen of Monro through the medial posterior choroidal artery. Furthermore, the medial posterior choroidal artery gave off the abnormal collateral vessels that penetrated the corpus callosum into the anterior cerebral artery territory, although their location did not correlate with the

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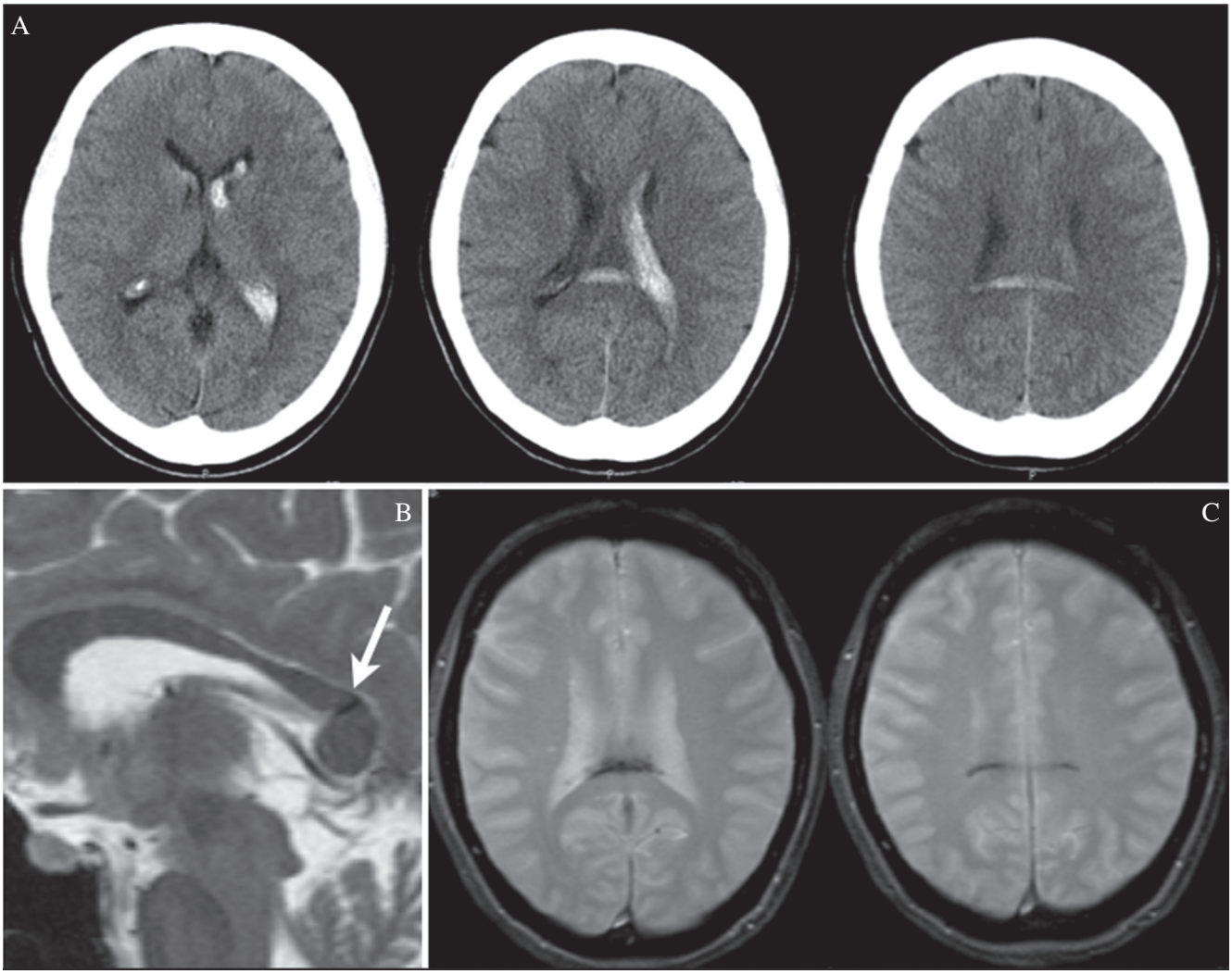

Fig. 1 Plain CT scans at the onset (A) demonstrated intracerebral hemorrhage in the splenium of corpus callosum, which is associated with intraventricular hemorrhage. T_2- (B) and T_2*-weighted MRI (C) on admission show that the bleeding occurred in the splenium of corpus callosum in this case.

site of bleeding (Fig. 3A, B). The data of MRI and time-of-flight MR angiography were employed to create three-dimensional computer graphic (3D-CG), using Amira version 5.0. As the results, 3D-CG technique clearly showed spatial relationship among the corpus callosum, the site of bleeding, and the abnormal collateral vessels (Fig. 4). ^{123}I-IMP single photon emission computed tomography (SPECT) demonstrated impaired cerebrovascular reactivity to acetazolamide in the bilateral frontal lobes. She underwent STA to middle cerebral artery (MCA) anastomosis combined with indirect bypass, encephalo-duro-myo-arterio-pericranial synangiosis (EDMAPS) on the right side, and then on the left side with an interval of 2 months.[15] Postoperative course was uneventful. Cerebrovascular reactivity completely recovered in the bilateral frontal lobes on repeated blood flow study 4 months after the second surgery. Follow-up cerebral angiography demonstrated that surgical collaterals widely covered the operated hemispheres, especially the frontal lobes. Abnormally dilated moyamoya vessels remarkably diminished on both carotid and vertebral angiograms. Abnormal collateral vessels originating from the medial posterior choroidal artery also disappeared on the left vertebral angiogram (Fig. 3C, D). On source images of MR angiography, the dilated medullary arteries from the lateral posterior choroidal arteries markedly decreased in their extent after surgery (Fig. 5). She has experienced no further episodes of ischemic and hemorrhagic stroke for 36 months after surgery.

Discussion

As aforementioned, there are two main sources of intracranial bleeding in moyamoya disease, including the dilated, fragile moyamoya vessels and the saccular aneurysms within the circle of Willis. For the former, the rupture is considered to occur due to persistent hemodynamic stress of the moyamoya vessels and occurs in the basal ganglia, thalamus, or periventricular region. Intraventricular hemorrhage is frequently complicated. Peripheral aneurysms in the

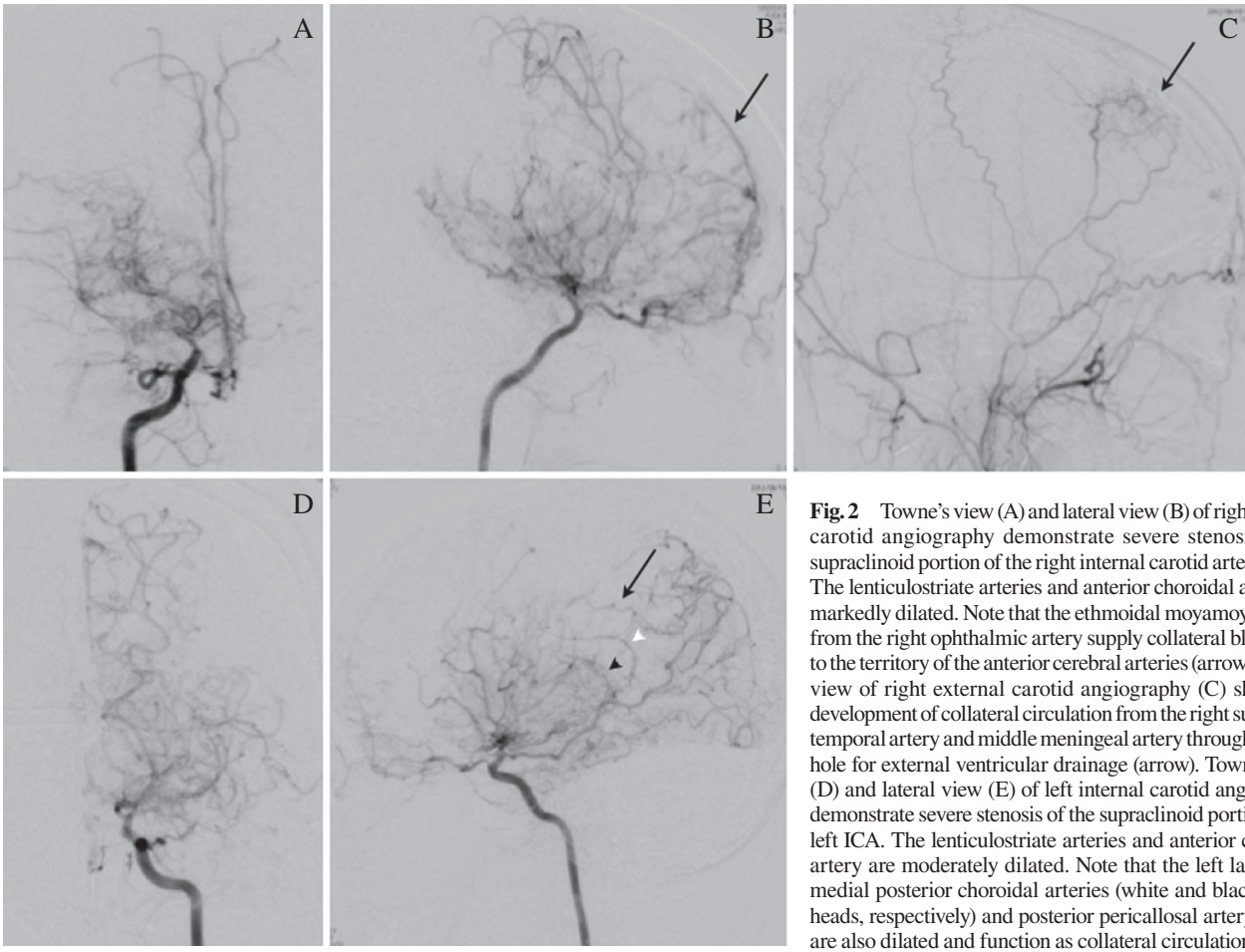

Fig. 2 Towne's view (A) and lateral view (B) of right internal carotid angiography demonstrate severe stenosis of the supraclinoid portion of the right internal carotid artery (ICA). The lenticulostriate arteries and anterior choroidal artery are markedly dilated. Note that the ethmoidal moyamoya vessels from the right ophthalmic artery supply collateral blood flow to the territory of the anterior cerebral arteries (arrow). Lateral view of right external carotid angiography (C) shows the development of collateral circulation from the right superficial temporal artery and middle meningeal artery through the burr hole for external ventricular drainage (arrow). Towne's view (D) and lateral view (E) of left internal carotid angiography demonstrate severe stenosis of the supraclinoid portion of the left ICA. The lenticulostriate arteries and anterior choroidal artery are moderately dilated. Note that the left lateral and medial posterior choroidal arteries (white and black arrowheads, respectively) and posterior pericallosal artery (arrow) are also dilated and function as collateral circulation.

moyamoya vessels might be identified on cerebral angiography in some patients.[1,3,4] Therefore, intracerebral hemorrhage in the splenium of corpus callosum is considered quite rare. To our best knowledge, there are only four reports that presented the cases that developed bleeding in the splenium of corpus callosum. Thus, Muraki et al. (1981) reported nine adult patients who experienced hemorrhagic stroke due to moyamoya disease. Of these, a 53-year-old female case presented with intracerebral hemorrhage in the corpus callosum, but no precise information was documented.[5] Miyamoto et al. (1984) reported a 55-year-old female case that repeated the bleeding in the corpus callosum due to unilateral moyamoya disease. The first bleeding occurred in the body of corpus callosum, but the second occurred in the genu. The authors concluded that these bleedings resulted from the rupture of the beads-like dilated arteries along the pericallosal artery in the non-involved side, which was considered as the collateral channels.[6] Irikura et al. (1996) reported 10 cases with hemorrhagic stroke due to moyamoya disease. Of these, two cases experienced splenial bleeding. They emphasized that the medullary arteries from the anterior and posterior lateral choroidal arteries showed a marked and irregular dilatation, but did not describe the etiology of splenial bleeding.[7] Very recently, Nah et al. (2012) analyzed the location of intracerebral hemorrhage in 93 patients with moyamoya disease. As the results, 4 (4.3%) of 93 patients presented the bleeding in the corpus callosum. In this report, plain CT scan showed that only one of these four patients developed the bleeding in the splenium of corpus callosum, but no other information on their demographic and radiological findings was described.[8] Therefore, totally eight patients have been reported to develop the bleeding in the corpus callosum due to moyamoya disease. Bleeding occurred in the body and genu in one patient, and occurred in the splenium in three. The bleeding site was not documented in the remaining four patients.

The present case experienced the bleeding in the splenium of corpus callosum. Cerebral angiography clearly showed that the collateral pathways from the vertebrobasilar system, that is, the posterior choroidal arteries and posterior pericallosal artery, were well developed on cerebral angiography. The medial posterior choroidal artery originates from the ambient portion of the PCA (P2 segment), encircles the midbrain medial to the PCA toward the pineal gland, and supplies the blood flow to the choroid plexus of the third ventricle. The lateral posterior choroidal artery originates from the quadrigeminal portion of the PCA (P3 segment) and passes laterally through the choroidal fissure and around the thalamus to

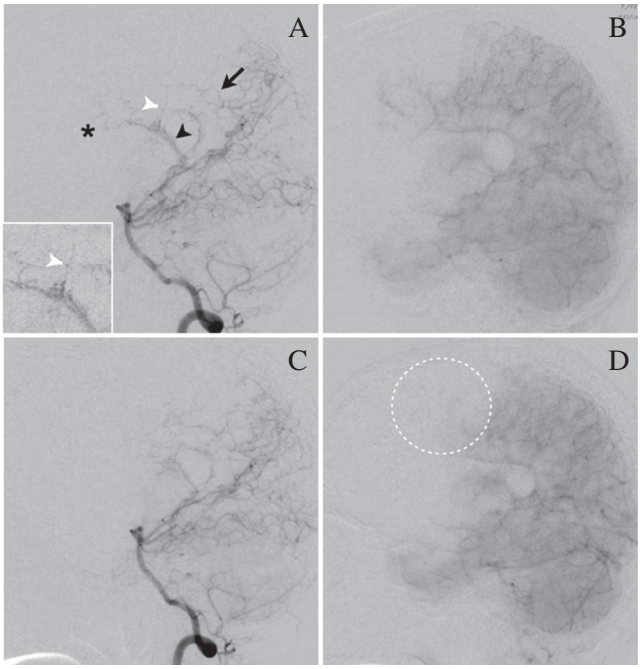

Fig. 3 Lateral views of pre (A, B) and postoperative left vertebral angiography (C, D). Panels A and C represent early arterial phase, and Panels B and D represent capillary phase. The right medial posterior choroidal artery (arrowheads) and posterior pericallosal artery (arrow) are markedly dilated before surgery (A), but diminish at 4 months after surgery (C). Note that the abnormal collateral vessels (A, white arrowhead) originate from the medial posterior choroidal artery and penetrate the corpus callosum to probably provide blood flow to the anterior cerebral artery territory. In addition, extensive collateral circulation is noted in the choroid plexus of the third ventricle near the foramen of Monro through the medial posterior choroidal artery (A, asterisk). The collateral pathways towards the territory of the anterior cerebral arteries decrease in their extent after surgery (D, dotted area).

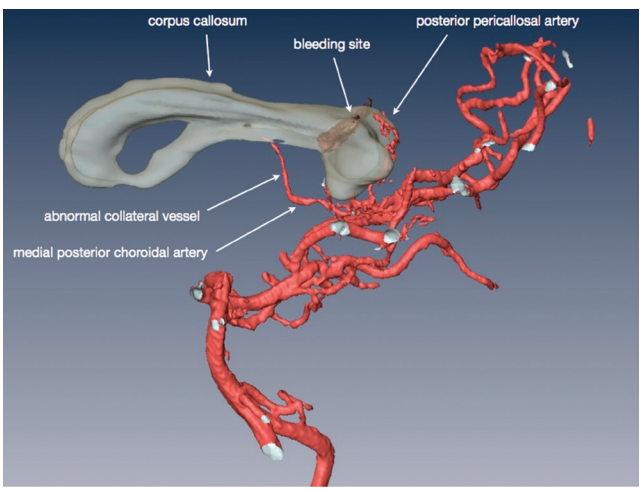

Fig. 4 Three-dimensional computer graphic shows a spatial relationship among the corpus callosum, the site of bleeding, and abnormal collateral vessels.

enter the choroid plexus of the lateral ventricle. On the other hands, the posterior pericallosal artery is the primary artery responsible for the splenium supply. According to recent anatomical study, the posterior pericallosal artery mainly

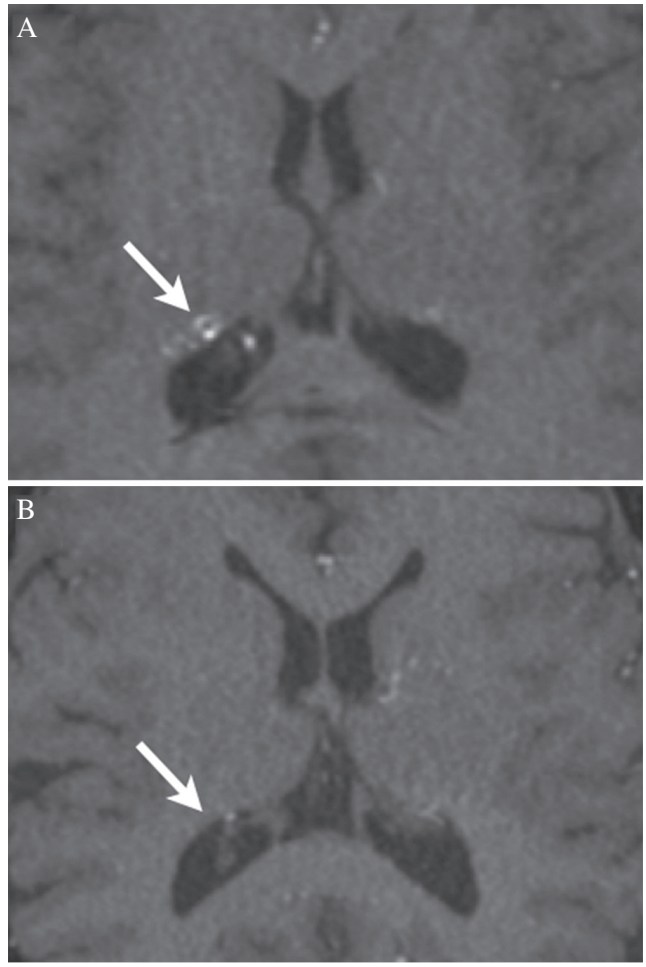

Fig. 5 Source images of time-of-flight MR angiography before (A) and after surgery (B) demonstrate that the abnormally dilated medullary arteries in the subependymal region of the right trigon clearly decrease in the extent after surgery (arrows).

originates from the parieto-occipital artery (28/53; 53%) or the P3 segment (12/53; 23%).[9] Considering radiological findings in the present case, the bleeding most likely occurred due to the rupture of the abnormal collateral vessels that originated from the medial posterior choroidal artery and penetrated through the corpus callosum (Fig. 3A). On 3D-CG, the abnormal collateral vessels did not correlate with the site of bleeding, probably because similar vessels at the site of bleeding became invisible due to thrombosis or collapse after bleeding (Fig. 4). This is the first report that may possibly indicate the etiology of bleeding in the splenium of corpus callosum in patients with moyamoya disease. In fact, previous studies have strongly suggested the causative role of these collateral pathways in the development of hemorrhagic stroke in adult moyamoya disease. Irikura et al. (1996) analyzed the findings on cerebral angiography in patients with moyamoya disease, and concluded that the collateral circulation via the choroidal and posterior pericallosal arteries was more prominent in the hemorrhagic group than in ischemic group.[7] Subsequently, similar results have been reported.[8,10]

Surgical revascularization is the most successful therapy to improve cerebral hemodynamics, and to reduce the risk of

subsequent stroke.[1] Surgical procedures for indirect bypass are specific for moyamoya disease.[11,12] However, the beneficial effects are not immediate because surgical collaterals require 3–4 months to develop. More importantly, collateral pathways through indirect bypass do not develop in about 40–50% of adult patients.[13] On the other hands, direct bypass procedures mainly include STA–MCA anastomosis and is quite useful to improve cerebral hemodynamics and to resolve ischemic attacks immediately after surgery.[14] The present case successfully underwent STA–MCA anastomosis combined with EDMAPS on both sides.[15] Recent randomized clinical trial, Japan Adult Moyamoya (JAM) trial, clearly showed that direct or combined bypass surgery significantly reduces the incidence of repeated hemorrhagic stroke in adult patients with hemorrhagic type moyamoya disease during a 5-year follow-up period.[3,4,16] More importantly, recent subgroup analysis of JAM Trial has demonstrated that patients with posterior hemorrhage due to the perforating arteries from the PCA and choroidal artery are at higher risk of rebleeding and accrue greater benefit from surgery than those with anterior hemorrhage due to the perforating arteries from the anterior and middle cerebral arteries.[17] Indeed, abnormal collateral circulation markedly diminished after surgery on cerebral angiography (Fig. 3) and the source images of MRA (Fig. 5).

Conclusion

We here report a rare case of adult moyamoya disease emerged with intracerebral hemorrhage in the splenium of corpus callosum probably due to the rupture of the abnormally dilated branches from the medial posterior choroidal artery. A 3D-CG technique would be useful to understand complex anatomy of collateral circulation in moyamoya disease. Direct or combined bypass surgery would be useful to prevent rebleeding in such adult patients with moyamoya disease.

Conflicts of Interest Disclosure

The authors have no conflict of interest or any financial disclosures. All authors who are members of Japan Neurosurgical Society (JNS) have registered online self-reported COI Disclosures Statement Forms through the website for JNS members.

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References

1) Kuroda S, Houkin K: Moyamoya disease: current concepts and future perspectives. *Lancet Neurol* 7: 1056–1066, 2008
2) Suzuki J, Takaku A: Cerebrovascular "moyamoya" disease: disease showing abnormal net-like vessels in base of brain. *Arch Neurol* 20: 288–299, 1969
3) Miyamoto S, Yoshimoto T, Hashimoto N, Okada Y, Tsuji I, Tominaga T, Nakagawara J, Takahashi JC, Investigators JAMT: Effects of extracranial–intracranial bypass for patients with hemorrhagic moyamoya disease: results of the Japan adult moyamoya trial. *Stroke* 45: 1415–1421, 2014
4) Yoshida Y, Yoshimoto T, Shirane R, Sakurai Y: Clinical course, surgical management, and long-term outcome of moyamoya patients with rebleeding after an episode of intracerebral hemorrhage: an extensive follow-up study. *Stroke* 30: 2272–2276, 1999
5) Muraki M, Kaneko M, Yamamoto T, Iwamoto K, Uemura K, Nakajima S: A clinical study of 9 cases of adult type of moyamoya disease associated with intracerebral hematoma. Proceedings of 10th annual meeting of surgery for cerebral stroke 10: 59–63, 1982
6) Miyamoto Y, Shiino A, Handa J: Recurrent callosal hematoma with a typical moyamoya disease. *Nippon Geka Hokan* 53: 667–671, 1984
7) Irikura K, Miyasaka Y, Kurata A, Tanaka R, Fujii K, Yada K, Kan S: A source of haemorrhage in adult patients with moyamoya disease: the significance of tributaries from the choroidal artery. *Acta Neurochir* (*Wien*) 138: 1282–1286, 1996
8) Nah HW, Kwon SU, Kang DW, Ahn JS, Kwun BD, Kim JS: Moyamoya disease-related versus primary intracerebral hemorrhage: [corrected] location and outcomes are different. *Stroke* 43: 1947–1950, 2012
9) Kahilogullari G, Comert A, Ozdemir M, Brohi RA, Ozgural O, Esmer AF, Egemen N, Karahan ST: Arterial vascularization patterns of the splenium: An anatomical study. *Clin Anat* 26: 675–681, 2013
10) Morioka M, Hamada J, Kawano T, Todaka T, Yano S, Kai Y, Ushio Y: Angiographic dilatation and branch extension of the anterior choroidal and posterior communicating arteries are predictors of hemorrhage in adult moyamoya patients. *Stroke* 34: 90–95, 2003
11) Matsushima Y, Fukai N, Tanaka K, Tsuruoka S, Inaba Y, Aoyagi M, Ohno K: A new surgical treatment of moyamoya disease in children: a preliminary report. *Surg Neurol* 15: 313–320, 1981
12) Scott RM, Smith JL, Robertson RL, Madsen JR, Soriano SG, Rockoff MA: Long-term outcome in children with moyamoya syndrome after cranial revascularization by pial synangiosis. *J Neurosurg* 100: 142–149, 2004
13) Mizoi K, Kayama T, Yoshimoto T, Nagamine Y: Indirect revascularization for moyamoya disease: is there a beneficial effect for adult patients? *Surg Neurol* 45: 541–548; discussion 548–549, 1996
14) Ishikawa T, Houkin K, Kamiyama H, Abe H: Effects of surgical revascularization on outcome of patients with pediatric moyamoya disease. *Stroke* 28: 1170–1173, 1997
15) Kuroda S, Houkin K, Ishikawa T, Nakayama N, Iwasaki Y: Novel bypass surgery for moyamoya disease using pericranial flap: its impacts on cerebral hemodynamics and long-term outcome. *Neurosurgery* 66: 1093–1101; discussion 1101, 2010
16) Houkin K, Kamiyama H, Abe H, Takahashi A, Kuroda S: Surgical therapy for adult moyamoya disease: can surgical revascularization prevent the recurrence of intracerebral hemorrhage? *Stroke* 27: 1342–1346, 1996
17) Takahashi JC, Funaki T, Houkin K, Inoue T, Ogasawara K, Nakagawara J, Kuroda S, Yamada K, Miyamoto S, Investigators JAMT: Significance of the hemorrhagic site for recurrent bleeding: prespecified analysis in the Japan adult moyamoya trial. *Stroke* 47: 37–43, 2016

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