 doi: 10.2176/jns-nmc.2023-0195

NMC Case Report Journal 11, 13-18, 2024

Moyamoya Syndrome Associated with Late-onset Idiopathic Aqueduct Stenosis Successfully Treated with Endoscopic Third Ventriculostomy

Taishi HONDA,[1] Masaki ITO,[1] Haruto UCHINO,[1]
Taku SUGIYAMA,[1] and Miki FUJIMURA[1]

[1]*Department of Neurosurgery, Hokkaido University Hospital, Sapporo, Hokkaido, Japan*

Abstract

Moyamoya disease (MMD) is a rare idiopathic cerebrovascular disorder that causes transient ischemic attack (TIA) and ischemic stroke in the pediatric population. Herein, we report an extremely rare case of Moyamoya syndrome (MMS) and late-onset idiopathic aqueduct stenosis, a unique form of non-communicating hydrocephalus. A 17-year-old female presented with an intractable headache and occasional faintness. Pertinent medical history included a fourth ventricle epidermoid cyst without any evidence of aqueduct stenosis, which was surgically removed when she was two years of age. The patient subsequently experienced a TIA and was diagnosed with MMD at 14 years of age. Under the definitive diagnosis of MMS associated with a brain tumor, the patient underwent surgical revascularization of the symptomatic right hemisphere without complications. Although the ischemic symptoms resolved postoperatively, a medically intractable headache with occasional faintness persisted. Serial magnetic resonance imaging ultimately revealed newly developed non-communicating hydrocephalus due to acquired aqueduct stenosis at the age of 17. After careful exclusion of the development of either or both a periventricular anastomosis and vault moyamoya vessels along the surgical route using cerebral angiography, we performed an endoscopic third ventriculostomy (ETV) via the right anterior horn without complications. A complete resolution of her chronic headache with the shrinkage of the third ventriculomegaly was observed postoperatively. In cases of MMS associated with symptomatic aqueduct stenosis, transdural collaterals on the cranial vault and periventricular collaterals should be meticulously evaluated preoperatively using cerebral angiography to safely perform an ETV.

Keywords: aqueduct stenosis, hydrocephalus, Moyamoya syndrome, endoscopic third ventriculostomy

Introduction

Moyamoya syndrome (MMS) is characterized by the progressive stenosis of the terminal portion of the internal carotid arteries and their proximal branches with a compensatory development of collateral vessels at the base of the brain associated with well-recognized conditions, which can cause stroke or transient ischemic attack (TIA) even in the pediatric population.[1-4] Cranial therapeutic irradiation, Down syndrome, and autoimmune diseases are well-recognized conditions potentially showing moyamoya angiopathy (i.e., Moyamoya disease (MMD) and MMS); however, late-onset idiopathic aqueduct stenosis (LIAS), a unique form of non-communicating hydrocephalus with an idiopathic obstruction at the level of the Sylvian aqueduct, is not well-recognized as an associated condition for MMS.[5,6] Herein, we report an extremely rare case of MMS in a female adolescent complaining of a medically intractable headache due to LIAS that was successfully treated with endoscopic third ventriculostomy (ETV).

Case Report

A 17-year-old female presented with an intractable head-

Received August 21, 2023; Accepted November 9, 2023

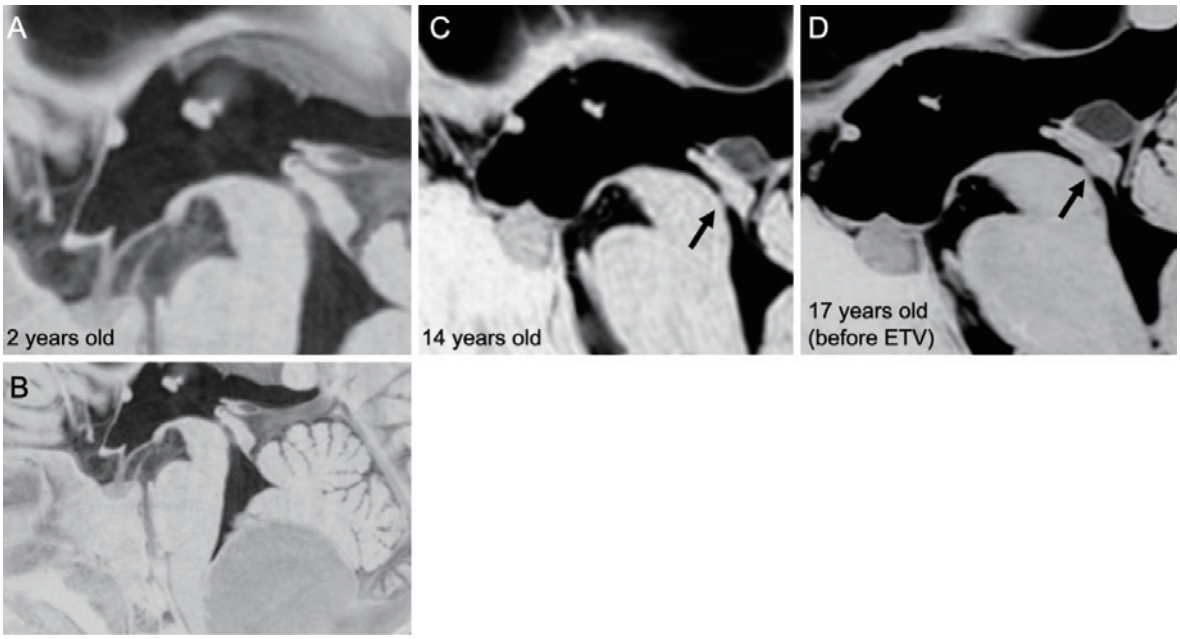

Fig. 1 Sagittal heavy T2 weighted images acquired at the age of 2, 14, and 17 years.
Serial MR imaging (A, C, D) shows a gradually enlarging third ventricle with chronologically evident aqueduct stenosis. Notably, the third ventriculomegaly was not evident in infancy (A) before resecting a fourth ventricular cyst (B). Afterward, no intracranial space-occupying lesions were observed except for the pineal cyst, which does not show a mass effect on the aqueduct (black arrow), and a comparatively small fourth ventricle with an enlarged third ventricle (C, D).

ache and occasional faintness. Relevant medical history revealed a fourth ventricular epidermoid cyst, a non-neoplastic brain tumor, and a pineal cyst without any evidence of aqueduct stenosis (Fig. 1A, B). When she was two years of age, the fourth ventricular cyst was surgically removed without complications. Although the clinical condition remained stable after the initial surgery, at the age of 14, the patient suffered from headaches associated with repeated transient weakness and sensory disturbances in her left extremities. Magnetic resonance (MR) angiography revealed moyamoya angiopathy in both hemispheres, with decreased cerebral blood flow (CBF) in the right hemisphere, as demonstrated by N-isopropyl-p-[^{123}I] iodoamphetamine single-photon emission computed tomography. Under the definitive diagnosis of MMS associated with a brain tumor, combined direct and indirect revascularization of the symptomatic right hemisphere was performed without complications. Ischemic symptoms were relieved postoperatively, as shown by a considerable improvement in CBF in the affected hemisphere. However, at the age of 17, the patient complained of an unresolved, persistent, medically intractable headache with occasional faintness. Serial MR imaging ultimately revealed newly developed non-communicating hydrocephalus due to acquired aqueduct stenosis (Fig. 1C, D), suggesting a surgical indication for ETV. To exclude the development of dangerous collateral vessels (either or both periventricular anastomosis and vault moyamoya vessels)[7-9] along the surgical route, we performed cerebral angiography via the surgical route of the ETV in the right hemisphere, which confirmed their absence (Fig. 2A, B). The superficial temporal artery, deep temporal artery, and middle meningeal artery (MMA) were well-developed on the right side as a result of the combined revascularization performed over three years (Fig. 2 C). Notably, the left anterior branch of the MMA was confirmed on the left cranial vault as a transdural collateral (Fig. 2C). Based on these angiographic findings, the patient underwent ETV under general anesthesia.[10] A burr hole was placed in the right frontal region near the cranial edge. A plastic sheath was inserted into the anterior horn of the right lateral ventricle, through which a flexible neuroendoscopic system (VISERA OLYMPUS VEF type V; Olympus, Tokyo, Japan) was introduced. The third ventricle was reached via the abnormally dilated foramen of Monro. Anatomical structures, such as the bilateral mammillary bodies, infundibular recesses, and the orifice of the Sylvian aqueduct, were promptly identified without encountering the marked development of periventricular anastomosis (Fig. 3A, B). There was no evidence of past intraventricular hemorrhage or past meningitis. ETV was completed without complications using endoscopic forceps and a balloon catheter (Fig. 3C, D). The postoperative course was uneventful, with a complete resolution of the patient's chronic headache and shrinkage of the third ventriculomegaly (Fig. 4). The patient was discharged without neurological deficits, and clinical status remained stable dur-

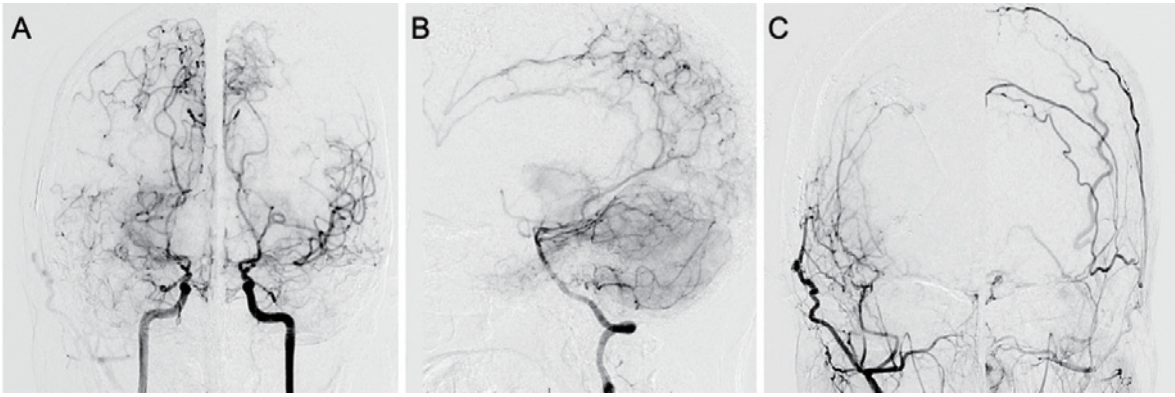

Fig. 2 Cerebral angiograms before endoscopic third ventriculostomy for comprehensive evaluation of the collaterals associated with moyamoya angiopathy.
Abnormally dilated perforators, including the lenticulostriate, choroidal, and thalamic arteries, are observed in the internal carotid (A) and vertebral angiographies (B); however, no periventricular anastomoses are observed, suggesting the possibility of safe access to the lateral ventricles. External carotid angiographies (C) showed a marked development of the transdural anastomosis via the anterior convexity branch of the middle meningeal artery (so-called vault moyamoya) in the left hemisphere, whereas in the right hemisphere, a marked ingrowth of the deep temporal and middle meningeal arteries as well as a patent direct superficial temporal artery to middle cerebral artery bypass but no vault moyamoya, were observed.

ing the one-year follow-up period.

Discussion

This case report describes an extremely rare case of MMS associated with a brain tumor and LIAS in a female adolescent who complained of an intractable headache with occasional faintness. Serial MR ultimately revealed newly developed non-communicating hydrocephalus due to acquired aqueduct stenosis. After careful examination of the development of either or both periventricular anastomosis and vault moyamoya vessels along the surgical route using cerebral angiography, we performed ETV via the right anterior horn under general anesthesia without complications. We would emphasize the importance of preoperative cerebral angiography in evaluating the collateral vasculature associated with moyamoya angiopathy, especially transdural collaterals on the cranial vault as well as periventricular collaterals (i.e., lenticulostriate arteries (LSAs) and thalamic and choroidal anastomoses)[7-9] before ETV. To our knowledge, there have been no reports of symptomatic non-communicating hydrocephalus due to LIAS in patients with MMS in the literature. Although such a case might rarely be experienced in clinical practice, intraventricular bleeding is a common clinical presentation of adult MMD,[1-3] which may cause non-communicating hydrocephalus requiring ETV. Therefore, we believe that our case report may provide valuable insights into such situations.

LIAS is a unique subgroup of non-neoplastic stenosis of the aqueduct in adolescents and adults that can be idiopathic, such as subarachnoid hemorrhage, meningitis, or neoplasm.[5,6] The clinical features of LIAS include 1) no intracerebral lesions except for hydrocephalus, 2) either or both enlarged lateral and third ventricles with a comparatively small fourth ventricle, 3) no apparent history of intracranial bleeding or meningitis, 4) no prior surgical treatment for hydrocephalus, and 5) undiagnosed in infancy.[5] The recent technical and material innovations in the neuro-endoscopy era enabled third ventriculostomy as a safe and effective treatment of choice for aqueduct stenosis[11,12] including LIAS. Fukuhara et al. reviewed 31 patients with LIAS who were treated with ETV, and 26 of 31 (83.9%) of these patients demonstrated favorable outcomes after ETV, with a mean follow-up duration of 26.2 months. Therefore, the authors concluded that ETV was considered the best surgical option for LIAS.[5] We concur with their findings, and even in our case, the patient presented with MMS and LIAS when the cranial vault and periventricular collaterals were less developed, allowing ventricular access for ETV.

When considering ETV in patients with moyamoya angiopathy (i.e., MMD and MMS), transdural and periventricular collaterals along the surgical route may pose a potential risk of bleeding during or after surgery. First, when accessing the lateral ventricle via the anterior horn, vault moyamoya collaterals[9] and periventricular LSA anastomosis[7] have a potential risk of bleeding. Recently, dilated abnormal collateral vessels originating from the medial posterior choroidal artery have been reported as the origin of bleeding in the corpus callosum in adult MMD.[13] Altogether, periventricular collaterals originating from the LSAs and choroidal arteries have a potential risk of bleeding when accessing the lateral ventricle. Second, when making a stoma on the third ventricle floor, the periventricular thalamic anastomosis[7] also has the potential risk of lethal

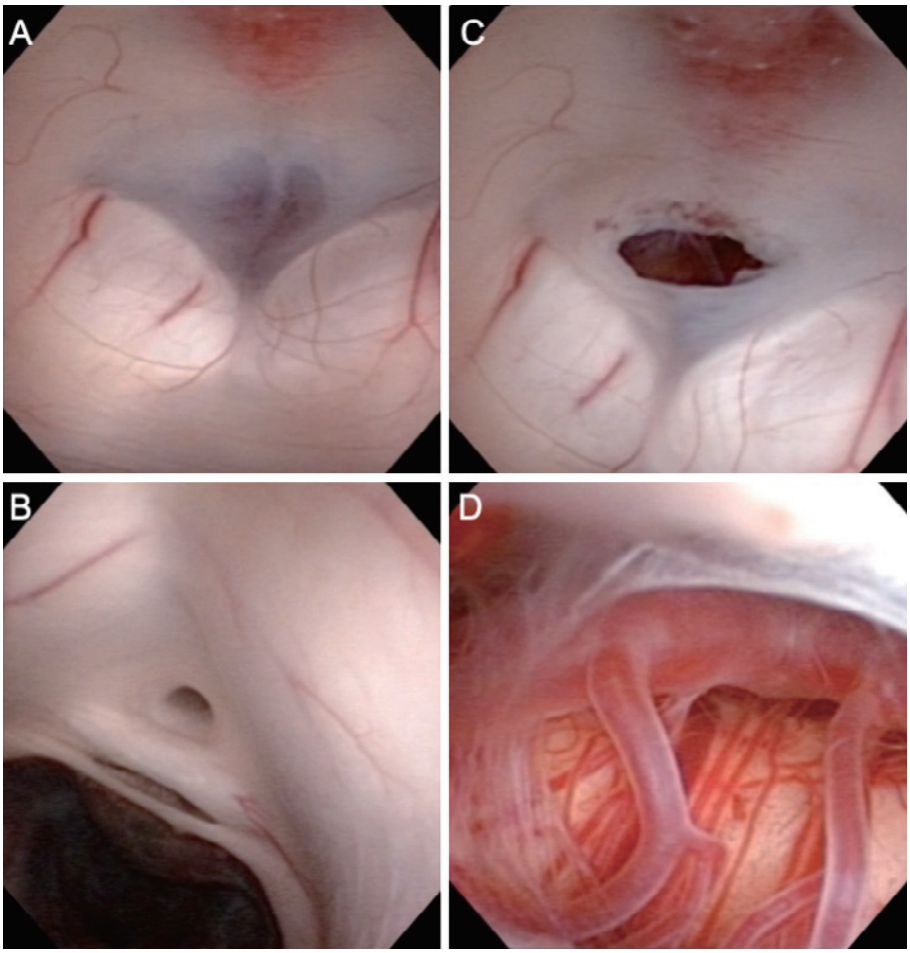

Fig. 3 Fiber video photographs during endoscopic third ventriculostomy.
A thinned third ventricle floor (tuber cinereum) is observed without a marked development of periventricular anastomosis (A). The orifice of the Sylvian aqueduct shows no evidence of past hemorrhagic nor inflammatory (i.e., membranous) obstruction (B). The thinned third ventricle floor is successfully fenestrated (C). Notably, perforators (thalamoperforating artery) dilated from the tip of the basilar artery are observed through the fenestrated stoma (D).

bleeding. In our case, we anticipated that during a single burr hole entry into the lateral ventricle, transdural collaterals on the cranial vault or periventricular collaterals (i.e., the LSAs and choroidal arteries) could be injured as the first step in ETV, followed by the thalamic anastomosis during fenestration of the third ventricle floor. Therefore, we carefully examined the vault and periventricular collaterals before ETV using cerebral angiography. Consequently, we confirmed transdural collaterals on the left anterior cranial vault, which should be secured, but there were no dangerous collateral vessels (either or both periventricular anastomosis and vault moyamoya vessels) via the surgical route of the ETV in the right hemisphere (Fig. 2A, B). Based on the angiographic findings, we safely entered the ventricular system via the right anterior horn with minimal bleeding and completed ETV under clear visualization of the third ventricle floor. The potential limitation of the surgical indication for ETV in our case is that if dangerous periventricular collaterals are found, including the LSAs and thalamic and choroidal anastomoses, another method of cerebrospinal fluid diversion, such as ventriculoperitoneal shunt insertion, should be considered, although a ventricular tap can still cause intraventricular bleeding.

In conclusion, when treating non-communicating hydrocephalus in MMS and LIAS with ETV, careful preoperative evaluation of the transdural collaterals on the cranial vault and periventricular collaterals using cerebral angiography is warranted.

Acknowledgments

This work was partially supported by JSPS KAKENHI (Grant numbers 19H03765 [M.I.] and 23H03015 [M.F.]).

Abbreviations

CBF, cerebral blood flow; ETV, endoscopic third ventriculostomy; ^{123}I-IMP-SPECT, N-isopropyl-p-[^{123}I] iodoampheta-

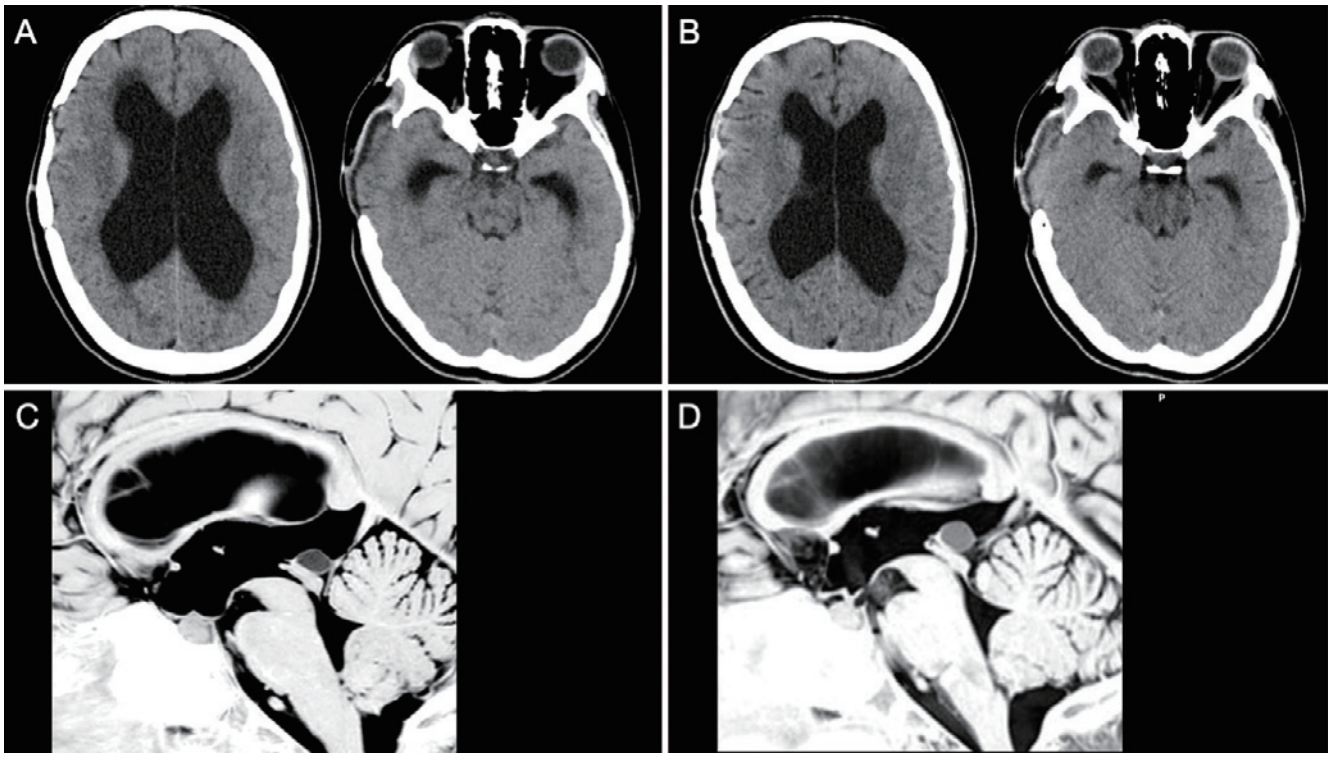

Fig. 4 Postoperative changes in the lateral and third ventricle system after successful endoscopic third ventriculostomy. Preoperative (A) and postoperative (B) computed tomography scans show comparative shrinkage of the lateral ventricles after endoscopic third ventriculostomy. Sagittal heavy T2 weighted images before (C) and immediately after surgery (D) clearly show shrinkage of the third ventriculomegaly and cerebrospinal fluid flow voids at the floor through the third ventricle stoma.

mine single-photon emission computed tomography; LIAS, late-onset idiopathic aqueduct stenosis; MMA, middle meningeal artery; MMD, Moyamoya disease; MMS, Moyamoya syndrome; MR, magnetic resonance; TIA, transient ischemic attack

Conflicts of Interest Disclosure

All authors who are members of the Japan Neurosurgical Society (JNS) have registered online self-reported COI Disclosure Statement Forms through the website for JNS members.

References

1) Fujimura M, Tominaga T, Kuroda S, et al.: 2021 Japanese guidelines for the management of Moyamoya disease: guidelines from the research committee on Moyamoya disease and Japan stroke society. *Neurol Med Chir* 62: 165-170, 2022
2) Kuroda S, Fujimura M, Takahashi J, et al.: Diagnostic criteria for Moyamoya disease - 2021 revised version. *Neurol Med Chir* 62: 307-312, 2022
3) Kuroda S, Houkin K: Moyamoya disease: current concepts and future perspectives. *Lancet Neurol* 7: 1056-1066, 2008
4) Scott RM, Smith ER: Moyamoya disease and Moyamoya syndrome. *N Engl J Med* 360: 1226-1237, 2009
5) Fukuhara T, Luciano MG: Clinical features of late-onset idiopathic aqueductal stenosis. *Surg Neurol* 55: 132-136, 2001
6) Martinoni M, Miccoli G, Riccioli LA, et al.: Idiopathic aqueductal stenosis: late neurocognitive outcome in ETV operated adult patients. *Front Neurol* 13: 806885, 2022
7) Funaki T, Takahashi JC, Houkin K, et al.: Angiographic features of hemorrhagic moyamoya disease with high recurrence risk: a supplementary analysis of the Japan Adult Moyamoya Trial. *J Neurosurg* 128: 777-784, 2018
8) Yamamoto S, Funaki T, Fujimura M, et al.: Development of hemorrhage-prone anastomoses in asymptomatic moyamoya disease - a comparative study with Japan adult moyamoya trial. *J Stroke Cerebrovasc Dis* 28: 104328, 2019
9) Suzuki J, Takaku A: Cerebrovascular "moyamoya" disease. Disease showing abnormal net-like vessels in base of brain. *Arch Neurol* 20: 288-299, 1969
10) Endo H, Fujimura M, Kumabe T, Kanamori M, Watanabe M, Tominaga T: Application of high-definition flexible neuroendoscopic system to the treatment of primary pineal malignant B-cell lymphoma. *Surg Neurol* 71: 344-348, 2009
11) Bouras T, Sgouros S: Complications of endoscopic third ventriculostomy. *World Neurosurg* 79: S22.e9-S22.12, 2013
12) Munekata Y, Sugiyama T, Ueda Y, Mizushima M, Motegi H, Fujimura M: Late-onset obstructive hydrocephalus associated with occipital encephalocele with large skull defect successfully treated by endoscopic third ventriculostomy. *Childs Nerv Syst* 39: 307-310, 2023
13) Yamamoto S, Akioka N, Kashiwazaki D, Tomita T, Kuwayama N, Kuroda S: Moyamoya disease emerged with corpus callosum hemorrhage: a 3D computer graphic analysis. *NMC Case Rep J*

16: 85-89, 2016

Corresponding author: Masaki Ito, M.D., Ph.D.

Department of Neurosurgery, Hokkaido University Hospital, North 14, West 5, Sapporo, Hokkaido 060-8638, Japan.
e-mail: cooky-co@med.hokudai.ac.jp