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Managing intervention for severe intraventricular hemorrhage casting in moyamoya disease: Report of two cases

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ABSTRACT

INTRODUCTION: When severe intraventricular hemorrhage (IVH) casting in moyamoya disease (MMD) is mentioned, experts advocate not to evacuate the IVH in acute phase. However, the devastating outcomes derived from this empirical rule have not been addressed. Herein, we report two MMD cases undergoing obliteration of ruptured aneurysm and early complete aspiration of severe IVH casting, and showed good outcomes.

PRESENTATION OF CASES: Case 1: A 55-year-old woman was admitted to our hospital. Her initial Glasgow Coma Scale (GCS) was 4 and a computerized tomography (CT) scan showed severe IVH. Cerebral angiography (CAG) revealed MMD and an aneurysm. Direct surgery was performed for the ruptured aneurysm, followed by endoscopic complete aspiration of residual IVH. The modified Rankin Scale (mRS) was 1 at discharge.

Case 2: A 44-year-old woman was admitted to our hospital. Her initial GCS was 4 and the CT scan showed a severe IVH. CAG revealed MMD and an aneurysm. Transcatheter arterial embolization was carried out for the aneurysm, followed by endoscopic complete aspiration of the residual IVH. The mRS was 2 at discharge.

DISCUSSION: Performing only external ventricular drainage in acute phase for such IVH casting is generally preferred way, however, there are no scientific data supporting this empirical rule. Early surgical removal of severe IVH casting is an approach for severe IVH casting in MMD.

CONCLUSION: Early aspiration of severe IVH casting is an option for managing MMD with critical intracranial pressure, and it has the potential to prevent poor outcomes in MMD patients with severe IVH.

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1. Introduction

Moyamoya disease (MMD) can lead to hemorrhagic stroke including severe intraventricular hemorrhage (IVH) with devastating consequences. However, few studies have shown the optimal timing for IVH removal. There are several cases with MMD experiencing severe IVH casting due to intraventricular aneurysm (IVA) rupture have been reported [1–11], though little information was provided regarding the management for IVH casting. Performing only external ventricular drainage (EVD) in acute phase and waiting for lysis of hematoma and patients' neurological recovery is

a generally preferred way for the reason one could avoid iatrogenic destruction of parenchymal tissue and collateral circulation [12], however, there are no scientific data supporting this empirical rule. Information regarding the timing of the surgical intervention for severe IVH casting associated with MMD is appallingly lacking. Herein, we report two MMD cases with severe IVH casting due to IVA rupture who underwent emergency obliteration of ruptured aneurysm and early complete endoscopic aspiration of the IVH, and showed relatively good outcomes at discharge. This work has been reported in line with the SCARE criteria [13].

2. Case reports

2.1. Case 1

A previously healthy 55-year-old woman experienced the sudden onset of headache followed by loss of consciousness. The patient's Glasgow Coma Scale (GCS) was 4 on arrival. Her blood pressure was 160/110 mmHg, heart rate 30 beats per minute. A computerized tomography (CT) scan showed severe IVH casting, mainly involving the right lateral ventricle, and CT angiography

Abbreviations: AchA, anterior choroidal artery; CSF, cerebrospinal fluid; CT, computerized tomography; EC-IC, extra-cranial and intra-cranial; EVD, external ventricular drainage; GCS, Glasgow Coma Scale; ICA, internal carotid artery; ICH, intracerebral hemorrhage; ICP, intracranial pressure; IV-An, intraventricular aneurysm; IVH, intraventricular hemorrhage; MMD, moyamoya disease; mRS, modified Rankin Scale; PchA, posterior choroidal artery; STA-MCA, superficial temporal artery-middle cerebral artery; VP, ventriculo-peritoneal.

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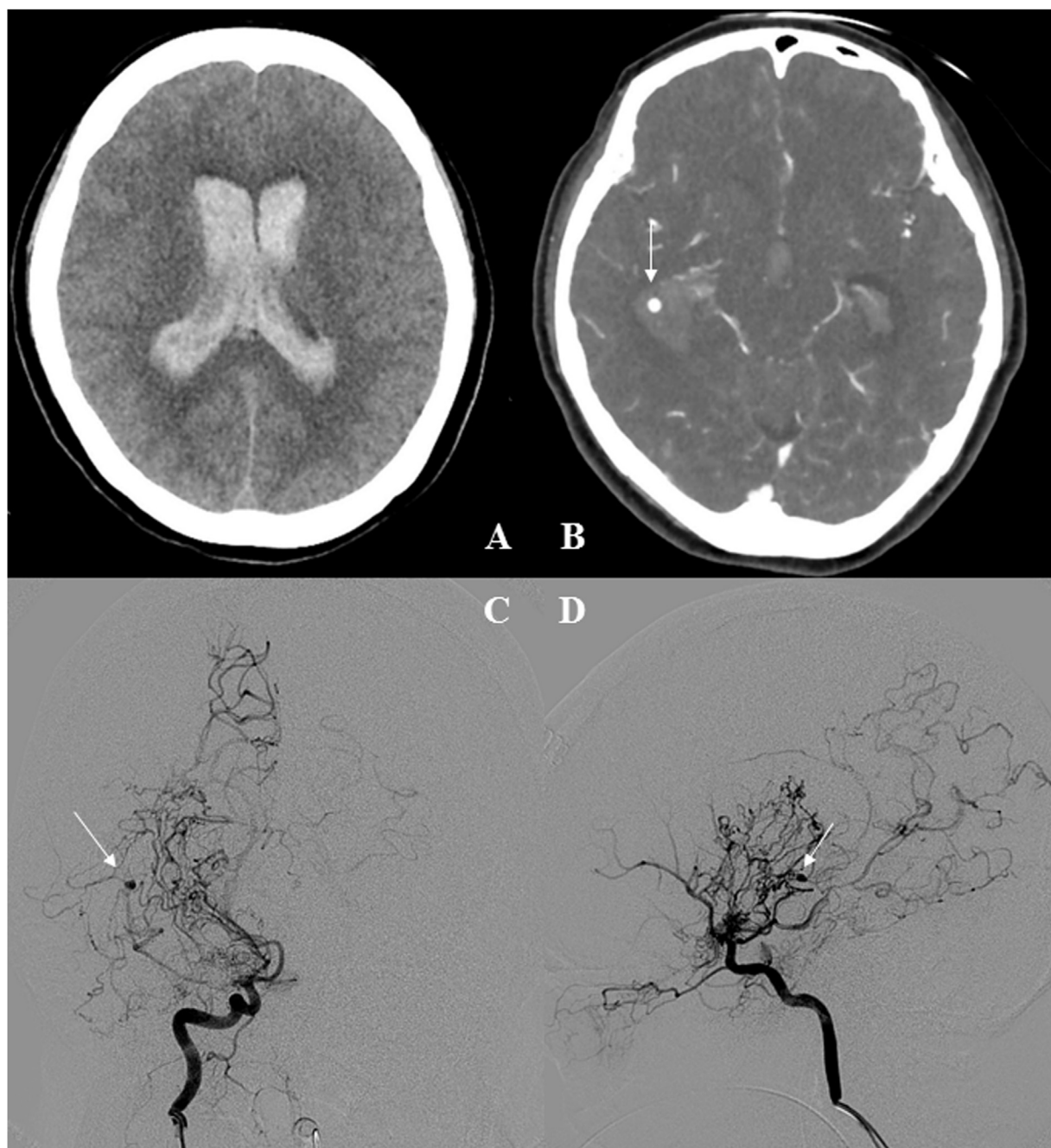


Fig. 1. Initial computerized tomographic images and cerebral angiographic images of case 1.

A: Computerized tomography (CT) revealed severe intraventricular hemorrhage. B: CT angiography showed a round 5 mm aneurysm in the right inferior horn of the lateral ventricle (white arrow).

C: Right carotid cerebral angiography revealed occlusion of the terminal portion of the right internal carotid artery with extensive moyamoya vessels, and an aneurysm in the distal portion of the anterior choroidal artery (white arrow). D: Right carotid cerebral angiography, lateral view, shows an aneurysm (white arrow).

showed a round 5 mm aneurysm in the right inferior horn of the lateral ventricle. Bilateral EVD was undertaken immediately after arrival, but most of the IVH persisted. Cerebral angiography (CAG) revealed occlusion of the terminal portion of the right internal carotid artery (ICA) with extensive moyamoya vessels, and demonstrated the characteristic appearance of MMD as well as an aneurysm in the distal portion of the anterior choroidal artery (AchA) (Fig. 1). We performed direct surgery via the right middle temporal gyrus and detached the aneurysm from the parent artery. After treating the IVA, we performed endoscopic surgery to remove the residual IVH in the left lateral ventricle, allowing nearly all of the IVH to be removed (Fig. 2). The patient's consciousness gradually

recovered, reaching GCS 15 in a month, and right superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis was carried out. The modified Rankin Scale (mRS) was 3 8 weeks after the onset, with recovery to 1 a year after the onset.

2.2. Case 2

A previously healthy 44-year-old woman experienced the sudden onset of headache followed by vomiting and loss of consciousness. Her GCS was 4 on arrival. A CT scan showed a cast-type IVH, mainly involving the right lateral ventricle. CT angiography revealed an aneurysm in the right body of the lateral ventricle.

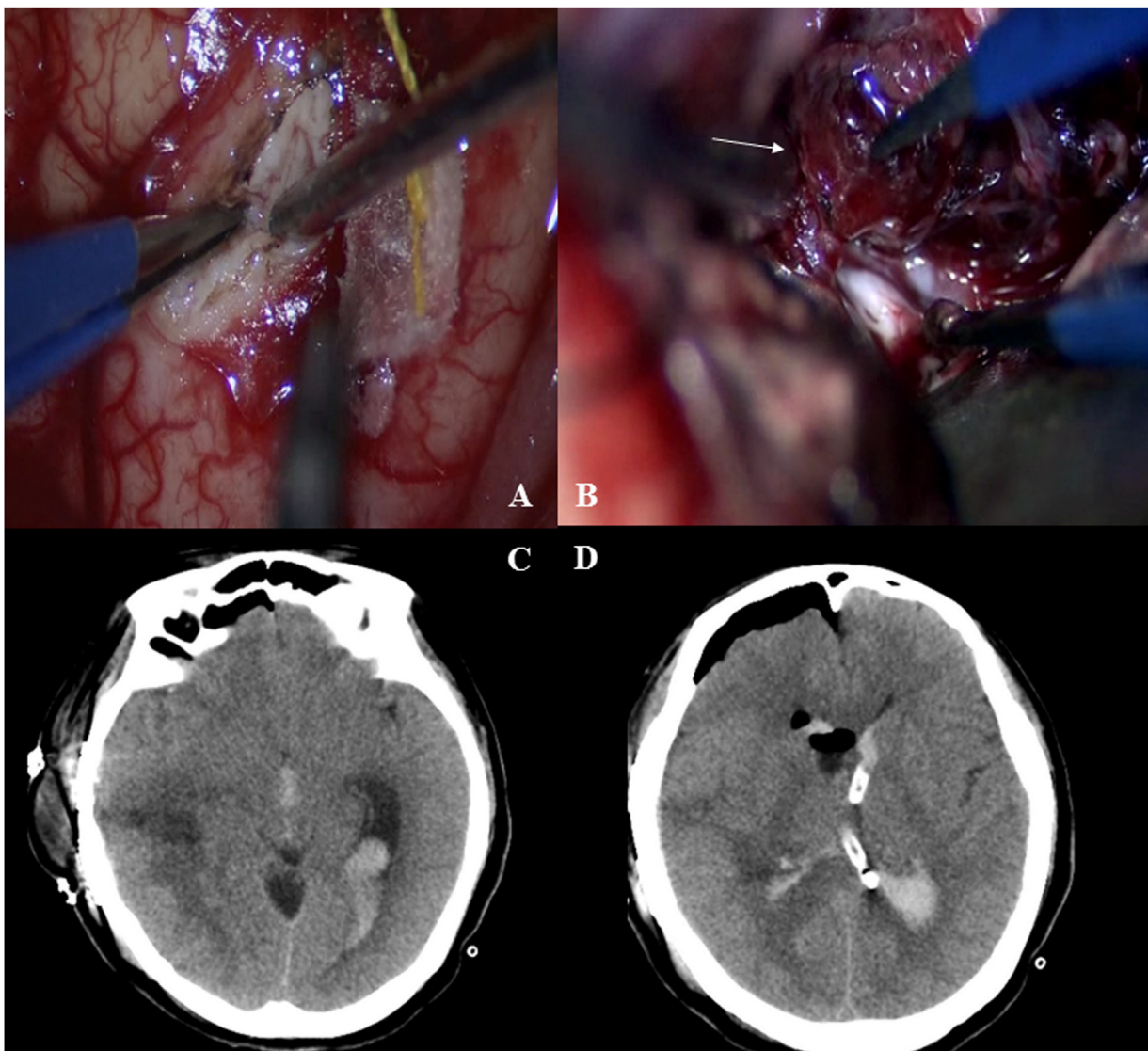


Fig. 2. Operative views and postoperative computerized tomographic images of case 1.

A: A small corticotomy was made in the right middle temporal gyrus. B: Intraoperative findings revealed an aneurysm in the inferior horn of the right lateral ventricle (white arrow). C: Computerized tomography (CT) after the direct surgical procedure confirmed removal of the aneurysm. D: CT scan obtained in the immediate post-treatment period after endoscopic removal of the intraventricular hemorrhage.

Bilateral EVD was performed immediately after arrival, but most of the IVH persisted. CAG showed occlusion of the terminal portion of the right ICA, with extensive moyamoya vessels. The characteristic appearance of MMD was noted, and an aneurysm in the posterior choroidal artery (PchA) was revealed, as well as blowout of contrast agent into the ventricle on the moving image (Fig. 3). We immediately performed transcatheter arterial embolization using Histoacryl (n-butyl-2-cyanoacrylate). After treating the IVA, we performed endoscopic surgery to eliminate the residual IVH, nearly all of which was removed (Fig. 4). Her consciousness gradually recovered, reaching GCS 10 in two months, and GCS 15 a year after tracheotomy closure. Right STA-MCA anastomosis was carried out a year after the onset. The patient's mRS was 4 at 8 weeks after the onset, with recovery to 2 a year after the onset.

3. Discussion

Several MMD cases with severe IVH casting due to IVA rupture have been reported [1–11]. Most had ruptures of a distal AChA or a distal PchA aneurysm, as in our cases. Early

reports described conservative treatment for IVA, and tended to show unfavorable outcomes due to IVA re-rupture [2,3,11]. Most cases reported in recent years received direct or endovascular surgery for the ruptured IVA [1,3,5–10]. Direct surgery should be considered when a surgical corridor would not destroy the collateral circulation associated with MMD, with endovascular surgery otherwise being preferable. If the parent artery supplies an important collateral route but the preservation of this artery is not possible, simultaneous cerebral revascularization including EC-IC bypass should be considered.

The optimal treatment for severe IVH casting in cases with MMD has not yet been established. Performing only EVD in acute phase and then observing the lysis of hematoma and patient for neurological recovery is a preferred approach still now [1]. This treatment has the advantages of allowing invasive procedures to be omitted in patients with unstable disease and avoiding iatrogenic destruction of parenchymal tissue and collateral circulation. However, the prognosis of the severe IVH patients in MMD would only become evident after the lysis of hematoma in this method.

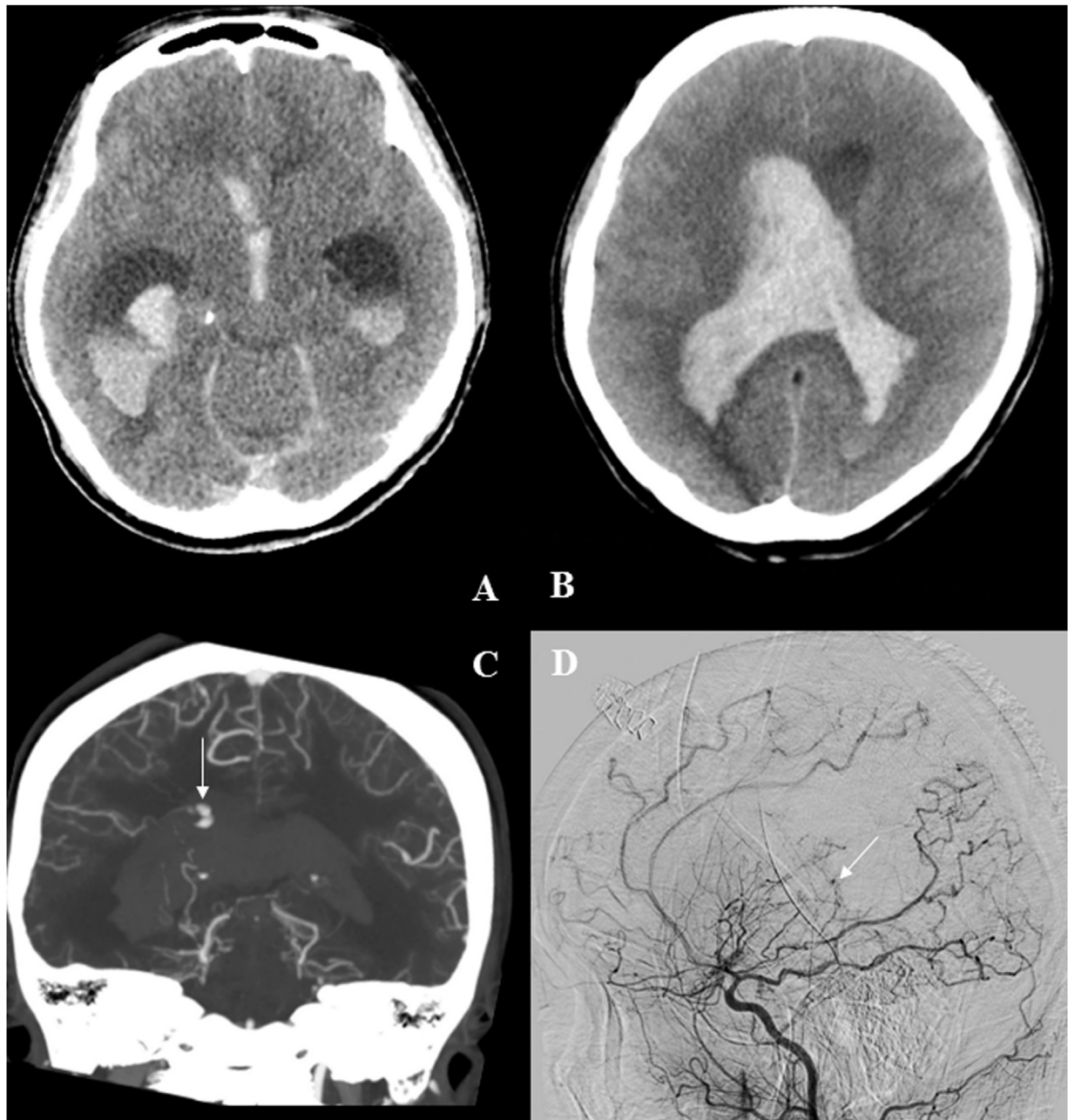


Fig. 3. Initial computerized tomographic images and cerebral angiographic images of case 2.

A: Computerized tomography (CT) showed acute hydrocephalus. B: CT revealed severe cast-type intraventricular hemorrhage. C: CT angiography showed an aneurysm in the right body of the lateral ventricle (white arrow). D: Right carotid cerebral angiography revealed occlusion of the terminal portion of the right internal carotid artery with extensive moyamoya vessels, and an aneurysm in the distal portion of the posterior choroidal artery (white arrow).

Early surgical removal of severe IVH casting is another approach for severe IVH casting in MMD. This treatment has the advantage of avoiding acute-phase critical intracranial pressure (ICP), which may cause irreversible tissue damage leading to late phase ischemic complications [14,15]. The patients who suffered severe IVH casting shows wide range of prognosis, and how the IVH casting has the harmful effect for specific patients has not been well presented. A study using a canine model of IVH showed periventricular blood infiltration to cause ependymal tissue disruption and cellular desquamation [16], such that early removal of severe IVH might prevent irreversible ependymal tissue damage.

Among the reported MMD cases with severe IVH due to IVA rupture, endoscopic procedures were performed in some case [8],

however, that report focused on the observation and description of ependymal tissue and did not describe the timing of the removal of IVH. Information regarding the timing of the surgical intervention for severe IVH casting associated with MMD is appallingly lacking. Performing only external EVD in acute phase for IVH casting is still generally preferred way, however this approach could ignore the potential to decrease the critical ICP and irreversible ependymal tissue damage.

4. Conclusions

Although only two cases, our cases showed relatively good outcomes despite initially severe neurological status. For a cast-type IVH remaining after EVD in MMD, early removal of IVH,

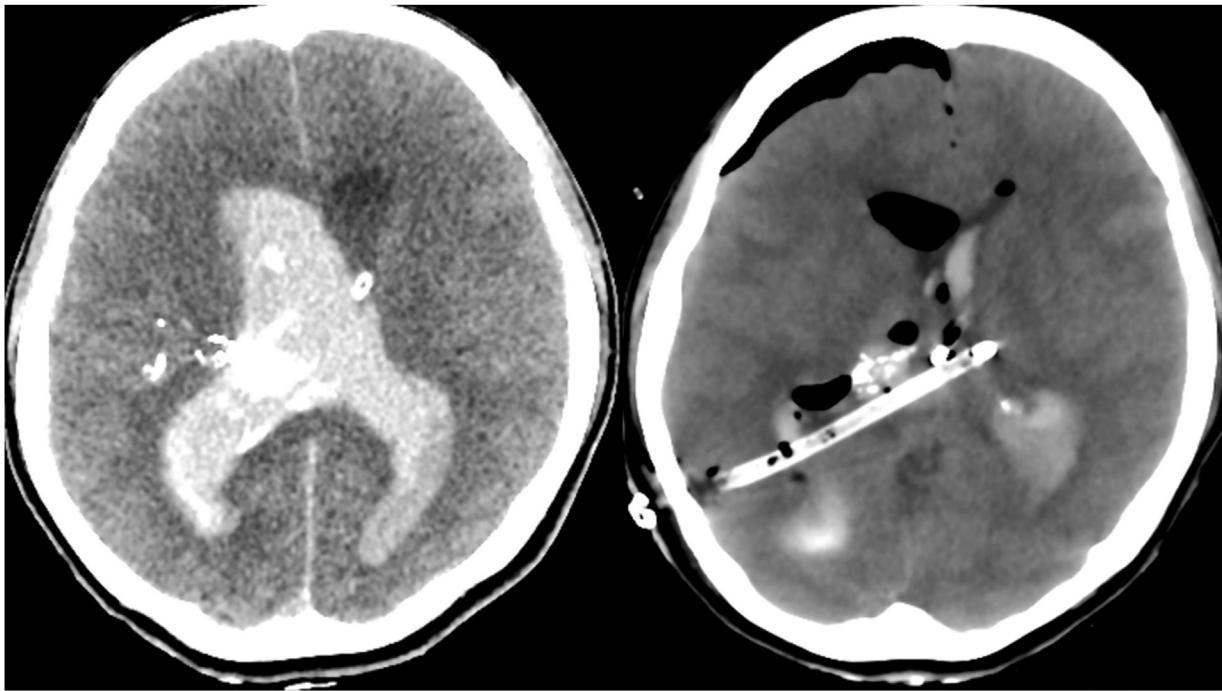


Fig. 4. Postoperative computerized tomographic images of case 2.

Left: Computerized tomography (CT) scan obtained immediately after transcatheter arterial embolization using Histoacryl (n-butyl-2-cyanoacrylate). Right: CT scan obtained immediately after endoscopic removal of the intraventricular hemorrhage.

as in our present two cases, may contribute to better outcomes.

5. Limitations

The major limitation of this case report is its small number of patients. Another limitation is the short follow-up period. A longer period is needed to assess the validity of our conclusions.

Declaration of Competing Interest

All authors have no conflict of interest and funding.

Funding

All authors have no conflict of interest and funding.

Ethical approval

This report has ethics committee approval in our institution. The IRB number is 366.

Consent

Written informed consent was obtained from all individual participants and relations in the first degree included in the study for publication of this case report and accompanying images.

All procedures performed in this study were in accordance with the ethical standards of our institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Author contribution

Conception and design: Goto. Drafting of the article: Goto. Treating the patients: Goto, Hino, and Oka. Analyzing the imaging data:

Goto and Hino. Figures: Goto. Critically revising the article: Hino. All authors read and approved the final manuscript.

Registration of research studies

Not applicable because this is purely observation study.

Guarantor

Corresponding author Yukihiro Goto is the guarantor of the work and the conduct of the study, had access to the data, and controlled the decision to publish.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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