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Case Report

Direct carotid-cavernous fistula presenting with intracranial hemorrhage without ocular symptoms ☆,☆☆

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ABSTRACT

Herein, we report a unique case of nontraumatic direct carotid-cavernous fistula presenting with intracerebral hemorrhage without any ocular symptoms. A 90-year-old woman was found unconscious and vomiting due to a subcortical hemorrhage in the temporal lobe. Magnetic resonance angiography revealed a direct carotid-cavernous fistula of Barrow type A. Extensive cortical venous reflux from the superficial middle cerebral vein was observed and identified as a probable contributor to the cerebral hemorrhage. We performed successful embolization using combined transarterial and transvenous approaches. We first occluded the dangerous venous drainage via the transvenous approach, followed by selective occlusion of the direct carotid-cavernous fistula via the transarterial approach. This strategy provided that the dangerous venous drainage was completely occluded first in case complete obliteration could not be achieved with the transarterial approach.

Introduction

Direct carotid-cavernous fistulas (CCFs) arise from trauma or aneurysm rupture in the cavernous sinus (CS) and lead to rapid and severe symptoms. Prompt intervention is critical in patients with progressive vision loss, cortical venous reflux, or intracerebral hemorrhage (ICH). However, ICH associated with direct CCF is rare, and treatment requires occlusion of the dangerous venous drainage [1,2]. This report presents a

Abbreviations: ACA, anterior cerebral artery; CCF, carotid-cavernous fistula; CS, cavernous sinus; ICA, internal carotid artery; ICH, intracerebral hemorrhage; IPS, inferior petrosal sinus; MCA, middle cerebral artery; MRA, magnetic resonance angiography; SMCV, superficial middle cerebral vein; SOV, superior ophthalmic vein; TAE, transarterial embolization; TVE, transvenous embolization.

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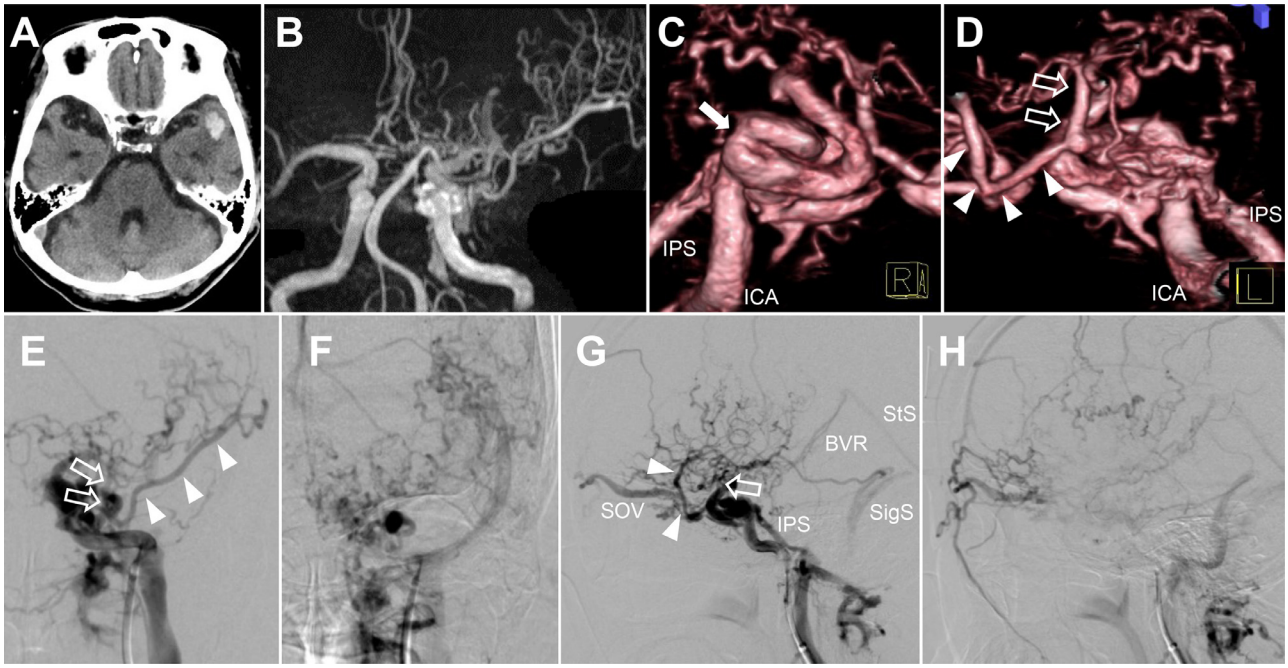

Fig. 1 – (A) Initial computed tomography revealed a subcortical left temporal lobe hemorrhage. (B) Magnetic resonance angiography demonstrating a left carotid-cavernous fistula (CCF) associated with cortical venous reflux. (C) Lateral view from the inside the three-dimensional rotational angiography (3D-RA) image suggest the presence of a fistula opening (arrow). (D) Lateral view from the outside the 3D-RA image reveals that the superficial middle cerebral vein ([SMCV], arrowheads) and the uncal vein (open arrows) converge and connect with the cavernous sinus (CS). (E) The frontal view of digital subtraction angiography (DSA) from the left internal carotid artery (ICA) in the early phase reveals no visualization of the artery beyond its terminal portion. The SMCV (arrowheads) and the uncal vein (open arrows) are confirmed. (F) The frontal view of DSA from the left ICA in the late phase reveals extensive cortical venous reflux from the SMCV to the left cerebral hemisphere. (G) The lateral view of DSA from the left ICA in the early phase reveals diverse venous outflows. The SMCV (arrowheads) and the uncal vein (open arrows) are confirmed. (H) The lateral view of DSA from the left ICA in the late phase reveals that the superior ophthalmic vein drainage is the last to be visualized.
Abbreviations BVR, basal vein of Rosenthal; ICA, internal carotid artery; IPS, inferior petrosal sinus; SOV, superior ophthalmic vein; SigS, sigmoid sinus; StS, straight sinus.

unique case of non-traumatic direct CCF with ICH, but no ocular symptoms managed through combined transvenous embolization (TVE) and transarterial embolization (TAE).

Case report

A 90-year-old woman presented with vomiting, reduced consciousness, and intermittent facial and right upper-limb seizures. She had no history of head trauma or significant ocular symptoms. Computed tomography revealed a 10 × 17 mm subcortical hemorrhage in the left temporal lobe (Fig. 1A). Magnetic resonance angiography (MRA) revealed left CCF (Fig. 1B). Diagnostic cerebral angiography confirmed no left external carotid artery lesions but identified a fistula at the C4 posterior genu of the left internal carotid artery (ICA), indicative of direct CCF of Barrow type A (Figs. 1C and D) [3]. Extensive cortical venous reflux from the superficial middle cerebral vein (SMCV) was observed (Figs. 1E-H). The left anterior cerebral artery (ACA) and middle cerebral artery (MCA) were not visible because of the steal phenomenon induced by the high-flow shunt.

Endovascular treatment was performed on the following day. After systemic heparinization, a 6Fr Cerulean DD6 catheter (Medikit, Tokyo, Japan) was coaxially inserted into a 7Fr Shuttle Guiding Sheath (COOK Medical, Bloomington, IN, USA) in the left ICA. An SL-10 microcatheter (Stryker Neurovascular, Fremont, MN, USA) was navigated through the ICA fistula into the CS for the TAE. Subsequently, a 7Fr Roadmaster catheter (Goodman, Aichi, Japan) was inserted into the left jugular vein. For TVE, another SL-10 microcatheter was guided through the ipsilateral inferior petrosal sinus (IPS) and the CS into the SMCV. To prioritize the embolization of dangerous venous drainage, coil embolization was targeted to the SMCV from the uncal vein bifurcation, proximal to the CS (Figs. 2A-C). The TVE microcatheter was repositioned near the fistula, and the first coil was deployed as a scaffold. Under shunt flow interruption using a transform occlusion balloon catheter (Stryker Neurovascular, Fremont, MN, USA), a second coil was inserted through the microcatheter for TAE and intertwined with the first coil to stabilize the frame. Additional coils were

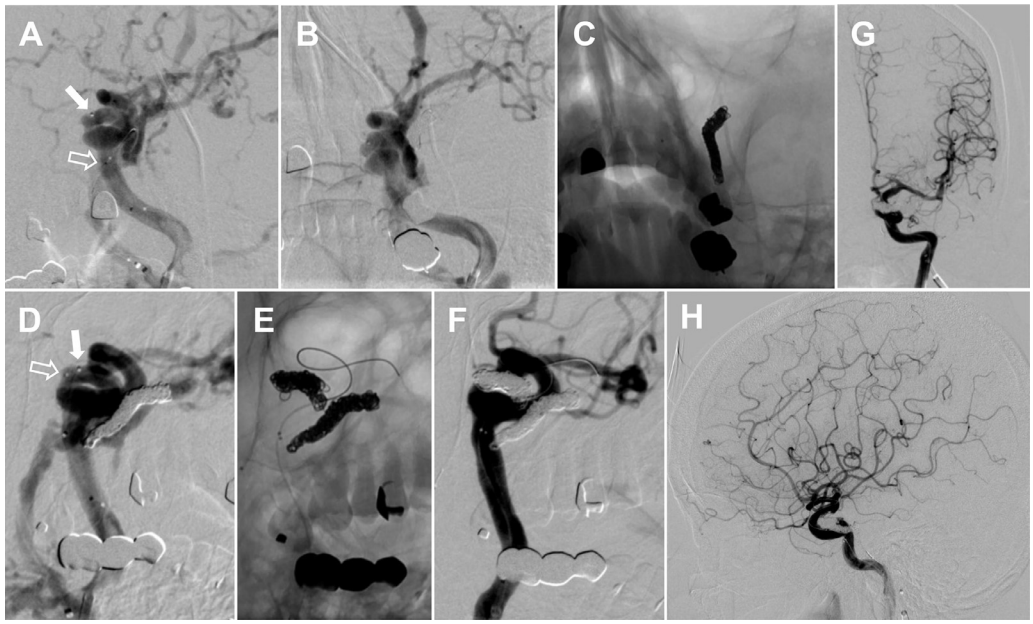

Fig. 2 – (A) The SL-10 (arrow) was positioned from the fistula of the internal carotid artery into the CS via an arterial route. Another SL-10 (open arrow) was guided through the inferior petrosal sinus and CS towards the SMCV via a venous route. (B) Confirmation of obstruction in the drainage to the SMCV. (C) The SMCV was coiled and embolized up to the confluence with the CS. (D) The SL-10 (open arrow) was guided venously and placed near the fistula. (E) Coil embolization was performed at the site of the fistula. (F) Confirmation of the disappearance of the shunt into the CS. (G) The frontal view of the final angiographic image reveals the complete resolution of the carotid-cavernous fistula (CCF) and the visualization of the ipsilateral anterior cerebral artery and middle cerebral artery. (H) The lateral view of the final angiographic image reveals the complete resolution of the CCF.

inserted through the microcatheter for TVE to achieve selective occlusion by covering the dorsal ICA with the lateral CS (Figs. 2D-F). Final angiography confirmed the complete resolution of the CCF and visualization of the ipsilateral MCA and ACA (Figs. 2G and H).

The patient's postoperative course was uneventful, and no ocular symptoms were observed. Two months after the surgery, the patient was discharged following rehabilitation. A 1-year follow-up MRA showed no CCF recurrence.

Discussion

Direct CCFs often present with ocular symptoms, such as proptosis, chemosis, and orbital bruit, and the incidence of ICH in direct CCFs is low, ranging from 0.9% to 3% [1,2,4]. The interval between the onset of ocular symptoms and the occurrence of ICH can range from several days to 6 years, with a median of 7 weeks [4]. As in the present case, CCF with ICH without ocular symptoms is rare. The hemorrhagic presentation is thought to be related to angiographic evidence of congestion in the sphenoparietal or petrous sinuses [2]. Intracranial hemorrhages caused by CCF arise due to retrograde flow into the cortical veins or rupture of the venous sinus walls, which is attributable to the pressure load of a high-flow shunt. In cases of ICH requiring hematoma evacuation, it is optimal to occlude the CCF endovascularly before surgical evacuation to minimize the risk of rebleeding or intraoperative venous hemorrhage. Furthermore, the initial step in endovascular treatment should prioritize the occlusion of dangerous venous drainage, considering the potential for incomplete shunt closure or inability to reaccess [5]. In this case, successful treatment was achieved by first occluding the SMCV, identified as the source of ICH, before shunt occlusion. Subsequently, complete occlusion of the CCF was accomplished using combined TVE and TAE approaches. This strategy effectively prevented rebleeding and ensured favorable outcomes. Various treatment methods have been reported, including covered stents, flow diverters, and liquid embolic agents [6–10]. Nevertheless, in managing direct CCFs associated with ICH, it is essential to adhere to the principle of initially occluding dangerous venous drainage, irrespective of the selected treatment modality.

In direct CCFs, ocular symptoms are typically observed during the acute phase. However, in cases with hemorrhagic onset, ICH precedes ocular symptoms in approximately 12% of cases [4]. Ocular symptoms result from progressive superior ophthalmic vein (SOV) congestion and cranial nerve palsy in CS. There are instances of cerebellar or brainstem hemorrhage without ocular symptoms in CCFs, predominantly involving posterior venous drainage. Furthermore, in CCFs with anterior venous drainage, shunt flow dispersion or low-flow lesions may reduce venous hypertension in the SOV, leading to the absence of typical orbital-ocular signs [2,11]. In this case, diverse venous outflows were noted, including anterior via the ophthalmic vein, inferior via the IPS, posterior via the basal

vein of Rosenthal, and superior via the SMCV (Fig. 1G). The SOV drainage was the last to be visualized and was likely the lowest flow among these drainage routes. These observations could be attributed to the absence of ocular symptoms in this rare clinical case.

Conclusion

In a rare case of direct CCF presenting with ICH but lacking ocular symptoms, successful treatment was achieved with a combination of TVE and TAE. In cases of direct CCF with ICH onset, preemptive embolization of dangerous venous drainage before shunt occlusion is essential to minimize the risk of rebleeding and improve treatment outcomes.

Submission statement

This manuscript is original and has not been submitted elsewhere in part or in whole.

Previous presentations

None.

Patient consent

Written informed consent was obtained from the patient to publish identifiable information or images within the manuscript.

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