

Coexistence of anterior cranial fossa dural arteriovenous fistula and arteriovenous malformation with the same drainage system: illustrative case

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BACKGROUND The authors report a rare case of coexistence of dural arteriovenous fistula (DAVF) and arteriovenous malformation (AVM), with a common trunk drainer from both DAVF and AVM in the left anterior cranial fossa (ACF) with simple DAVF in the right ACF.

OBSERVATIONS A 63-year-old female presented with seizure. Cerebral angiography showed bilateral DAVFs in the ACF and AVM in the left frontal lobe. A dilated frontal vein acted as a simple drainer of the right DAVF. In contrast, a dilated vein with large varix was the common drainer of both the left DAVF and the AVM. During surgery, indocyanine green videoangiography was performed with direct observation. In the left ACF, the drainer occlusion of the DAVF resulted in partial shrinkage of the varix and decreased distal blood flow. Additional main feeder occlusion of the AVM could decrease the blood flow further, but not completely because of the residual pial supplies for the AVM. Finally, the nidus of the AVM with varix was removed by en bloc resection.

LESSONS Neurosurgeons should be aware of the coexistence of DAVF and AVM with a common trunk drainer. Only simple occlusion of the drainer from DAVF is not sufficient, so removal of the AVM is essential.

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KEYWORDS anterior cranial fossa; arteriovenous malformation; coexistence; dural arteriovenous fistula; common drainage system; indocyanine green videoangiography

Intracranial dural arteriovenous fistulas (DAVFs) and arteriovenous malformations (AVMs) are relatively rare lesions, with detection rates of 0.15–0.29 per 100,000 per year and 1.12 per 100,000 per year, respectively.^{1–3} Coexistence of intracranial DAVF and AVM is extremely rare, with only three reported cases of intracranial DAVF and AVM in different regions. We experienced a case of DAVF and AVM coexisting in the same anterior cranial fossa (ACF), with a common trunk drainer from the shunt of the DAVF and AVM. During the surgery, we could visualize the hemodynamic changes during occlusion of the DAVF drainer and AVM feeder using stepwise indocyanine green (ICG) videoangiography. We also obtained complete histological examination of the AVM, and the common drainer with varix from both the DAVF and AVM.

Here, we describe the coexistence of DAVF and AVM in the ACF with the same drainage system. Neurosurgeons should not overlook this rare

occurrence because simple occlusion of the DAVF drainer as a result of misidentification as only DAVF will not achieve complete cure and allow the possibility of hemorrhage from the coexisting AVM.

Illustrative Case

A 63-year-old female was referred to our department without any appreciable past medical history. Computed tomography demonstrated old left frontal intracranial hemorrhage. Magnetic resonance imaging revealed apparent cortical venous dilation beside the hemorrhage and suspected the nidus of the AVM (Fig. 1).

Right and left external carotid angiography showed the arteriovenous shunt supplied by the bilateral infraorbital arteries, ethmoidal artery, and middle meningeal artery, which revealed bilateral DAVFs in the ACF (Fig. 2). The arteriovenous shunts were both located at

ABBREVIATIONS AVM = arteriovenous malformations; ACF = anterior cranial fossa; DAVF = dural arteriovenous fistula; ICG = indocyanine green; SSS = superior sagittal sinus.

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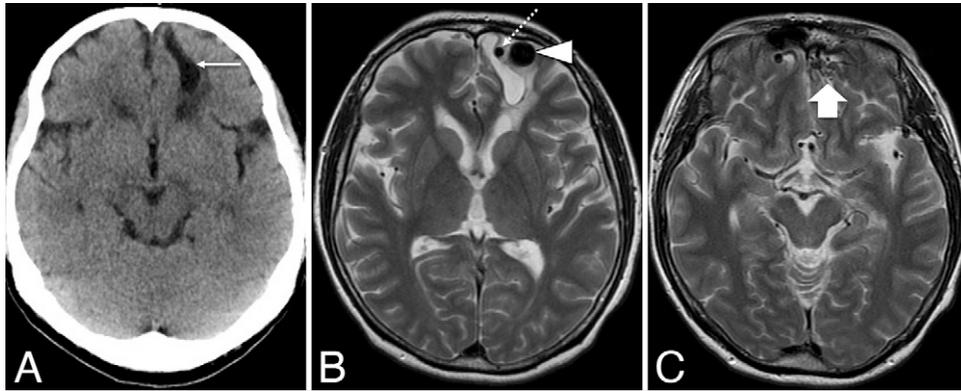


FIG. 1. Initial axial computed tomography scan (A) and T2-weighted magnetic resonance image (B and C) demonstrating the cavity caused by old intracranial hemorrhage (white arrow), dilated drainer (white broken arrow), varix (white arrowhead), and nidus (large white arrow).

the cribriform plate and drained into the bilateral cortical veins to the superior sagittal sinus (SSS). Left internal carotid angiography showed the arteriovenous shunt with nidus and intranidal aneurysm supplied by the left fronto-orbital artery, which revealed the AVM. The left draining cortical vein was a common trunk from the shunt of the left DAVF and AVM. Venous varix was observed at the bilateral drainers,

especially dilation in the left drainer (Fig. 3). We diagnosed bilateral ACF DAVFs, with coexistence of left ACF DAVF and AVM that had a common trunk drainer. We planned surgical intervention.

A bifrontal osteoplastic craniotomy with both side dural incisions was performed. The dilated cortical vein, venous varix, nidus, and shunting points of the bilateral DAVFs could be observed. The nidus and a proper feeder of the AVM in the left frontal lobe was identified between the shunting point of the left DAVF and dilated varix (Fig. 4A). We performed initial ICG videoangiography before inducing hemodynamic changes (Fig. 4B). The DAVF in the right ACF had simple architecture with a single drainer, so drainer occlusion could result in shrinkage of the dilated distal drainers (Fig. 4C and D). In the left ACF, drainer occlusion of DAVF at the cribriform plate was performed to reduce the blood flow to the drainer both from the left ACF DAVF and AVM. This simple drainer occlusion could result in shrinkage of the varix with residual shunting flow. Then, occlusion of the left fronto-orbital artery, which was a proper feeder of the nidus, shrank the dilated left varix immediately but still incompletely because of the remaining pial arterial supply from the AVM (Fig. 4C and D). Releasing the temporary occlusion of the left fronto-orbital artery caused the re-expansion of the varix (Fig. 4E and F). Therefore, we removed the

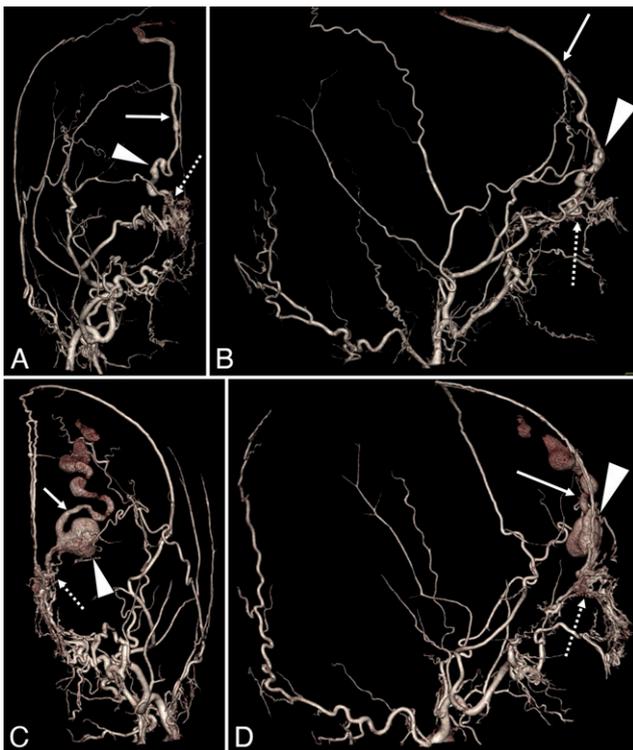


FIG. 2. Three-dimensional reconstruction images using rotational digital subtraction angiography of both right (A, anteroposterior view; B, lateral view) and left (C, anteroposterior view; D, lateral view) external carotid arteries depicting the bilateral dural arteriovenous fistulae (DAVFs) in the anterior cranial fossa (ACF), shunting point (white broken arrows), bilateral frontal draining veins (white arrows), and varix in the frontal lobe (white arrowheads).

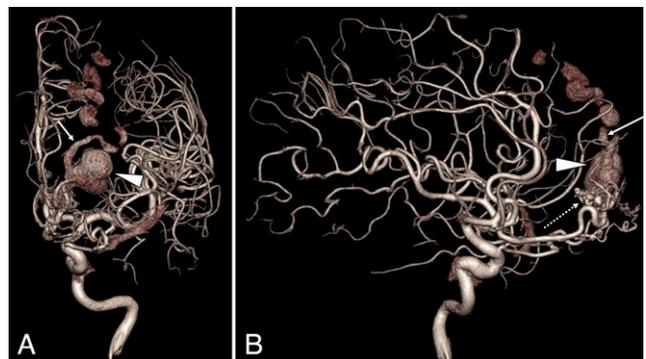


FIG. 3. Three-dimensional reconstruction images using rotational digital subtraction angiography of the left internal carotid artery (A, anteroposterior view; B, lateral view) depicting the nidus of the arteriovenous malformation (AVM) (white broken arrow) in the left frontal lobe and its draining vein (white arrows) with varix (white arrowheads) that was the common drainer of the left DAVF.

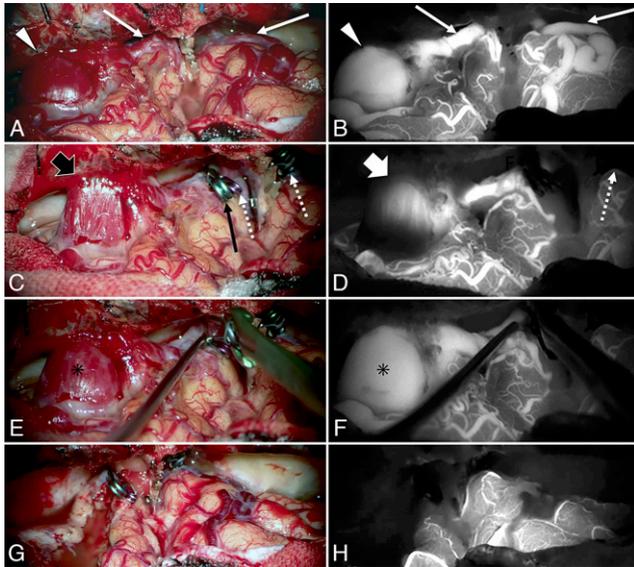


FIG. 4. Intraoperative initial microscopic view (A) and indocyanine green (ICG) videoangiography (B) showing the bilateral frontal dilated drainers (white arrows) and expanded varix in the left frontal lobe (white arrowheads). Microscopic view (C) and ICG videoangiography (D) after occlusion of the drainers of the bilateral DAVFs in the ACF (white broken arrows) and main feeder of the AVM (black arrow) revealing the shrinking varix (large black arrow) with residual shunting flow (large white arrow). Microscopic view (E) and ICG videoangiography (F) after occlusion of the bilateral drainers and release of the main feeder of the AVM demonstrating reexpansion of the varix (asterisk). Intraoperative final microscopic view (G) and ICG videoangiography (H) showing complete removal of the AVM and normalization of the bilateral frontal lobe hemodynamics.

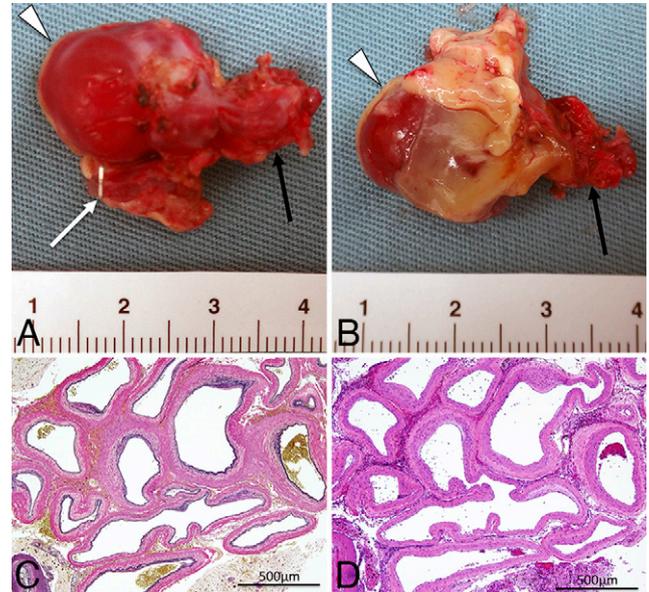


FIG. 5. Photographs of the surgical specimen (A and B) showing the varix (white arrowheads), drainer (white arrow), and nidus (black arrow). Photomicrograph (C, elastic van Gieson staining, original magnification $\times 40$) demonstrating the arterial and venous structures in the cerebral parenchyma. These findings corresponded to AVM. Photomicrograph (D, hematoxylin and eosin staining, original magnification $\times 40$) showing hemosiderin infiltrating around the vessels corresponding to old intracranial hemorrhage.

AVM and varix completely by coagulated the proper feeder with pial supply of AVM and coagulated drainer at the just distal to varix (Fig. 4G). Finally, we confirmed that no residual AVM and venous dilatation on the frontal lobe remained using ICG videoangiography (Fig. 4H).

Histological examination revealed vessels of various sizes in the brain parenchyma with hemosiderin, and the diagnosis was AVM (Fig. 5). In addition, intimal thickening with elastic lamina was partially observed, and the final diagnosis was varix. Postoperative angiography detected no residual DAVF and AVM. The patient's postoperative course was uneventful.

Discussion

The present case of DAVF coexisting with AVM using the same draining system in the ACF is unique. Only two cases of DAVF in the ACF with bilateral frontal drainers have been treated surgically.^{4,5} Therefore, the present bilateral case was certainly rare. In addition, our case featured coexistence of the intracranial DAVF and AVM in the same region. The three previous cases of coexistence of intracranial DAVF and AVM had these lesions located in different regions as follows: SSS DAVF and left occipital AVM; SSS DAVF and right temporal AVM; and tentorial DAVF and left cerebellopontine fissure AVM.⁶⁻⁸

Observations

In our case, the left DAVF in the ACF shared a common draining system with the AVM, as confirmed by cerebral angiography

and intraoperative visualization using ICG videoangiography during stepwise clipping of the drainer of the DAVF and feeder of the AVM. The concept of surgery for DAVF in the ACF is disconnection of the fistula which drains into a dilated vein.⁹ In the present case, the right DAVF was quite simple, so drainer occlusion could cure the right DAVF. However, the same procedure could not be effective on the left side, because the arterial supply for the AVM drained into the same dilated cortical vein as the left DAVF. These very interesting hemodynamics were confirmed by intraoperative ICG videoangiography using stepwise temporary clipping of the drainer of the DAVF and feeder of the AVM. This ICG videoangiography also demonstrated the pial supply for the AVM. Consequently, we could remove the AVM completely. If we had remained unaware of the complicated hemodynamic system in the left ACF and selected the surgical strategy of drainer occlusion only as a treatment for the DAVF, the patient might have suffered a miserable outcome.

Histological examination revealed that the AVM contained vessels of various sizes in the brain parenchyma with hemosiderin. The AVM is a lesion that is usually present at birth and grows proportionately with age.¹⁰ Consequently, venous hypertension following sinus thrombosis is the primary mechanism of the formation of DAVFs.^{6,11,12} Venous hypertension may promote the growth of microscopic arteriovenous shunts, which are found within the vasa vasorum of normal pachymeninges, and may stimulate the release of angiogenic factors in experimental models.¹²⁻¹⁴ In addition, head trauma is reported to be one of the etiologies of DAVF.^{2,15}

Our case had a history of head trauma 20 years previously. The present case can be summarized as bilateral DAVFs in the ACF coexisting with unilateral frontal AVM. The assumed cause of this

interesting clinical condition can be explained as follows: (1) left frontal AVM caused long-term venous hypertension of SSS; (2) subsequent increasing cortical venous pressure in the bilateral anterior frontal lobes; (3) acquired bilateral DAVFs developed in the ACF with expression of multiple angiogenic factors; (4) head trauma might contribute to the development of DAVF; and (5) asymptomatic left frontal intracranial hemorrhage occurred from the AVM or DAVF. Further clinical investigation may reveal more facts about this clinical condition.

Lessons

Neurosurgeons should be aware of the possibility of the coexistence of DAVF and AVM. Preoperative angiography to establish the angio-architecture in detail and plan the approximate operation strategies is important. ICG videoangiography during the stepwise occlusion of drainer of the DAVF and feeder of the AVM was quite useful to understand the complicated hemodynamics of DAVF coexisting with AVM.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Yamamoto, Shibahara, Kumabe. Acquisition of data: Yamamoto, Shibahara, Inukai, Niki, Usui, Kimura, Kumabe. Analysis and interpretation of data: Yamamoto, Inukai, Kumabe. Drafting the article: Yamamoto, Shibahara, Hide, Kumabe. Critically revising the article: Yamamoto, Kumabe. Reviewed submitted version of manuscript: Yamamoto, Hide, Kumabe. Approved the final version of the manuscript on behalf of all authors: Yamamoto. Administrative/technical/material support: Yamamoto, Koizumi, Hyakutake, Ishima. Study supervision: Shibahara, Kumabe.

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