A Case of Cavernous Sinus Dural Arteriovenous Fistula Presenting with Medulla Oblongata Dysfunction in Parallel to Thrombosis of a Varix on a Drainage Route after Transvenous Embolization

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Objective: We report a case of cavernous sinus dural arteriovenous fistula (CSdAVF) presenting with medulla oblongata dysfunction in parallel to thrombosis of a varix on a drainage route after transvenous embolization (TVE).

Case Presentation: A 76-year-old male presented with deep sensory disturbance. Cerebral angiogram revealed a right CSdAVF with retrograde venous drainage refluxing to the anterior medullary vein. A varix arising from the vein was buried in the medulla oblongata, and an edematous change was shown in the nerve tissue around the varix. TVE was successfully performed and the shunt flow completely disappeared. The neurological symptoms improved immediately after TVE. The following day after TVE, medulla oblongata dysfunction appeared again, which was more severe than that before TVE. MRI showed thrombosis and a volume increase of the varix, and an enlargement of edematous change in the nerve tissue around the varix. Administration of corticosteroids gradually improved neurological and imaging findings.

Conclusion: A varix on a drainage route can cause brainstem dysfunction in CSdAVF. Thrombosis of the varix may exacerbate neurological symptoms.

Keywords ► cavernous sinus dural arteriovenous fistula, transvenous embolization, varix, thrombosis, medulla oblongata dysfunction

Introduction

Dural arteriovenous fistula (dAVF) can develop brainstem dysfunction. Increased shunt flow is known as the mechanism,¹⁾ and no other mechanisms were mentioned. In addition, no study has clarified the influence on the

nerve tissue around a varix on a drainage route after thrombosis of the varix in dAVF.

We report a case of cavernous sinus dural arteriovenous fistula (CSdAVF) presenting with medulla oblongata dysfunction related to the varix on the drainage route, whose neurological symptoms exacerbate after transvenous embolization (TVE) in parallel to thrombosis of the varix.

Case Presentation

A 76-year-old male without medical or smoking history developed deep sensory disturbance of the right extremities. Consciousness was clear, and neither motor paralysis nor superficial sensory disturbance was observed. T2-weighted image showed many flow voids around the right Sylvian fissure (**Fig. 1A**). There was no parenchymal lesion in the bilateral cerebral hemispheres. An occupying lesion was observed in the ventral region of the medulla oblongata, which showed an iso intensity on T2-weighted image and a low intensity on diffusion-weighted image.

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Fig. 1 MRI before TVE. (A) T2-weighted image. Many flow voids were observed around the right Sylvian fissure. (B) T2-weighted image, (C) susceptibility-weighted image, and (D) diffusion-weighted image. Note a varix in the ventral region of the medulla oblongata. The varix showed an iso intensity on T2-weighted image and low intensity on diffusion-weighted image. The varix consisted of an iso intensity area and high intensity area on susceptibility-weighted image. Note an edematous change in the nerve tissue around the varix. TVE: transvenous embolization

Fig. 2 DSA before TVE. (A and B) Frontal view. (C and D) Lateral view. (A and C) Early phase. (B and D) Late phase. Note a dural arteriovenous fistula at the right cavernous sinus. A primary inflow vessel was the right artery of the foramen rotundum. Two outflow vessels from the right cavernous sinus were shown: the right superficial middle cerebral vein and the right superior petrosal sinus. Note a varix arising from the posterior wall of the anterior medullary vein (arrow). TVE: transvenous embolization

The lesion consisted of an iso intensity area and high intensity area on susceptibility-weighted image. An edematous change was shown in the nerve tissue around the lesion (**Fig. 1B–1D**). DSA revealed a right CSdAVF with a shunting point in the lateral, superior, anterior region of the right cavernous sinus (**Fig. 2**). The right artery of the foramen rotundum was a primary inflow vessel, and a small volume of blood was supplied from the jugular branch of the right ascending pharyngeal artery and the inferolateral trunk of the right internal carotid artery. As the venous drainage, two outflow vessels from the right cavernous sinus were shown. One is the right superficial middle cerebral vein, which drained to the superior sagittal sinus and to the right basal vein via the right deep middle cerebral vein. The other is the right superior petrosal sinus, which refluxed to the right inferior vermian vein via the right vein of the great horizontal fissure, to the left basal vein via the anterior pontomesencephalic vein, and to the anterior spinal

vein via the anterior pontomesencephalic vein and the anterior medullary vein. A varix arising from the posterior wall of the anterior medullary vein was observed. The varix was consistent with the occupying lesion in the ventral region of the medulla oblongata detected on MRI, shown to be buried in the medulla oblongata. The drainage via the varix had not contributed to normal venous return. The right superior ophthalmic vein and the right inferior petrosal sinus were not visualized. The normal venous drainage was not delayed either in the bilateral cerebral hemispheres or the brainstem. Borden classification was type II and Cognard classification was type IIa + b.

TVE was performed for the CSdAVF under local anesthesia. A 6Fr Roadmaster (Goodman, Aichi, Japan) was inserted into the right internal jugular vein via the right femoral vein. A 0.035-inch Radifocus guidewire (Terumo Corporation, Tokyo, Japan) was inserted to the right cavernous sinus through the thrombosed right inferior petrosal

Fig. 3 DSA after TVE (lateral view). **(A and B)** Embolization of the right superior petrosal sinus led to the disappearance of the reflux to the anterior medullary vein and the varix. **(C and D)** Embolization of the right superficial middle cerebral vein and anterior part of the right cavernous sinus led to the disappearance of the shunt flow. TVE: transvenous embolization

Fig. 4 MRI after TVE. **(A and D)** T2-weighted image. **(B)** Susceptibility-weighted image. **(C)** Diffusion-weighted image. **(A–C)** The following day after procedure. Note a volume increase in the varix and an enlargement of edematous change in the nerve tissue around the varix. The intraluminal space of the varix uniformly showed a low intensity on susceptibility-weighted image. There was no high intensity lesion around the varix on diffusion-weighted image. **(D)** 18 days after procedure. Note a volume reduction of the varix and a disappearance of the edematous change in the nerve tissue around the varix. TVE: transvenous embolization

sinus, then an Echelon (Medtronic, Dublin, Ireland) coaxial to a 0.012-inch Radifocus guidewire (Terumo) was inserted along the route. Selective embolization was impossible because the shunting point was difficult to access, so we performed outflow occlusion and sinus packing. First, the outflow to the right superior petrosal sinus was embolized using an ED COIL 10 ExtraSoft (Kaneka, Osaka, Japan), HyperSoft 3D (Terumo), and Target 360 Nano (Stryker, Kalamazoo, MI, USA), with a total length of 23 cm. After that, the reflux to the anterior medullary vein and the varix disappeared (**Fig. 3A and 3B**). Deep sensory disturbance improved at that point. Subsequently, the right superficial middle cerebral vein and anterior part of the right cavernous sinus were embolized using a HyperSoft 3D, VFC (Terumo), ED COIL 10 ExtraSoft, ED COIL infini 10 ExtraSoft, AXIUM 3D (Medtronic), DeltaPac (Micrus, Mountainview, CA, US), and Target 360 Ultra (Stryker), with a total length of 416 cm. Consequently, the

shunt flow on right external carotid angiography completely disappeared (**Fig. 3C and 3D**). In addition, right internal carotid angiography, left common carotid angiography, and left vertebral angiography showed no shunt flow and congestion around the brainstem. No additional neurological symptoms appeared after the procedure.

The following day after TVE, deep sensory disturbance of the bilateral extremities, hoarseness, impaired swallowing reflex, and hypoglossal nerve palsy appeared. Immediate DSA showed no different findings from those of the previous day. T2-weighted image showed a volume increase in the varix and an enlargement of edematous change in the nerve tissue around the varix (**Fig. 4A**). The intraluminal space of the varix uniformly showed a low intensity on susceptibility-weighted image (**Fig. 4B**), confirmed to be thrombosed. There was no high intensity lesion around the varix on diffusion-weighted image (**Fig. 4C**). Administration of corticosteroids gradually improved neurological

symptoms. Anticoagulation was not performed during the period. T2-weighted image obtained 18 days after TVE revealed a volume reduction of the varix and a disappearance of the edematous change in the nerve tissue around the varix (**Fig. 4D**). The patient was transferred to another hospital 21 days after TVE. Subsequent rehabilitation improved most neurological symptoms except slight discomfort on swallowing, and the patient was discharged 158 days after TVE with a modified Rankin Scale score of 1.

Discussion

The present case is a CSdAVF with deep sensory disturbance, which is an atypical neurological symptom for CSdAVF. TVE resulted in the disappearance of the shunt flow. Post-treatment course was also atypical; deep sensory disturbance disappeared immediately after TVE, then deep sensory disturbance, hoarseness, impaired swallowing reflex, and hypoglossal nerve palsy appeared the following day after TVE in parallel to thrombosis of the varix on the drainage route.

Each neurological symptom in the present case can be explained as a symptom of the medulla oblongata. The varix arising from the anterior medullary vein was buried in the medulla oblongata, caught between the right medial lemniscus and the left medial lemniscus. Deep sensory disturbance was consistent as a symptom due to dysfunction of the medial lemniscus. Furthermore, the hypoglossal nerve and the nucleus ambiguus were located lateral to the medial lemniscus. Hoarseness, impaired swallowing reflex, and hypoglossal nerve palsy were consistent as symptoms due to dysfunction of these sites. Thus, the present case can be regarded as CSdAVF presenting with medulla oblongata dysfunction which exacerbated after the shunt flow disappeared.

Increased shunt flow is known as a mechanism in which dAVF cause brainstem dysfunction. Concerning CSdAVF, 10 cases were reported.²⁾ In each of them, neurological and imaging findings improved after the shunt flow disappeared. On the other hand, in the present case, medulla oblongata dysfunction exacerbated and an edematous change enlarged after the shunt disappeared, which require the different mechanism from increased shunt flow.

Paradoxical worsening^{3,4)} and extensive venous thrombosis⁵⁾ are known as mechanisms in which neurological symptoms exacerbate in parallel to the disappearance of shunt flow. The former is venous return disturbance of the

superior ophthalmic vein mainly observed in CSdAVF. The latter is venous return disturbance of the spinal cord mainly observed in spinal dAVF. Anticoagulation is thought to be effective for both mechanisms because they are caused by thrombosis of the draining route. In the present case, thrombosis of the drainage route involving the varix may induce venous return disturbance causing neurological symptoms, and anticoagulation might have been effective.

Thrombosis of the varix may influence the surrounding nerve tissue in the present case because the neurological symptoms due to the surrounding area of the varix appeared in parallel to thrombosis of the varix and enlargement of an edematous change in the nerve tissue around the varix. To our knowledge, no study has reported that thrombosis of a varix induces neurological symptoms. On the other hand, thrombosis of an *aneurysm* was reported to have the potential to induce neurological symptoms in some case reports.^{6,7)} In each of the cases, symptoms appeared immediately or a few hours to days after thrombosis of an aneurysm in parallel to a volume increase in the aneurysm and an appearance of edematous change in the nerve tissue around the aneurysm. In addition, the administration of corticosteroids improved imaging findings, leading to complete or partial recovery from neurological symptoms after a few weeks to months. As the etiology for appearance of neurological symptoms due to the nerve tissue around the aneurysm, two mechanisms are discussed. One is a mechanical compression caused by volume increase in the aneurysm after thrombosis. The other is an edematous change for inflammatory responses induced by thrombosis. In the present case, the changes in neurological and imaging findings resembled those in these reports although the histological wall structure was different. As such, it is conceivable that thrombosis of the varix induced compression and/or edematous change of the medulla oblongata causing neurological symptoms in the present case. By the way, the neurological symptoms transiently improved immediately after the disappearance of the shunt flow in the present case. This may have been because a reduction in the shunt flow decreased the venous return pressure, relieving compression and/or edematous change of the medulla oblongata transiently.

This is the first report which refer to the pathology that thrombosis of a varix on the drainage route with occupying effects exacerbates local neurological symptoms in dAVF. Occupying effect for dilation of a drainage route is rare itself, except those with a vein of Galen dAVF. Iwamuro

et al.⁸⁾ reported a case of a dAVF at the tentorium, and Viñuela et al.⁹⁾ presented eight cases of pial and subpial arteriovenous fistulae. In all surgically treated cases among them, a procedure promoting thrombosis was performed for each varix, and no neurological exacerbation occurred after procedure. Furthermore, a rapid volume reduction of the varix was confirmed after procedure in each of cases for which the diameter of the varix was described. On the other hand, Kinouchi et al.¹⁰⁾ presented a case with a different course from them. According to their summary, outflow occlusion was performed for a symptomatic dAVF at the tentorium with a varix on the drainage route, and neurological symptoms exacerbated in parallel to thrombosis of the varix and an enlargement of the varix after procedure. The varix was resected after that, improving the neurological symptoms. Thrombosis of the varix may have influenced on the surrounding nerve tissue although the authors have not referred. In the nine cases presented by Iwamuro et al.⁸⁾ and Viñuela et al.,⁹⁾ each varix was consistent with the shunting point or connected to the shunting point through a dilated vein. On the other hand, in the case reported by Kinouchi et al.¹⁰⁾ and the present case, each varix was connected to the shunting point through the vein without dilatation. Differences in the structure of a drainage route may contribute to the appearance of local neurological symptoms related to thrombosis of the varix. This is the consideration with a small number of cases. More cases should be accumulated for further discussion.

In the present case, a varix on a drainage route of a CSdAVF caused medulla oblongata dysfunction, leading to an exacerbation of the neurological symptoms in parallel to thrombosis of the varix. From such a rare case, it is indicated that thrombosis of a drainage route can induce brainstem dysfunction in CSdAVF. Furthermore, it is suggested that a varix on a drainage route can influence on the surrounding nerve tissue causing local neurological symptoms in dAVF, and thrombosis of the varix may exacerbate the neurological symptoms in such a case. These conditions should be considered for perioperative management.

Conclusion

Thrombosis of a drainage route may induce brainstem dysfunction in CSdAVF. A varix on a drainage route may

influence on the surrounding nerve tissue causing local neurological symptoms in dAVF, and thrombosis of the varix may exacerbate neurological symptoms.

Disclosure Statement

There is no conflict of interest for the first author and coauthors.

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