Cervical Myelopathy Caused by Intracranial Dural Arteriovenous Fistula

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Intracranial dural arteriovenous fistula (dAVF) usually results in various problems in the brain. But it can be presented as a myelopathy, which may make early diagnosis and management to be difficult. We recently experienced a case of cervical myelopathy caused by intracranial dAVF. A 60-year-old man presented with a 3-year history of gait disturbance due to a progressive weakness of both legs. Neurological examination revealed spastic paraparesis (grade IV) and Babinski sign on both sides. Magnetic resonance imaging showed serpentine vascular signal voids at C2–T1 on T2-weighted image with increased signal intensity and swelling of spinal cord at C1–C4. We performed a brain computed tomography angiography and found intracranial dAVF with multiple arteriovenous shunts. Venous drainages were noted at tentorial veins and cervical perimedullary veins. After Onyx embolization, the patient showed gradual improvement in motor power and gait disturbance. The venous drainage pattern is a well-known prognostic factor of dAVF. In our case, the intracranial dAVF drained to spinal perimedullary vein, which seemed to result in the ischemic myelopathy. Although it is a rare condition, it sometimes can cause serious complications. Therefore, we should keep in mind the possibility of intracranial dAVF when a patient presents myelopathy.

Key Words: Brain · Arteriovenous fistula · Ischemia · Spinal cord diseases

INTRODUCTION

Intracranial dural arteriovenous fistula (dAVF) usually results in various problems of the brain like seizure or hemorrhage. But it sometimes can be presented with symptoms of myelopathy, which may make diagnosis and early management to be quite difficult. We recently experienced a case of upper cervical myelopathy caused by intracranial dAVF without any spinal vascular malformation.

CASE REPORT

A 60-year-old man visited Chung-Ang University Hospital with a 3-year history of progressive gait disturbance. Neurological examination revealed motor weakness (grade IV) of both legs with spastic gait and bilateral pathologic reflexes, Babinski sign and ankle clonus. Pain, temperature, and proprioceptive sensations were normal. Sphincter functions were well preserved. Cervical magnetic resonance imaging (MRI) showed serpentine vascular signal void from C1 to T1, and diffuse cord edema and swelling at C1 to C4 (Fig. 1). The first diagnostic impression was spinal arteriovenous fistula. We performed spinal angiography, but there was no evidence of vascular abnormality at the cervicothoracic spine. Then, we performed brain computed tomography angiography (CTA) to screen possible intracranial vascular abnormalities (Fig. 2). The brain CTA revealed dAVF at the posterior fossa, and subsequent transfemoral cerebral arteriography confirmed the vascular abnormalities (Figs. 2, 3). The dAVF received multiple arterial supplies from the meningohypophyseal artery of internal carotid...
Fig. 2. (A, B) Brain computed tomography angiography (CTA) showed abnormally dilated venous channels (white arrow) in the posterior fossa suggesting dural arteriovenous fistula. (C) Cervical perimedullary veins noted on Brain CTA (white arrow).

Fig. 3. (A) Arterial phase of right internal carotid angiogram showing arteriovenous fistula from the artery of the meningohypophyseal trunk (white arrow). (B) Arterial phase of right external carotid angiogram showing fistulas from the artery of foramen rotundum (black arrow) and the petrosal branch of the right middle meningeal artery (white arrow). (C) Venous phase of right internal carotid angiogram shows venous drainages to the prepontine (white arrow) and cervical perimedullary veins (black arrow). (D) Venous phase of right external carotid angiogram shows venous drainages to the cervical perimedullary vein (black arrow).

Fig. 4. Postembolization angiographs. (A) Arterial phase of right internal carotid angiogram shows complete occlusion of fistulas from the artery of the meningohypophyseal trunk (dashed circle). (B) Arterial phase of right external carotid angiogram shows complete occlusion of fistulas from the petrosal branch of the right middle meningeal artery (dashed circle) and residual fistula from the artery of the foramen rotundum (black Arrow). (C) Venous phase of right internal carotid angiogram shows complete occlusion of the prepontine and cervical perimedullary veins (dashed circle). (D) Venous phase of right external carotid angiogram shows complete occlusion of drainages to the cervical perimedullary vein (dashed circle).

We performed Onyx embolization under general anesthesia. We could not occlude all the feeding arteries, remaining 2 residual fistulas from the foramen rotundum artery. Venous drainages were connected to prepontine vein and cervical perimedullary vein (Fig. 3). We performed Onyx embolization under general anesthesia. We could not occlude all the feeding arteries, remaining 2 residual fistulas from the foramen rotundum artery. Venous drainages were connected to prepontine vein and cervical perimedullary vein (Fig. 3). We performed Onyx embolization under general anesthesia. We could not occlude all the feeding arteries, remaining 2 residual fistulas from the foramen rotundum artery. Venous drainages were connected to prepontine vein and cervical perimedullary vein (Fig. 3). We performed Onyx embolization under general anesthesia. We could not occlude all the feeding arteries, remaining 2 residual fistulas from the foramen rotundum artery. 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ticity. One year after the embolization, we performed brain CTA, there were decreased diameter of transverse pontine vein, aneurysmal dilatation in right mesencephalic vein and no other abnormal density in the brain. He still showed full motor power and mild spasticity.

**DISCUSSION**

Intracranial dAVF consists of pathological shunts connecting dural arteries with dural or cortical veins. Intracranial dAVF accounts for 10%–15% of intracranial arteriovenous malformations (AVMs). Intracranial dAVF may induce myelopathy by interruption of cervical spinal venous drainage. Cases of dAVF have been reported rarely since the first report in 1982. Our case also showed cervical myelopathy caused by an intracranial dAVF draining to the cervical perimedullary vein.

Symptoms of intracranial dAVF are known to largely depend on the patterns of venous drainage. There are 2 types of dAVF classification, Borden or Cognard classification systems, according to the patterns of venous drainage. The Borden classification divides the lesions according to the site of venous drainage and the presence of cortical venous drainage. The venous drainage were the dural sinus without cortical venous drainage and the presence of cortical venous drainage. Cases of dAVF with spinal cord lesion. Early diagnosis and early management are important for the favorable prognosis of dAVF especially in the case of high-risk types, Borden type III and Cognard type V.

Endovascular treatment is known to be the treatment of choice, and surgical treatment is a second option in cases the endovascular treatment is not feasible. The treatment goal is to solve the venous hypertension by complete occlusion of all arteriovenous shunts. If the fistulas and draining veins are not completely blocked, collateral vessels can be recruited and the bleeding risk will persist. However, when the complete occlusion is too difficult, selective occlusion of accessible fistulas may be the safer choice. We conducted an endovascular embolization with Onyx, a nonadhesive embolic agent composed of ethyl alcohol-dimethyl sulfoxide. It has been used for embolization of AVMs since the late 1980s and has been increasingly used in the recent years because of its ease of use and safety. The advantage of Onyx is that the operator can control its volume and rate of delivery. However, it requires longer time (7–100 minutes) to see the obliteration of the AVMs with fluoroscopic examination comparing to N-butyl-cyanoacrylate (less than 1 minute), which may increase the radiation exposure. In our case, we chose Onyx embolization because of the multiple feeding arteries to the lesion. Some feeders were too small or had too complicated curvature for selection, which made the endovascular occlusion to be incomplete. There were residual fistulas from artery of foramen rotundum after the embolization finished, but, fortunately, the perimedullary venous drainage was disappeared completely (Fig. 4).

**CONCLUSION**

The venous drainage pattern of intracranial dAVF seems to relate closely with the risk of complications. When the intracranial dAVF drains to the spinal perimedullary vein, it can inhibit the circulation of spinal cord and result in the ischemic myelopathy. Even though it is rare condition, we should keep in mind the possibility of intracranial dAVF when a patient shows cervical myelopathy without any spinal vascular abnormality.
CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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