Progressive Myelopathy Caused by Dural Arteriovenous Fistula at the Craniocervical Junction
—Case Report—

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Abstract
A 68-year-old male presented an unusual dural arteriovenous fistula (AVF) located at the craniocervical junction. Magnetic resonance imaging revealed dilated perimedullary veins around the spinal cord at C-1 and C-2 levels, as well as high intensity signals in the spinal cord on T2-weighted images. Vertebral angiography identified an AVF at the point where the right vertebral artery penetrates the dura. The fistula was a single and direct communication between the vertebral artery and the spinal vein. Surgical interruption of the fistula at its venous side resulted in prompt improvement of both motor and sensory signs and symptoms.

Key words: dural arteriovenous fistula, myelopathy, craniocervical junction, venous hypertension, single fistula

Introduction
Spinal dural arteriovenous fistula (AVF) is a rare condition but cannot be ignored in the differential diagnosis of myelopathy. Spinal dural AVF occurs mostly in the 5th and 6th decades of life with male preponderance, and most frequently occurs in the mid-thoracic to upper lumbar region. The radicular artery is the feeding vessel, with the nidus in the dura close to the root sleeve, and the venous drainage comes out intradurally into the perimedullary venous plexus, resulting in venous hypertension that is supposedly responsible for the myelopathic symptoms. We report a rare case of dural fistula with typical clinical features, caused by a direct, single communication between the vertebral artery and the spinal intradural veins at the craniocervical junction, not the multiple vascular channels imbedded in the dura.

Case Report
A 68-year-old male without significant history of medical disorder or trauma had been well until January 1996, when he started to complain of motor and sensory disturbances in his legs. The illness progressed gradually over 6 months, accompanied by difficulty in urination. Examinations at other hospitals failed to identify the definitive diagnosis, so he was finally transferred to Kobe City General Hospital on July 28 for further evaluation.

On admission, he was basically tetraparetic, more prominent in the lower than the upper extremities. He was hypesthetic in superficial sensations and anesthetic in deep sensations below the level of T-6, with urinary retention and slight dyspnea. Magnetic resonance (MR) imaging revealed numerous low intensity areas around the spinal cord at the C-1 and C-2 levels, suggesting dilated perimedullary veins. In addition, high intensity signals were found in the spinal cord at the C-6 and C-7 levels (Fig. 1 left). Vertebral angiography revealed an AVF, exactly at the spot where the right vertebral artery penetrates the dura (Figs. 2 and 3). The fistula was a single and direct communication between the right vertebral artery and the intradural spinal veins, with no fine vascular channels as seen in ordinary nidus-like structures. The drainage from the fistula was into the intradural perimedullary veins, with no contribution from the external carotid branches. Based on the neuroradiological findings, we speculated that the
venous hypertension caused by the AVF hampered the normal venous circulation in and around the spinal cord, leading to the development of progressive myelopathy.

With the patient in the prone position, a small suboccipital craniectomy was performed with removal of the C-1 and C-2 laminae. No abnormalities were found epidurally. Intradural investigation showed markedly dilated tortuous red veins on the dorsal surface of the apparently normal spinal cord. The fistula was located intradurally at the spot where the right vertebral artery penetrates the dura mater. However, there were no abnormalities recognized around the dura in the vicinity of the fistula. The fistula was successfully interrupted at its venous side with an aneurysm clip, resulting in the dilated veins on the surface of the spinal cord changing from arterial to venous color.

Postoperative angiography confirmed complete obliteration of the fistula (Fig. 4). MR imaging also showed disappearance of the flow void signs as well as the high intensity signals in the spinal cord (Fig. 1 right). Clinical signs and symptoms dramatically improved after surgery. The muscle strength of his upper extremities recovered to the normal range and the sensory disturbances descended to the level of the T-10 dermatome. His postoperative course was smooth and he was transferred to a nearby hospital for rehabilitation.

Discussion

Spinal dural AVF was first reported as early as in 1911 under the designation of “angioma venosus racemosum” of the dorsal thoracic spinal cord, but not until lately have the anatomical and pathophysiological details become well understood. In the 1940s and 1950s, pathologists had already noticed that the vessels on the dorsal surface of the spinal cord in the disease were dilated veins, not anomalous vessels. However, the introduction of the spinal angiography confused the essentials of the disease, although it clearly presented radiological evidence of arteriovenous shunt as a definite component of the disease. For example, Di Chiro, one of the pioneers of spinal angiography, regarded this as a variation of intradural arteriovenous malformations. The current concept of the spinal dural AVF was established by the observation that the intradural dilated vessels around the spinal cord are only draining veins, whereas the nidus is located in the dura around the root sleeve. The clinical characteristics of the disease have gradually become clear, with the accumulation of clinical reports.

Today, spinal dural AVF is known to be the commonest among spinal vascular malformations and is at a key position in the differential diagnosis of myelopathy without apparent causes. Our patient shares the common clinical features of typical spinal dural AVFs as far as age, sex, neurological manifestations, mode of onset, and draining pattern are concerned. This case is unusual in the location at the craniocervical junction, and the form of the fistula.

Only seven well-documented cases of dural AVFs located at the craniocervical junction manifesting as ascending myelopathy have been reported. All patients were in the 5th or 6th decades of life, all male except one patient, and presented with myelopathy of gradual onset. The ascending pharyngeal artery was involved in many cases as the feeding vessel, other than the vertebral artery. Motor and sensory disturbances in the legs were usually the initial signs of this disorder. The discrepancy between neurological, topographical findings and location of the AVF is likely to delay earlier diagnosis of the illness. Today, MR imaging is the first step to reach the diagnosis. Demonstration of the dilated perimedullary veins around the spinal cord and the abnormalities of the spinal cord (swelling and/or change of intensity) are the key points indicating this condition. However, angiography is essential to confirm the final diagnosis. Cerebral angiography is also important to rule out the presence of intracranial dural AVFs, because these can also cause...
myelopathy when the downward intradural venous drainage is significant enough to hamper the normal venous circulation in and around the spinal cord.4)

Typically, spinal dural AVF should have a nidus component imbedded in the dura around the root sleeve.14) In the present case, angiography failed to demonstrate any nidus-like component in the dura, which was confirmed during surgery. The fistula was a simple and single communication between the right vertebral artery and the intradural spinal vein. A similar case in location and form of fistulous connection has been reported.13) A new classification of dural AVFs based on the pattern of venous drainage, regardless of whether intracranial or spinal location, has been reported. Each type is further subdivided into two groups according to the presence of simple or multiple 3) Simple fistula is defined as a direct connection between the feeding artery and the draining vein as in our case. From the

Fig. 2 Preoperative right vertebral angiograms (anteroposterior view) showing the arteriovenous fistula draining into the perimedullary intradural veins.

Fig. 3 Preoperative right vertebral angiograms (lateral view).

Fig. 4 Postoperative right vertebral angiograms (left: anteroposterior view, right: lateral view) showing complete disappearance of the fistula.
therapeutic point of view, as far as spinal dural AVFs with only intradural drainage are concerned, an interruption of the fistulous connection at the venous side is adequate to both extirpate the fistula completely, and to relieve the myelopathic symptoms, irrespective of the form of fistulas. Clinical experiences also confirm inclusion of the simple type under the category of dural AVFs is reasonable, even when there are no dura-based nidus-like vascular channels as in this case.

With advancement of interventional techniques, embolization of spinal AVFs has become an alternative to surgery. Glue, such as N-butyl cyanoacrylate, is commonly used embolic material, expecting permanent cure, while particles such as polyvinylalcohol have been known to provide only temporary occlusion, since early recanalization is the rule. The main advantage of endovascular treatment lies in its less invasiveness, but it does not necessarily mean that the risk is lower. We think that surgical interruption of the fistula still could be the first choice, as far as spinal AVFs only with intradural drainage are concerned, since the surgical procedure is fairly simple, the risk involved remains within an acceptable range and, above all, it brings about instantaneous, complete, definitive cure.

References


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