Case Report

A 66-year-old female presented with progressive dysesthesia and hypalgesia ascending from both toes to the thighs on both sides. A neurological examination revealed that the cranial nerve, deep tendon reflex, strength, and coordination of both upper and lower extremities were almost normal and that the dysesthesia and hypalgesia occurred at less than the L3 level. Her symptoms were suspected to be due to a spinal dural AVF, inflammatory demyelinating disease, or a spinal tumor at the lumbar level. Thoracic and lumbar magnetic resonance imaging (MRI) with a T2-weighted image revealed no lesion. Her symptoms were not due to a pathological reason. She underwent a medical examination by several orthopedists and a neurologist. Cervical MRI with a T2WI revealed a high intensity lesion on the dorsal surface of the cervical cord between the C2 and C4 levels (Fig. 1A, B). Cervical enhanced MRI revealed serpentine vessels around the cervical spinal cord and medulla oblongata. She underwent angiography, and an intracranial dural AVF was suspected. Eight months after onset, she was referred to our institution. A neurological examination revealed normal cranial nerves, normal motor function of both upper and lower extremities, both dysesthesia and hypalgesia ascending from the toes, without brainstem signs. Moreover, we should perform cerebral angiography as early as possible because dural AVFs with slow-flow venous drainage can produce false negatives on magnetic resonance angiography.

Keywords: dysesthesia, intracranial dural AVF, myelopathy, cerebral angiography

Introduction

A spinal dural arteriovenous fistula (AVF) occasionally produces motor and sensory disturbances of the lower extremities. Generally, intracranial dural AVFs result in headache, pulsatile tinnitus, ocular symptoms (chemosis, exophthalmos, and diplopia) and focal signs, including hemiparesis and hemisensory disturbances. In the present case, we experienced a patient with an intracranial dural AVF that was identified by ascending dysesthesia and hypalgesia from both toes to the thighs on both sides, no motor paresis of the lower extremities, and no symptoms of the upper extremities. Therefore, we describe this case and review the past relevant literatures.

Case Report

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In order to reduce the venous hypertension and obliterate the arteriovenous shunt, transvenous embolization of the left sigmoid sinus and transverse sinus was performed. After
embolization, the left external carotid angiography showed obliteration of the fistula and disappearance of the spinal perimedullary venous drainage. Three months postoperatively, a cervical MRI with T2WI revealed a remarkably decreased extent of high intensity. The symptoms of dysesthesia and hypalgesia improved gradually (Fig. 1E–G).

Discussion

Myelopathy due to venous hypertension of the spinal cord that is induced by a spinal dural AVF is a relatively commonly encountered vascular malformation, and the fistula most often occurs in the thoracic region. Woimant et al. were the first to describe intracranial dural AVFs with perimedullary venous drainage in 1982. Since the first reports, seven cases of transverse-sigmoid sinus dural AVFs with perimedullary venous drainage have been reported. The findings of spinal cord swelling with abnormal MRI signals are non-specific and could be considered to be caused by a tumor, demyelination, myelitis, and/or trauma. Previous reports have emphasized that when symptoms suggest a spinal dural AVF and a spinal angiography is normal, we should examine the intracranial region because Cognard et al. have reported that type V dural AVFs produce progressive myelopathy in 50% of the cases and the other 50% of cases exhibit hemorrhage. Although type V dural AVFs are rare, they are rapidly progressive in 25% of the cases. It was extremely rare that the present case only manifested the sensory disorders of dysesthesia and hypalgesia. Therefore, an atypical dural AVF could lead to a delayed or incorrect diagnosis. The present case had another drainage route into the contralateral transverse-sigmoid sinus besides the spinal perimedullary vein, and therefore could avoid the rapid progression of myelopathy. In short, the presence of several drainage routes could reduce the venous hypertension by dividing the venous flow. Kwon et al. have reported that an intracranial dural AVF with slow venous flow was not visualized with magnetic resonance angiography (MRA) resulting in a false negative. Haryu et al. described contrast-enhanced dynamic MRA was more sensitive and useful in detecting enlarged spinal veins compared to T2-weighted MRI. By using digital subtraction angiography, visualization of the venous drainage pattern, as well as prompt and correct diagnosis, can be achieved. Therefore, when symptoms and MRI/MRA findings suggest a spinal dural AVF but spinal angiography does not show any abnormality, intracranial angiography should be performed because intracranial dural AVFs (Cognard type V) progressively produce myelopathy.

Aminoff et al. were the first to propose the theory of venous hypertension, and Merland et al. reported that venous hypertension might be produced by any cranial or spinal...
dural AVFs in 1974. Venous hypertension is considered to result in arterial flow into the spinal perimedullary veins and low perfusion, causing the stagnation of blood flow and ischemia of the spinal cord. Moreover, although the intradural spinal venous system is valveless and the venous perimedullary coronal plexus is in direct continuity with the veins of the posterior fossa, a venous congestive myelopathy is facilitated by the transmission of a high venous pressure to the spinal cord tissue, Suh and Alexander have suggested that there are valve structures between penetrating veins and medium-sized intramedullary veins because of the obstruction in a dye-injection experiment and that the valves of veins in the spinal cord are responsible for the tendency of higher venous pressure in the outer circumference. These reasons suggest the possibility that the sensory disorder was dominant in the lower extremities. In this case, the dysesthesia and hypalgesia except for the motor function, could account for the disorder of the nerve fiber of the lateral spinothalamic tract and the dorsal column in the leg area that ran up the outermost region.

In our review of previous reports, 60 cases of intracranial dural AVFs leading to myelopathy have been reported (Table 1). The patients ages ranged from 20 years to 79 years (mean, 57 years), with the male to female sex ratio of the 60 cases being 3:1 (Table 2). Most intracranial dural AVFs with spinal venous drainage cause myelopathy and

| Case | Age y/o, sex | Symptoms | Brainstem sign (+/−) | Feeder/Drainage | Site of shunt | Delay in diagnosis (months) | Outcome |
|------|--------------|----------|----------------------|-----------------|--------------|-----------------------------|---------|
| 1 (20) | 43, M | Tetraplegia, (−) | OA, MHA/Petrosal vein, | Petrous apex | 20 | GR |
| 2 (20) | 68, M | Paraplegia, (−) | OA, MHA/PMV | Torcular | 6 | SD |
| 3 (20) | 42, M | Paraplegia, (−) | OA, APA/Petrosal vein, PMV | Tentorial | 7 | MD |
| 4 (2) | 35, F | Tetraplegia, (+) | OA, MMA/PMV | Lateral sinus | 4 | GR |
| 5 (2) | 37, M | Tetraplegia, (+) | MMA/Petrosal sinus | Petrosian region | 9 | Death |
| 6 (2) | 53, M | Tetraplegia, (−) | MHA/PMV | Tentorial | 5 | SD |
| 7 (2) | 69, M | Paraplegia, (−) | APA, OA/SPS | Petrus sinus | 12 | GR |
| 8 (2) | 68, F | Tetraplegia, (+) | OA, APA, MMA/SPS | Petrous sinus | 4 | MD |
| 9 (18) | 69, M | Paraplegia, (−) | MHA/PMV | Torcular | Unknown | Unknown |
| 10 (20) | 40, M | Paraplegia, (−) | MHA/PMV | Tentorial | 12 | GR |
| 11 (20) | 63, M | Paraplegia, (−) | PMV/PMV | FM | 4 | GR |
| 12 (20) | 74, M | Paresthesia, (-) | PMA/PMV | FM | 6 | Death |
| 13 (7) | 50, M | Tetraplegia, (−) | MHA, OA/SPS, PMV | Petrous sinus | 7 | GR |
| 14 (7) | 71, M | Tetraplegia, (−) | OA, MMA/SPS, PMV | Petrous sinus | 4 | GR |
| 15 (19) | 78, F | Paraplegia, (−) | Unknown/PMV | Tentorial | 6 | SD |
| 16 (19) | 42, M | Paraplegia, (−) | Unknown/PMV | Tentorial | Unknown | Unknown |
| 17 (20) | 31, M | Paraplegia, (−) | MHA/TS, pontomesencephalic vein | Tentorial | 4 | MD |
| 18 (20) | 36, M | Tetraplegia, (−) | MMA, VA/Inferior vermain vein | Torcular region | 1 | SD |
| 19 (20) | 47, M | Tetraplegia, (−) | VA/Inferior vermain vein | Torcular region | 12 | SD |
| 20 (20) | 74, M | Tetraplegia, (−) | VA/PMV | FM | Unknown | SD |
| 21 (20) | 50, F | Paraplegia, (−) | OA, MMA/Lateral venous sinus | Lateral sinus | 6 | GR |
| 22 (20) | 67, M | Tetraplegia, (−) | APA, OA/PMV | Petrous sinus | 6 | MD |
| 23 (12) | 36, M | Tetraplegia, (−) | VA/PMV | Tentorial | 12 | SD |
| 24 (4) | 69, M | Tetraplegia, (−) | ICA meningeal branch/PMV | Tentorial | 36 | Death |
| 25 (4) | 53, M | Paraplegia, (−) | ICA meningeal branch/Petrosal vein, PMV | Tentorial | 6 | GR |
| 26 (4) | 40, F | Tetraplegia, (+) | ICA meningeal branch/ SOV, PMV, SPS | Cavernous sinus | 12 | Death |
| 27 (4) | 75, F | Tetraplegia, (−) | MMA/PMV | Petrous sinus | Few days | GR |
| 28 (4) | 51, F | Paraplegia, (+) | OA, MMA/right sigmoid sinus | Sigmoid sinus | 3 | GR |
| 29 (20) | 68, M | Tetraplegia, (−) | VA/PMV | CCJ | 6 | MD |
| 30 (15) | 70, M | Dysesthesia, (−) | VA/PMV, AMV | CCJ | 27 | GR |
| 31 (20) | 46, M | Paraplegia, (−) | APA/PMV | Unknown | Few days | GR |
| 32 (13) | 65, M | Tetraplegia, (+) | OA, PMA/Petrosal vein | Torcular region | 3 | GR |
| 33 (17) | 69, M | Paraplegia, (−) | APA, VA/PMV | CCJ | 48 | MD |
| 34 (17) | 53, M | Tetraplegia, (−) | APA/PMV | FM | 24 | GR |
| 35 (10) | 58, M | Tetraplegia, (−) | VA/PMV | CCJ | 6 | Death |
| 36 (18) | 64, M | Paraplegia, (−) | APA/PMV | Tentorial | 0.5 | SD |
| 37 (6) | 71, M | Paraplegia, (−) | ICA meningeal branch/SPS | Petrous sinus | Unknown | MD |
| 38 (6) | 47, M | Tetraparesis, (−) | ECA, VA/Petrosal vein | FM | 5 | SD |
| 39 (6) | 58, F | Tetraparesis, (−) | APA/Petrosal sinus | Skull base | Unknown | SD |

(Continued)
Table 1 (Continued)

| Case | Age y/o, sex | Symptoms | Brainstem sign (+/–) | Feeder/Drainage | Site of shunt | Delay in diagnosis (months) | Outcome |
|------|--------------|----------|----------------------|-----------------|--------------|-----------------------------|---------|
| 40 (20) | 57, M | Tetraplegia, (–) | APA/PMV | Transverse sinuses fissure | FM | 36 | SD |
| 41 (20) | 79, M | Tetraplegia, (–) | VA, APA, posterior auricular branch/PMV | Tentorium | 6 | MD |
| 42 (20) | 58, M | Paraplegia, (–) | MHA/PMV | Skull base | 0.5 | GR |
| 43 (16) | 68, M | Dysesthesia, (–) | APA/AMV, PMV | CCJ | 10 | MD |
| 44 (12) | 45, M | Paraplegia, (–) | OA, APA/PMV | Tentorium | 2 | SD |
| 45 (20) | 42, M | Tetraplegia, (–) | ICA meningeal branch/PMV | Tentorium | 15 | MD |
| 46 (1) | 60, M | Tetraplegia, (–) | MHA/vein of tentorium, cerebellomedullary fissure | CCJ | 1 | GR |

(Continued)

Table 2  Characteristic of epidemiology and clinical findings in 60 cases

| Mean age (y/o) | 57 |
| Location | Sex (M:F) | 45:15 |
| Tentorium | 20 (33%) |
| Foramen magnum | 8 (13%) |
| Craniocephalic junction | 8 (13%) |
| Petrous sinus | 7 (12%) |
| Transverse-sigmoid | 6 (10%) |
| Torcular region | 3 (5%) |
| Others | 8 (13%) |
| Clinical manifestation | Tetraparesis | 35 (58%) |
| Paraparesis | 22 (37%) |
| Dysesthesia | 2 (3.3%) |
| Hemiparesis | 1 (1.7%) |
| Brainstem dysfunction | 20 (33%) |
| Prognosis | Good recovery (GR) | 19 (32%) |
| Moderate disability (MD) | 17 (28%) |
| Severe disability (SD) | 19 (31%) |
| Death | 5 (8%) |
| Delay in diagnosis (mean months) | 3.9 |
| Brainstem symptoms (+) | 11.9 |
| Brainstem symptoms (–) | 10.3 |
| Good prognosis (GR, MD) | 11.3 |
| Poor prognosis (SD, death) | 0 |

motor function disorder, and the clinical manifestations included paraparesis in 22 cases, tetraparesis in 35 cases, and hemiparesis in one case. Moreover, the clinical outcomes of the reviews were unfavorable: 19 of 60 cases (32%) showed a good recovery, 17 cases (28%) showed moderate disability, and 24 cases (40%) showed a poor outcome or death after treatment. We conducted a quantity comparative statistical analysis using a Student’s t-test. All statistical results were considered significant if P < 0.05. The analysis was performed with EZR (Saitama Medical Center, Jichi Medical University, Saitama), which is a graphical user interface for R (The R Foundation for Statistical Computing). While it was a small population, the comparison of the delay in diagnosis from onset between the brainstem sign positive group and the negative group showed that brainstem sign positive group had a significantly shorter delay than the negative group did (3.4 months vs. 9.6 months, respectively; P = 0.021). However, there was no significant difference between the prognosis and the delay in diagnosis (good prognosis: 10.3 months vs. poor prognosis: 11.3 months, P = 0.35). These results suggest that intracranial dural AVFs with brainstem signs experience aggressive progression, but the correlation between the disease duration and prognosis is relatively low in literature review. We speculate that severity of venous hypertension due to venous drainage pattern is an important factor influencing prognosis. However, it is important to arrive at a prompt and accurate diagnosis because early
diagnosis offers the possibility of improving the reversible symptoms and avoiding the poor outcomes.

Conclusion

The clinical manifestations of dural AVFs are related to venous drainage pattern and, not to the fistula location. Therefore, the symptoms of intracranial dural AVFs with spinal perimedullary venous drainage (Cognard type V) are often related to the spinal dysfunction, rather than to the brain. We should perform cerebral and spinal angiography without delay and consider that angiography should be performed as early as possible because the intracranial dural AVFs with slow-flow drainage may produce false negative findings on MRA. Delayed and incorrect diagnosis might result in the poor outcomes of irreversibly severe spinal injury and Foix-Alajouanine syndrome, which is considered to be the result of progressive vascular thrombosis resulting in a necrotic myelopathy due to spinal-continuous venous hypertension.

Conflicts of Interest Disclosure

The authors declare that they have no competing interests and no financial support.

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