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Factors Associated with Rapidly Deteriorating Myelopathy in Patients with Spinal Arteriovenous Shunts

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Abstract

Spinal arteriovenous (AV) shunts are rare conditions that sometimes present with myelopathy symptoms. The progression of the symptoms is usually gradual; however, some cases show rapid deterioration. We retrospectively investigated the factors that induced the rapid deterioration of myelopathy symptoms in patients with spinal AV shunts. We treated 33 patients with myelopathy with spinal AV shunts at our institutions, eight of whom experienced rapid deterioration (within 24 hours: 24.2%). Of these, three were related to the body movement or particular postures associated with playing golf, 30 minutes of Japanese straight sitting, and massage care. One patient showed deterioration after embolization for a tracheal aneurysm. The remaining four patients received steroid pulse therapy (high-dose steroid infusion) shortly before the rapid deterioration. These symptoms stopped progressing after cessation of steroid use. While positional or physical factors contributing to myelopathy deterioration might exist, we could not identify specific factors in this study. Nevertheless, rapid deterioration was frequently observed after high-dose steroid use. We must take care not to administer high-dose steroids for myelopathy caused by spinal AV shunt disease.

Keywords: spinal arteriovenous shunts, myelopathy, steroid

Introduction

Spinal arteriovenous (AV) shunts or malformations (abnormal connections between arteries and veins of the spine) are rare, and they are classified with classification systems such as dural arteriovenous fistula (AVF), intradural intramedullary glomus arteriovenous malformation (AVM), intradural intramedullary juvenile AVM, perimedullary AVF, and extradural AVF.[1,2] Of these, spinal dural arteriovenous fistulas (SDAVFs) are the most common vascular malformations of the spinal cord and its coverings.[3] Generally thought to be an acquired lesion, SDAVFs consist of an abnormal and direct connection between the dural branch of a radicular artery and a radicular vein along the inner dural surface, typically lateral to the intervertebral foramen near the nerve root. Venous drainage occurs in a retrograde fashion from the radicular vein into the perimedullary venous plexus. Once these veins become "arterialized" and their pressure increases, the normal anterograde venous drainage of the spinal cord is compromised. Arterialization of the perimedullary venous plexus results in venous congestion of the spinal cord and myelopathy, and in some cases, spinal cord infarction.[3–5]

The precise diagnosis of myelopathy caused by spinal AV shunt disease remains very difficult because the clinical features and noninvasive imaging results may be nonspecific. Intramedullary high-intensity lesions in magnetic resonance (MR) images are often erroneously diagnosed as a variety of other

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diseases. As a result, it often takes a long time (some months or more) to diagnose. Myelopathy symptoms caused by AV shunt disease progress slowly in most cases. Symptoms include lower extremity weakness and sensory impairment, pain, gait disturbance, and urinary and bowel dysfunction. However, in some cases, there is rapid progression during the time course to final diagnosis. To avoid this rapid deterioration in future patients, we reviewed previous medical records and investigated the factors that caused rapid deterioration of myelopathy in patients with spinal AV shunts.

Patients and Methods

We retrospectively reviewed the medical records of patients with spinal AVF with myelopathy who were treated at Kurume University Hospital and Kumamoto University Hospital from January 2010 to May 2020. Patients with hemorrhagic events were excluded. Each patient's symptoms and clinical course recorded in summary and charts were investigated. The contributing factors to rapid deterioration were examined. We defined rapid deterioration of myelopathy as follows: within 24 hours after a definite event, there was remarkable deterioration manifesting as paresis, sensory disturbance, gait disturbance, or urinary and bowel dysfunction. We defined subacute deterioration of myelopathy as deterioration occurring within 1–3 days after a definite event.

Statistical analyses

Statistical analyses were performed between cases of rapid deterioration and other cases in terms of sex, lesion level (categorized into three groups: craniocervical junction and cervical, thoracic, lumbar and sacral lesion), age, worst manual muscle test (MMT) before surgical interventions, and symptom duration. All statistical analyses were conducted using commercially available software (JMP v16.0; SAS Institute, Cary, NC, USA), and the significance level was set at $p < 0.05$.

This clinical study was reviewed and approved by the institutional ethical committee of Kurume University (No. 2020-012) and Kumamoto University (No. 1760).

Results

Characteristics of patients

We found 33 patients with myelopathy caused by spinal AV shunt disease (ten at Kurume University Hospital and 23 at Kumamoto University Hospital). Of the 33 patients, the sex distribution was 28 men and five women. The age distribution was 43–84 years (mean 65.9 years). Spinal AV shunts were located at the craniocervical junction in four, cervical in one, thoracic in 18, lumbar in ten, and sacral in two (one patient had double spinal lesions, and this was reported previously).[6] The most frequent spinal AV shunts were SDAVFs (26/33, 78%); some patients had other types of spinal AV shunts, four cases of perimedullary AVF and three cases of extradural AVF. The most frequent symptom was motor weakness (93.9%), followed by gait disturbance (90.0%), sensory disturbance (81.8%), and urinary sphincter dysfunction (51.5%) (Table 1).

Factors contributing to deteriorations

Eight patients had episodes of rapid deterioration of myelopathy (Tables 1 and 2). Of these, three patients showed rapid deterioration after ordinary nonspecific activities: one after playing golf, one (a calligrapher) deteriorated after sitting erect with his legs folded (Japanese proper sitting position) for 30 minutes, and one case after massage care (shown in Table 3). The other four patients deteriorated after steroid pulse therapy. The remaining patient rapidly deteriorated immediately after embolization for tracheal aneurysm in another hospital, which may have been caused by altered flow that exacerbated the spinal AV shunt symptom. In this study, two institutes (Kurume University and Kumamoto University) cared for two patients who showed rapid deterioration after steroid pulse. After these experiences, no steroid pulse treatment was performed for patients suspected of having spinal AVF disease.

Three patients had episodes of subacute (1–3 days) deterioration of myelopathy after some events. Of these three patients, one deteriorated after low-dose steroid use, one deteriorated after administration of prostaglandin E_1 (PGE_1), and one deteriorated after climbing stairs.

The results of statistical analyses showed no significant differences in sex ($p = 1.000$), lesion level ($p = 0.7289$), age ($p = 0.6418$), and symptom duration ($p = 0.7503$) between patients who experienced rapid deterioration and those who did not. However, significant difference was found in worst MMT before surgical interventions ($p = 0.0354$).

Case presentations

Case 1, a 72-year-old man: The patient underwent laminoplasty previously for cervical ossification of posterior longitudinal ligament. Lower extremity weakness and urinary retention occurred 8 years later. The symptoms gradually worsened despite having received physical therapy at another hospital. Magnetic resonance imaging (MRI) revealed hyperintensity at the medulla and cervical cord (Fig. 1A).

Table 1 Clinical data of the 33 patients with myelopathy caused by spinal AVF disease

Sex (F:M)	5:28 (male: 84.8%)
Age (mean)	65.9 years (43–84 years)
Time to diagnosis (mean)	19.5 months (4 days–10 years)
Location of AV shunt	
Craniocervical junction	4 (11.4%)
Cervical	1 (2.8%)
Thoracic	18 (51.4%)
Lumbar	10 (28.5%)
Sacral	2 (5.7%)
Symptoms	
Weakness	31 (93.9%)
Gait disturbance	30 (90.0%)
Sensory disturbance	27 (81.8%)
Urinary sphincter dysfunction	17 (51.5%)
Deterioration pattern	
1) Rapid (<24 h), n = 8	
Ordinary activity	3
Golf	(1)
Japanese sitting 30 min	(1)
Massage care	(1)
Medical procedure	5
Other lesion embolization	(1)
Steroid pulse	(4)
2) Subacute (1–3 days), n = 4	
Ordinary activity	1
Climbing up stairs	(1)
Medical procedure	3
Steroid (low dose)	(1)
PGE_1 agent	(2)

AVF: arteriovenous fistula, F: female, M: male, AV: arteriovenous, PGE_1: prostaglandin E_1.

Table 2 Summary of four cases deteriorated during ordinary daily activities

Sex	Age (years)	Level	Symptoms	Induced activities	Deteriorating course
M	61	Th7	L/E weakness and sensory impairment, gait disturbance, urinary sphincter dysfunction	Golf	Rapid
M	74	L4	L/E weakness, gait disturbance	Massage	Rapid
M	75	Th6	L/E weakness and sensory impairment, gait disturbance	Sitting	Rapid
F	43	Th7, Th8, Th9, Th12, L3	L/E weakness, urinary sphincter deterioration	Climbing stairs	Subacute

M: male, F: female, L/E: lower extremity.

Table 3 Lower extremity MMT and other symptoms before and after the event of rapidly deteriorated patients

	Before		After	
	L/E MMT	Others	L/E MMT	Others
1 (Steroid)	4	S, P	3	S, P
2 (Tracheal aneurysm embolization)	4		3	U
3 (Steroid)	3	S, U	0–1	S, U
4 (Steroid)	4–5		3	U
5 (Steroid)	4	S, U	0–1	S, U
6 (Sitting)	4–5	S	3	S
7 (Golf)	5	S	4	S, U
8 (Massage)	4–5		3	

MMT: manual muscle test, L/E: lower extremity, S: sensory disturbance, P: pain, U: urinary impairment.

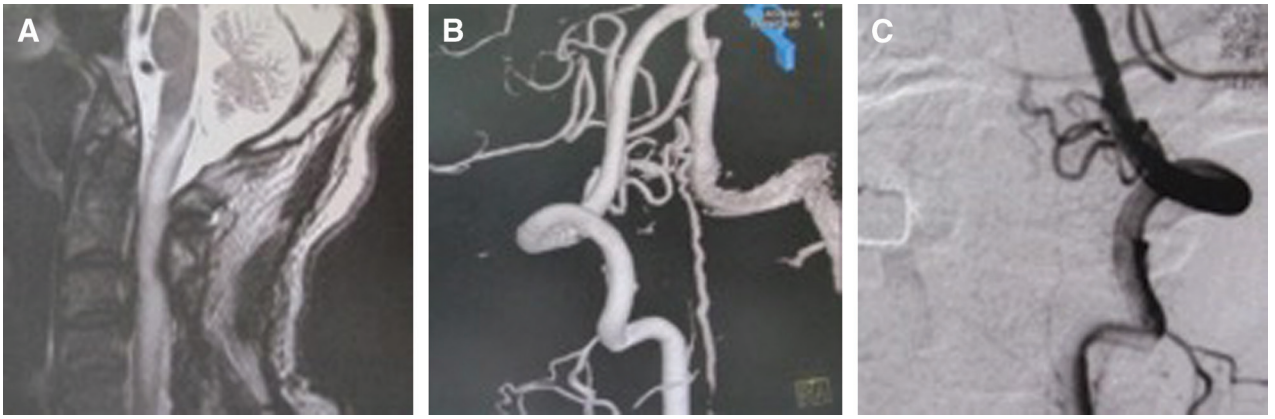

Fig. 1 (Case 1 in Table 4) (A) MRI T2WI revealing high-intensity lesion at the medulla and cervical cord. (B and C) CT angiography and digital subtraction angiography revealing dural AVF at the craniocervical junction. 72-year-old man: The patient had laminoplasty before cervical OPLL; 8 years later, R/E extremity weakness and urinary retention occurred. The symptoms gradually worsened despite having received physical therapy at another hospital. MRI revealed hyperintensity at the medulla and cervical cord. SDAVF was suspected, and the patient was referred to our service. The symptoms rapidly worsened after administration of steroids (hemiparesis MMT 3 ⇒ tetraparesis MMT 1). Emergent angiography revealed SDAVF at the cervico-medullary junction. The patient's symptoms gradually resolved after surgical treatment and physical therapy. AVF: arteriovenous fistula, CT: computed tomography, MMT: manual muscle test, MRI: magnetic resonance imaging, OPLL: ossification of posterior longitudinal ligament, SDAVF: spinal dural arteriovenous fistula, T2WI: T2-weighted image.

SDAVF was suspected, and the patient was referred to our service. The symptoms rapidly worsened (hemiparesis MMT 3 ⇒ tetraparesis MMT 1) after administration of steroids (betamethasone 1000 mg/day for 3 days). Emergent angiography revealed SDAVF at the cervicomedullary junction (Fig. 1B and 1C). The patient's symptoms gradually resolved after surgical treatment and physical therapy.

Case 2, a 45-year-old woman: The patient was hospitalized at another hospital because of motor weakness, pain, dysesthesia, and hyperreflexia in both lower extremities and gait disturbance; she was suspected of having multiple sclerosis. Initial MR T2-weighted images depicted intramedullary high intensity at Th8 and Th9 (Fig. 2). The patient's motor and sensory disturbance worsened (MMT 4 to 3) after administration of methylprednisolone (1000 mg/day for 3 days). Emergent reexamination of MRI was performed, and spinal AVF was suspected based on the findings of the small vessels around the spinal cord. The patient was referred to our hospital, and surgery was performed. The symptoms ameliorated after stopping the steroids and gradually relieved after surgical treatment and physical therapy.

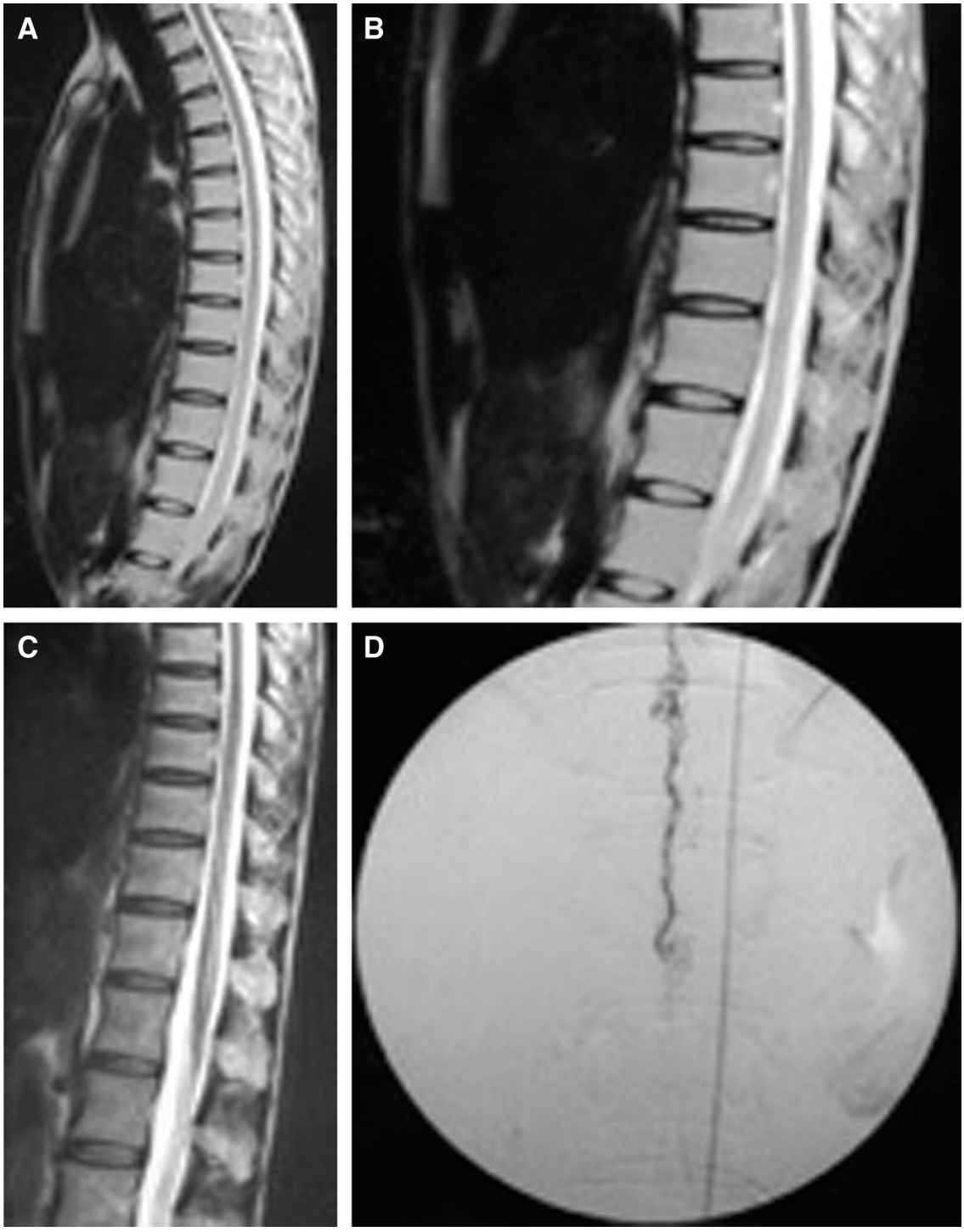

Fig. 2 (Case 3 in Table 4) (A and B) T2WI of MRI at first admission at the thoracic region. (C) Emergent MRI of T2WI after rapid deterioration following steroid pulse treatment. (D) Angiography revealing a perimedullary AVF. 45-year-old woman: The patient complained of motor weakness, pain, dysesthesia, and hyperreflexia in both lower extremities and gait disturbance, and MR T2WI showed intramedullary high intensity at Th 8 and 9 (A and B). In the first hospital, she was suspected of having multiple sclerosis and was administered methylprednisolone (1000 mg/day for 3 days); however, her motor and sensory disturbance worsened (MMT 4 to 3) the same day. Emergent reexamination of MRI was performed, and spinal AVF disease was suspected based on the findings of the small vessels around the spinal cord (C). The patient was referred to our hospital and surgery was performed. AVF: arteriovenous fistula, MMT: manual muscle test, MR: magnetic resonance, MRI: magnetic resonance imaging, T2WI: T2-weighted image.

Discussion

Pathological AV shunt leads to arterialization of the valveless perimedullary venous plexus, resulting in a reduction in the AV gradient, leading to reduced outflow from the radicular vein, retrograde venous drainage, and intramedullary edema due to venous hypertension.[3–5,7–12] Several studies have demonstrated that venous hypertension and hypervolemia at the DAVF level result in stagnation of blood flow and reduction in the AV gradient, which results in reduced intramedullary blood flow, edema, and ischemia.[13] This pathological condition causes myelopathy resulting in paresis, sensory disturbance, gait disturbance, or urinary and bowel dysfunction.

These symptoms gradually progress. Contributing factors to the deterioration include the following: prolonged standing and walking,[14] conditions related to the Valsalva maneuver,[15,16] and abdominal compression during lumbar surgery.[17]

Most patients show gradual or slow progression of these symptoms, although there are many opportunities for walking/standing and Valsalva maneuver-like conditions. Nevertheless, a few patients show rapid deterioration of these myelopathy symptoms, and the detailed mechanisms and inciting conditions remain unclear. Jellema et al. reported the clinical features of 80 cases of SADVFs. Four patients had an acute onset of symptoms, and the authors noted that acute deterioration occurred most often after exercise; however, in some cases, it occurred after a period of rest.[18] In our study, rapid and subacute deteriorations occurred after golf, sitting, climbing stairs, and massage care. These actions might induce an increased abdominal pressure; however, all actions were general activities of every person and were not specific.

The locations of these four cases were below the Th6 level. Increasing intra-abdominal pressure might play a role in contributing to the deterioration of symptoms.

The veins contain approximately 70% of the total blood volume and are 30 times more compliant than arteries. An increase in intra-abdominal pressure may increase intramural and transmural central venous pressure directly via a shift of blood volume from the splanchnic vasculature through the inferior vena cava to the right heart and indirectly (only intramural central venous pressure) via a shift of the diaphragm upward and associated increase in intra-thoracic pressure.[19]

Although deterioration does not occur in spinal AVF shunts even with other daily activities causing higher intra-abdominal pressure (e.g., cough, jumping),[20–22] we speculate that the flow of arterialized medullary veins further stagnated under increasing intra-abdominal pressure with commencement of ordinary daily activities, and some cases may deteriorate myelopathy. For these reasons, early diagnosis is necessary even when the symptoms are not particularly severe.

In clinical settings, unexplained progressive myelopathy should raise a suspicion of spinal AV shunt. The diagnosis is difficult because the presenting clinical features are often nonspecific and can mimic disorders such as spinal stenosis, demyelinating disease, and medullary tumors.[3,5,7,12,13] In situations where rapid progression occurs in patients with spinal AV shunt or undiagnosed intramedullary high-intensity lesions, steroid administration is sometimes considered because steroids are somewhat useful in various other diseases.

The main etiology of SDAVF is vasogenic edema, which is why clinicians hope that steroids will benefit the situation. However, steroids have the effects of inhibiting vascular permeability and fluid retention through leukotriene C/D, bradykinin, serotonin, and PGE_1. The usefulness of steroids for myelopathy in SDAVF has not been established; the issue is also controversial. In the present study, the rapid deterioration rate after steroid pulse therapy was 100% (four of all four cases). Seven case reports have been published about rapidly exacerbating symptoms after adrenocorticotropic hormone or steroid administration to SDAVF.[3,23–28] All 11 cases including our four cases are summarized in Table 4. All cases showed rapid deterioration. While most cases showed relatively good outcomes after steroid cessation and early surgery, two patients showed mild or poor improvement. The mechanism of steroid-induced deterioration hypothesizes that steroid administration causes oncotic shifts and/or altered renal dynamics leading to water reabsorption and preferential shunting of blood into the venous circulation.

This rapid deterioration and recovery might be considered a natural course and cured by surgical interventions, not related to the effect of steroids. In our series, there was no case of having steroid therapy without surgical intervention; therefore, such presumption was not completely excluded. However, previously reported cases[24,27] showed rapid deterioration with steroid use and immediate recovery after cessation of steroid use, without any surgical intervention. In this study, all of our cases in which steroid was used had a rapid and subacute deterioration. Hence, we highly postulate that steroid acts as a strong adverse effector to spinal AVF.

In addition, we also observed subacute deterioration after administration of PGE_1 in another patient (Fig. 3). Other five patients had also taken PGE_1

Table 4 Summary of previously reported cases and our cases showing rapid deterioration after steroid pulse treatment

No.	Age (years)/ sex	Level	Symptoms	Steroids	Symptoms after steroid use	Treatments	Postop. course
Söderlund[27]	63/M	Intracranial	Myelopathy including lower extremities	Methylprednisolone	Worsened, bilateral paraparesis	Steroid discontinuation	Recovery
Cabrera[24]	62/M	L2	Lower extremities weakness and sensory disturbance, dysuria, constipation	Methylprednisolone	Worsened, bilateral paraparesis, pain	Steroid discontinuation	?
McKeon[26]	52/M	Intracranial	Bilateral paraparesis, urinary retension	Prednisone/ methylprednisolone	Worsened, flaccid paraplegia, urinary retension	Steroid discontinuation, surgical obliteration	Improved
McKeon[26]	51/F	L3	Slowly progressive myelopathy	Methylprednisolone	Worsened, bilateral paraparesis	Steroid discontinuation, disconnection	Recovery, nonradiographic only
Mckeon[26]	58/F	L2	Progressive lower extremities weakness and sensory disturbance	Methylprednisolone	Worsened rapidly, bilateral paraparesis	Steroid discontinuation, surgical treatment	Recovery
Strowd[23]	45/M	L3	Allergic reaction, no neurological deficit	Prednisone/ methylprednisolone	Worsened rapidly, bilateral paraparesis, urinary retension, sexual dysfunction	Steroid discontinuation, surgical treatment	Recovery
Disano[28]	63/F	L2	Lower extremity sensory disturbance and clonus, urinary retention	Methylprednisolone	Worsened rapidly, paraparesis, sensory disturbance	Steroid discontinuation, surgical treatment	Mild improvement
Case1	72/M	Craniocervical junction	Rt. U/E weakness, urinary retension	Betamethasone (pulse)	Worsened rapidly the next day (hemiparesis MMT 3 ⇒ tetraparesis MMT 1)	Steroid discontinuation, shunt/feeder surgical occlusion	Recovery, MMT 1 ⇒ 4
Case2	72/M	Craniocervical junction	Tetraparesis, sensory disturbance, urinary incontinence	Methylprednisolone (pulse)	Worsened rapidly on the same day	Steroid discontinuation, shunt/feeder surgical occlusion	Recovery, MMT 0 ⇒ 4

Table 4 Continued

No.	Age (years)/ sex	Level	Symptoms	Steroids	Symptoms after steroid use	Treatments	Postop. course
Case 3	45/F	Conus (perimedullary)	Paraparesis, sensory disturbance, gait disturbance	Methylprednisolone (pulse)	Worsened rapidly on the same day	Steroid discontinuation, shunt/feeder surgical occlusion	Recovery, MMT 3 ⇒ 4
Case 4	60/M	Conus (extradural)	Paraparesis, sensory disturbance, gait disturbance, bladder–rectal impairment	Methylprednisolone (pulse)	Worsened rapidly on the same day	Steroid discontinuation, transarterial embolization	Recovery in sensory, MMT 0 ⇒ 1, hypesthesia below Th10 ⇒ below L4

Postop.: postoperative, M: male, F: female, Rt.: right, MMT: manual muscle test, U/E: upper extremity.

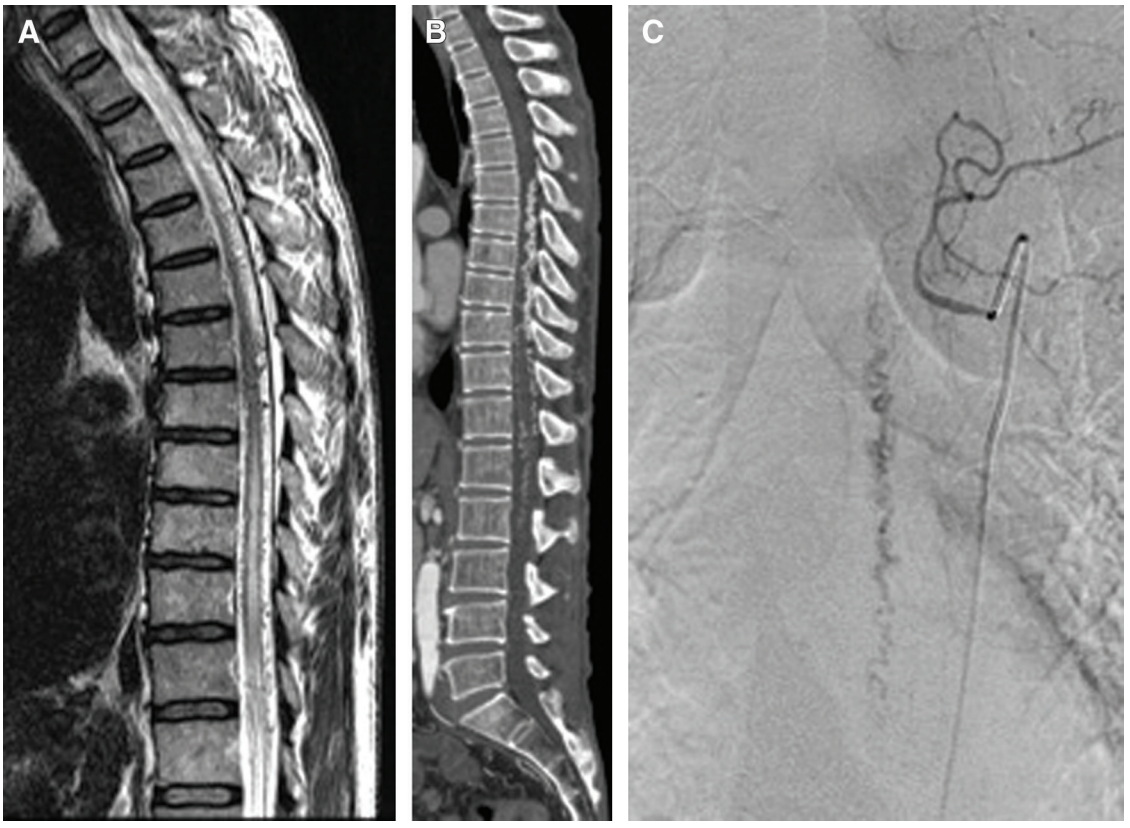

Fig. 3 (A) Intramedullary high-intensity lesion was found from thoracic to conus level on MRI T2WI. Abnormal flow voids were found around the cord. (B) Abnormal perimedullary vein was depicted with CT angiography. (C) Spinal AVF (epidural AVF) at Th7 was depicted with spinal DSA. 58-year-old man: He had a year history of bilateral foot paresthesia. Gait disturbance, mild urinary retention, and constipation occurred gradually. He visited the local clinic and suspected lumbar canal stenosis. The symptoms deteriorated with a few days course with the prescribed PGE_1 agent. Spinal AVF was suspected on lumbar MRI, and he was referred to our service. AVF: arteriovenous fistula, CT: computed tomography, DSA: digital subtraction angiography, MRI: magnetic resonance imaging, PGE_1: prostaglandin E_1, T2WI: T2-weighted image.

agent; however, the relevance of PGE_1 administration and the time course was not clear. As previously reported, PGE_1 has a vasodilating effect.[29–31] With PGE_1 vasodilating effect, it may be surmised that the mechanism similar to high-dose steroid induced deterioration. We treated two other patients who had taken low-dose steroids and did not experience such rapid deterioration; it may be assumed that the dose-dependent nature of steroid pharmacokinetics proportionally affects the spinal AV shunt.[32,33]

The presence of a significant number of smooth muscle fibers within the wall of the radicular vein before and behind the dural bottle neck has been reported, and dynamic regulation was suggested by van der Kuip et al.[34]

Thron et al. evaluated 104 specimens obtained from unselected autopsies and did not confirm the presence of valves in the radiculomedullary veins; nevertheless, they confirmed two main types of venous exit through the dura mater: slit type 60%, bulge or nodular type 35%, and the remaining 5% could not be assigned to either one of the main types.[35]

These anatomical studies were normal unselected autopsy cases; nevertheless, such anatomical structures and interactions with the steroid contractile effect smooth muscle,[36] steroid, PGE_1, and ordinary daily activities may be relevant to the deteriorating mechanism of spinal AVF.

Certainly, the same ordinary daily activities do not always cause the same deterioration reproductively shortly thereafter and in other patients with spinal AV shunt. Ordinary daily activities causing rapid or subacute deteriorations were not the first event for any patient. This may be due to individual variations. Further studies are needed to elucidate more precise mechanisms and factors.

Physicians should keep this pitfall in mind when treating these intramedullary high-intensity lesions until exclusion of the possibility of spinal AV shunt disease.

Furthermore, if spinal AV shunt is diagnosed, steroid use should be avoided, as well as activities that may further increase abdominal pressure. However, as demonstrated by our results and previous report, ordinary daily activities and even rest may sometimes deteriorate the symptoms. Therefore, once spinal AV shunt is diagnosed, early treatments are recommended to prevent progression to neurological deterioration.

Limitations

As with most retrospective studies, the lack of prospective enrollment limits the ability to established causality. The small sample size in our series may weaken the robustness of our observations; verification of our findings may ultimately require a large prospective multi-institutional study that would serve to reduce any selection bias associated with the two institutions study and would improve the generalizability of our results.

Conclusion

We reviewed patients with spinal AV shunts treated in our institutions and evaluated the factors leading to rapid worsening of myelopathy. Because ordinary actions such as sitting and climbing upstairs may induce rapid deterioration, early diagnosis is necessary. Furthermore, rapid deterioration was induced by the administration of steroid pulse therapy. Therefore, we recommend that high-dose steroid administration should be avoided, especially in patients with intramedullary high-intensity lesions on MRI T2 images, until the possibility of spinal AV shunt disease is excluded.

Conflicts of Interest Disclosure

All authors who are members of the Japan Neurosurgical Society (JNS) have registered self-reported COI disclosure statements through the website for JNS. The authors declare no conflicts of interest for this manuscript.

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