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Venous Congestive Myelopathy Caused by Spinal Vascular Malformation

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Vascular myelopathy can be result from either extrinsic or intrinsic spinal cord lesions. Extrinsic pathology results from spinal cord compression and/or injury associated with mass effect. Intrinsic pathology results from spinal cord infarction, hemorrhage and venous congestion.[1] Among intrinsic spinal cord pathology, spinal cord infarction and hemorrhage develop symptoms rather suddenly, whereas venous congestion develops various symptoms gradually. Although spinal vascular malformation is a rare neurovascular disease, it is related to all three intrinsic and even extrinsic pathologies by compressing the spinal cord due to dilated arterial aneurysm or veins.[2-4] Venous congestion tends to be neglected at the time of diagnosis because of its insidious and vague symptom onset.[5] When spinal dural arteriovenous fistula (SDAVF) develops symptoms, abnormal T2-wighted image or fluid attenuated inversion recovery signal intensity appears on magnetic resonance imaging in the midline scan along the spinal cord.

CAUSE OF VASCULAR MYELOPATHY RELATED WITH VENOUS CONGESTIVE MYELOPATHY

Vascular myelopathy caused by venous congestion is called venous congestive myelopathy (VCM). SDAVF is the most common cause of VCM.[3] Some other pathologies such as spinal pial arteriovenous shunt or spinal cord malformation may also be associated with VCM. SDAVF has the most typical presentation. SDAVF must to be suspected on the basis of a typical presentation history when an elderly man presents with progressive walking difficulty and various sensory changes in the lower extremities. Retrograde venous reflux into the intrinsic spinal cord veins *via* the radicular vein is the key pathology of VCM. Regardless of shunt lesions, it is necessary to identify the location of the shunt and the point of regurgitation at the radicular vein needs to be identified.[6] It may be direct or indirect, i.e., a shunt alone contributes to such regurgitation (direct), or other extrinsic venous drainage may be combined with the antegrade epidural or paravertebral venous drainage and retrograde regurgitation into the intrinsic spinal cord veins *via* the radicular vein (indirect). If it is direct, even a small shunt can cause a severe VCM.

PROGRESSION OF NEUROLOGICAL SYMPTOMS

Symptoms usually begin insidiously and

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progress gradually with varying degrees of sensory changes, gait instability/imbalance, extremity weakness, urinary urgency or hesitancy and bowel incontinence. It is therefore defined as having the presence of sensory, motor, or sphincter dysfunction as assessed by the Pain-Sensory-Motor-Sphincter score.[7]

The development of neurological symptoms usually begins with sensory changes. Pain is sometimes present in the early stage but is not common. Motor deficit appears slowly and progressively, especially as walking difficulty because the spinal vascular pathology usually involves the thoraco-lumbo-sacral levels. Deficit of upper extremities or other cervical and/or thoracic level problem like respiratory difficulty can also be seen when cervical spinal cord is involved. Sphincter problem occurs in the late phase of the SDAVF.[7]

SUDDEN SYMPTOM AGGRAVATION OF VENOUS CONGESTIVE MYELOPATHY

Patients with VCM usually have a typical history of symptom aggravation. Exercise, energy drinks, some food intake and steroid injections are known to be precipitating factors. Aggravation after exertional or vigorous exercise is a well-known symptoms of SDAVF. After walking for a long distance, the patient will feel weakness in the extremities and will sit down to rest to recover and then try to walk again. The patient is usually in better condition in the morning and feel worse in the afternoon. Such deterioration pattern becomes progressive day by day. During the course of the disease progression, patients sometimes experience a rather sudden worsening of symptom (rapid progression), which is associated with a poor prognosis.

On the other hand, an event of sudden reversible collapse of both legs can be observed soon after the intake of energy drink or some food intake. Taurine is known to be a representative one and is contained in some energy drinks and also in some seafood.[8] Aggravation lasts for about 3 hours, and then patients recover slowly after taking rest. The patients who enjoy such foods have a painful memory in the heart so that they try to avoid such foods and respond promptly if a question about such history was given. Steroid injection or even oral administration must be carefully monitored or abandoned.[9] Although patients usually recover from the event within 24 hours, some patients did not completely recover from the aggravated condition.

RECOVERY FROM VENOUS CONGESTIVE MYELOPATHY AFTER TREATMENT

If the causative vascular shunt lesion is removed and patients are managed appropriately, symptomatic recovery is expected. Surgical resection or embolization is the only way to remove the lesion. Regarding the progress after successful embolization, the patients usually feel better about their legs on the day after recovery from general anesthesia.[3,10-15] They say the legs are light (not heavy as before). And then sensory and motor deficits slowly improve over the next few days. Sphincter symptoms related to bladder and bowel function may recover but not as much as motor symptoms. Complete loss of sphincter function is usually associated with a much more severe motor deficit and then does not recover even with appropriate treatment.

During recovery with improved walking difficulty, sensory symptoms also improve. However, the patients usually complain of different patterns of diverse sensory symptoms that are difficult to categorize. Some patients develop intermittent involuntary kicking of legs, myoclonus, which is defined as a sudden, brief, lightning-like muscle contraction.[16] Myoclonus of the legs is also noted during sleep. Such myoclonus during recovery in patients with VCM does not appear to be classified among the various causes of myoclonus.[16]

Fund
None.

Ethics Statement
No approval from IRB nor informed consent is required for this editorial.

Conflicts of Interest
DCS has been the Editor-in-Chief of the *Neurointervention* since 2018. No potential conflict of interest relevant to this article was reported.

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REFERENCES

1. Vuong SM, Jeong WJ, Morales H, Abruzzo TA. Vascular diseases of the spinal cord: infarction, hemorrhage, and venous conges-

tive myelopathy. *Semin Ultrasound CT MR* 2016;37:466-481
2. Song Y, Kwon B, Al-Abdulwahhab AH, Nam YK, Ahn Y, Jeong SY, et al. Rare neurovascular diseases in Korea: classification and related genetic variants. *Korean J Radiol* 2021;22:1379-1396
3. Park JE, Koo HW, Liu H, Jung SC, Park D, Suh DC. Clinical characteristics and treatment outcomes of spinal arteriovenous malformations. *Clin Neuroradiol* 2018;28:39-46
4. Jung SC, Song Y, Cho SH, Kim J, Noh SY, Lee SH, et al. Endovascular management of aneurysms associated with spinal arteriovenous malformations. *J Neurointerv Surg* 2018;10:198-203
5. Rabinstein AA. Vascular myelopathies. *Continuum (Minneap Minn)* 2015;21:67-83
6. Suh DC, Kim HS, Baek HJ, Park JW, Kim KK, Rhim SC. Angioarchitecture of spinal dural arteriovenous fistula - evaluation with 3D rotational angiography. *Neurointervention* 2012;7:10-16
7. Suh DC, Song Y, Park D, Han M, Lim YM, Park JE, et al. New grading system for the clinical evaluation of patients with spinal vascular lesions. *Neuroradiology* 2018;60:1035-1041
8. Suh DC, Jeong S, Choi YH, Cho SM, Yun SY, Son AY, et al. Reversible symptom aggravation by intake of taurine-rich foods in patients with venous congestive myelopathy: controlled case series study. *Neurointervention* 2022;17:93-99
9. Lee CS, Pyun HW, Chae EY, Kim KK, Rhim SC, Suh DC. Reversible aggravation of neurological deficits after steroid medication in patients with venous congestive myelopathy caused by spinal arteriovenous malformation. *Interv Neuroradiol* 2009;15:325-329
10. Al-Abdulwahhab AH, Song Y, Kwon B, Suh DC. Embolization tactics of spinal epidural arteriovenous fistulas. *Neurointervention* 2021;16:252-259
11. Song Y, Cho SH, Lee DW, Sheen JJ, Shin JH, Suh DC. Osseous versus nonosseous spinal epidural arteriovenous fistulas: experiences of 13 patients. *AJNR Am J Neuroradiol* 2019;40:129-134
12. Cho SH, Suh DC. Transdural segment of the radicular vein in spinal dural arteriovenous fistula. *Neurointervention* 2017;12:57-58
13. Adrianto Y, Yang KH, Koo HW, Park W, Jung SC, Park JE, et al. Concomitant origin of the anterior or posterior spinal artery with the feeder of a spinal dural arteriovenous fistula (SDAVF). *J Neurointerv Surg* 2017;9:405-410
14. Suh DC, Cho SH, Park JE, Liu H, Jung SC. Induced-wedge technique to improve liquid embolic agent penetration into spinal dural arteriovenous fistula. *World Neurosurg* 2016;96:309-315
15. Shin HK, Suh DC, Jeon SR. Intraoperative direct puncture and embolization (IOPE) using a glue material for spinal cord arteriovenous fistula: a case report. *Eur Spine J* 2015;24 Suppl 4:S594-S599
16. Caviness JN. Myoclonus. *Continuum (Minneap Minn)* 2019;25:1055-1080