

Case Report

Histologically proven venous congestive myelopathy without concurrent vascular malformation: Case reports and review of the literature

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Abstract

Background: Venous congestive myelopathy is a progressive myelopathy that is generally caused by a spinal dural arteriovenous fistula.**Case Description:** We report a patient with histologically confirmed venous congestive myelopathy without concurrent vascular malformations in radiological and intraoperative findings.**Conclusions:** The definitive underlying etiology of this congestive myelopathy was unclear. However, this case report highlights the possibility of venous congestive myelopathy with etiology other than a dural arteriovenous fistula. Further, a systematic and elaborate examination should be undertaken to explore the underlying pathology whenever this type of spinal parenchymal lesion is detected.**Key Words:** Foix-Alajouanine syndrome, Spinal cord tumor, Spinal dural arteriovenous fistula, Venous congestive myelopathy**Access this article
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INTRODUCTION

Venous congestive myelopathy, generally known as Foix-Alajouanine syndrome, is clinically characterized by a progressive motor and sensory deterioration. The pathological finding of this condition is described as a necrosis in the spinal cord.^[2,3,5,6,10,13] It mostly results from impaired venous outflow secondary to a spinal dural arteriovenous fistula (dAVF).^[1,3,8,9]

There have been few reports of venous congestive myelopathy without concurrent dAVF or other vascular malformations.^[14,15,18,22] The underlying etiology is assumed to be an occult dAVF in most cases but is still unclear.

We report a patient with histologically confirmed venous congestive myelopathy without concurrent vascular malformation in radiological and intraoperative findings. Several possible mechanisms other than occult vascular malformation are discussed.

CASE REPORT

A 68-year-old man with no significant past medical history had suffered from paresthesia in his right lower leg. The area of paresthesia slowly extended rostrally up to the umbilicus in 6 months. At 5 months after the onset, he could not walk without cane and at 6 months, vesicorectal dysfunction appeared. On physical examination upon admission at 9 months after the

onset, a sensory disturbance below the level of T12, impaired vesicorectal function, and spastic gait were observed. Manual muscle testing showed no weakness in the lower extremities, but he could not walk without cane due to the spasticity and impaired deep sense in lower legs. The deep tendon reflex was promoted in the lower extremities. Blood cell and chemistry examinations revealed no abnormalities. Cerebrospinal fluid test was normal (cell 2/3, protein 21 mg/dl, glucose 65 mg/dl).

Magnetic resonance imaging (MRI) showed an intramedullary T2 hyperintensity and swelling of the spinal cord extending from the level of T5 to T7 [Figure 1]. And the lesion showed faint and partial Gadolinium enhancement. Findings of flow void or dilated pial venous plexus around the spinal cord were not observed. The spinal cord of this patient was about 45° rotated to the right in axial sections of the thoracic MRI, even though there was no rotation of the vertebral column.

Based on his progressing deterioration and radiological findings, the differential diagnosis included demyelination disease, transverse myelitis, sarcoidosis, Lyme's disease, spinal cord infarction, and spinal cord tumor. The patient was first admitted to the neurology department and completely examined. He had no history or finding of being bitten by a tick. Vitamin B12 and angiotensin converting enzyme was normal. HIV, syphilis, and hepatitis were negative. Immunoglobulin was also within normal level. The ophthalmological examination showed no abnormal finding. Chest computed tomography (CT) scan showed no bilateral hilar lymphadenopathy. A trial of steroid-pulse therapy (i.v. methylprednisolone 1000 mg for 3 days) resulted in

no clinical or radiological improvement. Based on the no efficacy of steroid-pulse therapy in addition to the slowly progressing clinical course transverse myelitis or demyelination disease was less likely suggested. It was thus suspected that the patient had a spinal cord tumor, and he was referred to our department for histological diagnosis.

We performed a midline myelotomy following T5 to T8 laminectomy. Because the spinal cord was rotated, it was difficult to find its midline. Thus we used dorsal column mapping at the midline myelotomy. A biopsy specimen was obtained at the T7 level on the enhanced area of the MRI. No dilated or tortuous vessels suggestive of vascular malformation were observed on the surface of the spinal cord or the dural sac [Figure 2].

The biopsy specimen showed an increased number of small hyalinized vessels, gliosis, vascular thrombosis, and hemosiderin deposition. Neither tumor cells nor signs of active inflammation were observed [Figure 3]. There were no signs of demyelination by either myelin basic protein or Klüver-Barrera stain. These histological findings were compatible with a venous spinal infarction caused by congestive myelopathy.

One week-postoperative MRI showed a marked decrease of the T2 hyperintensity area, spinal cord swelling and lesion enhancement [Figure 4]. Clinically, his gait disturbance became worse transiently, presumably due to the damage to the dorsal funiculus, but it improved with rehabilitation. The sensory disturbance was stable. No clinical and radiological recurrence was observed at one year after the operation.

DISCUSSION

DAVF has been reported to cause neurologic dysfunction on the basis of venous hypertension.

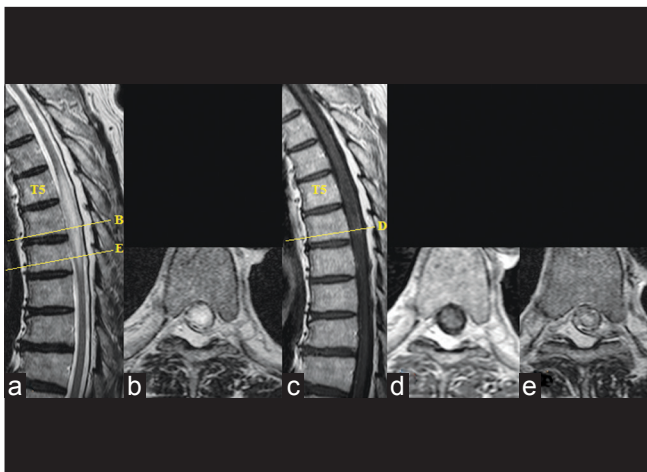


Figure 1: Preoperative magnetic resonance images of the thoracic spine. Sagittal (a) and axial (b) T2-weighted images show intramedullary hyperintensity and swelling of the spinal cord extending from the level of T5 to T7. Sagittal (c) and axial (d) T1-weighted MR images show faint and partial Gadolinium enhancement. Findings of flow void or dilated pial venous plexus are not observed. The spinal cord is rotated approximately 45° to the right in the axial section (e)

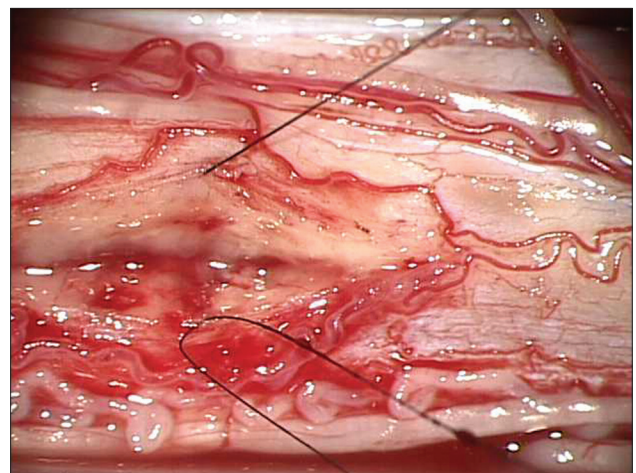


Figure 2: Intraoperative photograph showing the spinal cord rotating to the right. There are no dilated or tortuous vessels

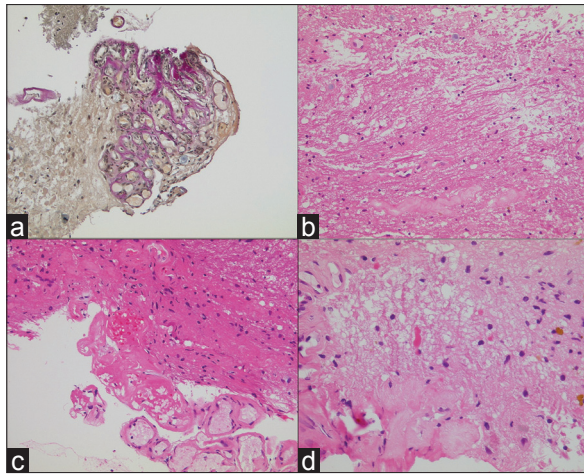


Figure 3: Photomicrographs of a biopsy specimen showing an increased number of small hyalinized vessels (a, b), gliosis (b), vascular thrombosis (c), and hemosiderin deposition (d). Neither tumor cells nor signs of active inflammation are observed. EVG stain, $\times 100$ (a); H and E, $\times 200$ (b, c) and 400 (d)

Direct communication between arteries and veins increases venous pressure, which causes an increase in intramedullary arterial vasodilation in order to maintain perfusion pressure. These changes then result in the increased tissue pressure, possibly associated with progressive exhaustion of the autoregulatory capacity.^[20] Ultimately, intramedullary edema develops, leading to a decrease in perfusion pressure and ischemic injury. Surgical studies have shown that the interruption of the shunt halts progression and can lead to functional improvement.^[16]

Pathological studies of congestive myelopathy associated with dAVF have shown marked white matter hypocellularity and small vessels with thickened and hyalinized walls. Extensive vascular sclerosis and gliosis of the white matter is demonstrated with disorganization of the white matter, patchy myelin loss, and concomitant axonal loss.^[4,11,21] The present case showed a similar pathology as the reported cases, but dAVF was not identified in our case.

There have been several reports of histologically confirmed venous congestive myelopathy or Foix-Alajouanine syndrome without concurrent dAVF [Table 1].^[14,15,18,19,22] All of the 14 reported cases showed a subacute progressing myelopathy. Two of these patients underwent spinal angiography, both showed no dAVF. All cases showed no abnormal vessels intraoperatively. Although the histopathological findings were typical of venous congestive myelopathy, a spinal dAVF was not demonstrated in any of the cases throughout their entire history and follow-up. There are several possible explanations for the absence of a demonstrable fistula, but the authors of these studies concluded that the most probable cause was an occult dAVF due to the insufficient

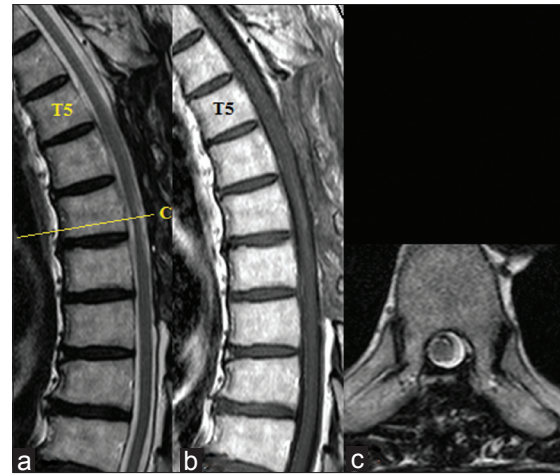


Figure 4: Postoperative magnetic resonance images of the thoracic spine. Sagittal (a) and axial (c) T2-weighted MR images, and sagittal (b) T1-weighted MR images show disappearance of the intramedullary T2 hyperintensity and contrast enhancement

diagnostic studies. Their reasoning was as follows. First, a spinal dAVF can be missed if a complete selective spinal angiography (i.e., entire thoracic and lumbar) is not performed. Second, the fistula may be found at a site distant from the cord signal abnormality, possibly even an intracranial site.^[7,17,23] Third, the demonstration of vessels supplying dAVF may be compromised by atherosclerosis of the segmental vessels^[23] or thrombosis of the draining veins.^[3]

However, we can state the absence even of occult dAVF with high probability for the following reasons. First, the preoperative MRI failed to show flow void around the spinal cord. Although we did not perform a spinal angiography, the intraoperative findings showed no abnormal, tortuous, or dilated vessels. Second, both his symptoms and MRI findings showed marked improvement just after the biopsy, despite the fact that he underwent no surgical intervention for a vascular malformation.

In the reported cases, the symptoms of 3 of the 14 patients, for whom descriptions of the clinical course were available, improved after biopsy like our patient, although the reasons were unknown. In our case, judging from the rapid radiological improvement after the biopsy, we can state that the decompression procedure together with the biopsy might have played a role in his improvement irrespective of the cause. Furthermore, the mechanical compression or distortion of a perimedullary vein could have caused congestion in our case.

A recent report described a patient who developed venous congestive myelopathy associated with a non compressive herniated cervical intervertebral disc at the same level.^[12] It was assumed that the disc herniation resulted in impaired drainage of the blood from the spinal cord

Table 1: Summary of reported case of venous congestive myelopathy without concurrent spinal dural arteriovenous fistula

Authors and year	Age/sex	Symptoms	Duration (months)	Level	Preoperative MRI	Surgical findings	DSA	Postoperative MRI	Clinical course
Rodriguez <i>et al.</i> , 2005	38/F	Chest and left arm pain, paresthesia	60	Lower C-upper T	T2 high, cord enlargement, enhancement	Cord enlargement, increased vascularity at midline	No fistula	n.d	Stable at 13 months
	62/M	Paraparesis, paresthesia, pain, incontinence	24	Conus	T2 high, cord enlargement, enhancement	Cord enlargement, discoloration, no abnormal vessels	n.e	Improved	Improved at 18 months
	61/M	Paraparesis, paresthesia, incontinence	n.d	T1-L-conus	T2 high, cord enlargement, no enhancement	n.d	No fistula	Stable	Completely paraplegic at 26 months
	54/F	Paraparesis, paresthesia, incontinence	27	T6-7	T2 high, no enhancement	Abnormal intramedullary tissue, no abnormal vessels	n.e	Stable	Slight improvement at 2 months
Minich <i>et al.</i> , 1991	58/M	Paraplegia, paresthesia, incontinence	24	T4-conus	T2 high, cord enlargement	Vein adherent to enlarged spinal cord	n.e	n.d	No change
	62/M	Paraplegia, paresthesia	48	Conus	T2 high	Vein adherent to enlarged conus, necrotic material	n.e	n.d	No improvement; bladder became atonic
	77/F	Paraplegia, paresthesia, incontinence	3	Conus	T2 high, enhancement	Enlarged conus, necrotic material	n.e	n.d	No change
	47/F	Brown-Sequard syndrome(C4), right-sided paresis	1	C2-8	T2 high, enhancement	Asymmetric enlarged cervical cord, white friable material	n.e	n.d	No change
Montine <i>et al.</i> , 1995	71/M	Paraparesis, paresthesia, incontinence	48	Conus	T2 high, conus enlargement, enhancement	Enlarged conus, no abnormal vessels	n.e	n.d	n.d
	69/F	Paraparesis, paresthesia	7	T8-conus	T2 high, enhancement	Enlarged conus, no abnormal vessels	n.e	n.d	n.d
	56/M	Paraplegia	n.d	Lower T	cord enlargement, enhancement	No abnormal vessels	n.e	n.d	n.d
Tsutsumi <i>et al.</i> , 2009	78/M	Paraplegia, paresthesia, incontinence	48	T12-L2	T2 high, enhancement	No abnormal vessels, necrotic material	n.e	Stable	Improved at 3 months
Schwartz <i>et al.</i> , 1997	62/M	Paraparesis, incontinence	120	L9-T1	T2 high, cord enlargement, enhancement	No abnormal vessels, necrotic material	n.e	n.d	Stable at 6 months, later subtle progression at 2 years after cessation of warfarin
	27/F	Paraparesis, incontinence	16	T6-9	T2 high, cord enlargement, enhancement	Small vessels, not hypervascular	n.e	n.d	Stable at 9 months
Present case	68/M	Paraparesis, paresthesia, incontinence	6	T5-8	T2 high, cord enlargement, enhancement	Cord enlargement, no abnormal vessels	n.e	Improved	Improved at 1 year

C: Cervical, T: Thorax, L: Lumbar, n.d: not described, MRI: Magnetic resonance imaging, DSA: Digital subtraction angiography, n.e: not examined

through compression of the venous plexus near the intervertebral foramen. This kind of venous compression could have been the cause of myelopathy in the present case, although no apparent compressive or spondylotic lesion was found in the MRI. One abnormal finding in the present case was the rotation of the spinal cord, but it is unclear whether this rotation could have caused the venous congestion.

In our case, rotation of the spinal cord or other unclear underlying etiology could have diminished venous return from the spinal cord and caused the venous congestive myelopathy. Although this mechanism of venous congestive myelopathy is different from that of cases with a dAVF, the pathophysiologic sequelae may be similar in both types of venous congestion.

CONCLUSION

We reported a case of histologically confirmed venous congestive myelopathy without concurrent vascular malformation. The definitive underlying etiology of this congestive myelopathy is still unclear. However, venous congestive myelopathy can be caused by a dAVF as well as by other etiologies, and thus a systematic and elaborate examination should be undertaken to explore the underlying pathology whenever this type of spinal parenchymal lesion is detected.

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