Lumbar Congestive Myelopathy Mimicking Neoplasia Without Concurrent Vascular Malformation
—Case Report—

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Abstract
A 78-year-old male presented with congestive myelopathy manifesting as progressive gait disturbance following conservative therapy for lumbar spinal canal stenosis, with suspected spinal cord tumor in the conus medullaris. His past medical history was unremarkable and he was not aware of any traumatic injury in the back or infectious disease. On admission, he had clumsy hand, moderate paraparesis, significant sensory disturbance below the L5 level, and severe vesicorectal dysfunction. The deep tendon reflex was promoted in the upper extremities, but poorly induced in the lower extremities. Blood examination found no abnormalities including values of tumor markers. Cerebral, cervical, and thoracic magnetic resonance (MR) imaging revealed no contributory pathology without spondylotic change at the C3-C6 levels. Lumbar MR imaging showed fusiform swelling of the cord from the T10 to T12-L1 levels, with rimlike enhancement at the T12-L1 levels by gadolinium. The patient underwent surgery. Intraoperatively, the dorsal surface of the affected cord was pale, not swollen, and sparsely vascularized without tortuous vessels. Midline myelotomy caused escape of creamy material that was identified as necrotic neural tissue. A collapsed vessel, located on the surface of the cord, was histologically identified as a thrombosed vein. The histological findings were compatible with spinal infarction caused by congestive myelopathy. Whole craniospinal and iliac angiography performed postoperatively failed to reveal any dural and paraspinal vascular malformation. His paraparesis, sensory disturbance, and vesicorectal dysfunction improved significantly after surgery. Congestive myelopathy may be caused by various angiographically occult etiologies other than dural arteriovenous fistula.

Key words: congestive myelopathy, Foix-Alajouanine syndrome, dural arteriovenous fistula

Introduction
Angiodysgenetic necrotizing myelopathy, generally known as Foix-Alajouanine syndrome, is considered to be the end result of spinal venous congestion associated with dural arteriovenous fistula (dAVF). However, the underlying pathophysiology is not fully understood because this entity is infrequent with variable clinical manifestations and inconsistent appearance on both spinal angiography and intraoperative examination. Only a few reports have described congestive myelopathy from biopsy specimens, instead of autopsy findings. Endovascular treatment of dAVF is thought to lead to favorable outcome at long-term follow up, but the severity of neurological deficits and patient's age are significant prognostic factors.

Here we describe a case of histologically verified lumbar congestive myelopathy mimicking tumor with subacute deterioration, but no concurrent vascular malformation was identified by neuroimaging.

Case Report
A 78-year-old male had suffered progressive gait disturbance for 2 months. The patient had been followed up by local orthopedic surgeons for 4 years under a diagnosis of lumbar spinal canal stenosis. However, his activity of daily life had gradually deteriorated and he was nearly bedridden when referred to our department, under suspicion of a spinal cord tumor. His past medical history was unremarkable and he was not aware of any traumatic injury in the back or infectious disease.

On admission, he was alert and well-oriented without detectable cranial neuropathy. He had
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clumsy hand, attributed to spondylotic change at C3-C6 levels confirmed by cervical magnetic resonance (MR) imaging. He had moderate paraparesis and could not walk even with assistance. The sensory disturbance was significant below the L5 level with impaired sphincter and vesicorectal functions. The deep tendon reflex was promoted in the upper extremities, but poorly induced in the lower extremities. Blood examination found no abnormalities with no unusual values of tumor markers. Cerebral and thoracic MR imaging revealed no contributory pathology. Lumbar MR imaging showed fusiform swelling of the spinal cord from the T10 to the T12-L1 levels after administration of gadolinium (Fig. 1).

The patient underwent surgery under a diagnosis of spinal cord tumor in the conus medullaris. Intraoperatively, the dorsal surface of the affected cord was pale, not swollen, and sparsely vascularized without tortuous vessels. Midline myelotomy 3 mm in length caused escape of creamy material (Fig. 2). The incised pia mater was not re-approximated. A collapsed vessel, located on the surface of the cord, was histologically identified as a vein with thickened wall and luminal thrombus associated with aggregation of lymphocytes. The creamy material consisted of fibrinoid material, histiocytes with hemosiderin deposit, and vascular proliferation (Fig. 3). These histological findings were compatible with spinal infarction caused by congestive myelopathy.

Postoperatively, the patient underwent an extensive whole craniospinal and iliac angiography, which failed to reveal any dural and paraspinal vascular malformation. His paraparesis, sensory disturbance below the L5 level, and vesicorectal dysfunction improved significantly after surgery and he began to walk with assistance in 2 weeks. MR imaging taken 2 months after surgery showed persistent intramedullary hyperintensity on the T2-weighted image (Fig. 4). The patient was transferred to a rehabilitation hospital.

Discussion

Spinal venous hypertension may be transmitted to the intrinsic veins of the cord, resulting in reduction of the arteriovenous pressure gradient within the cord, followed by decreased tissue perfusion and progressive hypoxia of the neural tissue. Intramedullary vasodilation and loss of autoregulatory capacity may also occur with associated cord edema. The end result is stagnation of blood flow and hypoxia from perfusion impairment leading to neurological dysfunction.

In the present case, the clinical presentation was typical of congestive myelopathy manifesting as subacute deterioration following gradually progressive neurological impairment. The histological findings were compatible with thickened venous wall with luminal thrombus, perivascular lymphocyte aggregation, and vascular proliferation and hemosiderin-laden histiocytes in the necrotic tissue, without signs of malignancy. On the other hand,
expansion by MR imaging and extensive whole craniospinal and iliac angiography detected no vascular malformation associated with the congestive myelopathy. Moreover, an evacuation of the intramedullary necrotic tissue combined with external decompressive maneuver resulted in remarkable improvement of the congestive myelopathy, although any potential vascular lesion remained untreated.

We assume that formation of perimedullary tortuous vessels on the dorsum of the spinal cord, depending on the degree of the shunt flow, might not suggest dAVF because a faint shunt may not cause tortuosity of the affected veins. In the present case, the dorsal surface of the affected cord showed sparse distribution of normal vasculature, instead of serpentine vessels, whereas the histological findings strongly suggested congestive myelopathy.

The histological findings indicated congestive myelopathy, but the absence of detectable vascular malformations can be explained by either the end result of an occult dAVF, thrombosed dAVF resulting from venous congestion isolated from the normal blood circulation, other underlying pathology causing venous congestion without concurrent vascular malformation, or a combination. Spinal dAVF was demonstrated in only three of seven cases and prebiopsy angiography found no abnormalities in two patients. Possible explanations included lack of complete selective spinal angiography, fistulas located distant from the site of the cord signal abnormality, and feeder vessels compromised by atherosclerosis of segmental vessels or thrombosis of the draining veins.

Therefore, negative angiographical findings cannot exclude dAVF. Repeat spinal angiography might be recommended for patients presenting with congestive myelopathy and angiographically unverified dAVF.

Recognition of congestive myelopathy by surgical exploration is important because it may lead to the correct identification of a non-neoplastic lesion or

**Fig. 2** Intraoperative photographs showing (A) sparsely vascularized dorsal surface of the cord without concomitant tortuous vessels, and (B) escape of whitish and creamy material following midline myelotomy (arrowhead).

**Fig. 3** Photomicrographs of (A) a vein resected from the dorsal surface of the cord demonstrating thickened wall (arrowhead), luminal thrombus, and aggregating lymphocytes (arrow), (B) cyst content involving the fibrinoid material, histiocytes (arrows), and vascular proliferation (arrowheads), and (C) histiocyte with hemosiderin deposit (arrowheads). Hematoxylin and eosin stain, × 40 (A), × 100 (B), and ×600 (C).

**Fig. 4** Sagittal T2-weighted magnetic resonance image taken 2 months after surgery demonstrating persistent intramedullary hyperintensity (arrowheads).
surgically treatable disease. In the present case, performing only the internal decompression maneuver lead to a significant improvement of the neurological deficits, although an angiographically undetectable vascular lesion might have been present. Endovascular therapy is indicated for favorable outcome to treat angiographically verified dAVF.

Congestive myelopathy may be caused by various etiologies other than dAVF, so a systematic survey should be undertaken for exploring the underlying pathology as well as comprehensive management.

References


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