

# Rapidly Progressed Spinal Dural Arteriovenous Fistula Mimics Transverse Myelitis

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Spinal dural arteriovenous fistula (SDAVF) is a rare cause of myelopathy. SDAVF can be explained by an acquired arteriovenous shunt between a dural artery and medullary venous system which accounts for venous hypertension and congestion.<sup>1</sup>

## CASE

A 67 year- old- man came to our ER complaining of paraparesis and urinary incontinence. Two weeks prior to the admission, he recognized slowly progressing weakness in both lower legs along with urinary incontinence. On his first spinal magnetic resonance imaging (MRI) examination at an outside institution, there was a high signal lesion from the T5 level to the conus on T2- weighted images and contrast enhancement around the cord. Longitudinally extensive transverse myelitis was their initial diagnosis. The patient was transferred to our hospital for further evaluation.

Initial neurological examination revealed alert mental status. Muscles of lower extremities showed symmetrically increased tone with exaggerated ankle and knee jerk reflexes. The Babinski sign was positive on both feet. Muscle

power was assessed as grade 4 out of 5 according to Medical Research Council (MRC) grading. All modalities of sensation were decreased below L1 dermatomal level. Not only urinary incontinence but also decreased anal sphincter tone was detected. General and systemic examination of upper extremities were proved normal.

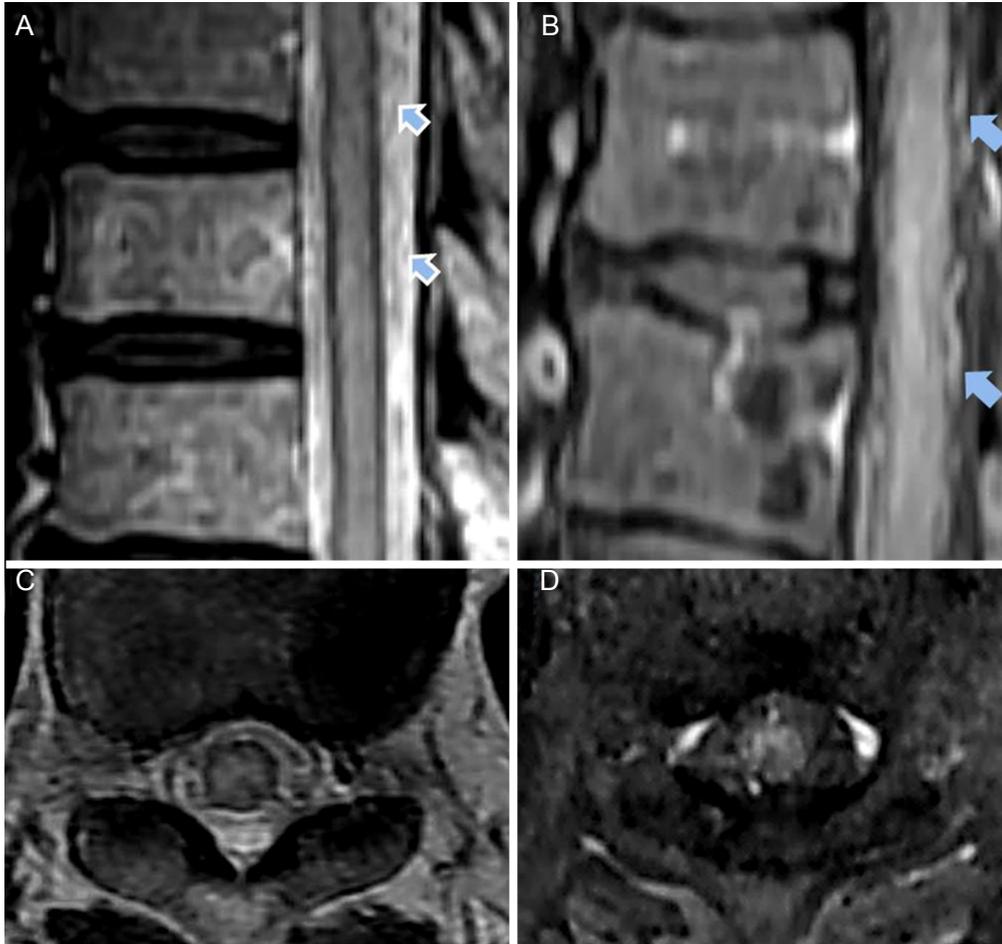
A cerebrospinal fluid (CSF) study revealed a CSF protein level at 52.1 mg/dL and glucose level at 56 mg/dL. The number of cells in CSF was 1 white blood cell/ $\mu$ L (1 polymorphonucleocyte) and 0 red blood cell/ $\mu$ L. Oligoclonal band was negative. Negative results were obtained for sedimentation, C-reactive protein, hepatitis markers, vasculitis markers, and human immune deficiency virus. Although steroid pulse regimen was started immediately, neurologic deterioration was progressed and motor power of both the legs downgraded to MRC 2 only after 5 days.

Follow-up MRI revealed dilated perimedullary veins of the spinal cord seen as flow voids on T2 weighted imaging and as enhanced structures on post-contrast T1 weighted imaging (Fig. 1). Diagnosis of SDAVF was presumed based on MRI findings. Spinal angiography was performed to demonstrate a SDAVF. It showed anterior spinal artery, muscular branch, a feeder arising from right pedicular artery, and localized the AVF between the arterial feeder and perimedullary venous system (Fig. 2A). Although selective embolization of the arterial feeder drained from same trunk with anterior spinal artery was challenging, post embolization image showed significantly decreased flow to the venous system (Fig. 2B).

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**Figure 1.** Spine magnetic resonance imaging (MRI); (A),(B) sagittal, axial T2 weighted image, (C), (D) sagittal, axial enhanced T1 weighted image. Sagittal T2 weighted image shows diffuse enlargement and continuous high signal intensity in lower thoracic and lumbar spinal cord. Enlarged sagittal T2 weighted image and enhanced T1 weighted image with enhance show tortuous vascular structures in perimedullary space at thoracic spine level (A and C, arrows).

There was no progression of neurologic deficits after embolization. The patient was transferred to the rehabilitation center, and received physical therapy for 3 months. The patient showed improvement in muscle strength (MRC 2), but some sequelae remain such as difficulty in ambulation and urinary dysfunction.

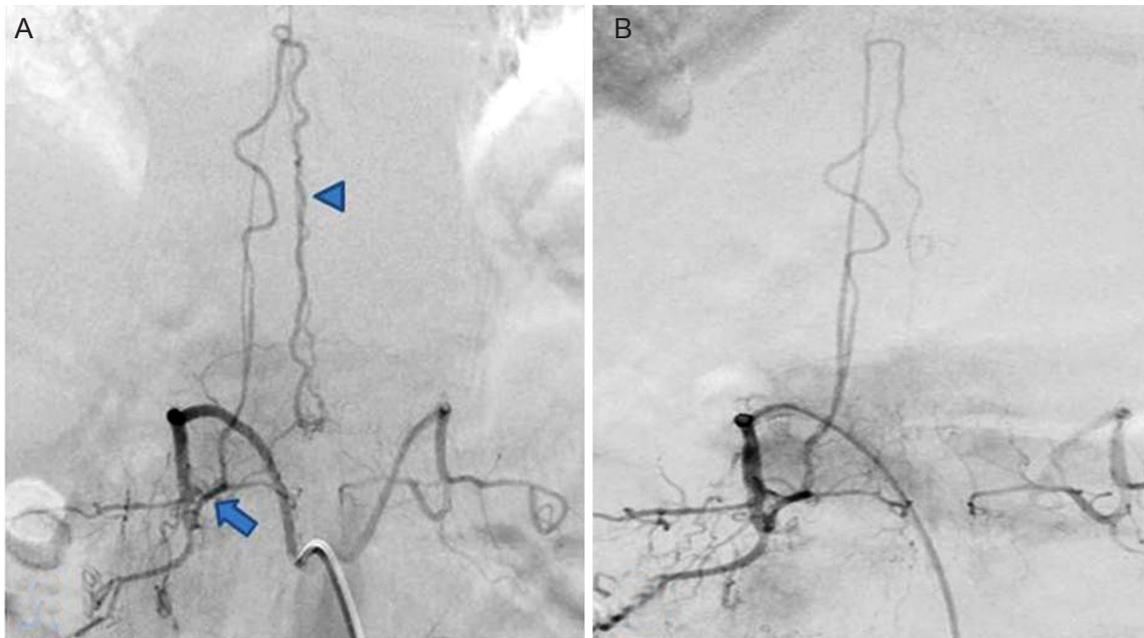
## DISCUSSION

Clinical manifestations of SDAVF typically include lower extremity weakness exacerbated by exercise, bowel or bladder disturbance, gait disturbances, and sensory symptoms without involvement of upper extremities. However, SDAVF is not easily diagnosed since there are more common etiologies with combination of nonspecific symptomatology such as degenerative disc disease, spinal stenosis,

and peripheral neuropathy.<sup>2,3</sup>

In a retrospective review of 326 patients with SDAVF, 265 patients were initially misdiagnosed in prior to the diagnosis of SDAVF.<sup>4</sup> The symptoms of SDAVF are usually progressive with an insidious development of disability over a period of time, and the time between the onset of symptoms and diagnosis has been reported to be between 12 and 44 months, with a mean duration of 22.9 months.<sup>5</sup> In our case, deterioration occurred very rapidly and the patient became severely disabled only within 3 weeks.

Three weeks after the onset, treatment of intravenous steroid pulse did not show any improvement in symptoms, so we had to review all previous studies from initial outside MRI. After evaluating all the data, the SDAVF was made as a final diagnosis. Four weeks after the onset, our patient underwent embolization of the fistula.



**Figure 2.** Spinal digital subtraction angiography. (A) Initial angiography at the right L1 segment shows a spinal dural arteriovenous fistula (arrow) fed from right pedicular artery and a dilated intramedullary vein (arrow head). (B) Follow-up angiography shows partial occlusion of the fistula after the embolization.

Enhancement of the dilated perimedullary veins is relatively specific manifestation of SDAVF and has been reported in as many as 88% of patients.<sup>6</sup> In this case, it would be more difficult to identify SDVAF without the MRI findings because of rapid progression. With recent advances in magnetic resonance angiography (MRA) and computed tomography angiography (CTA), diagnosis of SDAVF using noninvasive modalities is now attainable. Sensitivity and specificity of MRA in the diagnosis of SDAVF are 91% and 78%, respectively.<sup>1</sup> Multi-detector computed tomography angiography (MDCTA) has also led to the detection of abnormal perimedullary veins and an accurate localization of the fistula in 73% of patients.<sup>1</sup>

Since SDAVF can only be treated by intervention or surgery. Delay in diagnosis could lead to permanent disability. In accordance with one previous study, an estimated 50% of untreated patients were severely disabled within 3 years of the onset of lower extremity weakness.<sup>7</sup>

Although the patient received interventional treatment relatively early, some sequelae still remained. As imaging modalities have advanced, neurologists should be able to consider a possibility of SDAVF and diagnose the disease at an early stage.

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