Clinical Reasoning: A 49-Year-Old Woman With Isolated Sinus Intracranial Dural Arteriovenous Fistula With Perimedullary Drainage

Hui Lu, MD, Yong-jie Ma, MD, Heng Liu, MD, and Hai-Feng Li, MD, PhD

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Correspondence
Dr. Li
drlhf@163.com

Abstract

While demyelination is the most common etiology of longitudinally extensive myelopathy, other causes are important to recognize. In this study, we present the case of a longitudinal cervical lesion with a very rare cause. We discuss the approach to the differential diagnosis and workup for longitudinal myelopathy. This clinical reasoning case also illustrates the anatomical relationship between symptomatic spinal cord lesions and nonsymptomatic intracranial etiologies.

From the Department of Neurology (H. Lu, H.-F.L.); Department of Neurosurgery (Y.M.), Xuanwu Hospital, Capital Medical University, Beijing; and Department of Neurology (H. Liu), The Affiliated Hospital of Chengde Medical College, Hebei, China.

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Section 1

A 49-year-old woman noticed difficulty squatting beginning 9 months before presentation. She gradually developed numbness and cold in both legs and began to fall due to limb weakness. Two months before admission, the numbness spread to the lower abdomen, and she reported mild bowel and bladder dysfunction. Her medical and family histories were unremarkable.

On examination, she was alert and oriented. Cranial nerves were intact. The strength of bilateral hip flexor (iliopsoas), thigh adduction (adductor), knee extensor (quadriceps), knee flexor (hamstrings), foot dorsiflexor (tibialis anterior), and foot plantar flexion (gastrocnemius) was rated as 5-/5, while the strength in her upper extremities was 5/5. Abdominal reflexes were absent despite hyperreflexia of biceps and patellar tendons bilaterally. Pinprick and vibration were diminished below the sixth thoracic level. The Chaddock sign was present bilaterally, suggesting corticospinal tract damage.

Questions for Consideration:

1. What is the localization of the lesion?
2. What investigations would you initiate?

GO TO SECTION 2

Section 2

Physical examination suggested a lesion involving bilateral corticospinal tracts (bilateral lower extremity weakness, 4-limb hyperreflexia, absence of abdominal reflexes, and the Chaddock sign), spinothalamic tracts, posterior funiculus (diminished pinprick and vibration below T6), and intraspinal autonomic tracts (sphincter dysfunction).

Bilateral biceps hyperreflexia localized the lesion to above the 5/6th cervical segment. In addition, no signs related to corticobulbar tract (palmar mandibular reflex, snout reflex) or evidence of cranial nerve dysfunction were found, indicating a lesion below the medulla oblongata. Given that the examination suggested a transverse cervical lesion, a cervical spinal MRI was obtained, which revealed a swollen lesion extending from the first to the fourth cervical vertebrae (Figure 1, A–C).

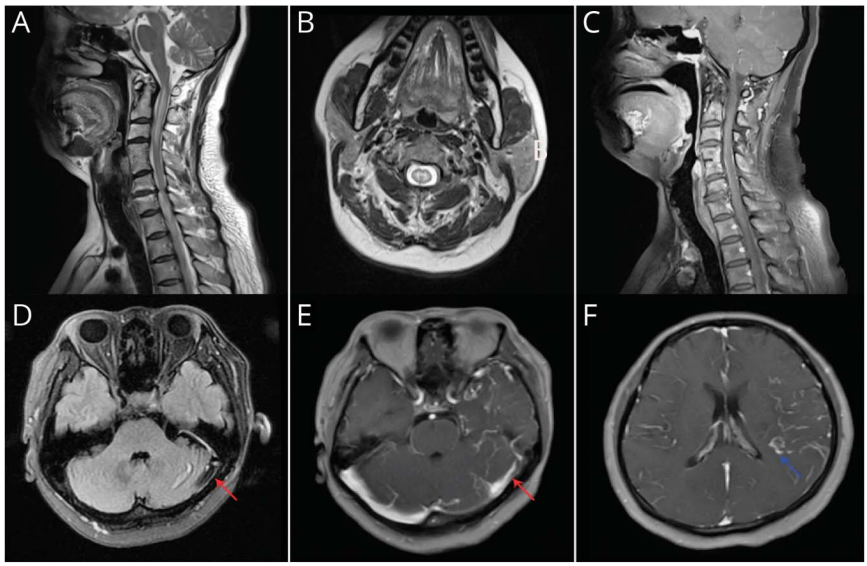
Based on the images from MRI, longitudinally extensive transverse myelitis (LETM) was initially considered as the most likely diagnosis. Possible etiologies for chronic progressive LETM include demyelination, chronic infection, sarcoidosis, or other rheumatism. Laboratory workup addressing the abovementioned etiologies was performed. The following serum investigations were unremarkable: complete blood count, erythrocyte sedimentation rate, C-reactive protein, infectious panel (hepatitis, HIV, tuberculosis, brucellosis, and syphilis), antinuclear antibodies (ANA), antineutrophil cytoplasmic antibodies (ANCA), and angiotensin-converting enzyme (ACE). CSF opening pressure was 80 mm H_2O, with normal cell count, protein, glucose, chloride, and oligoclonal bands. The following serum and CSF antibodies also showed negative results: anti–aquaporin-4 antibodies and anti–myelin oligodendrocyte glycoprotein (MOG).

Demyelinating disease, such as neuromyelitis optica spectrum disorder (NMOSD), is the most common cause of LETM among Chinese.[1] However, a chronic progressive course without remissions, spared optic nerves, and normal CSF profiling with negative antibodies ruled out NMOSD, MOG antibody–associated disease, or multiple sclerosis. The patient lived in north China where human T-lymphotropic virus type 1 infection is rare. Other chronic infections, such as syphilis or neuroborreliosis, were excluded based on negative test results reported earlier. Normal ACE level, normal pulmonary CT, and spared dorsal subpial enhancement argued against sarcoidosis. Because ANA and ANCA showed negative results, other rheumatisms were unlikely.

Questions for Consideration:

1. What is the differential diagnosis at this stage?
2. What would you do to uncover the etiology?

Figure 1 Cervical Spinal and Brain MRI



Sagittal T2WI (A), axial T2WI (B), and enhanced (C) cervical spinal MRI revealed a swollen lesion extending from the first to the fourth cervical vertebrae. T2 Flair (D) and enhanced brain MRI (E&F) showed left transverse sinus abnormality (red arrow) with multiple tortuous vessels in the left frontotemporal region (blue arrow).

GO TO SECTION 3

Section 3

Because no evidence for inflammatory or infectious cause was identified, alternative diagnoses should be considered. Given the chronic course, normal CSF profiling, and cervical spine MRI findings (Figure 1, A–C), differential diagnosis was expanded to include arteriovenous fistula (AVF), subacute combined degeneration, and copper deficiency. Spinal dural arteriovenous fistula (DAVF) is a well-known cause of myelopathy, generally located on thoracic or lumbar dura mater[2] with typical tortuous vessels and perimedullary flow voids seen on MRI. While the absence of flow voids in the patient's spinal MRI initially argued against spinal DAVF, they can be absent if shunt volume is small. The patient was screened for serum homocysteine, vitamin B_{12}, and copper, which were normal.

While waiting for spinal magnetic resonance angiography to investigate AVF, cranial MRI was performed to exclude intracranial lesions. Her cranial MRI showed left transverse sinus abnormality, with multiple tortuous vessels in the left frontotemporal region (Figure 1, D–F). Multiple tortuous vessels are highly suggestive of intracranial DAVFs. Rarely, intracranial DAVFs might drain into the perimedullary venous system and cause myelopathy.[3] Intracranial DAVFs are usually chronic, but acute onset has been reported in 25% cases.[4] Because symptoms are not related to the fistula site but the anatomical distribution of draining veins, early diagnosis of intracranial DAVFs is challenging.

Question for Consideration:

1. What would you do to clarify the diagnosis?

GO TO SECTION 4

Section 4

Magnetic resonance vein angiography (MRV) and digital subtraction angiography (DSA) are necessary to confirm the diagnosis. Her MRV demonstrated dilation of the left temporal drainage vein and partial left sigmoid sinus and stenosis of the left transverse sinus, the left internal jugular vein, and partial left sigmoid sinus. DSA confirmed DAVF with early visualization of the left transverse sinus. The DAVF was supplied by the meningeal branch of the left ascending pharyngeal artery, the left middle meningeal artery, the left posterior meningeal branch of the vertebral artery, the stylomastoid branch, and the lateral meningeal branch of the left occipital artery, with retrograde cortical venous reflux (CVR) from the left sigmoid sinus and the transverse sinus. There was retrograde blood flow through the superior petrosal sinus to the petrosal vein and then to the perimedullary veins (Figure 2A), which induced congestive myelopathy. Proximal and distal ends of the left transverse sinus were invisible, suggesting an isolated sinus. Intracranial DAVF with an isolated sinus was diagnosed and classified as Borden III and Cognard V.[5]

Because sinus thrombosis might be the cause of isolated sinus and DAVF, thrombotic predisposition including coagulation function test (prothrombin time, activated partial thromboplastin time, thrombin time, and fibrinogen), D-dimer, protein C&S, anticardiolipin antibody, and anti–β-2 glycoprotein 1 antibody were evaluated and were normal.

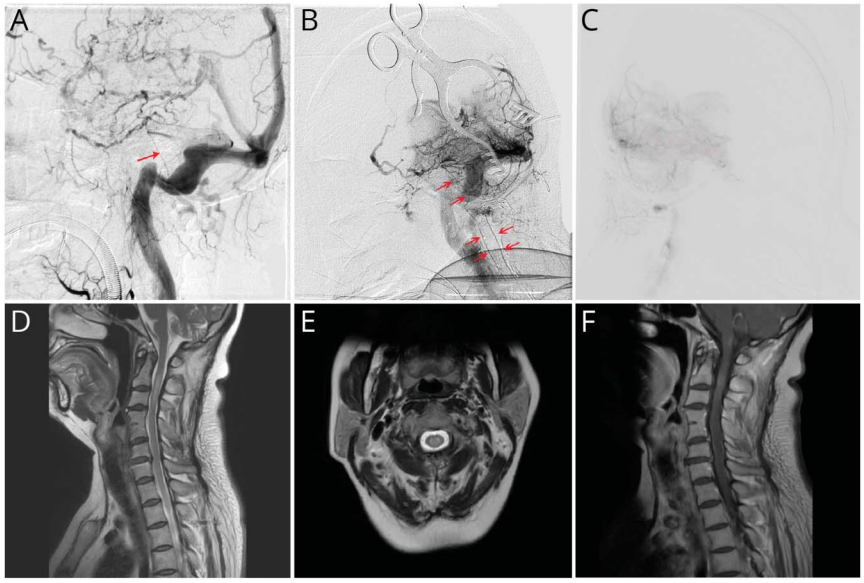
Onyx embolization through the left middle meningeal artery was performed, causing blood flow to reflux from the occluded left transverse sinus into the supplying artery. Next, embolization of the DAVF was conducted through the left ascending pharyngeal artery and left occipital artery. After partial embolization, the display of perimedullary veins was more obvious (Figure 2B). A follow-up angiogram revealed that supply arteries had mostly disappeared, but a complete occlusion of the fistula was not achieved. The residual fistula was surgically occluded through a retro-sigmoid approach. Postsurgery angiography revealed disappearance of the fistula (Figure 2C). Her postoperative course was uneventful. One month later, her strength of bilateral lower extremities returned to normal, with mild hypesthesia at the 12th thoracic level. Two months after the surgery, an MRI demonstrated relieved edema of the cervical cord (Figure 2, D–F).

Discussion

Isolated sinus is defined as dural venous sinus with thrombosis on both sides of the diseased sinus segment, which does not have any antegrade or retrograde venous access.[6] It has been reported that DAVF can develop secondary to sinus thrombosis.[7] Arteriovenous shunts preexist within the walls of the venous sinus and can become enlarged and recanalized after sinus thrombosis.[8] DAVF forms with a network of small vessels opening into the walls of isolated sinus.[7] Frequently, the venous flow refluxes into the cortical venous system,[9] resulting in venous congestion. Occasionally, the venous drainage redirects blood flow from the isolated sinus into perimedullary drainage veins, leading to spinal cord edema and myelopathy.[10] Most intracranial DAVFs draining into spinal veins are on the dura of the posterior cranial fossa, such as transverse and sigmoid sinuses, superior petrosal sinus and petrous apex, tentorium cerebelli, foramen magnum, anterior condylar foramen, and torcular herophili.[4,10]

The rare location of the intracranial DAVF usually leads to misdiagnosis because the symptoms are nonspecific. It requires critical time-to-treatment considerations because delays in

Figure 2 Digital Subtraction Angiography



(A) There was retrograde blood flow through the superior petrosal sinus to the petrosal vein and then to the cervical perimedullary veins (red arrow), which induced the hypertensive myelopathy of cervical spinal cord. (B) After partial embolization, the perimedullary veins (red arrow) were more obvious. (C) Postsurgery angiography revealed disappearance of the fistula. Sagittal T2WI (D), axial T2WI (E), and enhanced (F) cervical spinal MRI demonstrated relieved edema of the cervical cord 2 months after the surgery.

diagnosis can be potentially life-threatening.[6] However, it is difficult for neurologists to distinguish intracranial DAVFs from other causes of myelopathy. Longitudinal myelopathy without cord surface flow voids is prone to be misinterpreted as antibody-negative NMOSD. Physicians should be aware of vascular myelopathies early because glucocorticoid administration might cause acute deterioration in 50% patients with DAVF.[11,12] Fluid retention and increased venous congestion are proposed to be potential mechanisms for deterioration after steroid.[12] Characteristics of NMOSD to differentiate from vascular myelopathy include intramedullary hypointensity on T1WI, bright spot on T2WI, and gadolinium enhancement of the lesion, which might be absent in vascular myelopathies.[13] Diagnostic clues for intracranial DAVF are engorgement of upper spinal veins and lower brainstem veins, serpentine signal voids, or extramedullary enhancement, although these indicators might be absent, as the case in this patient.[14] The golden standard imaging for DAVF is DSA. If vascular myelopathy is highly suspected, cerebral angiography should be considered to avoid misdiagnosis when spinal angiography fails to reveal an AVF.

The Borden and Cognard classification of DAVF is based on venous drainage.[5] High-grade fistulas with CVR are proven to be aggressive, indicating an increased risk of intracranial hemorrhage and neurologic deficits.[5] A multimodal approach (embolization, surgery, or both) is often required to achieve complete occlusion of the fistulous zone and closure of both distal feeding arteries and proximal draining venous pedicles.[6,15] The optimal treatments remain debated and controversial.

In summary, we reported a rare cervical myelopathy case caused by an isolated sinus with intracranial DAVFs. Intracranial DAVF should be considered in differential diagnosis of myelopathy because steroid administration would cause acute deterioration.

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Appendix Authors

Name	Location	Contribution
Hui Lu, MD	Department of Neurology, Xuanwu Hospital, Capital Medical University, Beijing, China	Drafting/revision of the article for content, including medical writing for content; major role in the acquisition of data
Yong-jie Ma, MD	Department of Neurosurgery, Xuanwu Hospital, Capital Medical University, Beijing, China	Drafting/revision of the article for content, including medical writing for content; major role in the acquisition of data
Heng Liu, MD	Department of Neurology, The Affiliated Hospital of Chengde Medical College, Hebei, China	Major role in the acquisition of data
Hai-Feng Li, MD, PhD	Department of Neurology, Xuanwu Hospital, Capital Medical University, Beijing, China	Drafting/revision of the article for content, including medical writing for content

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