Case Report/Series

Intracranial dural arteriovenous fistula presenting like longitudinally extensive transverse myelitis

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ABSTRACT

Intracranial dural arteriovenous fistula (DAVF) that drains into spinal perimedullary veins can generate longitudinally extensive transverse myelitis (LETM)-like lesion, which often represents a significant diagnostic and therapeutic challenge. This is a case report of a 50-year-old male referred with all extremity weaknesses. Despite receiving high-dose intravenous steroids for suspected myelitis, no improvement was recorded. Spinal imaging showed abnormal hyperintensity extending from the T6 vertebral level to the medulla, and a flow void lesion from the cervicomedullary junction up to the L3 level. Angiography confirmed a Cognard type V spinal DAVF, which was treated with transarterial embolization of the feeding vessel. Follow-up angiography showed complete occlusion of the fistula without any backflow. Flow voids are no longer visible on MRI conducted 3 weeks post-procedure. Unfamiliarity with these disorders often leads to delays in diagnosis and treatment. Therefore, it is essential to consider intracranial DAVF as a differential diagnosis for LETM-like lesions.

KEYWORDS endovascular procedures, dural arteriovenous fistula, transverse myelitis

Intracranial dural arteriovenous fistulas (DAVFs), a unique subtype of vascular malformations of the central nervous system, are characterized by pathological connections between the meningeal arteries and dural venous sinuses or cortical veins, constituting 10–15% of all brain vascular malformations.[1,2] Its clinical presentation and classification are influenced by the arterial supply, arteriovenous (AV) shunting level, and venous drainage pattern.[1]

Borden et al[3] classified DAVFs into three groups based on the characteristics of venous drainage: type I directly drains into a sinus or meningeal vein; type II drains into a sinus but exhibits retrograde drainage into other sinuses (IIa) or cortical veins (IIb); type III drains directly into cortical veins. Cognard et al[4] expanded the classification into five groups, adding type IV, which drains into the cortical vein with a large venous ectasia, and type V, with spinal venous drainage. The presence of cortical venous drainage in Borden type II and III, and Cognard type IIb–V is associated with a high-risk profile for DAVFs.[5]

Cognard type V DAVF is a rare vascular anomaly that is challenging to diagnose because of its various clinical presentations. It may be localized to the spinal cord despite the intracranial location of the fistula. Cortical venous drainage often appears more aggressively due to spinal vein congestion, with symptoms including severe headaches, intracranial hemorrhage, and non-hemorrhagic neurological deficits, such as seizure and myelopathy.[6] Common misdiagnoses of type V DAVFs have included myelitis (16%), spinal DAVF (13%), tumors (9%, mainly lymphoma and glioma), and strokes (5%). Initial symptoms typically involve motor deficits (78.9%, either paraparesis or quadriparesis), followed by sensory symptoms (38.9%), sphincteric disturbances (25.6%), dizziness (17.4%), ataxia (12.6%), and brainstem

symptoms (8.9%).[7] A recent simplified classification divides intracranial DAVFs into two groups: sinus fistula, where dural arteries shunt directly into a dural sinus, and non-sinus fistula, with the pathological shunt present in the dural leaflet without communication with the sinus, allowing the sinus to continue functioning in brain venous drainage.[8] This report presents the case of a patient diagnosed with intracranial Cognard type V DAVF treated with transarterial embolization (TAE). This study examined the diagnostic characteristics and identified clues that may help in the early differentiation from transverse myelitis.

CASE REPORT

A 55-year-old male was referred from the department of neurology with a 4-week history of gradually worsening walking difficulties, burning, and decreased sensation in both of his upper and lower extremities. He experienced progressive weakness in his lower extremities for 3 weeks before admission, which worsened in the last week, followed by weakness in the upper extremities, urinary retention, and constipation.

Physical examination revealed that his motor power grades varied across muscle groups. In the right lower extremity, the hip flexors and extensors, and knee extensors and flexors were graded 2/5, while the ankle plantar flexors were graded 1/5. All muscle groups in the left lower extremity were graded 0/5. The bilateral ankle dorsiflexors, toe flexors, and extensors were graded 0/5. The right upper extremity showed various strengths, with shoulder abduction and adduction graded as 4/5 and elbow and wrist movements graded as 5/5. For the left upper extremity, both shoulder abduction and adduction, and elbow flexion and extension were graded 5/5, while wrist and finger movements varied slightly in strength. Sensory examination showed a reduced sensation of light touch and vibration around the acromioclavicular joint or C4 dermatome, extending distally and involving both lower extremities. Proprioception in the bilateral foot was decreased. The patient exhibited hyperactive deep tendon reflexes, clonus, and positive Babinski signs bilaterally. The cranial nerves were grossly intact, with bulbar symptoms, including swallowing difficulties and postural hypotension.

Magnetic resonance imaging (MRI) of the spinal cord, both with and without contrast, revealed an intramedullary hyperintensity signal spanning three vertebral segments from the medulla oblongata to the T6 level. Initially, the patient was diagnosed with a longitudinally extensive transverse myelitis (LETM) lesion and myelopathy secondary to an inflammatory etiology. T2 flow voids and T1 post-gadolinium imaging revealed serpentine-dilated perimedullary veins extending from the cervicomedullary junction to the L3 level (Figure 1).

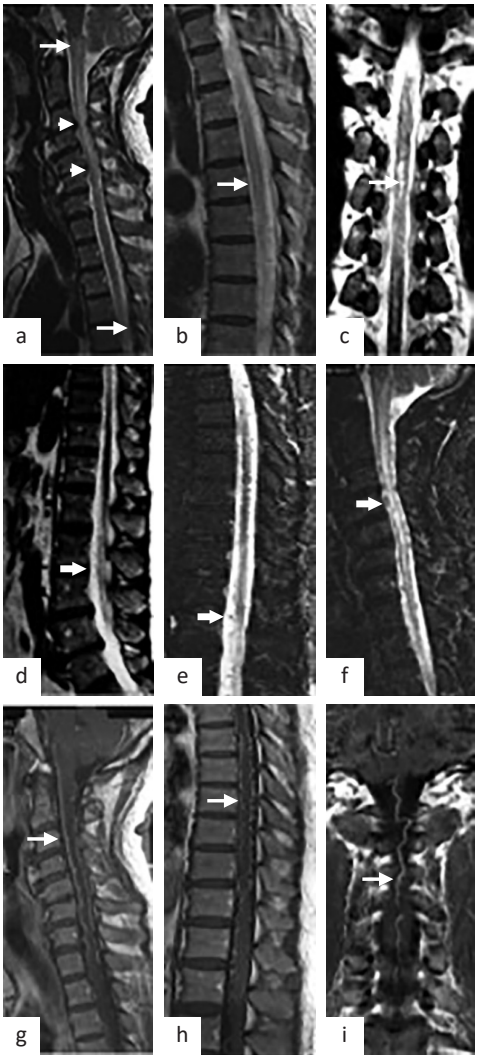

Figure 1. Spinal cord MRI. Spinal cord T2-weighted MRI sequence showing hyperintensity starting from the CMJ down to T6 (thin arrows) (a–c); cervical spondylosis was seen (white arrowheads), but not significant as the cause of the current symptoms. Numerous flow voids with edema and swelling along of the spinal cord down to lumbar level (arrows) (d–f). Spinal cord sagittal (g and h) and coronal (i) gadolinium-enhanced T1-weighted sequence showing enlargement of the spinal veins in the anterior and posterior epidural space (thin arrows). CMJ=cervicomedullary junction; MRI=magnetic resonance imaging

Brain MRI revealed hyperintensity on T2 and isointensity on diffusion-weighted imaging involving

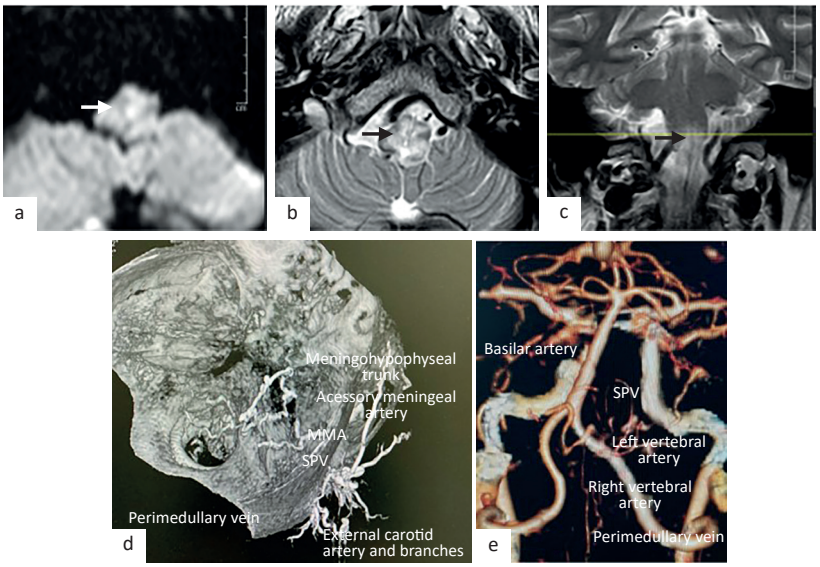

Figure 2. Cerebral imaging. Brain MRI with normal intensity signal (white arrow) at axial DWI (a) and high intensity signal (black arrow) at T2-weighted sequence (b) at the level of medulla oblongata, as showed at coronal section (black arrow) (c). Cerebral CT angiography with right anterior oblique view of 3D bone reconstruction (d) and 3D CT angiogram (e) showing early filling of perimedullary vein during arterial phase. CT=computed tomography; DWI=diffusion-weighted imaging; MMA=middle meningeal artery; MRI=magnetic resonance imaging; SPV=superior petrosal vein

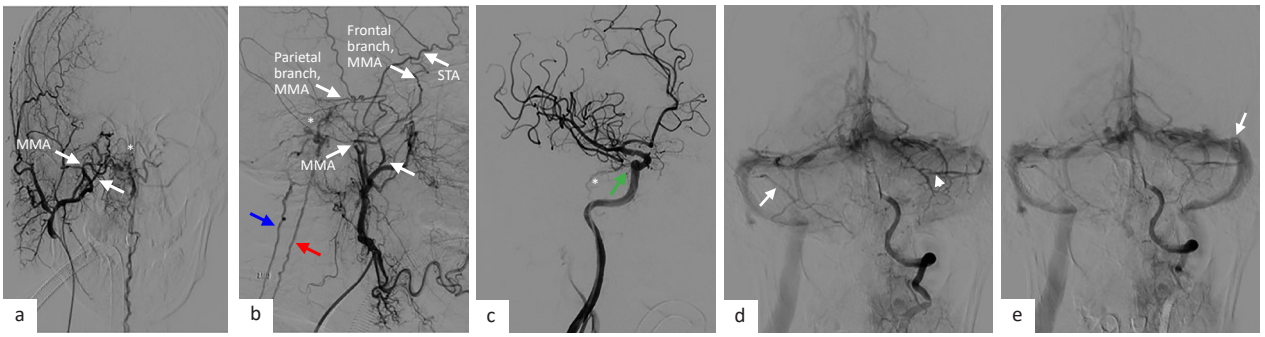

Figure 3. Angiographic study on right external carotid artery on anteroposterior (a) and oblique projection (b) demonstrate a network of dural collateral arteries to the fistula point. Although the main supply to the shunt is derived from the branch of right acessory meningeal artery (white arrow) drainage to perimedullary veins (anterior [red arrow] & posterior [blue arrow] spinal veins), there is an additional supply from posterior side of MMA and meningohypophyseal trunk of internal carotid artery (green arrow) (c). The fistula point is denoted by an asterisk (*). Frontal view of the left vertebral angiography at venous phase demonstrated left SPS (white arrowhead) and partially fills the right one (white arrow) (d). Transverse sinus appears more prominent on the left side (white arrow) (e). MMA=middle meningeal artery; SPS=superior petrosal sinus; STA=superficial temporal artery

the medulla oblongata and upper cervical region, indicating vasogenic edema without ischemia and no post-contrast enhancement. Cerebral computed tomography angiography revealed early contrast filling of the vein during the arterial phase at the left petrous apex (Figure 2).

Despite daily intravenous administration of methylprednisolone (1 g), no clinical improvement was observed until the fifth day of treatment. Comprehensive testing included metabolic, hematological, and autoimmune investigations for neuromyelitis optica IgG and anti-dsDNA, along with an infectious workup covering viral, bacterial, parasitic, and fungal causes. Cerebrospinal fluid analysis from a lumbar puncture that included protein, glucose, and cell counts yielded unremarkable results. Despite steroid administration, progression of motor weakness was noted in the right upper extremity, with a power grade of 4/5. Because the progression of symptoms extended beyond 4 weeks, another diagnosis other than LETM had to be considered.

A super-selective cerebral angiogram of six vessels indicated the presence of a right posterior fossa DAVF supplied by the accessory meningeal artery and the petrosal branch of the right middle meningeal artery (MMA), with an additional small feeder from the tentorial branch of the meningohypophyseal trunk (Figure 3). Venous drainage occurred in the right superior petrosal sinus through the superior petrosal vein (SPV), anterior pontomesencephalic vein, and

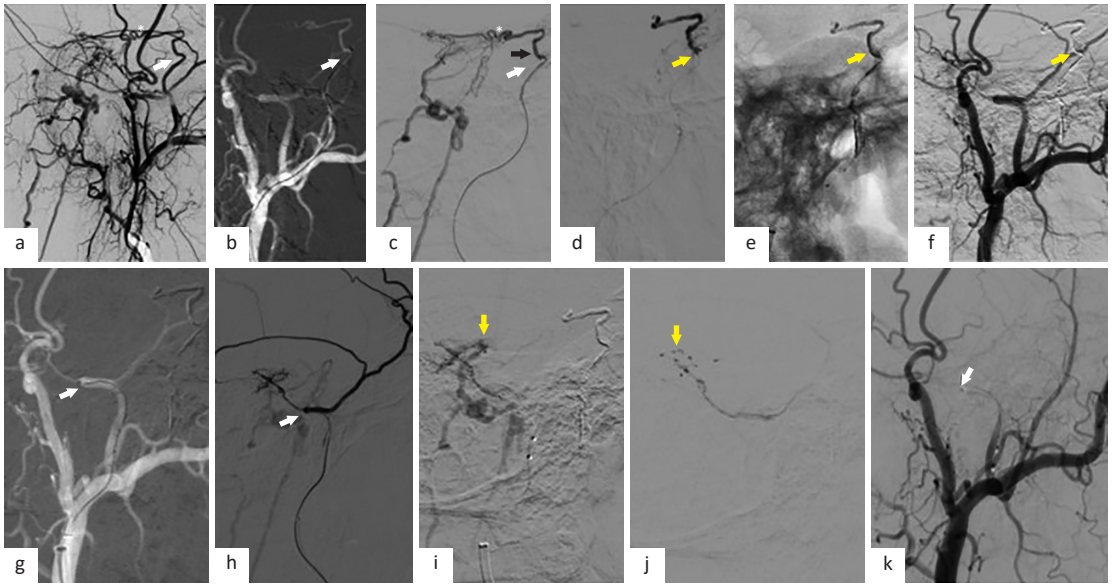

Figure 4. TAE processes. Accessory meningeal artery embolization (a–f). Initial angiography shows dural AV fistula (*) feeded from right accessory meningeal artery (white arrow) (a). At the most distal branch of accessory meningeal artery, a microcatheter (black arrow) was directed to fistula point (b and c). The fistula point is denoted by an asterisk (*). Squid® 12 injection is done through a microcatheter under fluoroscopic control (yellow arrows) (d and e). After embolization, flow to fistula point decreased significantly (yellow arrow) (f). Posterior branches of MMA embolization (g–k). A microcatheter (Sonic 1.2 F; Balt Montmorency, France) was advanced toward the fistula (white arrow) (g) contrast media injection confirmed the fistula point (white arrow) (h). Embolization process using Squid® was performed (yellow arrows) (i and j). External carotid artery angiography showed complete obliteration of the fistula (white arrow) on post-embolization (k). AV=arteriovenous; MMA=middle meningeal artery; TAE=transarterial embolization

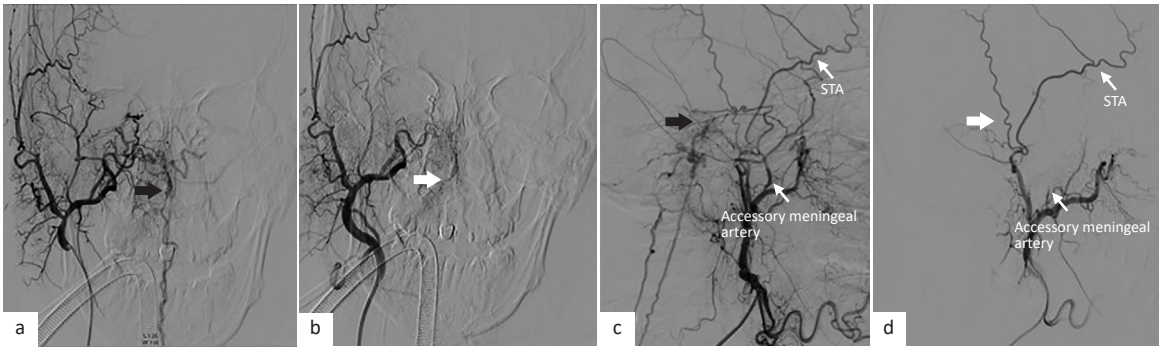

Figure 5. Angiographic comparison before and after embolization treatment. Anteroposterior projection pre-embolization (black arrow) (a) and post-embolization (white arrow) (b) fistula point. Lateral projection pre-embolization (black arrow) (c) and post-embolization (white arrow) (d) showing complete obliteration of fistula point. STA=superficial temporal artery

spinal perimedullary veins (anterior and posterior spinal veins).

After obtaining informed consent from the patient and his family, we used an endovascular approach for treatment. Diagnostic angiography and TAE were performed under general anesthesia using a digital subtraction angiography machine (AlluraClarity FD20; Philips Healthcare, United Kingdom) via femoral access. Heparin was administered intravenously at an initial dose of 3,000 units, followed by 1,000 units per hour, to maintain an activated coagulation time of 250–300 sec. A 6 Fr guiding catheter was navigated to the external carotid artery, and the MMA was accessed using a Sonic 12 microcatheter (Balt Group, France) to reach the fistula site. Squid® (Balt, USA), a liquid embolic agent composed of ethylene-vinyl alcohol copolymer dissolved in a dimethyl sulfoxide solution with micronized tantalum powder for radio-opacity (Figure 4), was used to occlude the venous channels proximally at a controlled rate of 0.1 to 0.2 ml per minute to prevent vasospasm and angionecrosis.[9,10] Subsequent embolization sessions were conducted

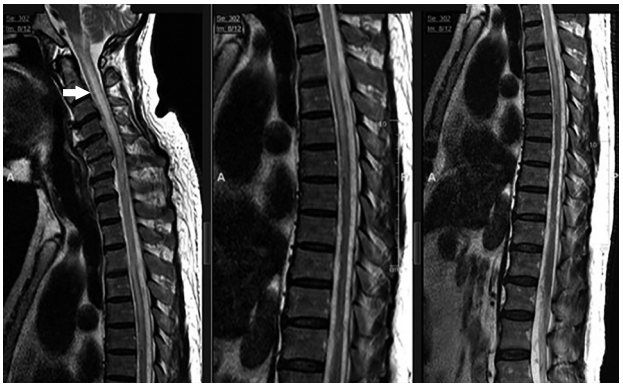

Figure 6. Postprocedural magnetic resonance imaging (MRI) T2 sequence showing improved cord signal, centromedullary hyperintensity (arrow) remained the same

through other feeders until complete disconnection of the fistula was achieved, confirmed by angiographic obliteration (Figure 5).

After 2 weeks, clinical assessment revealed a slight improvement in the patient's upper extremity power, while his lower extremity strength remained the same. The 3-week follow-up MRI revealed the absence of flow voids but persistent spinal cord edema (Figure 6). Three months after the procedure, both of his upper limbs had improved to grade 5, enabling him to achieve a modified Rankin scale score of 4 and sit independently.

DISCUSSION

Intracranial DAVF with spinal venous drainage account for 5.9% of all intracranial DAVFs. Typically located in the posterior cranial fossa, it may resemble a spinal DAVF[11–13] and is frequently misdiagnosed as another neuromuscular disorder due to nonspecific symptoms. In some cases, patients may undergo surgery for disc protrusions that are incidentally discovered on imaging.

Whittam et al[14] reported six cases of intracranial DAVF causing cervical cord or brainstem edema over 5.5 years, with four patients experiencing acute deterioration and clinical worsening after steroid treatment. All of the patients' symptoms lasted for 1–8 months. Similarly, a systematic review by De Grado et al[7] discovered that most patients of type V intracranial DAVF were middle-aged males, with 65% showing progressive symptoms, 79% showing initial motor symptoms, and 58% experiencing an initial misdiagnosis. Su et al[13] found a similar case of type V DAVF with draining veins to the cerebellar cortical veins, veins of the ventral brainstem, and perimedullary veins, which also presented initially with lower limb weakness. In contrast, Iampreechakul et al[11] reported a cavernous sinus DAVF connected to the perimedullary vein that led to mild migraine proptosis and diplopia without motor weakness. Moreover, Hou et al[15] analyzed 84 patients with intracranial DAVFs complicated by brainstem engorgement. The Cognard classification of DAVFs was II, III, IV, and V in 9 (10.7%), 1 (1.2%), 1 (1.2%), and 73 (86.9) patients, respectively.

The initial presentation of myelopathy poses diagnostic challenges. When a suspected myelitis lesion appears as a parenchymal T2 hyperintensity extending over three or more vertebral segments on sagittal spinal cord sequences, it is classified as longitudinally extensive.[16] Multiple studies have revealed that 50% of the clinical presentations and angiographic features of intracranial DAVFs with spinal venous drainage present with progressive myelopathy. This phenomenon is often attributed to extensive venous drainage towards the lower thoracic, lumbar, and conus medullaris levels.[3]

In this case, the fistula was supplied by branches of both the external and internal carotid arteries, mainly draining into the perimedullary venous system. The redirected arterial flow into the SPV leads to venous congestion in the spinal cord, affecting the spinal perimedullary veins. Post-gadolinium T1-weighted MRI imaging revealed dilated leptomeningeal and medullary vessels, venous ectasia, and parenchymal enhancement. T2-weighted and fluid-attenuated inversion recovery sequences highlighted the hyperintensities, which indicated vasogenic edema caused by venous hypertension.[17] Additionally, T2-weighted MRI scans displayed a serpentine vascular signal void from arterialized veins and diffuse edema in the cervical spinal cord, both are indicative markers of vascular abnormalities.[7,18–20] An analysis of 72 publications by De Grado et al[7] found that the MRI findings for Cognard type V DAVF include flow voids, T2 hyperintensities, and swelling in approximately 81.6%, 80.5%, and 56.3% of cases, respectively.

The treatment objective is to isolate the fistula from the draining vein using either surgical methods, endovascular techniques (including transarterial and transvenous), radiosurgery, or a combination of these strategies.[21] Generally, while spontaneous resolution may occur,[11] all treatment options remain justifiable.

However, the selection of approach depends on factors such as patient presentation, lesion characteristics, surgeon preferences, resources, and available skills.

Detailed assessment of the angioarchitectural features is vital for safe and efficient treatment planning.[22] In this case, the endovascular approach was chosen because of its less invasive nature. If selective catheterization of the dural feeding vessels is impossible, surgical intervention may be performed. Common surgical strategies include coagulation or clipping of the draining vein, obliteration of the arterial feeders or fistulas, and removal of the involved dural sinus. It is crucial to differentiate between sinus and non-sinus types of intracranial DAVFs. Sinus fistulas exhibit a direct connection between an arterial dural branch and the dural sinus, and sometimes involve subsequent cerebral veins known as "red" or "arterialized veins" due to their abnormal blood flow. These non-functional veins should be cut off from the circulation. In their analysis of treated DAVFs, Kim et al[22] found that the transarterial route in a single endovascular embolization session was safe and efficient for isolated sinus cases, achieving complete or near-complete closure. In our case, TAE was selected to eliminate the fistula due to the isolation of the left petrosal sinus from thrombosis or stenosis.[22]

A DAVF is an acquired lesion with two leading etiological theories linked to sinus thrombosis. The first theory suggests that increased local venous pressure can cause the enlargement of AV shunts between the meningeal arteries and dural venous sinuses, resulting in a pathological shunt. The second theory proposes that venous hypertension stemming from outflow obstruction leads to reduced cerebral perfusion and stimulates neoangiogenesis.[23]

In the context of intracranial DAVF with spinal perimedullary vein drainage, three hypotheses outline the pathophysiology of myelopathy: venous hypertension, arterial steal, and direct compression by enlarged veins. Among them, venous hypertension is the most widely acknowledged. Blood stagnation in the intramedullary veins reduces microcirculation perfusion, leading to vasogenic edema and myelopathy. In our case, there was a decrease in venous flow through the right transverse and petrosal sinuses, leading to predominant drainage through the perimedullary venous plexus.[24]

Initially misdiagnosed with longitudinal transverse myelitis, the patient received steroids, which led to clinical deterioration. This adverse effect is likely due to transient mineralocorticoid-induced hypervolemia and resultant venous hypertension, a potentially life-threatening condition, particularly with high cervical cord involvement. Most patients with venous hypertensive myelopathy remained disabled post-treatment.[14]

This case management perspective was based on a retrospective review of published case reports and other literature. Treatment results were influenced by several factors, including delays in diagnosis and treatment. Moreover, the follow-up period was only 3 months, making further follow-up impossible to initiate due to a lack of patient adherence.

In conclusion, Cognard type V DAVFs, which have historically been challenging to diagnose, may resemble myelitis. These findings highlight the need to consider suspected cases of LETM with central myelopathy and dilated perimedullary veins. A lack of familiarity with this condition and its pathophysiology, along with discrepancies between clinical symptoms and lesion locations, may lead to diagnostic delays and hinder appropriate treatments. Given the potential for severe complications, clinicians should consider the possibility of an intracranial vascular anomaly as a cause of myelopathy.

Conflict of Interest
The authors affirm no conflict of interest in this study.

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