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Case Report

Precision Mapping: Intraoperative Indocyanine-Green Video Angiography in Thoracic Spinal Dural Arteriovenous Fistula, The Surgical Management: A Case Report

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Abstract

Background: The sudden or gradual onset of bilateral lower limb weakness, coupled with sensory deficits and alteration in reflex activity, should give rise to significant concern for potential emergent spinal conditions. The common spinal pathologies that we usually encounter are spinal cord compression, trauma, cauda equina syndrome, and Guillain-Barré syndrome. Conversely, there is a rare condition known as Foix-Alajouanine syndrome, which manifests with a wide spectrum of neurological symptoms originating from spinal vascular malformations. The formation of spinal dural arteriovenous fistula (dAVF) represents a prominent manifestation of Foix-Alajouanine syndrome.

Case Presentation: We report an uncommon case of thoracic spinal dural arteriovenous fistula (dAVF) managed through microsurgery with intraoperative Indocyanine-Green (ICG) video angiography assisted in a 37-year-old gentleman. This is a complex case, as the usual endovascular approach was not feasible due to the torturous configuration of the arterial feeder vessels. Our patient showed no neurological improvement during the 1st and 3rd-month follow-up evaluations, presumably owing to the delayed onset of the condition. Prolonged monitoring may unveil amelioration in our patient's symptoms.

Conclusion: Foix-Alajouanine syndrome, albeit uncommon, merits attention in cases of progressive myelopathy. Microsurgical intervention for spinal dural arteriovenous fistula (dAVF), complemented by intraoperative ICG video angiography, is an efficacious treatment strategy. Timely intervention is crucial for favorable outcomes.

Keywords: Spinal dural arteriovenous fistula (dAVF), Foix-Alajouanine syndrome, Microsurgery, Spinal vascular malformations

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Introduction

Acute or insidious development of bilateral lower limb weakness, accompanied by sensory loss, hypo- or hyperreflexia, and bladder dysfunction, should prompt a high level of suspicion for emergent spinal pathology. Examples, to name a few, include spinal cord compression or trauma, cauda equina syndrome, and Guillain-Barré syndrome. Additionally, there exists a rare condition called Foix-Alajouanine syndrome, characterized by a diverse array of neurological manifestations stemming from spinal vascular malformations. This syndrome is characterized by spinal vascular malformations, which can lead to venous congestion and ischemic myelopathy.[1] It predominantly affects the thoracic and/or lumbosacral regions, with spinal dural arteriovenous fistula (dAVF) representing a notable manifestation.[2] Here, we present a case of thoracic spinal dAVF managed through microsurgery with intraoperative Indocyanine-Green (ICG) video angiography assistance. The clinical presentation and treatment strategy are discussed in order to enhance disease awareness and facilitate early diagnosis.

Case Presentation

A 37-year-old gentleman with no significant past medical history presented with progressively worsening bilateral lower limb pain and weakness, persisting for three months. He had sought medical attention in emergency care multiple times over nine months. A physical examination of the patient revealed that he had no signs of external trauma along the spinal column. However, there was diminished strength in lower limbs, with Medical Research Council (MRC) scores of 2 for hip and knee movements and 1 for ankle movements. He lacked the ability to stand or walk unaided without risking safety and exhibited reduced sensory perception extending bilaterally from the L2 dermatome downwards, along with perineal anesthesia and absent reflexes. The rectal tone was also absent, with complaints of overflow incontinence. White cell counts, hemoglobin, platelet count, and renal profile were within normal limits. Anterior-posterior and lateral lumbosacral radiographs showed no abnormalities. Immediate magnetic resonance imaging (MRI) of the entire spine revealed central hyperintensity from the level T5 till the conus with dilated perimedullary veins on T2-weighted sagittal sequences (Figure 1A), indicative of spinal dAVF.[3] A magnetic resonance imaging (MRI) is a valuable tool for the initial assessment of spinal cord pathologies because it can precisely outline any abnormalities. Following that, the goal standard procedure, which is a spinal digital subtraction angiography (DSA), was done. The spinal digital subtraction angiography (DSA) revealed a Type IB thoracic dAVF supplied by left T5 and T6 radiculomedullary arteries and tortuous draining veins (Figure 1B) with long-segment tortuous dilated veins in the thoracic spinal column (Figure 1C). Endovascular treatment, with an up to 89.5% success rate and low morbidity, has emerged as the primary treatment for dAVF.[2] However, due to the narrow and tortuous vessel course (Figure 1B), endovascular treatment was deemed unfeasible for this case. Following multidisciplinary consultation, microsurgical occlusion of the spinal dAVF was performed. Under general anesthesia, the patient was placed prone, and a midline linear incision was made, followed by subperiosteal dissection. Intraoperative fluoroscopy confirmed vertebral levels, and a T4-T6 laminectomy was performed. Dura was opened midline, revealing severely dilated tortuous vessels at the dorsum of the spinal cord (Figure 2A). ICG administration enabled visualization of the arterialized vein early in the arterial phase (Figure 2B), followed by the dilated draining veins in the venous phase (Figure 2C). A temporary clip was applied to the arterialized vein near the fistulous point adjacent to the left T6 exiting nerve root (Figure 2D). Post-clip application ICG run demonstrated the obliteration of the arterialized vein while maintaining draining vein flow, albeit less intense than before (Figure 2E). The arterialized vein was coagulated and sectioned, followed by watertight fascia duraplasty and lamina replacement using titanium plates and screws. Following surgery, he stayed hospitalized for 1 month to undergo rehabilitation and physiotherapy. During this period, he experienced an uncomplicated recovery. However, lower limb motor strength remained unchanged, and urinary catheter dependency persisted. Finally, a rehabilitation program for outpatient care was started as soon as he was discharged. The program consisted of both passive and active assisted range of motion exercises, muscle strengthening routines, neuromuscular electrical stimulation, and balance training. Postoperative MRI showed resolved cord edema and dilated perimedullary vessels (Figure 3A), while DSA revealed no residual dAVF (Figure 3B).

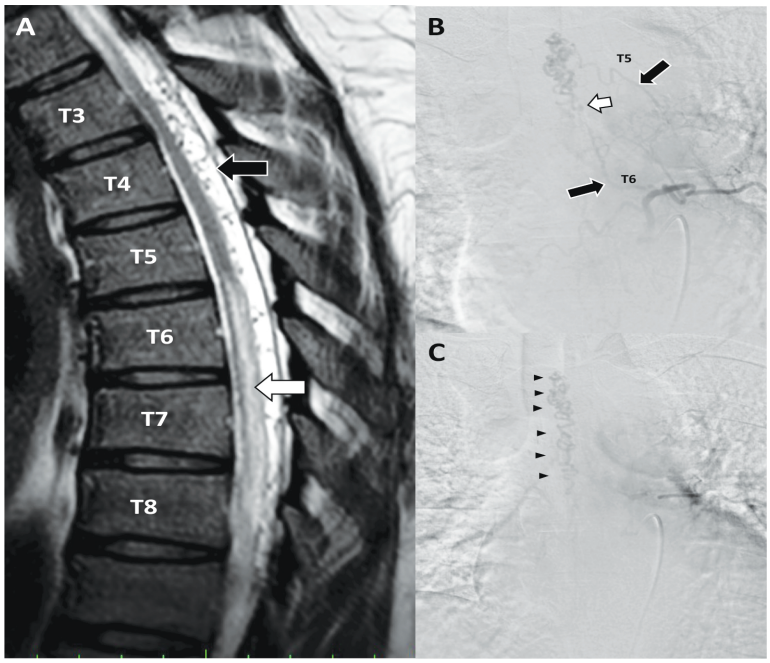

Figure 1 A: T2-weighted MRI of the sagittal thoracic spine revealed centromedullary hyperintensity extending from T5 to the conus (white arrow), accompanied by dilated perimedullary vessels (black arrow). These findings are characteristic of spinal dAVF.

B, C: DSA, following injection of the left T6 intercostal artery, demonstrated a thoracic spinal dAVF supplied by the left T5 and T6 radiculomedullary arteries (black arrows). The arterialized vein (white arrow) and dilated draining veins (multiple black arrowheads) were also observed.

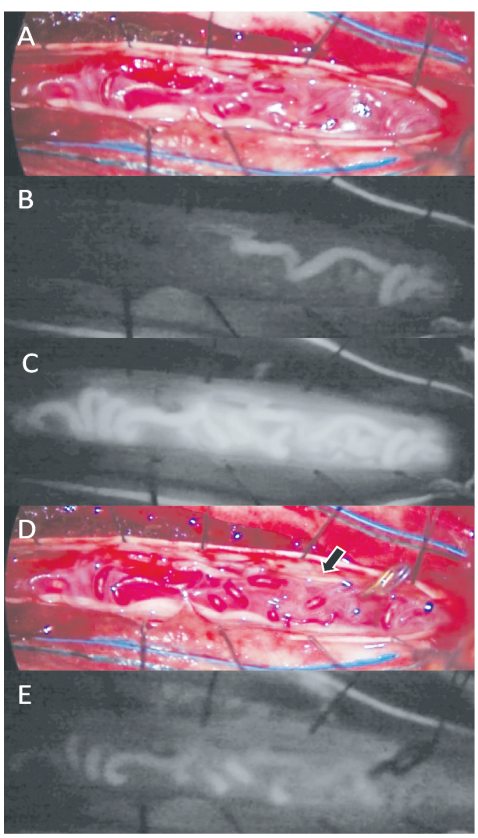

Figure 2 A - Upon durotomy, tortuous dilated vessels were observed at the dorsum of the spinal cord. Gross visualization revealed significant difficulty discerning the arterialized vein from the dilated draining veins.

B - Indocyanine-green was administered, highlighting the arterialized vein early in the arterial phase.

C - Indocyanine-green revealed the dilated draining veins during the venous phase.

D - A temporary clip was placed on the arterialized vein near the fistulous point, adjacent to the left T6 exiting nerve root (black arrow). Typically, the arterialized vein is supplied by the dura superficial to the nerve root.

E - Another indocyanine-green run demonstrated a disruption of blood flow to the arterialized vein while still maintaining flow to the draining veins, albeit with less intensity compared to before the fistula was obstructed.

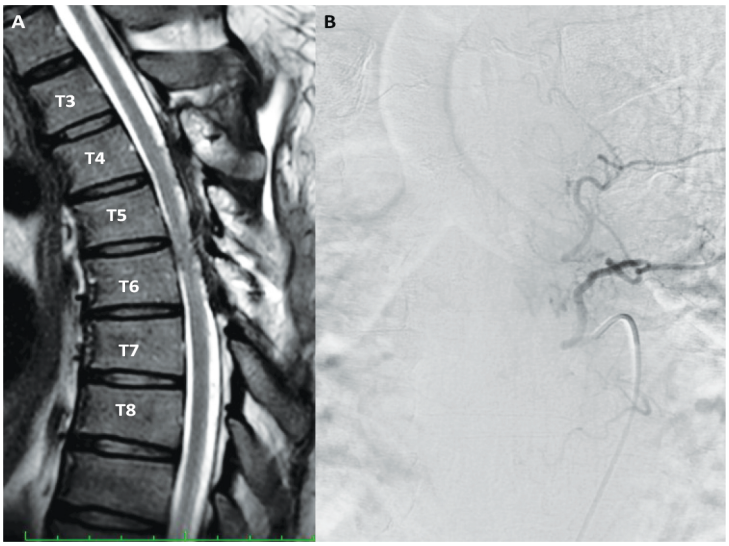

Figure 3 A: Post-operative MRI showed resolved cord oedema and dilated perimedullary vessels. B: DSA post-surgical occlusion revealed no residual dAVF.

Discussion

Spinal dural arteriovenous fistulas (dAVFs) are characterized by anomalous connections between arteries and veins without a distinct nidus. Despite their rarity, they are the most common type of spinal vascular malformation.[2] Foix-Alajouanine syndrome is not a standalone condition but rather a complication stemming from spinal dAVFs. Initially described by Foix and Alajouanine in 1926, this syndrome presents as necrotizing myelopathy, with further elucidation provided by Lhermitte in 1931. Aminoff and Logue proposed in 1974 that spinal arteriovenous fistulas (AVFs) lead to elevated intramedullary venous pressure, resulting in reduced arteriovenous pressure gradients and subsequent cord perfusion decline. Typi cally, the lower portion of the cord is predominantly affected, demonstrating signal alterations on MRI scans.[4] However, as evidenced by our case, misdiagnosis upon presentation is not uncommon, necessitating a high index of suspicion due to the exclusion of other urgent spinal pathologies during emergency department assessments. Patients with Foix-Alajouanine syndrome may present with progressive unilateral or bilateral weakness, sensory disturbances, and bladder, bowel, and sexual dysfunction, which can progress over several years before a diagnosis is established.[5] In this current case, the patient has reported experiencing bilateral lower limb weakness and pain that started nine months ago and has since shown a gradual deterioration, notably over the past three months. Radiological evaluation, including MRI and digital subtraction angiography (DSA), was crucial for distinguishing Foix-Alajouanine syndrome from other causes of progressive myelopathy and facilitated the formulation of an appropriate management strategy to impede disease advancement. MRI findings may initially appear normal but progress to show inflammation and hypointensity on T1 sequences and hyperintensity on T2 sequences, accompanied by dilated perimedullary vessels.[3,6] Phase-contrast MRI often reveals enlarged, tortuous vessels within the subarachnoid space. However, diagnosing spinal arteriovenous lesions can be challenging, with studies suggesting significant delays in accurate diagnosis.[7] The optimal treatment strategies for spinal dAVF encompass a spectrum of approaches, including endovascular therapy involving embolization, surgical ligation of the arteriovenous fistulas (AVFs), or a combination of both modalities in certain cases.[2] While surgery traditionally offers a potential cure for the lesion, endovascular treatment has gained momentum recently due to its high success rates (up to 89.5%), lower morbidity, and shorter hospital stays.[2,8] Following consultation with our neuro-interventional radiologist, microsurgical occlusion was determined to be the most suitable treatment option, given the narrow and tortuous nature of the arterial feeder, making endovascular treatment more challenging. Surgical intervention typically results in a higher rate of fistula obliteration compared to endovas-

cular techniques.[2] It is imperative to highlight that successful microsurgical occlusion of dAVF necessitates the occlusion of the arterialized vein rather than the arterial feeder, as failure to do so may lead to the recruitment of new feeding arteries and subsequent recurrence.[2] During the intraoperative phase, distinguishing between the arterialized and draining veins can pose challenges. Furthermore, there is a risk of catastrophic hemorrhage if the dilated draining veins are inadvertently occluded instead of the arterialized vein. Thus, intraoperative ICG video angiography serves as a valuable adjunct in identifying the arterialized vein, aiding in precise treatment delivery. Surgical treatment carries potential complications, including wound infection, cerebrospinal fluid (CSF) leaks, pseudomeningocele, hematoma, neurovascular injury, worsening myelopathy, and pressure sores.[8] Our patient experienced a post-operative pseudomeningocele, managed conservatively. The primary goal of treatment for spinal dAVFs is to prevent further neurological deterioration and promote functional recovery. A meta-analysis conducted by Steinmetz et al. demonstrated that surgical intervention resulted in improvement or stabilization of symptoms in up to 89% of patients with spinal dAVF, with 55% experiencing improvement, 11% experiencing worsening, and 34% remaining stable. However, only approximately one-third of patients reported improvement in urinary function following treatment.[9] Our patient did not exhibit neurological improvement at the 1st and 3rd-month follow-up assessments, likely due to the delayed presentation of the condition. A longer follow-up may reveal improvements in our patients' symptoms.

Conclusion

Foix-Alajouanine syndrome, although rare, warrants consideration in cases of progressive myelopathy. Microsurgical treatment of spinal dAVF, supplemented by intraoperative ICG video angiography, represents an effective management approach. Early intervention is pivotal for favorable outcomes, necessitating heightened clinical vigilance for prompt diagnosis and treatment initiation. The time elapsed between the onset of the neurological deficit and treatment can impact the patient's prognosis. The likelihood of regaining functional ambulation in patients with dAVF is closely linked to the timing of treatment. Delayed endovascular or surgical intervention results in poor prognosis, even in extended rehabilitation, as witnessed in this case.

Ethical statement

The authors are accountable for all aspects of the work and ensure that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's parents to publish this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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