Case Report

Spinal Dural Arteriovenous Fistula in a Pediatric Patient with History of Endovascular Therapy Failure: A Case Report

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ABSTRACT

Spinal Dural Arteriovenous Fistula (SDAVF) cases in children are extremely rare and pose a high risk for intraoperative hemorrhage. The clinical manifestation and imaging results may be vague and deceptive, frequently mistaken for other conditions such as demyelinating or spinal degenerative illnesses. SDAVF’s cause is not well understood. Here, we present the case of a 10-year-old male patient with SDAVF who did not improve after endovascular therapy. The patient complained of weakness in the lower extremities, skin thickness, tingling sensations, and painful bowel movements and urination. The patient underwent endovascular embolization due to spinal AVF from the 9th thoracic vertebrae until the sacral vertebrae one month earlier. But no significant clinical improvement was found. The vital signs of the patient were within normal limits. An MRI showed a flow-void lesion with tortuosity in the dorsal spinal area at the 9th and 10th thoracic vertebrae. Because an embolization procedure was performed on the patient, which resulted in no significant improvement, it was planned for the patient to undergo an MRI and MRA evaluation. An MRI and MRA later showed the formation of an extramedullary intradural cyst at levels T9 to T10 of the thoracic vertebrae. Decompression surgery (left hemilaminectomy) and tumor extirpation were thereafter carried out on the patient after the routine laboratory test was performed. After the procedure, the patient showed improvement and could carry out everyday activities independently at 10 months post-operatively. The failure of endovascular therapy can be attributed to several factors, such as the surgeon’s experience, tools and embolization technique, and follow-up treatment by surgery.

Keywords: AVF, AVM, treatment, endovascular therapy, cyst
INTRODUCTION

Arteriovenous Malformation (AVM) of the spinal cord occurs in approximately 120 cases in the US every year.\(^1\) AVM also accounts for 3–4% of all intradural mass lesions. This disease is rare in pediatric patients (1% of all spinal AVF) and usually only symptomatic during adulthood.\(^2\) Specifically, Foix and Alajouanine first defined Spinal dural arteriovenous fistula (SDAVF) in 1926, and its anatomical and clinical characteristics have been known ever since. However, it wasn't until 1974 that the fundamentals of therapy were first laid out.\(^3\) The SDAVF is a variety of spinal vascular abnormalities. The most popular categorization of congenital abnormalities, out of the many that have been presented to date, divides them into four groups depending on the anatomic location of the fistula and the arterial arteries involved.\(^4\)

Radiculomedullary and pial arteries supply blood to spinal vascular abnormalities that are inside or on the cord or at the proximal nerve root, while radiculodural and radiculomeningeal arteries supply blood to those that are on the dura. Understanding the pathogenesis, presentation, indications for treatment, and management of these lesions requires knowledge of their angiographic structure.\(^4\) Segmental arteries that originate at each level supply blood to the spine's bony components, dura, nerves, and soft tissues. These arteries come from the ascending cervical artery and vertebral artery at the cervical spine, the lumbar and intercostal arteries at the thoracic and lumbar spines, and the iliolumbar branches at the sacral region.\(^5\)

The pathological arteriovenous shunt in SDAVF is situated between the intradural radicular vein and the radiculodural artery(ies) within the dura mater. It typically sits at the dorsal wall of the dural root sleeve, where the radicular veins pass the dura at the posterior wall of the spinal nerve root sleeve, next to the radicular artery, within the intervertebral foramen.\(^6\) In a healthy person, 80–90% of the time, the blood that leaves the spinal cord drains to the venous plexus and medullary veins in a dorsal direction, while some people experience a combination of ventral and dorsal drainage. The radicular vein becomes arterialized when a shunt is present, and blood flow to the cord venous system and the perimedullary venous plexus is reversed.\(^7\)

The time between the development of symptoms and diagnosis might range from a few days (acute) to many years (unaggressive and slow-flow fistulas). A delayed or missed diagnosis may be caused by the vague nature of the symptoms, the mixed presentation, which occasionally includes indications of both upper and lower motor neuron deficiency (in 60% of cases), the unpredictable illness history, and deceptive imaging study findings. Given that patients tend to be older and that neurosurgeons, neurologists, orthopedic surgeons, and radiologists are unaware of SDAVF, this issue is made worse by the existence of prostatic, spinal degeneration, or vascular insufficiency comorbidities.\(^8\)

Each spinal vascular malformation is a unique lesion. The SDAVF may be safely and easily blocked surgically. Hemilaminectomy is required, along with exposing the dura, opening the dura to locate the radicular vein, tracking the vein to the fistula point and its dural connection, and then coagulating or cutting the vein at the attachment to remove it.
from the dura. An instantaneous change in color of the arterialized intradural veins indicates effective occlusion of the dura. The little feeders on the outside of the dural wall are often additionally coagulated by surgeons, but we don't think this is required because it might harm any adjoining radiculomedullary branches and result in cord infarction. Only the sacral area presents a challenge for surgical occlusion.\textsuperscript{2} Current surgical treatment options include endovascular occlusion, open surgical ligation or resection, spinal radiation, or both.\textsuperscript{10,11} Endovascular occlusion commonly has a success rate of more than 60\%.\textsuperscript{7} This case describes a child who underwent surgery for his spinal AVM after the failure of endovascular therapy.

**CASE REPORT**

A 10-year-old male patient had a chief complaint of weakness, numbness, and tingling in both lower extremities. The patient also complained of painful bowel movements and urination, with no history of trauma. The patient had undergone endovascular embolization due to spinal AVF from the 9\textsuperscript{th} thoracic vertebrae until the sacral vertebrae one month previously.

A physical examination found an increase in physiological reflexes as measured by the National Institute of Neurological Disorders and Stroke (NINDS) grading and evident Babinski reflex. Motoric examination revealed weakness from L2 to S1 (manual muscle testing grade of 1) and anesthesia from Th11 downward. A thoracolumbar MRI found a flow-void lesion with tortuosity in the dorsal spinal area from the 9\textsuperscript{th} thoracic vertebrae until sacral vertebrae (Figure 1).

\textbf{Figure 1}. (A) MRI of the thoracolumbal T2 sequences before embolization. Sagittal slice. (B) Axial view of the T2 sequences MRI of the thoracolumbal. (C) MRI of thoracolumbal T2 sequences before embolization of coronal slice. (D) Congestive perimedullary

The DSA was also found an AVF feeding to the segmental arteries of VTh 8 and VTh 11 (Figure 2). N-butyl cyanoacrylate glue was slowly and continuously injected into the patient's VTh 8 and VTh 10 segmental arteries as a portion of an embolization treatment. Angiogram results after the embolization revealed that the fistula had closed, and the draining vein had been attached by the glue.
After embolization, no significant clinical improvement was found. One month later, the patient experienced a decline in neurological function, such as defecation and urination disorders. Consequently, thoracolumbar MRI and MRA were performed to recheck for the presence of other pathologies. An intramedullary cyst was found as an extramedullary intradural tumor at the 9th thoracic vertebrae (Figure 3). The formation of cysts near the embolization site may occur due to the diversion of flow into nearby arteries, which consequently results in arterial occlusion and the formation of cysts or larger AVF. This new cyst was uncovered in the second MRI. As it was larger than the previous finding, this cyst hindered clinical improvement after the embolization.

To treat this newly found AVF, a decompression surgery, left hemilaminectomy, and tumor extirpation were planned (Figure 4). Before the operation was carried out, the patient was prepared by going through laboratory test to avoid other conditions that might affect the surgery. One week after the surgery, there was an improvement in motor and sensory functions. In the third month after surgery, the patient could stand up independently. Bowel and bladder functions also improved. Eight months after surgery, a thoracolumbar MRI for evaluation was performed, and there was no recurrence (Figure 5). Ten months after surgery, the patient was able to carry out daily activities independently.

Case reports that include data from fewer than three patients are not required to undergo approval from the institutional review board at our center. Only written consent from the patient is mandatory, and we have asked for the patient’s consent.
DISCUSSION

SDAVF’s initial symptoms are not specific, making it difficult to diagnose early, and it is often misdiagnosed. Common presenting symptoms are weakness of both limbs, paresthesia (50%), and back pain with mild intensity that radiates to the lower extremities. Progressive myelopathy symptoms are common but rarely acute (5%). The last symptom to appear is usually urinary incontinence or retention. These symptoms commonly appear and are exacerbated after activities that increase intramedullary venous pressure (e.g., exercise, Valsalva maneuver, singing, and certain postures) and are improved by rest.12,13 In this case, the patient had played football five days before the episode; therefore, exercise was thought to trigger increased abdominal venous pressure, which worsened venous congestion and its symptoms.

The SDAVF position in this patient is similar to the general population’s, which is between V Th 6 and VL 2 (80% of all cases). Other sites are less common: the sacrum (4%), high cervical lesions (2%), and low cervical lesions (between VC 2 and V Th 1, which are extremely rare).15 The prevalence in this area is due to the location of radicular veins, and the vascular system in these areas is more susceptible to hemodynamic changes.16

The aim of SDAVF treatment is to cause occlusion in the shunting zone. To the authors’ knowledge, there have been no published guidelines on the treatment of SDAVF. However, it is well established that there are two therapeutic options for SDAVF: surgery and endovascular therapy.15 The endovascular therapy success rate varies between 25% and 75%, while the surgical success rate is 98%. Should the glue not reach the draining vein (i.e., failed embolization), early surgical intervention is warranted, as a delay in the switch to surgical therapy has been found to have negative outcomes.15,17 In our case, the first endovascular embolization did not result in full clearance of the AVF and instead resulted in the formation of a cyst. This usually occurs when there is a diversion of embolization material to nearby anastomosing arteries, which have higher flow. The diverted material then causes embolization in another artery, which consequently results in the formation of a cyst. Several factors might have contributed to this event; in this case, it was probably due to the patient’s young age and the flow back through the anastomosis with the radicular artery above and below the lesion level.15

Between one week and three months after surgery, the patient routinely went to an orthopedic outpatient clinic for physiotherapy, ROM exercise, and muscle strengthening. In addition, the patient was educated to reduce any possible precipitating factors that might worsen the fistula, such as activities that increase abdominal venous pressure like frequent coughing, or delaying treatment should symptoms reappear.18 Normalizing motor functions was advised, and improvement was found not only in the motoric aspect but also in the sensory. The sensation from the umbilicus area to both legs improved, and the patient can now control bowel movements and urination (Table 1). Ultimately, ten months after surgery, the patient can carry out everyday activities independently. (Table 2).
Table 1. Results of pre-operative neurological examination, one week and three months post-operatively

<table>
<thead>
<tr>
<th>Level</th>
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<th>1 Week Post-operative</th>
<th>3 Months Post-operative</th>
</tr>
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<tr>
<td>L2</td>
<td>0/0</td>
<td>2/2</td>
<td>2/2</td>
</tr>
<tr>
<td>L3</td>
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<td>2/2</td>
<td>2/2</td>
</tr>
<tr>
<td>L4</td>
<td>0/0</td>
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</tr>
<tr>
<td>L5</td>
<td>0/0</td>
<td>2/2</td>
<td>4/4</td>
</tr>
<tr>
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<td>2/2</td>
<td>4/4</td>
</tr>
<tr>
<td>Sensory</td>
<td>Anesthesia 11th Thoracic Vertebrae</td>
<td>Hypoesthesia 11th Thoracic Vertebrae</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Table 2. Results of neurological examination six months, eight months, and ten months post-operatively

<table>
<thead>
<tr>
<th>Level</th>
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<th>Post-operative 8 Months</th>
<th>Post-operative 10 Months</th>
</tr>
</thead>
<tbody>
<tr>
<td>L2</td>
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<td>4/4</td>
</tr>
<tr>
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<td>3/3</td>
<td>4/4</td>
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<tr>
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<tr>
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<tr>
<td>Sensory</td>
<td>Normal</td>
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</table>
Spinal arteriovenous fistula (AVF) and arteriovenous malformation (AVM) are rare and complex, and they tend to be underdiagnosed because of their elusive pathology. This is because of a lack of awareness among clinicians, leading to severe disability and even death. Due to the nature of the disease, an individual treatment algorithm must be designed for each patient. The failure of endovascular therapy could be attributed to several factors, such as the surgeon’s experience, tools, and embolization technique, requiring follow-up treatment with surgery.\textsuperscript{19,20} Further research is needed to help identify and solve these obstacles and optimize the outcomes of this disease.

There are no reported cases of the formation of intradural cysts after embolization of AVF. Other literature discusses the formation of cysts long after a laminectomy.\textsuperscript{21} Other commonly known causes of cysts include neoplasms, trauma, lumbar puncture, arachnoiditis, or as a post-surgery complication.\textsuperscript{22}

Open surgery and endovascular embolization have not been directly prospectively randomized against one another. In SDAVF, open surgery had a 98% success rate, while endovascular embolization had a 46% success rate, according to a meta-analysis by Steinmetz et al. However, reported success rates for endovascular surgery have been increasing, reaching 70–80%, as a result of technical advancements in angio suites and microcatheters and increased practice with the procedure.\textsuperscript{7} If the embolization is unsuccessful, the glue or onyx that was injected may be used as a marker for the fistula to be located fluoroscopically before the skin is removed during surgery. Patients are instead directed straight to open surgery in certain hospitals due to the increased cure rate of surgery compared to embolization.\textsuperscript{8}

Currently, liquid embolization agents (LEAs) are the main tool for endovascular embolization of brain AVMs and AVFs. Polyvinyl alcohol (PVA) particles had been used sporadically but have been replaced almost completely by LEA recently due to high recurrence rates.\textsuperscript{23} The coil may be used as an adjunct to LEA embolization for certain techniques, such as pressure cooker technique.\textsuperscript{24–27} There are two major groups of LEAs available for embolization of cerebrovascular malformations: Cyanoacrylates (often colloquially called glue) and copolymers, or non-sticky embolic agents (described as having lava- or rubber-like properties).

The difference between sticky and non-sticky emboli refers to the possible complication of having the microcatheter tip stuck in concentrated acrylic adhesive. Making it hard to maneuver after the tip has been trapped. This perhaps unreported complication has prompted the development of the non-sticky LEA, ethylene vinyl alcohol (abbreviated as EVAL or EVOH), which is built to prevent such occurrences.\textsuperscript{28} When combined with dimethyl sulfoxide (DMSO), EVOH became significantly less sticky; however, to allow injection of large amounts of LEA into the vascular malformation, Jacques Moret influenced the development of a "plug and push" technique that required trapping the tip of a microcatheter.\textsuperscript{29,30} Similar to cyanoacrylate embolization, this results in an inability to remove the microcatheter when the reflux time exceeds a certain distance or the entrapping time exceeds several minutes. The high rate of microcatheters getting stuck or vessels bursting after forced removal using so-called non-adhesives has led to the development of microcatheters with removable tips, such as the Sonic Microcatheter (Balt, Montmorency, France).\textsuperscript{31} Therefore, a classification based on the chemical structure of LEAs seems more appropriate than a classification based on adhesion.

Initially, the application of cyanoacrylate was for wound closure. Only bucrylate was available for endovascular embolization. Since IBCA was no longer available (during the 1980s), it was replaced
by n-butyl cyanoacrylate (nBCA). Its toacryl has been widely used to embolize a variety of vascular disorders, including cerebrovascular malformations, such as AVM and SDAVF, for more than 40 years.\textsuperscript{32-35} Histoacryl is the only acrylic with the addition of a dye that allows the syringe to contain the embolizer to be clearly identified. Although widely used to this day, Histoacryl has not yet been approved for vascular use due to its lack of formal approval. nBCA is what is used in this case.

LEA is especially important as an embolic agent in endovascular diseases. The two most common disease for which LEA is used are Cerebral AVM and cranial SDAVF. These active ingredients have their own unique properties and relative benefits over others. The treating interventionist must be familiar with the specific properties of these agents used in the procedure to achieve optimum endovascular embolization for cerebrovascular malformations. Long used as the primary LEA used for AVM and SDAVF embolization, the use of cyanoacrylates has declined over the past decades but is still effective in certain settings.\textsuperscript{24,25} In addition to their embolic properties, cyanoacrylates are much cheaper compared to copolymers.

Significantly cheaper cyanoacrylate (compared to He Onyx, Squid, or PHIL) makes cyanoacrylate more popular than other LEAs. Various subtypes of cyanoacrylates are available from different manufacturers, but the limited regional availability in most countries limits the choice. Certain subtypes may be advantageous in certain situations. For example, glubran 2 has low vascular toxicity, and magic glue has low adhesion to microcatheters. However, these idiosyncrasies have not yet been fully studied scientifically.

All of the above LEAs can cause vascular toxicity and inflammatory effects on embolized vessel.\textsuperscript{36,37} For cyanoacrylates, the level of toxicity is relatively high and is attributed to the cyanoacrylate itself, whereas for copolymer LEAs, the toxic effects are often mixed and mild. Caused by DMSO. For cyanoacrylates, the reaction is driven by an exothermic type of polymerization reaction and the release of highly toxic components (acrylic acetate and formaldehyde).\textsuperscript{37} Administration of anti-inflammatory drugs such as corticosteroids before the operation can reduce the development of embolism-related inflammatory reactions.\textsuperscript{34}

A recognized clinical and imaging-based differential diagnosis is available for the uncommon, underdiagnosed condition known as SDAVF. Clinically, the search for a fistula is sparked, especially in middle-aged patients, by signs or symptoms of ascending motor or sensory illness coupled with lower, upper, and motor neuron indications. The primary diagnostic technique is MRI, with MRA or CTA used to pinpoint the lesion's site.\textsuperscript{3} While the fistula may be effectively treated in a significant portion of patients with the tools we have today, the instruments available do not allow for treatment of the drainage impairment itself. We think embolization should be used whenever it can be done safely. Endovascular surgery has a success rate of 70–80\% when carried out by skilled teams using cutting-edge methods and tools. Although surgery is more intrusive than this, the rate is lower than what has been stated.\textsuperscript{7}

CONCLUSION

Clinicians should be made aware of the potential for SDAVF since prompt treatment can enhance the prognosis and stop symptoms from getting worse to the point of being irreversible.

DISCLOSURES

Acknowledgment

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient gave his consent for his images and other clinical information to be reported in the journal. The patients
understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Conflict of interest**

The authors declare no conflict of interest.

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None

**Author Contribution**

**Primadenny**: Conceptualization, Methodology, Validation, Formal Analysis, Visualization, Supervision; **Rizal**: Project Administration, Investigation, Resources, Writing – Original Draft, Writing – Review & Editing; **Aries**: Methodology, Validation, Formal Analysis, Supervision.

All authors have contributed to all processes in this research, including preparation, data gathering and analysis, drafting and approval for publication of this manuscript.

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