
UNRAVELING THE COMPLEXITY: A CASE REPORT ON SPINAL DURAL ARTERIOVENOUS FISTULA

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Abstract: Spinal Dural Arteriovenous Fistulas (SDAVFs) represent a distinctive category of spinal vascular malformations, characterized by abnormal connections between arteries and veins within the spinal cords dura mater. This rare condition, often overlooked, can lead to severe morbidity if left untreated. In this case report we are going to discuss a 52-year-old male who presented with a month-long onset of lower extremity weakness, gait disturbance, and urinary/stool incontinence, emphasizing the complexities in SDAVF diagnosis and underscoring the significance of early intervention.

On examination, he exhibited an inability for active movements in his right arm and both legs, global muscle hypotonia, dysphonic speech, and inability to walk. Sensibility testing revealed transverse sensory loss at Th6-Th7 levels bilaterally, and reflexes were hypoactive with a positive Babinski response. No meningeal signs were noted, and sphincter control was lost. The rest of the physical examination was within normal ranges. Neuroimaging was performed.

MRI showcases spinal cord edema and dilated and tortuous perimedullary veins on the cervical and proximal thoracic segment, notably concentrated on the right side. Simultaneously, angiography delineates the presence of a spinal dural arteriovenous fistula (AVF), originating predominantly from the meningeal branches of the vertebral arteries, with a pronounced emphasis on the right side.

The effects of SDAVFs manifest as spinal cord congestion and edema, with impact on spinal cord function and development of avascular necrosis. This progression leads to irreversible dysfunction. Timely intervention in SDAVFs through prompt endovascular embolization or surgical procedures hold promise for alleviating neural damage and positive recovery process.

Keywords: Spinal Dural Arteriovenous Fistulas, Vascular malformation, Endovascular embolization.

1. INTRODUCTION

Spinal Dural Arteriovenous Fistulas (SDAVFs) are a unique type of vascular malformation found within the spinal cord. These irregularities occur as unusual connections between arteries and veins, specifically within the dura mater—the outer protective layer of the spinal cord. In the complex world of blood vessels, SDAVFs bring a notable change—a redirection of blood flow from arteries to veins, all happening within the safeguarding layers of the spinal cord's dura mater. Spinal dural arteriovenous fistulas are the most common type of spinal vascular malformations, mostly found in the thoracolumbar region and often diagnosed among middle-aged and elderly individuals. Despite being the most prevalent spinal vascular anomaly, their appearance is rare and might frequently go undetected.

In this text we will describe a case of a 52-year-old male who presented with a gradual onset of lower extremity weakness, gait disturbance, and urinary and stool retention over a month. The symptoms initially manifested as paresthesia in the lower extremities, followed by short periods of transient mild weakness in the legs in the period of two weeks before the examination. On the day of the examination, the weakness progressed to the point where he was unable to walk. He has no significant medical history and denies any allergies.

During the physical examination, there was an inability for active movements in his right arm and both legs. Some movements were observed in his left arm, albeit with a smaller range, speed, and precision. There was evident global hypotonia of the muscles. In the Mingazini examination position, his right arm fell abruptly, while his left arm fell quickly (within less than 10 seconds). Speech was dysphonic, and walking was impossible. Sensibility testing revealed transverse sensory loss at the Th6-Th7 levels bilaterally. The Sciatic Stretch Test was negative. Reflexes of his upper and lower extremities were hypoactive and symmetrical. A positive Babinski response was present bilaterally. No meningeal signs were noted, and he did not have control over his sphincters. The rest of his physical examination was within normal physiological ranges.

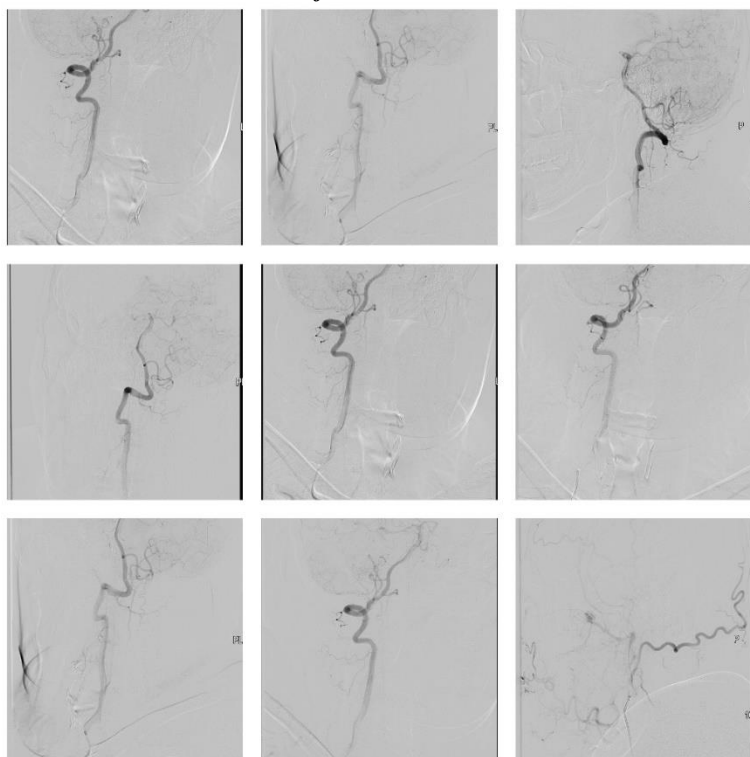
2. MATERIALS AND METHODES

Neuroimaging, including MRI and MRA, was conducted to assess the spinal cord. MRI and MRA are highly effective and sensitive methods for diagnosing spinal spinal pathological changes due to their ability to provide detailed visualization of the spinal cord and surrounding vasculature without invasive procedures. MRI can detect characteristic features such as spinal cord edema and dilated perimedullary veins, aiding in the identification of SDAVFs or spinal pathological changes. MRA complements MRI by offering dynamic imaging of blood flow, enabling the precise localization and assessment of arterial feeders and venous drainage patterns associated with SDAVFs or spinal pathological changes.

3. RESULTS

The MRI revealed spinal cord edema and dilated, tortuous perimedullary veins primarily on the right side in the cervical and proximal thoracic segments. Additionally, angiography confirmed the presence of a spinal dural arteriovenous fistula (AVF), predominantly fed by meningeal branches of the vertebral arteries, with notable prominence on the right side.

Figure 1 Angiogram images showing spinal dural arteriovenous fistula (AVF), predominantly fed by meningeal branches of the vertebral arteries.



Source: (Joveva et al., 2024)

4. DISCUSSION

Spinal dural arteriovenous fistulas (SDAVFs) constitute about 70% of all spinal vascular malformations, with an estimated incidence of 5–10 cases per million people annually (Thron, 2001). Typically underdiagnosed, SDAVFs

mostly affect elderly men, with a mean age of 55–60 at diagnosis. They are rare in individuals under 30. Predominantly found in the thoracolumbar region (T6 to L2), sacral lesions occur in around 4%, high cervical lesions in 2%, and low cervical lesions are extremely rare.

Spinal dural arteriovenous fistulas (SDAVFs) are usually acquired without a known cause. These abnormal connections between arteries and veins are located within the dura mater near spinal nerve roots. When arterial blood from the radiculomeningeal artery enters a radicular vein beneath the vertebral pedicle, it raises pressure in the spinal veins (Jellema, 2006). This increased pressure reduces the flow of blood in normal spinal veins, causing congestion and swelling inside the spinal cord. This chronic lack of oxygen can lead to progressive damage to the spinal cord (Tadić, 1985). Because there are fewer ways for blood to exit the lower part of the spine compared to the upper part, swelling is likely to move upward, explaining why symptoms of spinal cord dysfunction can occur even if the problem is further down the spine. This highlights the complexity of SDAVFs and their potential for significant neurological impairment over time.

Classification of Spinal Vascular Malformations in General and Dural AV Shunts in Particular: Spinal vascular malformations are classified into three groups: genetic hereditary lesions, genetic nonhereditary lesions, and single lesions. A common classification is based on spinal cord vascular anatomy, distinguishing between pial and dural arteriovenous (AV) shunts (Alkhaibary, 2024). Spinal cord AV malformations are fed by intrinsic arteries, while spinal dural AV fistulas (SDAVFs) are fed by radiculomeningeal arteries. Dural AV shunts can be categorized into ventral, dorsal, and lateral epidural groups based on embryologic venous drainage development. The ventral group drains structures from the notochord, leading to symptomatic compression, while the dorsal group relates to poorly developed veins causing spontaneous epidural hematomas. Lateral epidural DAVFs, the most common type, develop in the junction of bridging veins. (Krings, 2009), (Sim, 2022).

Early signs of venous congestion are not specific and may manifest as difficulty climbing stairs, gait disturbances, and sensory issues like paresthesias or patchy sensory loss. Radicular pain can occur in one or both lower limbs initially. Lower back pain without radicular distribution is also common. These neurological symptoms tend to worsen over time, often progressing upwards. Late-stage symptoms may include bowel and bladder incontinence, erectile dysfunction, and urinary retention, upper motor neuron involvement with clonus and positive Babinski signs, coexisting with lower motor neuron involvement. While the typical progression is slow, an acute onset and intermittent improvements are also possible (Aktürk, 2020), (Kang , 2011).

The varied neurological symptoms in SDAVF can be mistaken for polyneuropathy, tumor, or degenerative disk diseases (Sucuoğlu, 2020). Patients often consult orthopedic surgeons, urologists (thinking urinary retention is related to prostate hypertrophy), or psychologists (for erectile dysfunction) before seeing a neurologist. On MRI, cord edema and dilated perimedullary vessels without an intramedullary nidus suggest SDAVF. The main alternative is another spinal vascular malformation. If an SDAVF drains solely into anterior spinal veins, cord hypersignal on T2 alone may occur, resembling glioma, inflammatory lesion, or spinal ischemia in the differential diagnosis (Jeng, 2015).

Treatment options for spinal dural arteriovenous fistulas (SDAVFs) include surgical closure of the intradural vein, typically safe but with risks for sacral fistulas, or endovascular therapy using liquid embolic agents after catheterization of the feeding radiculomeningeal artery (Oh Y, 2021) (Krings, 2005). The goal is to close the shunting zone, though proximal arterial occlusion may lead to temporary symptom relief, recurrence is common due to collateralization. Treatment aims to stop disease progression, with prognosis influenced by symptom duration and pretreatment disability. Complete fistula closure can stop disease progression, yet only two-thirds of patients experience motor symptom regression and one-third sensory improvement (Takai, 2020). Impotence and sphincter disturbances are often irreversible, with persistent pain in some cases, and rare instances may worsen despite treatment. (Jellema, 2005).

5. CONCLUSION

Spinal Dural Arteriovenous Fistulas (SDAVFs) present as rare vascular abnormalities within the spinal cord, typically affecting elderly males. They often manifest with insidious neurological symptoms, including weakness, sensory disturbances, and urinary retention, potentially leading to significant disability if left untreated. Diagnosis can be challenging, with symptoms mimicking other conditions. Treatment options, including surgical closure or endovascular therapy, aim to halt disease progression, although outcomes vary, and some deficits may persist. Vigilance in recognizing symptoms and prompt intervention are crucial in managing SDAVFs to prevent further neurological deterioration and improve long-term outcomes.

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