Imagining findings can be misleading; clinical correlation is must: A Rare case of Spinal Cord A-V Malformation Masquerading Radiculopathy

Karan M Patel, Gautam Das, Amit A Sequeira, Praneet Singh

ABSTRACT
Spinal dural arteriovenous fistulas (SDAVF), is a clinical condition that can pose a challenge to the clinicians both in the diagnosis as well as treatment. This rare disorder can present with certain nonspecific symptoms and signs, including gait abnormality or lower-limb sensory or motor weakness. Symptoms may progress gradually or in some cases decline over a period. It can be treated surgically or through an endovascular approach involving disconnection of the fistula to make a prompt diagnosis, detailed history and proper neurological examination along with correlated imagine findings is the right way. We encountered a patient having long-standing bilateral lower limb weakness and sensory disturbances for which he was operated twice at different levels, with partial improvement. By a combination of detailed neurological examination and appropriate diagnostic imaging, we concluded that an arteriovenous (AV) malformation at the mid-thoracic spinal cord level was the causative factor for the same which is proposed below.

Keywords: Clinical examination, Fistula, Spinal cord AV malformation.

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INTRODUCTION
Berenbruch first reported spinal vascular malformation in 1890 in autopsy findings and two decades later, Krause recognized spinal vascular malformation during a surgical operation. SDAVF which are amongst the most commonly encountered vascular malformation of the spinal cord is found mostly amongst elderly males in the thoracolumbar region. These can be considered a treatable cause for progressive paraplegia or tetraplegia. These malformations are found within the dura mater in a location close to the spinal nerve root where radiculomeningeal artery meets a radicular vein.

Tadié et al. in their review found that radicular veins in the lower thoracic and lumbar region are fewer and smaller in caliber, making the lower thoracic and lumbar spinal cord more vulnerable to hemodynamic changes. These findings have also been concluded in a study by Vasdev et al.

Tadié et al. demonstrated a narrowing of the intervertebral veins at the point where it crosses the dura mater wherein it loses its vascular wall. The vascular wall gets replaced by an arachnoid cuff by the dura mater, forming a zigzag fashion on exiting the dura. It was hypothesized that the zigzag fashion and the narrowing of the intervertebral vein prevent blood flow going back to the intradural space, only permitting blood flow in a physiological direction. According to this observation, the “protective anti-flow back system” may be damaged in cases of SDAVF by congenital or acquired factors.

CASE REPORT
A 52-year-old male patient non-diabetic, non-hypertensive and having no other medical illness came to our clinic one month back. He was mainly complaining about burning sensations and weakness of bilateral lower limbs with NRS 8/10 since last 2 to 3 years. He was wheelchair bound, not able to walk or stand at all.

Before 15 years, he had low back pain which was non-radiating, non-dermatomal, continuous in nature, moderate on intensity, increased on physical activity and walking which was conservatively managed and over a period of 6 to 7 months.

Then again 7 years back, he started low back pain radiating up to ankle on the left side, continuous, electrical shock-like sensations, moderate in severity, increased after physical activity and walking and decreased by lying down and sitting, with NRS 7–8/10. After 1 month
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complains of pain also started on the right side with all the same above characteristics.

MRI was suggestive of a desiccated disc with central and bilateral paracentral annular disc bulge intending ventral thecal sac and compression of traversing nerve roots at L3-L4 and L4-L5 level, bulge disc, facet and flavum hypertrophy obliterating the lateral recess and narrowing neural foramina inferiorly on both sides with central canal stenosis. He was advised to undergo surgery, but he denied. Taking medications and aggressive physiotherapy, but over a period there was no improvement and gradually deteriorating.

In May 2015, he underwent posterior decompression by laminectomy at L3-4, the L4-5 level for same complains. After surgery, there was a partial improvement. He took medications, went for physiotherapy and after one month he was totally bedridden. Then underwent for serial MRI to find out any technical matter or inappropriate thing that was hurting him. But all investigations showed no any abnormality.

In October 2015, MRI showed central and paracentral disc protrusion causing thecal sac indentation, moderate spinal canal stenosis, bilateral neural foraminal narrowing at C4-5, C5-6 level. He underwent for surgical intervention for same. ACD by median corpectomy of C5 and discectomy at C4-5, C5-6, and fusion by titanium mesh cage done in the same month. But after surgery, there was no any improvement and deteriorating gradually. He had taken so many consultations of all other specialties, but not improving and became wheel-chair bound.

After completing history part, we did generally as well neurological examination (Table 1).

<table>
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<tr>
<th>Neurological Examination</th>
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<td>• Higher function: Normal</td>
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<td>• Cranial nerves: Normal</td>
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<tr>
<td>• Sensory: Fine touch, crude touch, temperature, vibration sensations are absent below T10 dermatome on both sides.</td>
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<tr>
<td>Two-point discrimination, pin-prick, pain sensations are diminished below T10 dermatome on both sides</td>
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We observed one clue that absence of knee-jerk and hyperreflexia of ankle jerk as well clonus is present, causing lower motor neuron (LMN) and upper motor neuron (UMN) type lesion respectively; correspond to some mid-thoracic spinal cord pathology. So we send him for MR angiography that shows dural A V malformation at the level of left-sided T9-10.

DISCUSSION

SDAVM though amongst the most commonly encountered spinal vascular malformation, are very frequently mis- or under-diagnosed. During the early course of the disease, the clinical symptoms can be nonspecific and can be frequently misdiagnosed as a case of spinal muscular atrophy/medullary tumor/sensory polyneuropathy or chronic inflammatory demyelinating polyneuropathy. A number of patients with SDAVF misdiagnosed as lumbar or cervical disc herniation have undergone surgical interventions resulting in failed back surgery syndromes. The condition though treatable in the majority of cases, however, if left undiagnosed, can result in paraplegia or paraparesis.

Oxygen-rich blood normally enters the spinal cord through arteries, which branch into smaller capillaries. Spinal cord uses the oxygen from the blood in those capillaries. The oxygen-depleted blood then passes into veins that drain blood away from the spinal cord. The oxygen-enriched blood entering the cord via arteries after consumption of the capillary oxygen passes into veins that drain the blood away from the spinal cord. In an SDAVF, the blood bypasses the capillaries entering directly into veins via arteries. This leads to arteri- lization of the intervertebral veins thereby increasing venous pressures culminating into wall thickening and tortuosity of radial veins (referred to as intramedullary veins). The resulting chronic venous hypertension and stagnation decreases tissue perfusion leading to edema of the spinal cord and progressive myelopathy.

SDAVF show a gradually progressive clinical course with nonspecific symptoms such as tingling, weakness, gait disturbance, paresthesia, and diffuse or patchy sensory loss. Nonradicular back pain may occur with cramping or burning sensation. The neurological
deficits are gradually progressive and worsening over time. Sometimes it may mimic an anterior spinal artery syndrome with acute deterioration after exercise or prolonged standing with partial relief of symptoms on rest. One of the dreaded complications of spinal AVM is rupture and bleeding in the spinal cord. Though intraspinal hemorrhage is rare, however, a unique case report described a patient with SDAVF at L4 suffering from spinal subarachnoid hemorrhage. Patients with a dural arteriovenous fistula at the craniovertebral junction may present with an acute headache due to intracranial subarachnoid hemorrhage.

Details of the normal intrathecal vasculature and the AVM’s relationship with the spinal cord cannot be ascertained in detail via contrast-enhanced computed tomography (CT) scan. In a case report by Guinto and colleagues, showed combined intrathecal metrizamide and arterial injection to successfully demonstrate the morphology of the malformation to assess operability better which however is an invasive and not a routinely performed.

The MRI being a noninvasive diagnostic used more frequently in diagnosing SCAVM. However, DiChiro et al. after their one year’s of experience with MRI found that it is not an accurate method for the diagnostic demonstration of dural AVM as intramedullary AVM. Poor delineation of feeders and drainers were found. Subsequent investigations by Doppman and associates suggested that T2 weighted enhancement may demonstrate the characteristic serpentine filling defects, but if the flow is slow, or if the lesion is small, then MRI may miss it. Dormont et al. investigated 36 SCAVM patients with MRI and found this technique particularly valuable for diagnosis of complications of SCAVM such as ischemic damage, hematoma, thrombosis, and hemorrhage. Minami et al. suggested in a similar way the reliability of MRI scan.

Symon et al. found that 51/55 patients with SCAVM had dilated and tortuous vessels on myelography. Gulliver and Noakes reviewed 940 myelograms and compared myelographic findings in patients with proven SCAVM with non-AVM patients who had filling defects and found that myelography may be normal in patients with AVM.

A case report of Marshman et al. showed only prominent epidural veins were noted in SCAVM patients. However, because of having precise accuracy and specificity, Selective angiography is considered the gold standard for the diagnosis of SCAVM. This is necessary not only to confirm the diagnosis but to distinguish the type of AVM and its blood supply. Such information is crucial to decide upon treatment.

Spinal cord edema and dilated and tortuous perimedullary veins on MRI are important to confirm the diagnosis. Early filling of radicular veins, delayed venous return, and an extensive network of dilated perimedullary venous plexus is the classic findings of SDAVM on angiography. One needs to have a thorough knowledge of SDAVM to avoid misdiagnosing the condition and prevent the occurrence of any irreversible complications.

Foix-Alajouanine syndrome a syndrome associated with spinal cord vascular malformations is described as an acute or subacute neurological deterioration attributed to spinal cord venous thrombosis related to an arteriovenous malformation, resulting in necrotic myelopathy.

Treatment options include surgical excision and therapeutic embolization. Surgical resection is generally considered the treatment of choice for dural AVMs, although embolization may be effective if there is no common origin of blood supply to the AVM and the cord.

The surgical prognosis is better in patients with dural than intradural AVMs since the intradural AVM has its nidus in the pia. Because of the surgical difficulties involved with resection of intradural AVMs, embolization is commonly used.

**CONCLUSION**

SCAVM though still considered as an overall a rare disorder affecting mostly middle-aged men, typically presenting with insidious symptoms of myelopathy and with either a gradual progressive or stepwise decline. Our patient was transferred to a higher specialist center with neurosurgical and interventional radiologist facilities to minimize the morbidity of unknown etiology. In this case, the diagnosis of dural AVM was initially overlooked by coincidental lumbar and cervical disc pathology on image findings but not matching with clinical presentation. So, careful history taking, detailed general and systemic examination and co-relation with appropriate investigations always make a better diagnosis. These are important for patients to have the best possible chances of favorable recovery.

**REFERENCES**


