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Case Report

Things are not what they seem neurologically and radiologically: An apt descriptor for spinal dural arteriovenous fistula (SDAVF) ☆

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ABSTRACT

Spinal dural arteriovenous fistulas (SDAVF) are the most common vascular malformations affecting the spinal cord. It is infrequently encountered in clinical practice and is believed to be acquired, predominantly affecting middle-aged and elderly men with unknown etiology. It is usually misdiagnosed despite presenting with conventional clinical findings and radiological features. Insidious onset of myelopathic findings is seen in addition to pathognomonic findings of cord edema and intrathecal flow voids on MRI. We present a case of SDAVF that was missed by the treating orthopedic surgeon and underwent spinal decompression with subsequent persistence of myelopathic symptoms. Angiography is required to confirm the diagnosis location of the fistula. Treatment is with embolization using liquid embolic agents or surgical through ligation of the draining vein. Endovascular techniques are minimally invasive, safe, and effective. Knowledge of the characteristics and advantages/disadvantages of each agent helps in planning and appropriate selection of agents for the patient. We report successful embolization with improved clinical outcomes for the patient using precipitating hydrophobic injectable liquid (PHIL) embolic agent. The outcome and prognosis of SDAVF depend on the duration of symptoms, severity of neurological symptoms, and successful occlusion of the fistulous draining vein. Awareness of this rare condition amongst clinicians and radiologists, would enable an earlier diagnosis and avoid morbid outcomes of this treatable condition.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Introduction

A Spinal Dural Arteriovenous Fistula (SDAVF) is a rare disease that accounts for 1%-2% of vascular neurological disease [1]. It shows a male preponderance with a male: female ratio of 5:1 [2]. This vascular fistula creates an abnormal arterialization of the venous plexus that subsequently results in venous hypertension, spinal cord edema, and eventually ischemia [3]. Commonly, patients present with chronic manifestations, however, interestingly few cases of acute presentations have been reported [1,4]. SDAVF is a treatable condition despite its rarity; however, the delay of diagnosis leads to unfavorable outcomes [4]. The differential diagnoses are broad and the clinical presentation bears semblance to degenerative spine disease symptoms [4]. In this case report, we aim to delineate a case of SDAVF discovered after spinal decompression surgery and the persistence of symptoms thereafter. Ethics committee approval has been obtained from the research committee for government hospitals Bahrain (approval serial number 62060623)

Case presentation

Patient 1 - A 61-year-old female, with comorbid diabetes, hypertension, and dyslipidemia presented to the emergency room with a history of lower back and bilateral lower limb pain and progressively increasing weakness impeding her ability to ambulate and walk. She had a history of progressive lower limb weakness and decreased sensation including the abdomen for 7 months. Weakness was more pronounced proximally than distally in the lower limbs. She had fecal and urinary incontinence as well. She denied any stigmata of connective tissue disease, any recent febrile illness, or vaccine intake. Her systemic review was negative.

In private care, her initial MRI showed spondylolisthesis at L5-S1 level with spinal canal stenosis (see Fig. 1). She underwent spinal decompression and fusion with pedicular screws and a cage. On retrospective review of her MRI done externally, we found cord edema as well that was missed by her treating orthopedic surgeon.

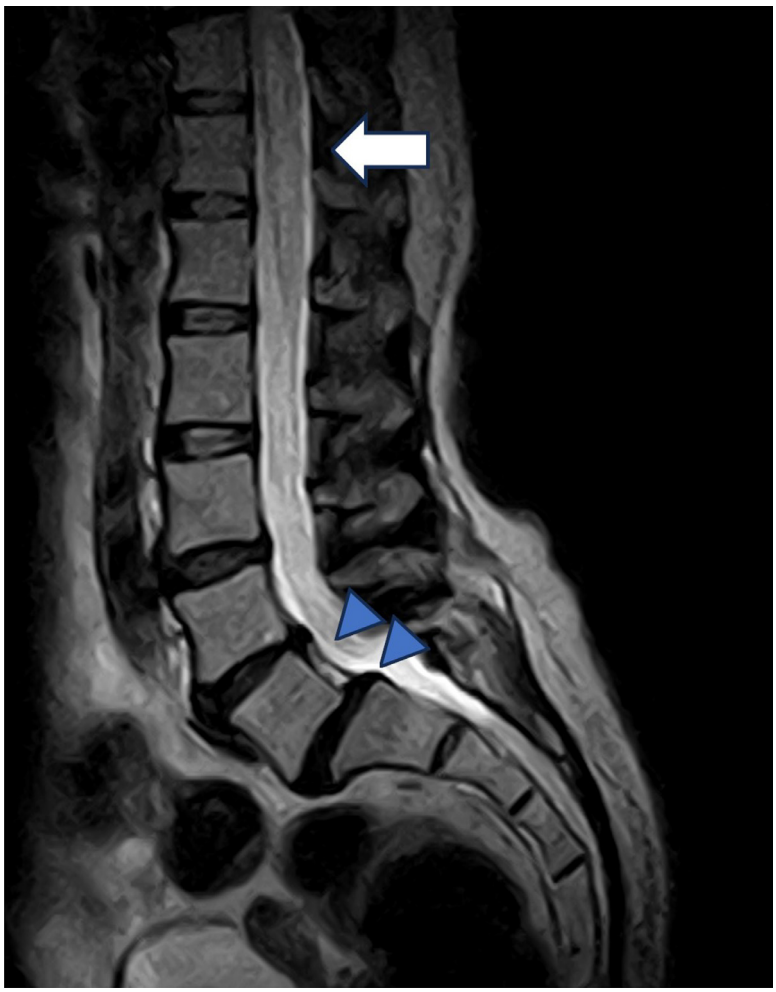

Fig. 1 – Preoperative sagittal T2 weighted imaging of the lumbosacral spine showing cord edema of the conus medullaris and lower thoracic spinal cord (*white arrow*). The study also shows grade II spondylolisthesis of L5 over S1 and central zone protrusion of the L4/5 intervertebral disc (*blue arrows*).

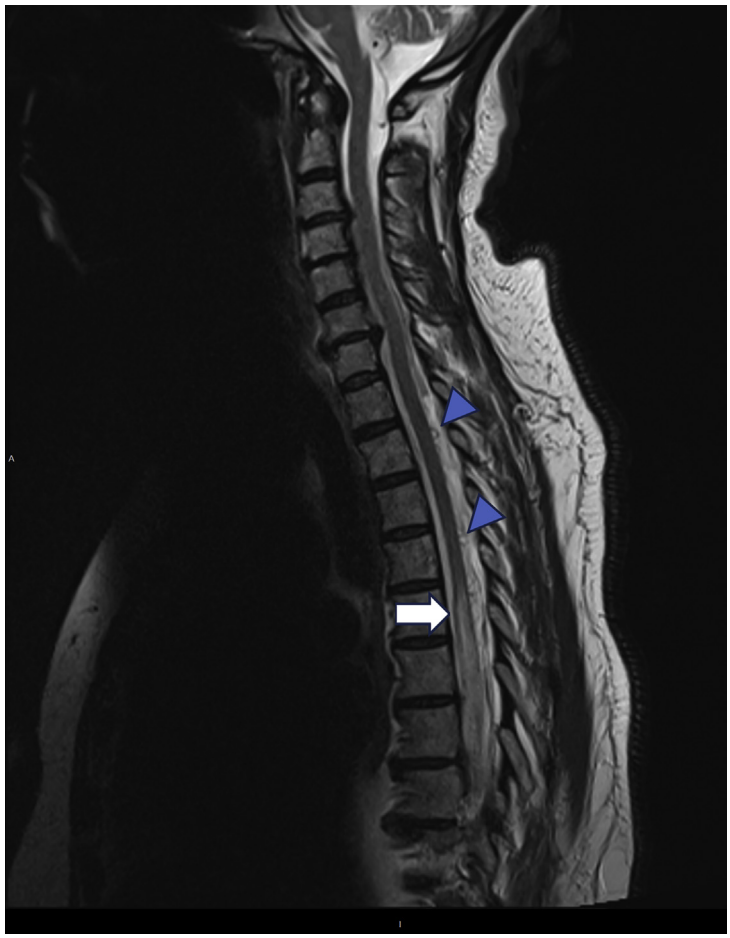

Fig. 2 – Sagittal T2 weighted MRI image showing central cord edema in the lower cord (white *arrow*) along with multiple vascular flow voids (blue *arrows*) pathognomonic of spinal dural arteriovenous fistula (SDAVF).

She decompensated 2 days after the spinal laminectomy operation for suspected lumbar spondylosis and presented 3 weeks later at our hospital.

On physical examination, power in the upper limbs was 5/5 (MRC Medical Research Council Scale) and reflexes were 2+. However, power in the proximal lower limbs was 3/5 and the distal lower limbs power was 4^{-}/5. Cranial nerves were intact. Cerebellar signs were absent in the upper limbs and were difficult to assess in the lower limbs due to weakness. Gait could not be assessed.

MRI spine was done that showed a postlaminectomy status with transpedicular screws and persistent postoperative fluid with edema indicating features of longitudinally extensive transverse myelitis with cord edema. Spinal cord swelling was seen starting from T5 to the conus with diffuse central height T2 signal density with no diffusion restriction. Anterolisthesis of L5 over S1 was observed. In addition, multilevel disc osteophyte complexes were seen at the cervical spine levels C3-4, C4-5, and C6-7 causing mild central canal stenosis and foraminal narrowing. The most likely differential diagnosis was transverse myelitis and she was started on steroids. Pregabalin was also started due to a reported electric shock-like sensation in the lower extremity, with improvement subsequently. An initial impression of transverse myelitis was made. Malignancy and arteriovenous malformation were kept as differential diagnoses based on the imaging findings.

A lumbar puncture was not performed as the patient declined. After receiving the methylprednisolone course, the patient's condition declined further and her power deteriorated gradually with a power of 2/5 in proximal muscles and 3/5 in distal lower limb musculature. The rest of her examination including the cranial nerve exam were unaffected and she was bedbound.

Two weeks later, MRI spine was repeated after clinical worsening, and this time it revealed a significant spinal cord swelling from T5 to the level of conus with no significant diffusion restriction and a pathognomonic finding of SDAVF causing venous hypertension (see Fig. 2).

Spinal angiography was done under general anesthesia. Through a right common femoral artery access, a spinal dural fistula with extensive venous extension arising from the L1 feeder (see Fig. 3). Artery of Adamkiewicz was identified from the left T9.

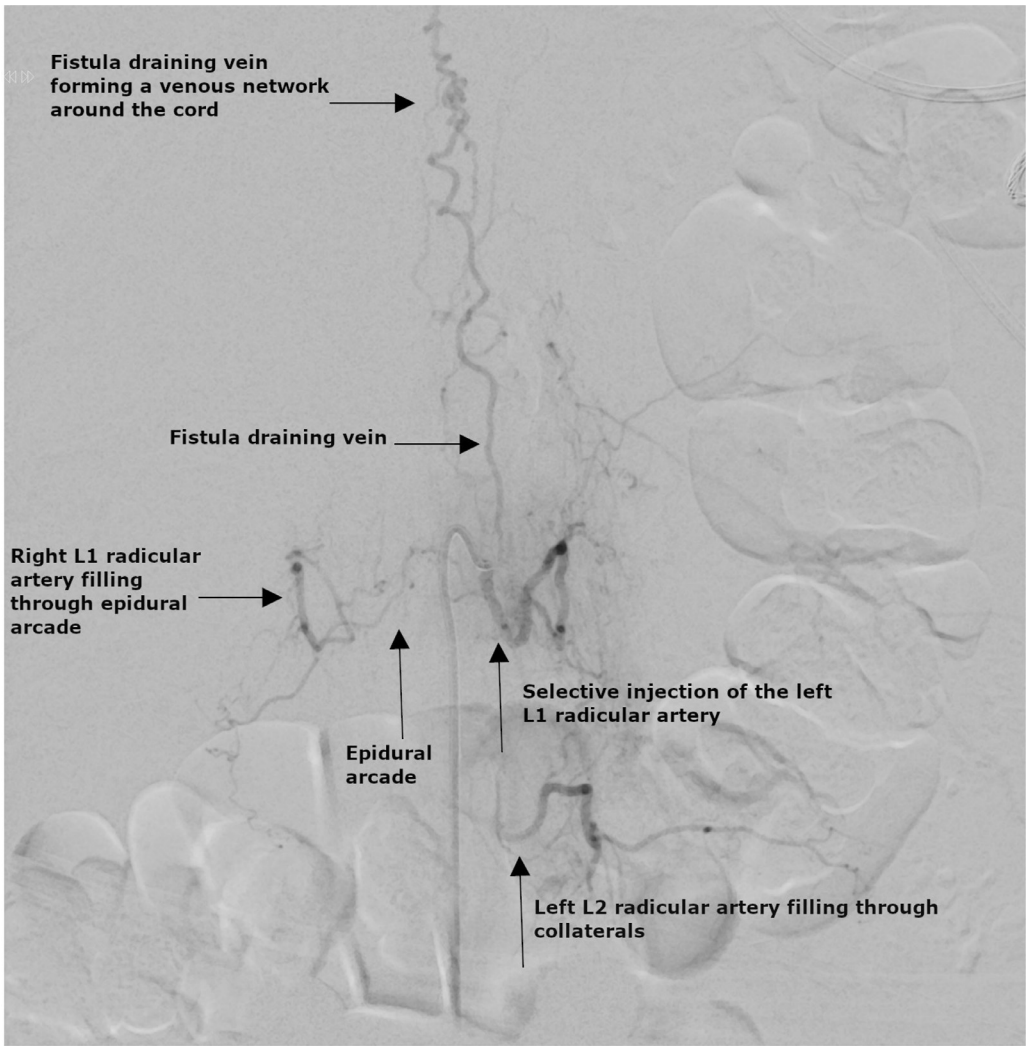

Fig. 3 – Selective angiogram of the left L1 radicular artery showing the fistulous vein arising at the L1 level and extending into the spinal canal and anastomosing with other perimedullary veins.

The patient was planned for embolization the subsequent week as the patient's history was complicated with renal impairment. Under general anesthesia, through left common femoral artery ultrasound guidance, L1, L2, and L3 level angiograms were done to exclude radiculomedullary feeders. The fistula was embolized using an Apollo detachable tip microcatheter with good venous penetration of Precipitating Hydrophobic Injectable Liquid (PHIL) (Microvention, Inc., CA) (see Fig. 4). Reflux into the right L1 pedicle through the epidural plexus and left L2 pedicle through paraspinal anastomosis was identified but the reflux had to be accommodated to achieve venous penetration of the fistula.

Postembolization, the patient stayed in the hospital and underwent physical rehabilitation. The plan was to transfer the patient to a rehabilitation center. She didn't fulfill the requirements for transfer due to hospital-acquired infections and was discharged home where she had ongoing physiotherapy care. One month after the procedure the patient's examination revealed a power of 4^{-}/5 proximally and 4/5 distally and she could now walk with a walker. She still reported urinary and fecal incontinence. 3D Digital Subtraction Angiography of the fistula draining vein and selective injection of the L1 radicular artery is shown in Fig. 5.

Discussion

Spinal dural AV fistula is a rare treatable condition that is encountered infrequently in clinical practice. It is frequently misdiagnosed with patients undergoing erroneous treatment prior to a confirmatory diagnosis. The rate of misdiagnosis is as high as 81% as per reported studies [5]. This is due to many factors notwithstanding non-specific clinical manifestations and imaging findings. The disease is predominantly encountered in middle to elderly aged patients 54-63 years of age [6]. Our patient fell within this range.

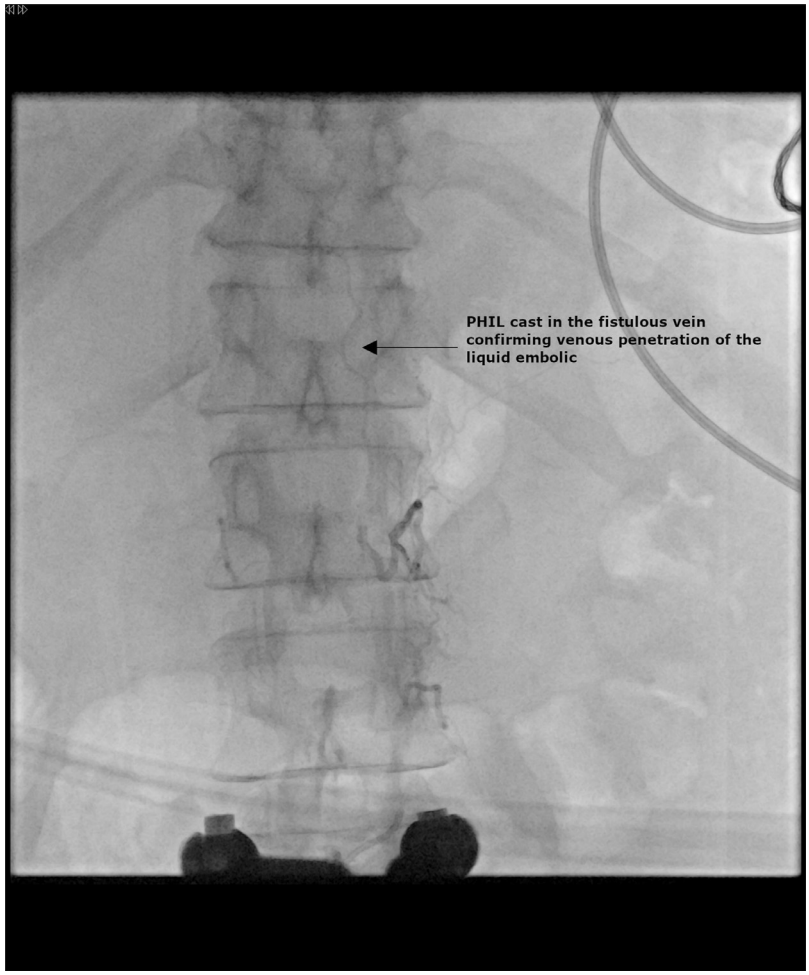

Fig. 4 – Un-subtracted angiographic image showing the embolic cast in the draining vein to confirm venous penetration of the liquid embolic agent PHIL (Precipitating Hydrophobic Injectable Liquid).

The similarity of clinical features and occurrence of intervertebral disc disease and benign prostatic hypertrophy in this age group adds to the initial diagnostic difficulty. Presenting symptoms of insidious myelopathic lower limb weakness and sensory loss, radicular pain, and sphincteric dysfunction in SDAVF confounds the diagnostic accuracy. Our patient had concomitant spondylolisthesis with urinary incontinence.

Classic MRI findings include cord edema represented by T2-weighted hyperintensity in the central cord with peripheral sparing, intrathecal flow void phenomena due to engorged tortuous veins and T1-weighted cord enhancement [7,8]. The patient showed cord swelling extending from T5 to the conus medullaris. An initial impression of myelitis was made due to venous dilatation and cord edema. Myelitis is a common misdiagnosis of SDAVF accounting for 22% of misdiagnoses [5]. Conus swelling with hyperintensities extending over 5-7 segments is seen in over 80% of cases [8]. The shunt in the patient was at the L1 level though conus swelling was present. This is attributed to lower venous drainage and proneness to congestion in the thoraco-lumbar spine compared to the cervical region [6]. It is worth noting that the presence of T2-weighted hyperintensity has a sensitivity of 100%, whilst the presence of T2-weighted hyperintensity and flow voids is 97% [9].

In a seminal paper, a characteristic finding of abrupt non-enhancing segment within an adjacent area of intense cord enhancement termed “missing piece sign” [10]. Identification of the sign could lead to an earlier diagnosis of fistulas through confirmation by angiography [11]. The presence of MRI findings does not yield clues to the location of the fistulas necessary for subsequent embolization. Most frequently, fistulas are located in the lower thoracic and upper lumbar regions [6]. Angiography is the gold standard for diagnosis and helps in determining the location of the fistula based on the early filling of the radicular vein. In our patient, the fistula was located in the L1 feeder. In a series of 49 patients, 94% of SDAVFs were located at or below the T5 root level, with only 6% located at level L1 [12]. Along the same vein, in another large study of 326 patients, 82.5% of SDAVFs were found between T5 to L5, with L1 feeder involvement in 8% of cases [5].

Therapeutic approaches include minimally invasive endovascular procedures and surgical techniques. With modern advances, a panoply of liquid embolic agents are available for embolization – magic glue, Onyx, Squid, and PHIL. Whilst each

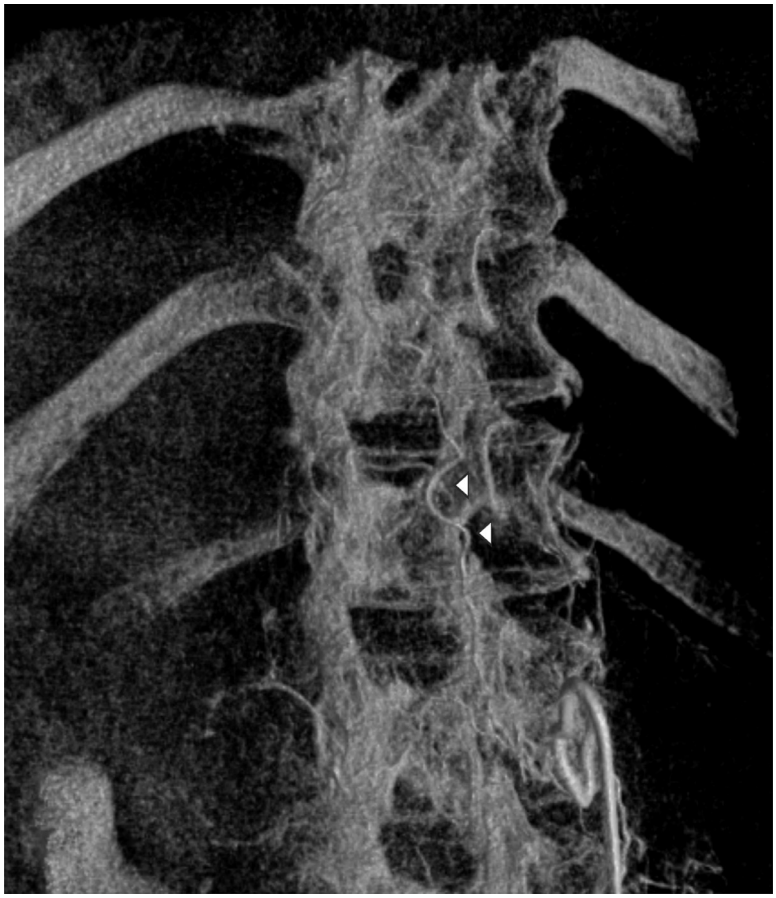

Fig. 5 – 3D DSA (digital subtraction angiography) showing fistula draining vein forming a venous network around the cord and selective injection of the left L1 radicular artery (*white arrows*).

embolic agent has its respective advantages/disadvantages, newer agents like Squid and PHIL offer additional advantages over Onyx and cyanoacrylates (glue) through stable visibility of the fistula network during longer injections and also a lower rate of artifacts on cross-sectional imaging [13]. In addition, the low viscosity of squid enables a deeper penetration into the target vessel [13]. PHIL requires no prior preparation unlike other agents and is ready to use [13]. We report a successful outcome of the embolization of SDAVF using newer liquid embolic agents.

Prognostic outcomes depend on the duration and severity of the illness and successful occlusion of the radicular vein from the dura at the shunt site [8,14].

In conclusion, typical clinical and imaging presentations of SDAVF can be missed by treating physicians. Awareness of the condition as a potential differential diagnosis would enable earlier treatment with improved outcomes.

Drug and instrument names - Precipitating Hydrophobic Injectable Liquid (PHIL) (Microvention, Inc., CA).

Patient consent

I confirm that a written confirmed consent for the publication has been obtained

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