TRIPLE THREAT: SPINAL DURAL ARTERIOVENOUS FISTULA IN SPINAL DYSRAPHISM AND TETHERED CORD SYNDROME, A CASE REPORT WITH LITERATURE REVIEW

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ABSTRACT

Introduction: The incidence of spinal dural arteriovenous fistula (SDAVF) in patients with spinal dysraphism is exceedingly rare with fewer than 30 cases reported. We report such a case and discuss the etiology, diagnostic, and treatment challenges of such patients.
Case Presentation: This is a 37-year-old gentleman with a history of myelomeningocele (MMC) repair at the age of 2 years who presented to us with worsening bilateral lower limb weakness and numbness. Clinical examination demonstrated affected motor and sensory functions from the spinal cord level of L2 and below. A Magnetic Resonance Imaging (MRI) of the Lumbosacral region revealed S3/S4 dysraphism with a tethered low-lying conus. A Computed Tomography (CT) angiogram of the spine followed by a spinal angiogram confirmed the diagnosis of a SDAVF with feeders from the median sacral artery and other branches of the left internal iliac artery. He underwent angioembolization, following which we noted a residual SDAVF and hence underwent a difficult second-stage angioembolization to the right lateral sacral artery branch. The patient remained status quo on follow-up.
Conclusion: SDAVFs occurring in spinal dysraphism requires a high index of suspicion to

ensure the diagnosis is not missed or masked by co-existing tethered cord and dysraphism. These lesions should be managed in a multidisciplinary multimodal individualized fashion.

MeSH Keywords: Dural arteriovenous fistula, tethered cord syndrome, spinal dysraphism, interventional radiology, angioembolization

Introduction:

The occurrence of a spinal dural arteriovenous fistula (SDAVF) in patients with spinal dysraphism and tethered cord syndrome (TCS) is extremely rare with fewer than 30 cases published. The authors discuss the likely etiology of the formation of SDAVFs in spinal dysraphism and TCS and its diagnostic and treatment challenges.

Case Presentation:

A 37-year-old gentleman with a history of a myelomeningocele (MMC) which was operated on at the age of 2 years presented to us with worsening bilateral lower limb weakness and numbness for the past 6 months. The patient also noted worsening urinary incontinence but did not report any change in bowel motions. Prior to this presentation, the patient has had a baseline of reduced pain and light touch sensation from the distribution of S1 caudally with some occasional stress urinary incontinence when performing heavy work such as lifting heavy objects, but otherwise ambulated with no assistance since childhood. For the past 6 months, the patient noticed worsening weakness of his hip and calf muscles, the most obvious of which, is his drastic decline in the ability to flex his hips which caused him to ambulate with a walking frame for the past two weeks. Furthermore, he noticed urinary incontinence even at rest. Clinical examination at the time of presentation revealed reduced pinprick and light touch sensation from spinal cord level of L2 and below with an almost-complete absence of sensation from S1 onwards. Motor examination revealed a motor power grading of 4/5 of the hip flexors (L2) and 2/5 of the knee extensors (L3) and 0/5 for the ankle dorsiflexors (L4), plantarflexors (S1) and the long toe extensors (L5). Babinski was bilaterally and the reflexes hyporeflexic. Anal tone was lax. A Magnetic Resonance Imaging (MRI) of the lumbosacral spine showed spina bifida at sacral vertebral level three and four (S3 and S4). The spinal cord was low-lying and tethered at the level of S2/S3. The spinal cord appears incorporated into the adjacent fatty tissue and lipoma-placode interface within the spinal canal at this level. In addition, the normal conus medullaris is not visualized due to this incorporation. Furthermore, there was a long-segment T2 hyperintense signal in the intramedullary thoracolumbar spinal cord from T10 to L3 which raised concerns of a possible arteriovenous shunting process such as an arteriovenous malformation (AVM) (Figure 1). There were no abnormal flow voids seen on MRI. After discussion with the radiologist, this patient underwent a CT angiography (CTA) of the whole spine. It was preferred over a Magnetic Resonance Angiography (MRA) of the spine as a CTA can image the entire spine with one contrast bolus. CTA of the whole spine confirmed the presence of a

spinal dural arteriovenous fistula (SDAVF) with the feeder likely to be a branch from the median sacral artery. There were multiple left sided S2/S3 level tortuous vessels which courses cranially as the mildly dilated posterior spinal vein, just posterior to the spinal cord which appeared to communicate with the median sacral artery at the vertebral level of L5 (Figure 2).

A spinal angiogram confirmed this finding of a SDAVF with the arterial contribution from the median sacral artery and branches of the left internal iliac artery. The patient was offered surgical vs endovascular treatment of the SDAVF. He opted for angioembolization and underwent an uncomplicated angioembolization to the distal median sacral artery which fed the SDAVF. Post embolization run demonstrated successful complete embolization of the SDAVF with coiling, ethyl alcohol and histoacryl glue. Unfortunately, post embolization day 2, the patient reported an increase in numbness and weakness of his bilateral lower limbs. As the patient had a successful first angioembolization procedure, concerns that the weakness and numbness could be due to cord edema rather than a residual SDAVF, hence, a CTA of the spine was performed on the same day which demonstrated a residual SDAVF likely contributed by the branch of the left internal iliac artery. The previously visualized communication from the median sacral artery was demonstrably obliterated postembolization on CT angiogram. A repeat thoracolumbosacral MRI spine showed similar T2 hyperintensity from T10 to L3 level with similar dilated tortuous vessels at the posterior aspect of the spinal cord as seen on the second CTA. A second-stage angioembolization performed 5 months later demonstrated SDAVF supplied by the lateral sacral arteries from both the left and right internal iliac artery. Angioembolization was successful to the right lateral sacral artery feeders. Post second-stage embolization, he reported slight worsening of the lower limb numbness however our clinical examination did not differ than that was found during his initial presentation. There was no worsening of urinary incontinence. He was planned for a repeat CT angiogram of the spine with a possibility of a third-stage angioembolization; however, the patient has declined for further intervention. During follow-up at one-year post second-stage angioembolization, his neurology has remained the same as the initial assessment.

Discussion:

Although the existence of SDAVFs have been observed in the 19th century, subacute myelopathy due to pathological vessels of the spinal cord or Foix-Alajouanine syndrome was first described in 1926 [1]. Various classifications of such vascular lesions of the spinal cords have been proposed with the most widely accepted classification proposed in 2002 by Spetzler et al [2]. The fortuity of an SDAVFs occurring in a patient with spinal dysraphism is extremely rare, with less than 24 cases reported in literatures. Table 1 lists these concurrences identified in published literatures. Although scarce in literature, it is highly likely that the concurrence of SDAVFs in spinal dysraphism are under-reported as these patients are often treated for tethered cord syndrome (TCS) and other pathologies [3,4,5].

Whilst spinal dysraphism are congenital in origin, the etiology of SDAVFs is unclear. Some argued that these vascular fistulas occurring in spinal dysraphism result from incomplete regression of mesenchymal tissue, hence congenital in nature [3,6,7]. However, the onset of presentation of SDAVFs in the adult age group suggests that these lesions are likely acquired rather than congenital [4,5,7,8]. The formation of SDAVFs in patients with spinal dysraphism has been postulated to *occur as a* result of the release of angiogenic factors from local lipomatous tissue [4,9,10,11]. In addition, it has been suggested that chronic venous hypertension due to the mass effect of cord lipomas and cord tethering result in angiogenesis, hence precipitating the formation of SDAVFs. Scrutiny into previously published cases as listed in Table 1 show that the formation of SDAVFs in spinal dysraphism and TCS are unlikely to be congenital given the age of diagnosis and unlikely to be due to prior surgery as only a third of these patients had prior surgery. Furthermore, the formation of these SDAVFs within the same vertebral levels as the spinal dysraphism and TCS suggests that locally-released angiogenetic factors are likely to be causative.

Table 1: Previously reported cases of SDAVFs associated with dysraphism
LMC: lipomyelomeningocele, MMC: myelomeningocele AVF: arteriovenous fistula, LSA: lateral sacral artery, IIA: internal iliac artery, RA: radicular artery, LA: lumbar artery

Authors, year	Age	Type of Dysraphism	Cord Tethering	Prior Surgery	AVF feeder/origin	Treatment of SDAVF
Srinivasan et al, 2016 [7]	21	L3-L5 LMC	Yes	No	L3 LA	Embolization
Iampreechakul et al, 2020 [10]	55	S2 spina bifida with angiomyolipoma	Yes	No	S3-S4 branches of IIA	Embolization followed by delayed surgery
Whitaker-Lea et al, 2018 [11]	57	L5-S2 LMC	Yes	No	LSA	Embolization
Przepiorka et al,2018 [13]	30	Filum terminale lipoma	Yes	No	Left S3	Surgery (Origin of feeder not mentioned)
	33	Filum terminale lipoma	Yes	No	S2-S3 level LSA and branches of IIA	Failed embolization x3→ surgery
Djindjian et al, 1989 [12]	53	Filum terminale lipoma	n/a	No	LSA	Embolization then excision of lipoma
Giordan et al, 2018 [3]	46	L3-S4 LMC	Yes	No	LSA	Embolization
	44	L2-S1 MMC	Yes	Yes	LSA	Embolization (Worsened due to

						spinal cord infarction)
	65	L2-L3 Diastematomyelia	Yes	Yes	LSA	Detethering then embolization
	64	LMC	Yes	No	LSA	2 of these patients underwent embolization, 1 not treated. *2 patients with 2 feeders
	57	MMC	n/a	Yes	LSA	
	64	n/a	Yes	Yes	LSA	
Horiuchi et al, 2017 [4]	51	L5 LMC	n/a	No	LSA	Embolization + surgery + Neuromonitoring
	53	L2/3 LMC	n/a	No	2nd LA	Embolization then surgery (multiple feeders)
Mavani et al, 2014 [9]	29	Lumbar LMC	Yes	Yes	L4 RA	Surgery
Sato et al, 2013 [8]	72	L3/4 LMC	Yes	No	2nd LA	Embolization
Talenti et al, 2017 [5]	19	Lumbosacral LMC	Yes	Yes	Middle sacral and lateral sacral branches of IIA (bilateral)	Embolization
	53	Sacral LMC	Yes	Yes	Right sacral segmental branch	Embolization then surgery (partial occlusion on embolization)
Cheung et al, 2005 [14]	42	Sacral LMC	Yes	No	S1/S2 anterior spinal artery	Surgery
Erdogan et al, 2007 [16]	40	L2/3 LMC	Yes	No	2nd LA	Embolization
Konig et al, 1999 [18]	50	Lumbosacral LMC	n/a	No	3rd LA	Surgery
Krisht et al, 2015 [15]	58	L4/5 LMC	Yes	Yes	Bilateral sacral arteries	Embolization, New feeder 6 months later requiring surgery
Rajeev et al, 2005 [17]	44	L1/2 LMC	Yes	No	L1 RA	Surgery
Weon et al, 2005 [6]	30	L4/5 LMC	Yes	No	Left L3/4 RA, right L3 RA from LA	Embolization

Coexistence of these pathologies pose diagnostic challenges as it is extremely difficult to determine the cause of myelopathy progression [9]. Symptoms are often attributed to the mass effect of the local tumor, TCS and chronic venous hypertension [6,12]. Some propose that increasing symptoms of neurological deficits should be attributed to the SDAVFs rather than the cord lipoma or tethering of the cord, whilst others report that presence of a spinal lipoma may worsen the symptomatology of such SDAVFs [13,14].

Clinical signs and imaging directed us to attribute this patient's symptoms to the SDAVF, hence the decision for angioembolization first. Nonetheless, this was a rather complex case. Although he successfully underwent angioembolization to the feeding vessel originating from the median sacral artery, other feeder vessels opened up, namely from the left lateral sacral artery branches. Difficult decision-making process was handled via a multidisciplinary meeting involving the radiologist, interventional radiologists as well as the neurosurgeon, and finally, the decision was made to reattempt angioembolization as this was less invasive and if successful, could reduce the morbidity for the patient should he undergo a second surgery to the spinal cord. It was technically challenging to perform a second embolization to the left lateral sacral artery branches due to the histoacryl material from previous embolization which caused the vessel to be stenotic with subsequent perforation of the feeder vessel. Interestingly, this phenomenon of other feeder vessels opening up post angioembolization is also observed in the other cases as mentioned in Table 1. In comparison to most of these cases, we attempted a second angioembolization rather than to proceed with surgery. Only one other case reported a similar strategy [13].

Of the cases published, majority of patients either had improving or non-worsening of symptoms whereas only one patient developed spinal cord infarction as the feeder vessel also supplied the anterior spinal artery [3]. The treatment challenges of these patients are conceivably outstanding due to two main factors. Firstly, it is often impossible to determine the exact cause for the symptoms in patients who have SDAVFs, spinal dysraphism and TCS. Secondly, due to the multiple pathologies present, often these patients benefit from a combination of treatment modalities rather than a single modality [15]. Treatment of the SDAVF should be aimed at disconnection of the AVFs from the venous drainage to relieve venous pressure and hence improve cord perfusion [14]. Although embolization cannot tackle the tethered cord or spinal lipoma, embolization can make resection and detethering easier [8,16]. Furthermore, embolization reduces blood loss and can help preop planning [4]. It is noteworthy that embolization may only be feasible in about three-quarters of patients and as demonstrated in this patient, embolization may be challenging in SDAVFs with multiple feeder vessels and may become more complicated in repeat procedures [1]. Some consider surgery to be superior as it has been reported to carry higher success rates in occluding the feeder vessel [17]. Surgery is advantageous as it enables all three pathologies to be tackled simultaneously by detethering the cord, excising the spinal tumor, and ligating the feeder vessels [10]. Additionally, surgery may be superior

compared to embolization as surgery may serve as a rescue in patients who failed embolization.

In hindsight, subjecting this patient to surgery after the first angioembolization could have potentially benefited him for a few reasons. Firstly, by detethering the cord, there would be a theoretical reduction in the release of angiogenetic factors hence reducing the occurrence of a new SDAVF formation. Secondly, surgery would be helpful as it could directly ligate all feeder vessels as well as be able to excise the cord lipoma. However, it is noteworthy that this would be a second surgery, which means adhesions could complicate the patients' recovery. Adhesion could make identification of feeder vessels much more difficult and may cause injury to the already-strained spinal cord. Non-improvement in his symptoms is similar to results found in other papers. Long-term prospective data on these cases are virtually non-existent, hence prognosis is unclear. Nevertheless, this patient could return in the future with worsening neurology. Should the patient return with worsening symptoms, perhaps the best course of action would be to re-map the feeder vessels with a CTA and a spinal angiogram followed by surgery to ligate the feeders, excise the lipoma and detether the cord. A preoperative angioembolization may be beneficial to reducing blood loss intraoperatively as well.

Patients with spinal dysraphism, TCS and SDAVFs should be managed in a multidisciplinary individualized approach whereby an informed discussion and decision-making is done with consideration of resources, clinical scenario, feeder vessel variabilities and patients' preferences.

Conclusion:

The serendipitous occurrence of spinal dural AVFs in spinal dysraphism and cord tethering is likely to be under-reported and poses diagnostic challenges. High index of suspicion of vascular anomalies should be present when a patient with spinal dysraphism presents with a new onset neurological deterioration. The authors propose multimodal and multidisciplinary approaches in diagnosing and managing such patients.

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Figure Legends:

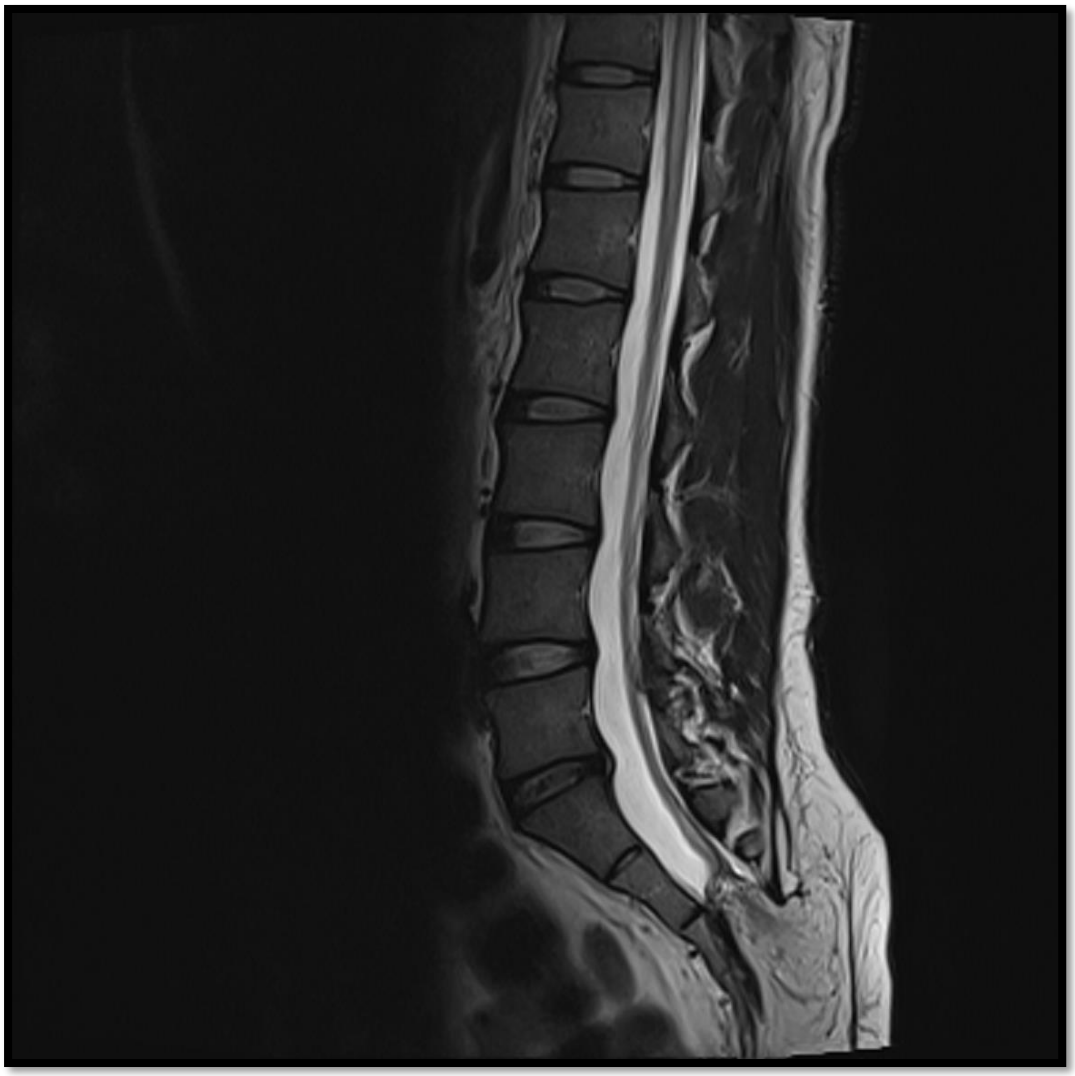

Figure 1: MRI Spine image Sagittal T2 MRI image demonstrating low lying tethered cord with high intensity T2 signals within the cord up till the level of L2.

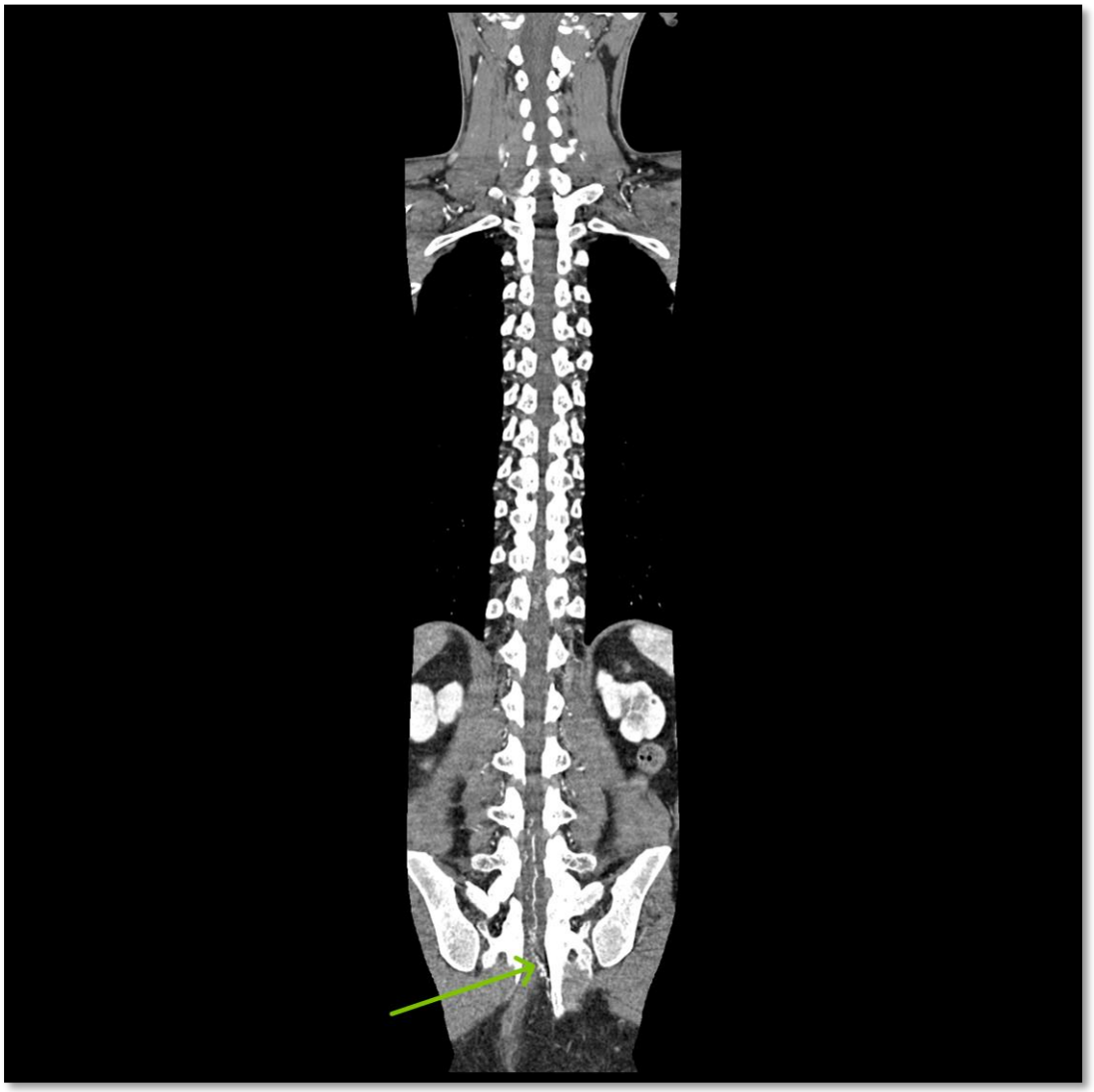

Figure 2: CT Angiogram image of the spine.
Coronal images of the patient's first CT angiogram demonstrating tortuous vessels at the spinal canal which courses cranially from S2/S3 level (green arrow).

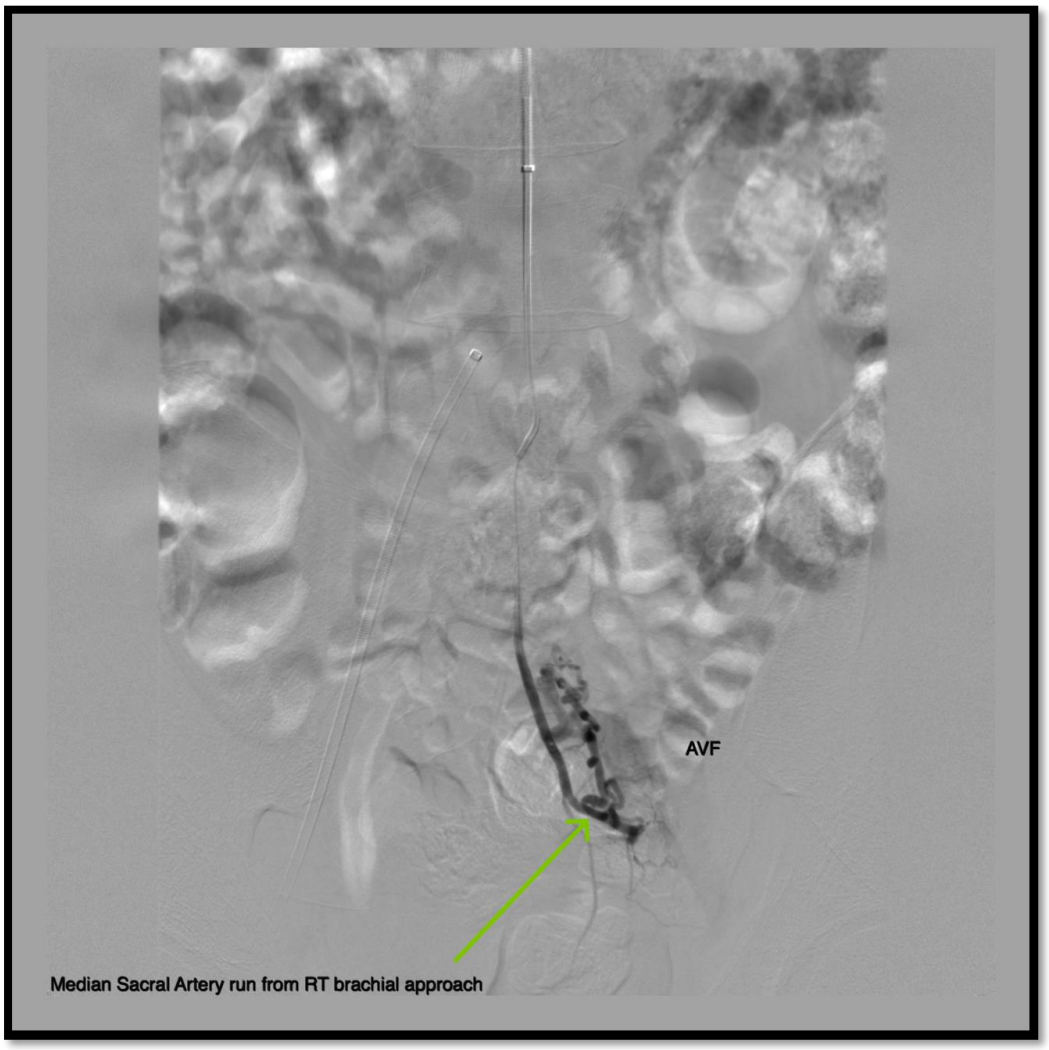

Figure 3: Spinal Angiogram image.

This was the median sacral artery run done via the right brachial approach. The arteriovenous fistula is demonstrated via the green arrow as the tortuous vessel flowing cranially from a branch of the median sacral artery.

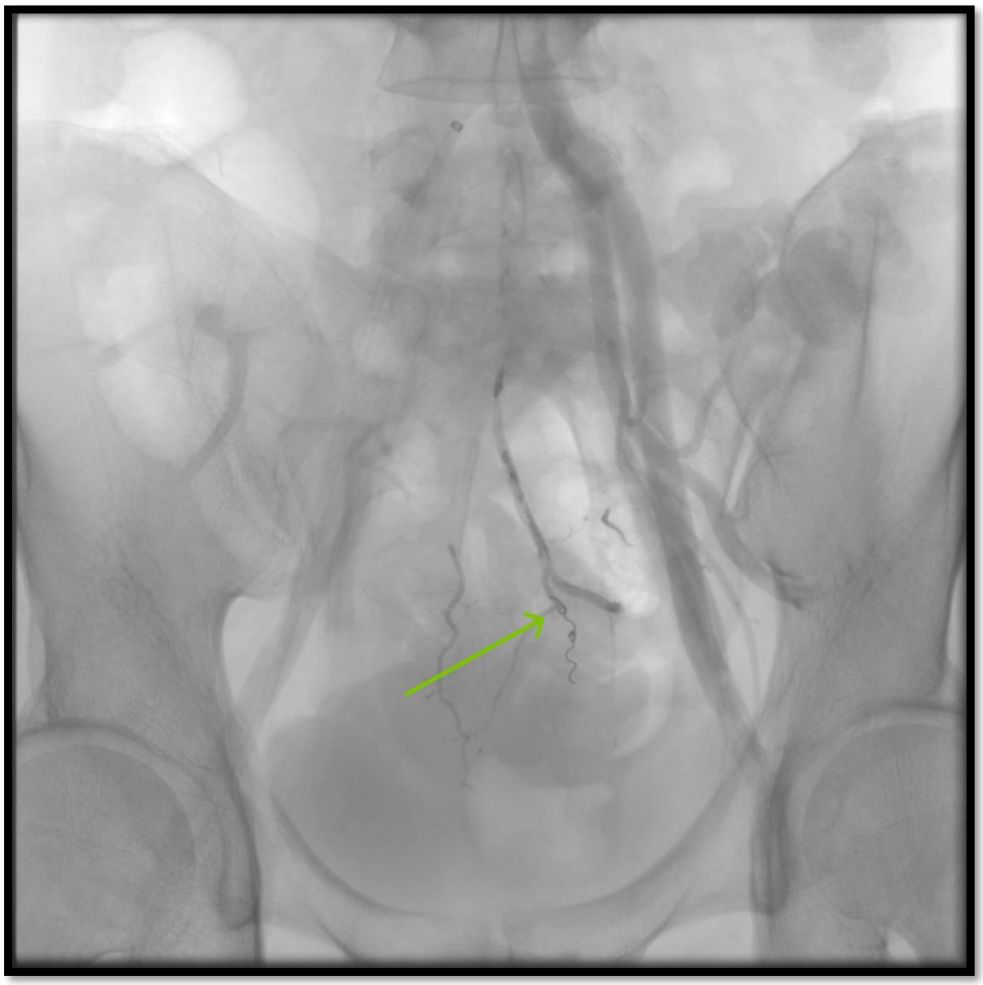

Figure 4: Post First Embolization Angiogram image.
Post coiling, ethyl alcohol and histoacryl glue embolization image shows non opacification of the SDAVF as seen in figure 3. Note the coil seen (green arrow). Note that the distal abdominal aorta is seen together with the both iliac arteries opacified with no residual or other possible feeder vessel as this point in time.