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Case Series

Radiology Section

Sacral Dural Arteriovenous Fistulas, a Rare and Underdiagnosed Subtype of Spinal Dural Arteriovenous Fistulas: A Case Series with Review of Literature

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ABSTRACT

Spinal Dural Arteriovenous Fistulas (SDAVFs) are the most common type of spinal vascular malformations with arterial feeders usually from the intercostals, vertebral, subclavian and lumbar arteries and rarely from the internal iliac arteries. Authors report three cases of SDAVFs at our institution where arterial feeders were from internal iliac artery. The endovascular embolisation of arterial feeders was done using n-BCA (n-butyl cyanoacrylate) embolic agent in one case and with Ethylene Vinyl Alcohol (Onyx) in two cases with complete obliteration of the fistula. This resulted in marked clinical improvement in all the patients. The lateral sacral arteries are not a common site of origin of arterial feeders to the SDAVFs and can be easily overlooked if thorough spinal Digital Subtraction Angiography (DSA) is not performed. Through these three cases, authors want to highlight the importance of performing complete spinal DSA.

Keywords: Endovascular embolisation, Internal iliac artery, Spinal vascular malformation

INTRODUCTION

The SDAVFs are the commonest spinal vascular malformations; however they are still rare and are often misdiagnosed or underdiagnosed. SDAVFs are usually located dorsally in the low dorsal and lumbar regions with 80% occurring between T6 and L2 [1]. The incidence is highest in the 5th and 6th decades of life with males getting affected more often. The symptoms are insidious in onset and gradually progressive with significant delay between presentation and diagnosis. The aetiology of SDAVF is unclear. However, it is considered to be an acquired disease in view of majority of patients becoming symptomatic in the middle age. Most of the cases are spontaneous and history of trauma present only in 4% of the patients [2]. They are never located within the spinal parenchyma unlike spinal arteriovenous malformations and hence rarely cause intra-medullary haemorrhage. They are abnormal, direct communication between a radicular/radiculomeningeal artery and a radicular/pial vein in the dura of an adjacent nerve root sleeve. The arterial feeders are usually from the intercostals, vertebral, subclavian and lumbar arteries. However, more rarely the internal iliac arteries can give feeders to the SDAVFs which can be easily missed on DSA if the internal iliac artery angiograms are not taken. Nishio A et al., recommended that in case of suspected SDAVF if the lesion is not identified in the dorsal and lumbar spine, an angiographic examination of the internal iliac arteries should be performed [3]. Authors present three cases of SDAVFs with arterial feeders from internal iliac arteries with patient presenting with progressive lower limb weakness and difficulty in walking with associated bladder disturbances.

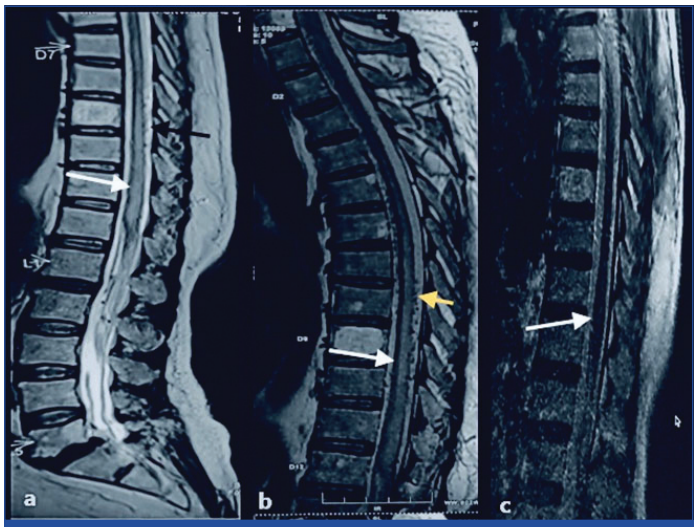
CASE SERIES

Case 1

A 65-year-old, male, presented with complaints of difficulty in walking since two years which was insidious in onset and gradually progressive. There was associated low backache since two years and jerky movements in bilateral lower limbs since eight months. There was urinary incontinence since four months.

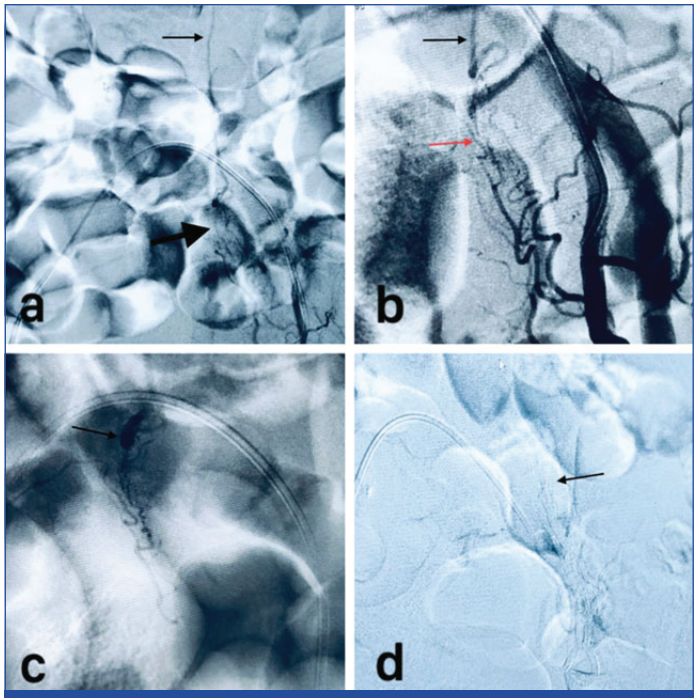
Patient was conscious, cooperative and well oriented to time, place and person with intact higher mental functions and cranial nerves examination. The motor examination was abnormal for bilateral lower limbs which showed spasticity and reduced power of 3 out of 5. Deep tendon reflexes were 3+ for knee and ankle. Plantar reflexes were extensor bilaterally. On sensory examination there was decreased touch and pain below L1 level.

On Magnetic Resonance Imaging (MRI) long segment intramedullary T2 hyperintensity within the dorsal spinal cord extending from D5-6 intervertebral disc level to lower end of D12 vertebra, multiple flow voids on the surface of the spinal cord with associated cord expansion and oedema [Table/Fig-1].



[Table/Fig-1]: a) Sagittal T2-weighted (T2W) Magnetic Resonance Image (MRI) of the lower dorsal and lumbar spine showing T2 hyperintense signal in the visualised dorsal spinal cord extending till the conus medullaris with associated expansion (denoted by white arrow) and multiple flow voids on the cord surface (denoted by black arrow). b) Follow-up MRI after six months showing more prominent cord edema, expansion (denoted by white arrow) with more prominent intradural flow voids (denoted by yellow arrow). c) Sagittal T2W MRI done six months after the endovascular intervention showing normal cord signal intensity with no expansion or flow voids on the cord surface (denoted by white arrow).

The DSA showed a type I SDAVF with feeding artery arising from anterior division of left internal iliac artery [Table/Fig-2].



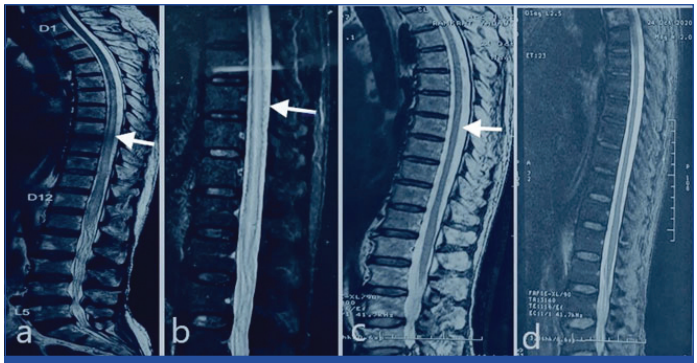
[Table/Fig-2]: a) Selective left lateral sacral artery angiogram showing the site of SDAVF (denoted by thick black arrow) and early draining vein (denoted by a thin black arrow).b) Left internal iliac artery (IIA) angiogram showing SDAVF with arterial feeders from left lateral sacral artery (denoted by a red arrow) with early venous drainage into a tortuous perimedullary vein (denoted by black arrow). c) DSA image showing embolisation of the fistula using onyx (denoted by black arrow). d) Post-embolisation image showing complete obliteration of fistula with visualisation of onyx cast (denoted by black arrow).

The angiogram was taken after selective catheterisation of the left internal iliac artery which delineated the arterial feeders from the left sacral artery and venous drainage into the perimedullary veins. Subselective catheterisation of the left lateral sacral artery was done using Dimethyl Sulfoxide (DMSO) compatible marathon microcatheter (Marathon™). Selective angiogram was taken to delineate the location and anatomy of the fistula explicitly. Further onyx 18 containing 6% ethylene vinyl alcohol was used as liquid embolic agent and was administered under fluoroscopic visualisation via the marathon microcatheter after coating its lumen with DMSO. Postembolisation check angiogram showed no connection between arterial and venous components of fistula. On three months follow-up postembolisation, the patient showed remarkable improvement in difficulty in walking and lower backache. However, urinary incontinence persisted.

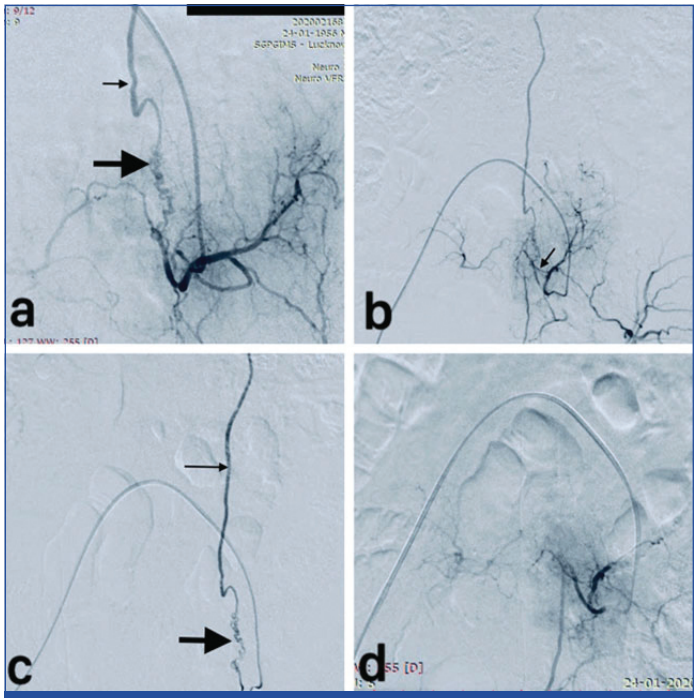
Case 2

A 63-year-old male patient presented with complaints of gait imbalance of eight months duration and gradually progressive bilateral lower limb weakness of six months duration. There was history of sensory loss over abdomen and urinary incontinence since two months. Patient was conscious, cooperative and well oriented to time, place and person with intact higher mental functions and cranial nerves. The motor examination of lower limbs showed increased tone with decreased power of 2 out of 5 and deep tendon reflexes of 4+ at knee and ankle. The plantar reflexes were extensor bilaterally. Sensory examination revealed pain and sensory loss below the D9 level.

On MRI, a long segment intramedullary cord hyperintensity was seen extending from D6 level till the conus medullaris with multiple prominent flow voids along the dorsal and ventral aspect of the dorsal cord, more marked in the lower dorsal cord region [Table/Fig-3]. DSA showed type I SDAVF in lower lumbar spinal canal with feeding artery from lateral sacral branch of left internal iliac artery. The venous drainage was into perimedullary vein. The artery of Adamkiewicz was seen arising from radiculo-medullary branch of left D10 posterior intercostal artery [Table/Fig-4].



[Table/Fig-3]: a) Standard sagittal T2W MRI of dorsolumbar spine showing long segment hyperintensity (denoted by white arrow) in dorsal and lumbar cord till the level of conus medullaris, multiple flow voids along cord surface and cord expansion. b) Sagittal T2W MRI with fat suppression showing cord hyperintensity extending from D6 till the conus medullaris with cord expansion and multiple flow voids (denoted by white arrow). c) Standard sagittal T2W image of the dorsolumbar spine post embolisation showing radiological improvement evident as non visualisation of flow voids on cord surface, normalcy of cord intensity and caliber (denoted by white arrow).d) T2 sagittal fat suppressed image showing the post treatment changes mentioned in figure c.



[Table/Fig-4]: a) Left IIA angiogram showing SDAVF with arterial feeders, fistula site (denoted by thick black arrow) and early draining vein (denoted by thin black arrow).b) Left IIA angiogram showing fistula with arterial feeders from left lateral sacral artery (denoted by black arrow). c) Angiogram showing embolisation of the fistula with liquid embolising agents (denoted by thin and thick black arrows at the perimedullary vein and fistula sites respectively). d) Postembolisation angiogram showing complete obliteration of the SDAVF.

Selective catheterisation of left internal iliac artery was done delineating arterial feeders from left sacral artery and venous drainage into the perimedullary veins. Subselective catheterisation of the left lateral sacral artery was done using Magic 1.2 Fr flow dependent microcatheter (Balt USA) having a length of 165 cm. Further, the embolisation of the fistula was done using mixture of glue (N-butyl cyanoacrylate) and lipiodol in a ratio of 1:2 under fluoroscopic visualisation.

Postembolisation selective angiogram of left sacral artery showed no residual connection between arterial and venous components of fistula. On three months follow-up postembolisation, the patient showed remarkable improvement in the lower limb weakness and difficulty in walking. However, pain sensation and bladder dysfunction persisted.

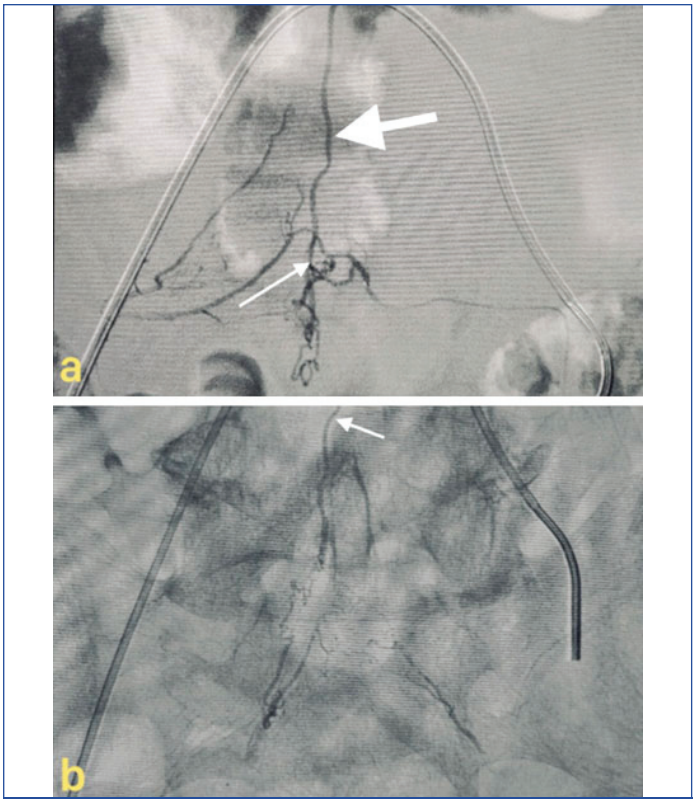
Case 3

A 51-year-old male patient presented with band like compressive sensation in lumbar region with radiation of pain into bilateral lower

limbs since two years. There was history of gradually progressive bilateral lower limb weakness since last 18 months with associated diminished sensation below L3 level. There was associated history of constipation and urinary incontinence for one year.

Patient was conscious, cooperative and well oriented to time, place and person with intact higher mental functions and cranial nerves. The motor examination of lower limbs showed increased tone with decreased power of 2 out of 5 and deep tendon reflexes of 4+ at knee and ankle. The plantar reflexes were extensor bilaterally. Sensory examination revealed pain and sensory loss below the D8 level.

The MRI dorsolumbar spine showed long segment intra-substance hyperintensity in the dorsolumbar region cord parenchyma with multiple flow voids on cord surface. Spinal DSA showed a SDAVF with feeders from lateral sacral arteries [Table/Fig-5a]. Under strict aseptic precaution and under general anaesthesia, right common femoral artery access was secured using 6Fr arterial sheath. A 5Fr Renal Double Curved (RDC) catheter was used to selectively cannulate the left internal iliac artery. An angiogram was taken which showed arterial feeders from right as well as the left lateral sacral artery. The left lateral sacral artery was selectively cannulated using marathon microcatheter and traxcess guide wire and 0.5 mL of glue and lipiodol mixture in a ratio of 1:2.5 was injected. The postembolisation angiogram showed complete obliteration of the fistula [Table/Fig-5b].



[Table/Fig-5]: a) Left lateral sacral artery microcatheter angiogram showing the site of SDAVF (denoted by thin white arrow) with feeders from bilateral lateral sacral arteries and an early draining perimedullary vein (denoted by thick white arrow). b) Postembolisation angiogram showing complete fistula obliteration and glue cast in both lateral sacral arteries and reaching the draining vein.

On three months follow-up postembolisation, the patient showed significant improvement in the lower limb weakness and pain symptoms. However, the bladder dysfunction did not show much improvement. The patient was lost to follow-up due to prevailing COVID conditions.

DISCUSSION

The spinal vascular malformations are divided into four subtypes which include type I (SDAVF), type II (glomus arteriovenous malformation), type III (juvenile arteriovenous malformations) and type IV (intradural arteriovenous fistula) [4]. The SDAVF are the most commonly encountered vascular malformation of the spinal cord accounting for ~70% of all such lesions and are one of the treatable causes of progressive myelopathy [1,2]. It was first described by Foix and Alajouanine in 1926 [5]. They are rare and underdiagnosed with reported annual incidence of 5-10 cases per million [2]. As per the available literature the average delay is 15-24 months between symptom onset and diagnosis confirmation [6,7]. The elderly males in the 5th and 6th decades of life are usually affected [8]. The most common location in spine is the dorsolumbar region with more than three quarter of cases located from mid-thoracic to the upper lumbar spine levels [1].

These fistulae are acquired, low-flow abnormal direct connections between a radicular/radiculomeningeal artery and a radicular/pial vein in the dura covering spinal nerve roots [2]. Approximately, 85% of SDAVFs consist of a single transdural arterial feeder; however, there are cases with many arterial feeders originating from either a single or multiple levels that may be either unilateral or bilateral [8].

The SDAVFs are mostly supplied by intercostals and lumbar arteries. However, arterial feeders have been reported from vertebral arteries, costocervical trunk, thyrocervical trunk, presacral and external carotid arteries. Rarely, fistulas located in the sacral region, are supplied by lateral sacral and iliolumbar arteries as in index cases [9]. The reported incidence of SDAVFs in the sacral region was 12.5% in a series by Nishio A et al., [3]. Larsen DW et al., [9] recommended that an angiographic examination of the internal iliac arteries be performed in patients with suspected SDAVF when the lesion has not been detected in the dorsal or lumbar region as in index cases where the intercostals and the lumbar artery angiograms did not show any fistula.

The presence of a fistula leads to arterialisation of the spinal veins resulting in spinal venous hypertension, cord congestion, cord oedema with untreated cases progressing to cord ischaemic and infarction [5]. The patients usually present with progressive lower limb weakness, pain or sensory changes. The sphincteric involvement can also be seen. The symptoms are generally insidious in onset and progress over many months to years.

Usually, the initial radiographic diagnosis is made on MRI. It helps in excluding the common causes of paraplegia and reveals characteristic appearance suggestive of SDAVF which include abnormal long segment T2 hyperintensity within the cord, cord expansion and flow voids on the dorsal and/or ventral aspect of cord. Spinal cord atrophy and some T1 postcontrast enhancement can be seen in chronic SDAVFs [5]. In acute presentation, the cord enhancement is due to capillary leak phenomenon secondary to venous hypertension. Diffuse multilevel intramedullary hyperintensity is the most sensitive finding [10]. Prominent serpiginous intradural extramedullary flow voids are the most specific finding. They usually span more than three segments. T2 peripheral cord hypointensity can be seen [11]. It is thought to represent pial capillaries containing deoxyglobin secondary to venous hypertension.

The segmental level of cord enlargement and signal change does not correlate with the location of the fistula [12]. Alternatively if MRI is contraindicated, Computed Tomography (CT) angiography with 75% successful localisation of fistula can be used [12]. CT myelography can also be used showing fistula as filling defects due to dilated veins [12]. DSA confirms the diagnosis in cases of clinical or radiological suspicion. However, it is an invasive procedure with potential risk of cord ischaemic secondary to dissection.

The treatment in SDAVF is aimed at occluding the shunting zone (i.e., the most distal part of the artery together with the most proximal part of the draining vein) [13], either by superselective embolisation with a liquid embolic agent or by a neurosurgical approach.

Surgery consists of targeted laminectomy and intradural exploration with coagulation or disconnection of the draining vein; occlusion rates as high as 98% have been reported [12]. However, high occlusion rates to the tune of 85% have been reported with endovascular embolisation [12]. The endovascular treatment showed results comparable to surgery when the fistula point was correctly disconnected as per single centre experience documented by Gioppo A et al., [14]. It is performed either with glue or onyx as in index cases after superselective catheterisation of the radiculomeningeal artery supplying the fistula which in two of the cases was left lateral sacral artery and bilateral sacral arteries in the third case. It is contraindicated if the radiculomeningeal artery also supplies the Anterior Spinal Artery (ASA) which was not the scenario in any of index cases. The risks and benefits of the endovascular embolisation were explained to the patients and their families. The risks are usually due to inadvertent embolisation leading to ischaemic sequelae and permanent neurological deficits [15].

Following obliteration of the fistula, the disease progression can be stopped and symptomatic improvement is usually seen. If treated early, motor and sensory functions recover better than pain and bladder dysfunction which are only reversed in a minority of patients as in index case which did not show improvement of bladder dysfunction.

A differential diagnosis of filum terminale Arteriovenous Fistula (AVF) and very rarely a cauda equina AVF should also be kept in mind as rarely they can also have a similar clinical and imaging picture. Filum terminale AVF usually has a single feeding artery with ASA continuing as the feeding artery [16] with normal calibre and course at the level of conus medullaris. The venous drainage is via filum terminale vein which runs parallel to the feeding artery giving a characteristic feeder drainer pattern.

The cauda equina AVF feeder could be radicular, spinal or both [17]. The ASA shows changes in calibre reflecting the transition of the ASA into a radicular artery in cauda equina AVF. Unlike the parallel feeder drainer patter seen in the filum terminale AVF, the cauda shows wavy drainer pattern [18].

CONCLUSION(S)

The SDAVFs are uncommon lesions and more so in sacral location and can be easily overlooked if high degree of clinical suspicion is not there. The initial diagnosis is usually made on an MRI with lesions manifesting as abnormal long segment cord hyperintensity with associated expansion and flow voids with or without cord enhancement. The definitive diagnosis is usually made on DSA. The three cases discussed above highlight the importance of doing a complete spinal DSA including evaluation of internal iliac arteries. This becomes more important if the patient has MRI findings suggestive of SDAVF and routine spinal angiography fails to demonstrate the fistula. The failure to perform complete spinal DSA in such cases would result into unnecessary delay in diagnosis leading to delayed treatment resulting in poor patient outcome.

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