

MANAGEMENT OF SPINAL DURAL ARTERIOVENOUS FISTULA, A REVIEW WITH ONE CASE REPORT**Mohamed El Hussein[@], Hussein Mouawia[#], Adnan Mrad^{\$} & Taghrid Chaaban^{*}**[@]MD, PhD, Neurosurgeon, Hôpital Libano Français, Zahle, Lebanon. [#]Director, Lebanese University, 4th Branch, Lebanon. ^{\$}Dean, Islamic University of Beirut. ^{*}Vice dean, Islamic University of Beirut, Lebanon.**Abstract**

Spinal dural arteriovenous fistulas (SDAVFs) are rare acquired vascular malformations of the spinal cord which if not treated properly, can lead to inevitable severe morbidity with progressive spinal cord symptoms. The management is still at high interest among specialists. If microsurgical treatment is still considered as a treatment of choice for SDAVFs, endovascular treatment is increasingly growing in interest with the development of endovascular techniques and new embolization materials.

In this article, a short discussion is made about the spinal dural arteriovenous fistulae in respect to anatomy, etiology, diagnosis and treatment. Careful patient selection, a multidisciplinary approach and standardized surgical techniques can lead to excellent results with virtually no complications.

Introduction

Vascular malformation of the spinal cord represents rare clinical condition characterized by a difficult diagnosis and complex management. Spinal dural arteriovenous fistulas (SDAVF) are the most common condition in these pathological entities with important clinical implications. These direct communications between radicular artery and medullary vein usually results in myelopathy due to venous hypertension. Assessment of a SDAVF is often difficult because of non-specific findings on non-invasive imaging modalities. With the advances in neuroimaging, micro neurosurgery and neuro endovascular techniques, the complete treatment of these pathological situations is very feasible with the possibility of complete remission of clinical symptomatology. Endovascular embolization was reported as an effective therapy in the treatment of SDAVFs that can be used as singular and definitive intervention in some particular cases. We

present a particular case with SDAVF treated by endovascular embolization and discuss the treatment possibilities to more fully understand the optimal management of these lesions.

Vascular Anatomy: Spinal cord vascularization is provided by the anterior spinal artery (ASA) and the paired posterior spinal arteries (PSA). The ASA consists of the junction of two branches originating from the two vertebral arteries proximal to the vertebra basilar junction. On its path, it receives contributions from branches of vertebral and ascending cervical arteries in the cervical region as well as from intercostal and lumbar arteries at the corresponding levels. The spinal meningeal arteries are branches of the segmental arteries founded at almost every spinal level supplying the dura in the spinal canal. Unlike these, spinal medullary arteries, which exist only at some levels, are implicated in spinal cord vascular perfusion. The artery of Adamkiewicz (great anterior spinal

medullary artery) is the dominant thoracolumbar segmental artery with variable origin from T8 to L1 vertebral segments that connects to ASA and supplies the spinal cord (figures 1, 2a&b). The posterior spinal arteries arise from either the posterior inferior cerebellar or

vertebral arteries (V3 or V4 segments) and as they descend on either side of the dorsolateral cord surface, they are reinforced by segmental/radicular branches. It make anastomoses with its fellow and with the anterior spinal artery^{1,2}.

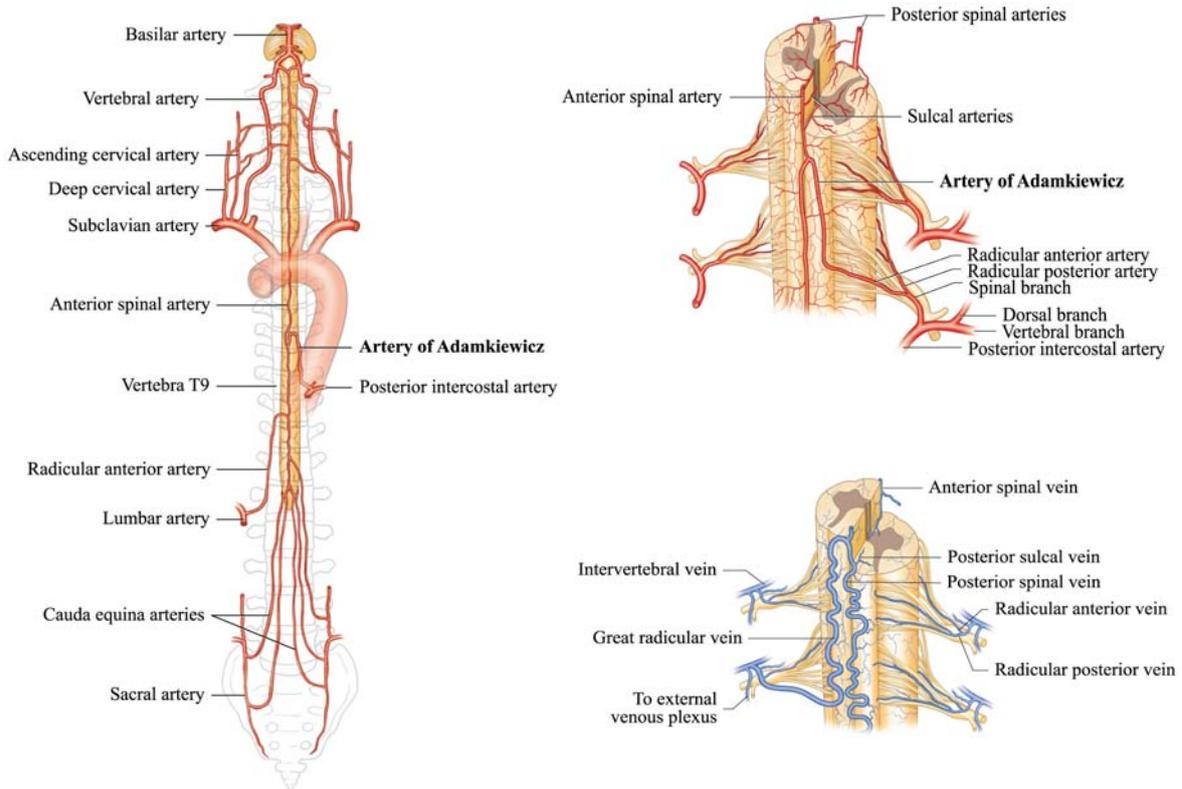


Fig.1, 2 a&b: Vascular anatomy of spinal cord Sited from: J.Vasc.Bras.2015,14(3):248-252

Etiology, Epidemiology and Pathophysiology: The etiology of vascular malformations of the spinal cord has not been clearly defined. Intradural parenchymal malformations arises in younger patient population and are believed to be congenital. However, spinal arterial dural fistulas commonly arises in elderly population and are believed to be due to trauma. These AVF malformations develops near a spinal dural artery, forming an abnormal arteriovenous communication with the venous circulation³.

SDAVF represents 70% of spinal arteriovenous shunts that commonly occur in the thoracic and lumbar spines of middle aged men^{2,4}. However, the disease

seems to be underdiagnosed^{5,6}. The majority of SDAVFs occurs spontaneously, but a post-traumatic etiology cannot be excluded in a significant proportion of them. Typically, this disease affects male patients (in 80% of cases).

The pathophysiology in spinal cord venous hypertension is due to one or a few small low-flow arteriovenous shunts between a spinal meningeal artery and a spinal medullary vein, typically located in the intervertebral foramen within the dura^{2,4,7}. The retrograde venous drainage circuit in SDAVF is represented by a spinal medullary vein into the peri medullary venous system and finally the medullary veins. The venous drainage of

the SDAVF is slow and expansive, and may reach the cervical spinal canal or cauda equina by ascending or descending blood reflux. Because the spinal medullary veins are not anatomically numerous, the presence of SDAVF is often associated with epidural veins congestion and thrombosis. That explains why the low-flow arteriovenous shunt of SDAVF induces a rises of venous pressure (74% of the mean arterial pressure) which leads to decreased arteriovenous gradient, segmental spinal cord edema that may progress into congestive ischemia and necrotizing myelopathy. Venous hypertension can be confirmed with angiography of the artery of Adamkiewicz by demonstrating severe prolongation of the venous phase². The caudo-cranial progression is favored by a valveless venous system of the cord resulting in 'arterialization' of these veins with thickened and tortuous walls. The pressure in the draining vein also varies with arterial pressure and may lead to an exacerbation of symptoms. Because the SDAVFs is a slow-flow fistulae, hemorrhage is a rare clinical manifestation. Subarachnoid hemorrhage is rarely encountered especially in high cervical localization^{2,7}.

Classification: Many classification systems were reported and changed over the time in the literature. Between 1971 and 2011, seven major classification

systems have been enunciated based on the evolution of diagnostic methods and treatments for spinal AV shunts. The most used is the division of the vascular spinal lesions into SAVFs and SAVMs. SAVFs are further subdivided based on their extradural versus intradural location. The intradural SAVFs were divided in ventrally or dorsally due to their relation to the spinal cord. In turn, intradural ventral SAVFs are further divided into types A, B and C depending on the number and size of feeding branches^{1,2}. Extradural SAVFs represents direct connection between a branch of a radicular artery and the epidural venous plexus (figure 3). These are rare entities characterized by enlargement of epidural veins with medullary venous congestion that may cause compression of the spinal cord or nerve roots. More recently, these lesions are divided into; type A (SAVFs drain into both the epidural venous plexus and perimedullary venous plexus) and type B (SAVFs drain only into Batson's plexus). Type B1 lesions compress the thecal sac due to an enlarged epidural venous plexus and type B2 lesions lack such compression^{1,6,7}. Intradural dorsal SAVFs are the most common type of spinal vascular malformation consisting in a direct connection between a dorsal spinal medullary artery and a medullary vein at the dural nerve root sleeve as shown in figure 4.



Fig.3 SAVF: The connection between radicular artery and the epidural venous plexus



Fig.4 SAVF: The connection between dorsal spinal medullary artery and medullary vein

Progression of venous hypertension to the coronal venous plexus leads to venous congestion and progressive myelopathy. Intradural ventral SAVFs are typically high-flow direct fistulas between the ASA and coronal venous plexus. The lesions develop in the ventral subarachnoid space and can be further categorized into three subtypes according to their size; Type A fistulas, are single-feeder lesions with slow blood flow and mild venous hypertension. Type B fistulas are progressively high-flow lesions with multiple minor feeders. Type C fistulas are usually large fistulas with a markedly enlarged venous drainage¹.

Clinical symptoms: Patients with AVFs are typically older than 40 years. These AVFs occur much more frequently in males than in females. Most clinical reports showed a delay between the onset of clinical symptoms and diagnosis of these vascular lesions (between 12 and 44 months), largely due to nonspecific clinical presentation⁶. The symptoms usually include a combination of unilateral or bilateral lower extremity motor weakness that is worsening by intense movements. Gait disturbance, sensory symptoms (pain, paresthesias, diffuse or irregular sensory loss, hyperesthesia) and sphincter/bladder disturbances are also seen and commonly leads the clinicians to consider or exclude many other disorders before considering SDAVFs. Often misleading, mono or poly radiculopathy and low back pain are encountered and contribute to the difficulty of true diagnosis. Bowel and bladder incontinence, sexual dysfunction and urinary retention are seen late in the course the disease process. The symptoms are typically progressive and the natural evolution of untreated patients is to severe aggravation over a period of 6 months to 2 years. Spontaneous recovery has not been reported so far as sudden worsening

has been more and more common^{1,7}. Misdiagnosis usually includes degenerative spine diseases, spinal cord tumours, neuromuscular diseases, peripheral vasculopathy or neurogenic claudication.

Imaging Diagnosis: Typically, a spinal MRI is ordered as a first-line screening method for the evaluation of myelopathy and diagnosis of SDAVFs. On T2-weighted sequences, the cord edema is depicted as a centromedullary not well-delineated hyper intensity over multiple segments that is often accompanied by a hypo intense rim, most likely representing deoxygenated blood within the dilated capillary vessels surrounding the congestive edema⁸. The abnormal shunts are much better outlined after administration of intravenous gadolinium-based contrast agents, increasing the sensitivity and specificity of MRI exam for SDAVFs diagnosis (figure 5A). The MRA sequences permits a better imaging characterization of the abnormal enlarged perimedullary vessels concerning their relative size, number and tortuosity. Magnetic resonance angiography is a useful tool for planning the surgical treatment and following monitoring (figure 5B). Other imaging signs like spinal cord edema, enlargement, gliotic or atrophy were reported depending on the stage of venous hypertension².

Conventional catheter angiography is the gold standard investigation for the diagnosis and classification of the SDAVFs. The selective catheterization and evaluation of each individual segmental arteries at thoracic and lumbar levels have to be performed for SDAVF identification. Once fistula is identified, a prolonged angiographic imaging acquisition has to be performed for a complete characterization of the venous drainage (figure 5C).



Fig.5A- MRI-T2

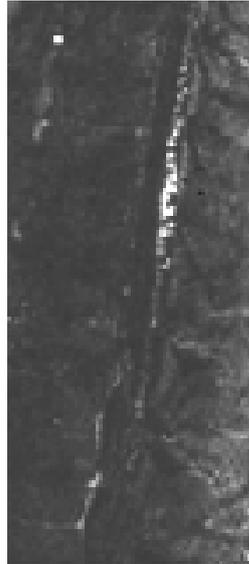


Fig. 5B- MRA

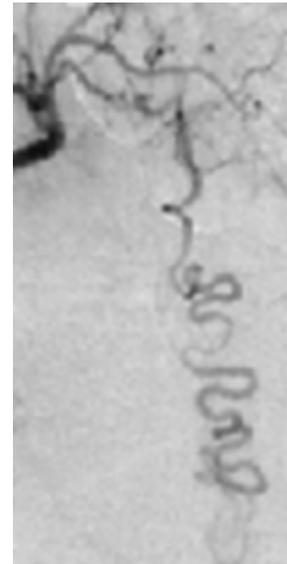


Fig. 5C- Catheter angiography

In patients with severe venous hypertension and myelopathy at thoracolumbar area the venous drainage is delayed or even absent. For suspected cervical or lumbosacral SDAVFs, the angiographic investigation of vertebral and ascending cervical arteries or the internal iliac and iliolumbar arteries should be performed. The major drawback of conventional angiography is the small risk of major complications due to its invasive nature and the use of iodinated contrast. Spinal angiography should also be used to evaluate the venous drainage after injection of the artery of Adamkiewicz. In patients with severe venous hypertension and myelopathy involving the thoracolumbar area, the venous drainage is prolonged or absent. If venous hypertension is demonstrated during spinal angiography, the cause must be found.

Treatment: Treatment options are dictated by the location of the lesion, the patient's medical condition, and the risk-versus-benefit ratio. It must be performed as soon as possible. It could be microsurgical ligation, endovascular obliteration, or both. Although the microsurgical treatment is considered the gold standard technique, the endovascular treatment could be a feasible and safe option. The microsurgical treatment is

performed by a posterior approach with a midline laminectomy one level above and below the fistula origin. The dura is opened in the midline and the radiculo-meningeal artery shunt must be identified extradural. In the case of a fistula with multiple small arterial pedicles, these should be carefully identified by dissection along the dural root sleeve (figure 6). The draining arterialized vein must be exposed and clearly identified against dilated perimedullary veins. The microsurgical technique consists in cauterization and micro scissor interruption of the fistula. Postoperative angiography is indicated to confirm complete surgical obliteration. Most of the studies have reported an improved disability scores and lower recurrence rates after microsurgery treatment compared with endovascular obliteration. Surgical management of SDAVF was also necessary when an incomplete endovascular obliteration or recanalization were the final results^{9,10}. because of the improvement in endovascular technique and embolic materials, the endovascular treatment of SDAVFs has more largely used.

The success of endovascular treatment was considered when a complete occlusion of the proximal radiculomedullary draining vein and the

site of the fistula itself were obtained. The procedure consists of right transfemoral access by 6F sheath placement. After the identification of the arterial supply of SDAVF, a guiding catheter is placed at the ostium of the corresponding segmental artery to offer more support for navigation into the often tortuous feeders. Then a micro catheter is advanced under road-mapping over a micro wire in order to reach the closest point to the fistula. If the embolization is performed too proximally in the radicular feeding artery,

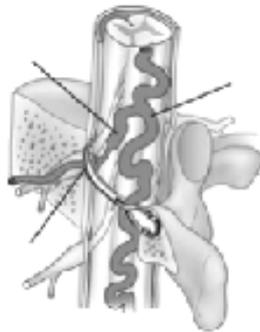


Fig.6: Microsurgical clipping and disconnection of artery feeder

Onyx (EV3, Irvine, California) has also been used to treat spinal cord AVMs. In a series of 17 patients with symptomatic intramedullary AVMs (15 exclusively intramedullary and 2 with additional perimedullary components) treated exclusively by Onyx, total obliteration was achieved in 6 patients; subtotal obliteration (tiny remnant), in 5; and partial obliteration, in 5¹⁰. Considering the relatively poor penetration of Onyx, for selected SDAVF with a low blood flow rate, Glubran 2 was the embolic agent targeting for curative embolization¹¹.

Finally, controlled angiography is performed by selective catheterization of the segmental arteries arising at least two levels above and below the SDAVF site. If there is no complete obliteration of the fistula, usually the patient is addressed for a microsurgical approach as soon as

collateral feeders could develop distally and re-permeabilized the fistula. A micro catheter angiography is recommended before starting the embolic agent injection in order to ensure if the anterior spinal, posterior spinal or a radiculomedullary artery are not directly connected to fistula. Endovascular occlusion is performed by slowly injecting Glubran 2 or Onix into the proximal draining vein while occluding the fistula site and feeding arterial vessels (figure 7).

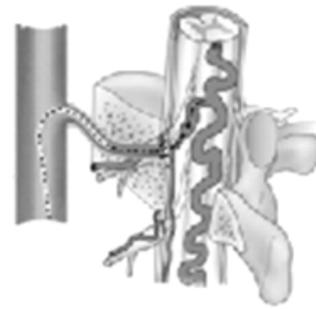


Fig.7: Endovascular glue injection in artery feeder and proximal radiculomedullary draining vein

possible. Also, if there are doubts concerning the complete occlusion of the proximal radiculomedullary draining vein, a control angiographies are performed at one, three and six months later^{2,9,12}.

Complications: The complications that results from open surgical ligation or resection are; meningitis, cerebrospinal fluid leak, and wound dehiscence. The endovascular technique is associated with higher incidence of recurrence, femoral hematoma, pseudoaneurysms, thrombosis, arterial dissection, and incomplete fistula occlusion⁴.

Illustrative Case

A 65 year old female presented to a neurologic department for progressive gait instability and weakness. MRI of the spine showed multiple abnormal blood vessels surrounding the spinal cord

(figure 8A). Based on the suspicion of a spine AVM, the patient was admitted to our neurosurgical department and her neurological examination revealed bilateral lower-extremity motor weakness and sensory deficits, with the left side more severely affected. The upper extremities had no motor or sensor deficit. Selective spinal angiography at left T-7 intercostal artery showed SDAVF supplied by its radiculomeningeal branch

and draining into a tortuous proximal radiculomedullary vein (figure 8B). We performed a posterior approach with a midline T7-T8 laminectomy. The dura was opened in the midline, the radiculo meningeal artery shunt was identified and we applied a clip on it. If during surgery, the collapse of the drainage vein is observed with the change of blood flow arterial to venous, the shunt is cauterized and disconnected (figure 8C, 8D).

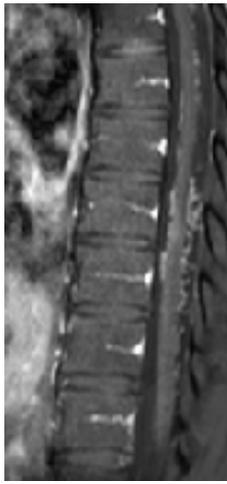


Fig.8A: MRI-T1/
TSE/enhancement

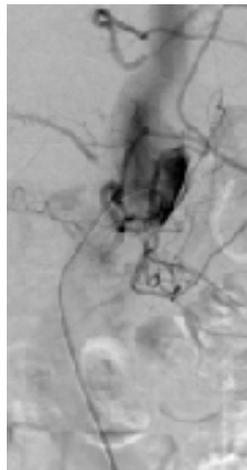


Fig.8B: Catheter
angiography



Fig.8C,8D: Shunt cauterization
and micro scissor disconnection

Postsurgical angiography was performed for fistula interruption documentation. The patient was discharged to a local recovery center. She continued to improve and was almost back to her baseline neurological and ambulatory status at the 8 month follow-up.

Discussion

The successful treatment of SDAVF requires a detailed understanding of clinical presentation and imaging findings to allow precise treatment. Owing to the rarity of the condition, clinicians must continue to share their experiences to advance their knowledge. SDAVFs are defined as abnormal direct connection between a radicular extradural artery and an intradural vein. The cause of SDAVF is not yet known. SDAVFs are more

common in men than in women, and most common in older adults. SDAVFs usually occur in the mid to lower spine (the thoracic and lumbar spine) Most of studies show a predominance of the lesion in the thoracic spine and to male gender.

The vast majority of patients presents with different degrees of neurological impairments usually correlated with the level of venous hypertension and its time occurrence. Symptoms of SDAVF are generally nonspecific, meaning they are similar to symptoms of many other problems that affect the spinal cord. These symptoms may include back pain, numbness and weakness in the legs, clumsiness, difficulty in walking or climbing stairs, impairment in bladder or bowel function, and sexual dysfunction.

Symptoms may develop slowly and steadily, or they may progress and then stay the same for a while before progressing again.

On MRI, the combination of cord edema, perimedullary dilated vessels, and cord enhancement is characteristic.

Treatment options consist of microsurgical exclusion and/or endovascular embolization. Embolization has the main advantage of being a less invasive procedure, which is particularly appealing in patients with SDAVFs who often have significant disability by the time they are correctly diagnosed¹³. There is a major limitation of embolization for SDAVFs as many of the patients are already neurologically impaired and need immediate complete and permanent obliteration of the fistula. Many authors still consider the surgical obliteration of SDAVF to be the gold standard for management of these lesions. However, improvements in endovascular technique and development of new embolic materials have made a greater number of patients with such vascular lesions to be treated for this type of management. The literature presents rates of successful endovascular therapy that vary between 25% and 90%^{4,7}. The advantages of this treatment have been associated with shorter time hospitalization, minimal procedural morbidity and earlier initiation of rehabilitation programs. Contraindications for endovascular occlusion of SDAVF are represented by the spinal cord supply from the same arterial trunk as the feeding artery of the fistula, difficulties of a distal catheterization of the feeder artery due to its anatomical

particularities, or recanalization of fistula after a previous embolization session. All experts have agreed that the success of endovascular treatment is closely related to the complete occlusion of both the arterial feeder and the proximal radiculomedullary vein. Recent comparative studies between microsurgical and endovascular treatment of SDAVFs on larger series of patients have shown that there are no statistically significant differences in post-interventional neurological recovery. Early diagnosis and successful treatment of the fistula were demonstrated to be strongly correlated with improvement in clinical symptoms. It was also found that improvement in motor function after treatment is more likely to occur than improvement in urinary dysfunction. The patients must be postinterventionally monitored by clinical examination and at least MRI imaging. In endovascular treated SDAVFs a catheter angiography control is recommended.

In conclusion, the SDAVF is a rare, treatable pathology, but can progress into serious morbidity and irreversible damage pathology. Treatment is essential because an untreated SDAVF can irreversible handicap. Endovascular treatment of SDAVFs represents a good and effective option for management of these vascular lesions. However, some limitations on the possibility of applying this type of treatment have been described. Timely diagnosis and treatment is essential to maximize the chances for recovery. For most cases, the surgical treatment is still considered to be the first intention treatment.

References

1. Ducruet, A. F., Crowley, R. W., McDougall, C. G., & Albuquerque, F. C. (2012). Endovascular management of spinal arteriovenous malformations. *Journal of neurointerventional surgery*, neurintsurg-2012.
2. Patsalides, A., Santillan, A., Knopman, J., Tsiouris, A. J., Riina, H. A., & Gobin, Y. P. (2010). Endovascular management of spinal dural arteriovenous fistulas. *Journal of neurointerventional surgery*, jnis-2010
3. James S Harrop, Brian H Kopell, (2016) Vascular Malformations of the Spinal Cord, Medscape, Feb.2016
4. Koch, M. J., Stapleton, C. J., Agarwalla, P. K., Torok, C., Shin, J. H., Coumans, J. V., & Patel, A. B. (2017). Open and endovascular treatment of spinal dural arteriovenous fistulas: a 10-year experience. *Journal of Neurosurgery: Spine*, 26(4), 519-523
5. Grandin C, Duprez T, Stroobandt G, et al. Spinal dural arterio-venous fistula: an underdiagnosed disease? *Acta Neurol Belg* 1997;97:17–21
6. Jellema K, Tijssen CC, Sluzewski M, et al. Spinal dural arteriovenous fistulas: an underdiagnosed disease—a review of patients admitted to the spinal unit of a rehabilitation center. *J Neurol* 2006;253:159–62
7. Sivakumar, W., Zada, G., Yashar, P., Giannotta, S. L., Teitelbaum, G., & Larsen, D. W. (2009). Endovascular management of spinal dural arteriovenous fistulas: a review. *Neurosurgical focus*, 26(5), E15
8. Gilbertson JR, Miller GM, Goldman MS, et al. Spinal dural arteriovenous fistulas: MR and myelographic findings. *AJNR Am J Neuroradiol* 1995;16:2049–57
9. Gemmete, J. J., Chaudhary, N., Elias, A. E., Toma, A. K., Pandey, A. S., Parker, R. A., & Robertson, F. (2013). Spinal dural arteriovenous fistulas: clinical experience with endovascular treatment as a primary therapy at 2 academic referral centers. *American Journal of Neuroradiology*, 34(10), 1974-1979
10. Corkill RA, Mitsos AP, Molyneux AJ. Embolization of spinal intramedullary arteriovenous malformations using the liquid embolic agent, Onyx: a single-center experience in a series of 17 patients. *J Neurosurg Spine* 2007;7:478–85
11. Liu, J., Lv, M., Lv, X., He, H., Liu, A., Qian, Z., & Li, Y. (2014). Curative Glubran 2 Embolization of Cerebral Arteriovenous Malformations Patient Selection and Initial Results. *Interventional Neuroradiology*, 20(6), 722–728
12. Gross, B. A., Albuquerque, F. C., Moon, K., & McDougall, C. G. (2016). Validation of an ‘endovascular-first’ approach to spinal dural arteriovenous fistulas: an intention-to-treat analysis. *Journal of neurointerventional surgery*, neurintsurg2016
13. Gokhale S, Khan SA, McDonagh DL, Britz G. Comparison of surgical and endovascular approach in management of spinal dural arteriovenous fistulas: A single center experience of 27 patients. *Surg Neurol Int* 21-Jan-2014;5:7