Spinal Dural Arteriovenous Fistula, a Rare and Frequently Misdiagnosed Cause of Mielopathy – Case Report and Review of the Literature*

Fístula arteriovenosa dural espinhal, uma causa de mielopatia rara e frequentemente não diagnosticada – Relato de caso e revisão da literatura

Thiago Magalhães de Souza[1] Paulo Gabriel Sacramento da Silva[1] Fellipe de Paula[1] Enrico Ghizoni[1] Andrei Fernandes Joaquim[1]

[1] Division of Neurosurgery, Department of Neurology, Universidade Estadual de Campinas, Campinas, SP, Brazil

Arq Bras Neurocir 2024;43(4):e355–e359.

Address for correspondence Thiago Magalhães de Souza, MD, Division of Neurosurgery, Department of Neurology, Universidade Estadual de Campinas, Rua Flavio Luis Ramos Fontes Cabral, 191, ap.18, Cidade Universitária, Campinas, SP, Brazil (e-mail: souza.tmagalhaes@gmail.com).

Abstract

Spinal dural arteriovenous fistulas (SDAVFs) are rare vascular malformations with unspecific clinical manifestations, which often lead to misdiagnosis and delay in the establishment of effective therapies. Since the neurological prognosis depends on symptom duration and pretreatment neurological status, rapid identification and obliteration of the fistula are key to provide patients with a better quality of life. We herein describe an illustrative case of an elderly patient presenting with progressive myelopathy, with a definitive diagnosis of SDAVF after 18 months of the initial symptoms. We also review the core concepts regarding this condition and discuss strategies to prevent misdiagnosis and worsening of the neurological outcome.

Keywords

- arteriovenous fistulas
- dural
- myelopathy
- spinal cord disease

Resumo

Fístulas arteriovenosas durais espinhais (FADEs) são malformações vasculares raras e com manifestações clínicas pouco específicas, o que frequentemente leva a erros diagnósticos e ao atraso no estabelecimento de terapias efetivas. Considerando que o prognóstico neurológico depende da duração dos sintomas e do *status* neurológico prévio, a agilidade na identificação e obliteração da fístula são essenciais para prover melhor qualidade de vida aos pacientes. Neste estudo, descrevemos um caso ilustrativo de um paciente idoso com quadro de mielopatia progressiva, com diagnóstico definitivo de FADE após 18 meses do início dos sintomas. Realizamos ainda uma revisão sobre os conceitos principais da doença e discutimos estratégias para prevenir atrasos diagnósticos e piora neurológica.

Palavras-chave

- fístulas arteriovenosas
- dural
- mielopatia
- doença da medula espinal

* *Study conducted at the Teaching Hospital of Universidade Estadual de Campinas, Campinas, SP, Brazil.*

received
August 1, 2023
accepted
May 16, 2024

DOI https://doi.org/10.1055/s-0044-1788603.
ISSN 0103-5355.

Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

Introduction

Spinal dural arteriovenous fistulas (SDAVFs) are acquired vascular malformations characterized by anomalous connection of a radicular artery and a medullary vein, with no capillaries between them.[1–3] The higher pressure from the shunt elevates intradural venous pressure, causing venous congestion, which leads to medullary edema and ischemia.[1,2] Chronic hypoxia results in progressive myelopathy.[1,2]

The manifestations of SDAVF myelopathy are unspecific and follow a variable course, potentially delaying diagnosis and proper management.[2] In fact, one study[4] has found that more than 81% of the affected patients were initially misdiagnosed, and 62% were submitted to incorrect treatments. Although the treatment is effective in obliterating the fistula, the prognosis depends on symptom duration and pretreatment neurological status,[1,2] highlighting the importance of an accurate and rapid diagnosis.

We herein describe a well-documented case of SDAVF with an important diagnostic delay, exemplifying the impact of misdiagnosis; we also discuss strategies to prevent misdiagnosis and worsening of the neurological outcome.

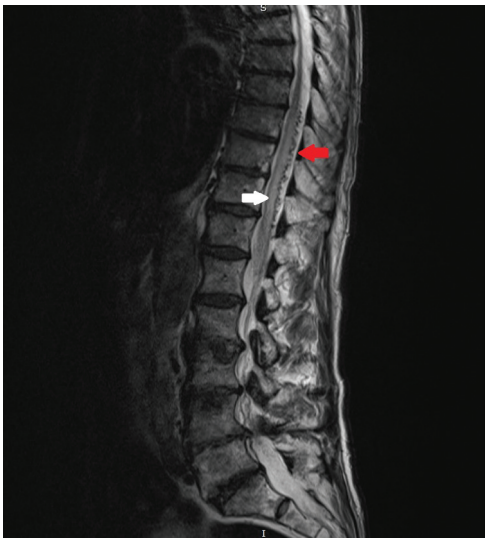

Fig. 1 Typical characteristics of a spinal dural arteriovenous fistula (SDAVF) on magnetic resonance imaging (MRI), as represented by the case herein reported. Images of the T2-weighted sequence: centromedullary edema represented by spinal cord hyperintensity (**white arrow**) and dilated posterior veins shown as serpiginous flow voids (**red arrow**). In the case herein described, alterations were observed from T7 to the conus medullaris.

Case Report

An 80-year-old man presented to our hospital's emergency room complaining of difficulty in walking that had worsened in the past year, with progressive shortening of walkable distance. Since the previous month, he had been unable to ambulate without assistance. Lower mechanical back pain and burning sensation in the lower limbs were associated; the symptoms were more prominent on the left side and were not responsive to a previous therapeutic test with gabapentin. The upper limbs were asymptomatic. Furthermore, he complained of urinary and fecal incontinence that had initiated one year and a half and four months before arriving at our service, respectively. He had a medical history of arterial hypertension, benign prostate hyperplasia, and had had previous diagnoses of syphilis and tuberculosis (both allegedly fully treated). He was also a former smoker and alcoholic. Before reaching our institution, he was first referred to an orthopedic surgeon, who diagnosed degenerative spinal disease, and to a neurologist, who established the syndromic diagnosis of spastic paraparesis and also prescribed baclofen.

After the initial assessment, he was referred to the Neurology Department for further investigation. A neurological examination showed impairment in strength in the lower limbs, almost symmetrical (discreetly worse on the right side). There was also tactile hypoesthesia in the right leg and hypopallesthesia in both lower limbs. The tone was normal.

The main primary hypotheses were neurosyphilis, subacute combined degeneration of the spinal cord, polyneuropathy, and compressive myelopathy. A cerebrospinal fluid examination did not detect any significant pathological change, and the serum levels of B12 were within the normal range. Magnetic resonance imaging (MRI) was performed to evaluate cord compression, and indirect signs of SDAVF were observed: spinal cord edema in the thoracic level as well as flow voids suggestive of a vascular lesion (►**Fig. 1**). Because of the MRI findings, a digital subtraction angiography (DSA) was performed and confirmed a dural fistula from the intercostal arteries entering just below the right pedicle of the T9 level and the radicular vein (►**Fig. 2**).

After discussion with the Neuroradiology Division, we decided to refer the patient to surgical occlusion of the dural fistula. A T9 laminoplasty was performed, with dural opening. The right T9 nerve root was found, and the main artery entering the neural foramen was coagulated—after a brief period of transient clipping. The medullary veins just after artery ligation decreased their flow. The dura mater was

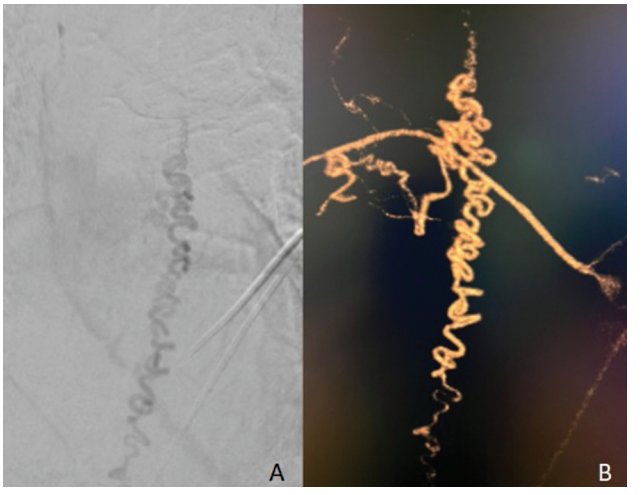

Fig. 2 Digital subtraction angiography (DSA) findings (**A**) and three-dimensional (3D) reconstruction (**B**): SDAVF at the level of T9, draining to a dilated and tortuous posterior medullary vein, with both ascending and descending flow; signals suggestive of venous congestion.

tightly closed, and the T9 lamina was fixed with miniplates. The surgical technique used in the procedure is shown and described in ►**Fig. 3**.

Standard postoperative care was offered, and the patient was in discharge conditions 48 hours after surgery, reporting an immediate improvement in lower-limb strength (from grade 2 proximally just before surgery to grade 3). A postoperative MRI scan showed resolution of the posterior flow void and decrease in the centromedullary hypersignal intensity on the T2-weighted imaging (►**Fig. 4**).

Discussion

Spinal vascular malformations are mainly classified based on anatomic or angioarchitecture features. The first descriptions were made by Di Chiro[7] according to angiographic characteristics, and he divided this group of pathologies into three categories: type I – single-coiled vessel; type II – glomus; and type III – juvenile.[8] With posterior actualizations including a fourth type, intradural perimedullary arteriovenous fistula,[5,6] this classification remains one of the most widely accepted (►**Table 1**).[7,8] More recently, extensive surgical experience gave rise to an anatomical categorization by Spetzler et al.[9] based on the location of the disease and its relation to the spinal cord and the spinal dura mater (►**Table 2**). An endovascular classification by Rodesch et al.[10] is also frequently used when considering endovascular treatment, and it divides these entities into arteriovenous malformations and fistulas, subdividing fistulas into macro and micro types, and adding a genetic classification as hereditary, non-hereditary or single lesions.

Among spinal vascular malformations, SDAVFs are the most common comprehending 60 to 80% of all cases.[2,11] They correspond to Di Chiro's type I or Spetzler et al.'s dorsal

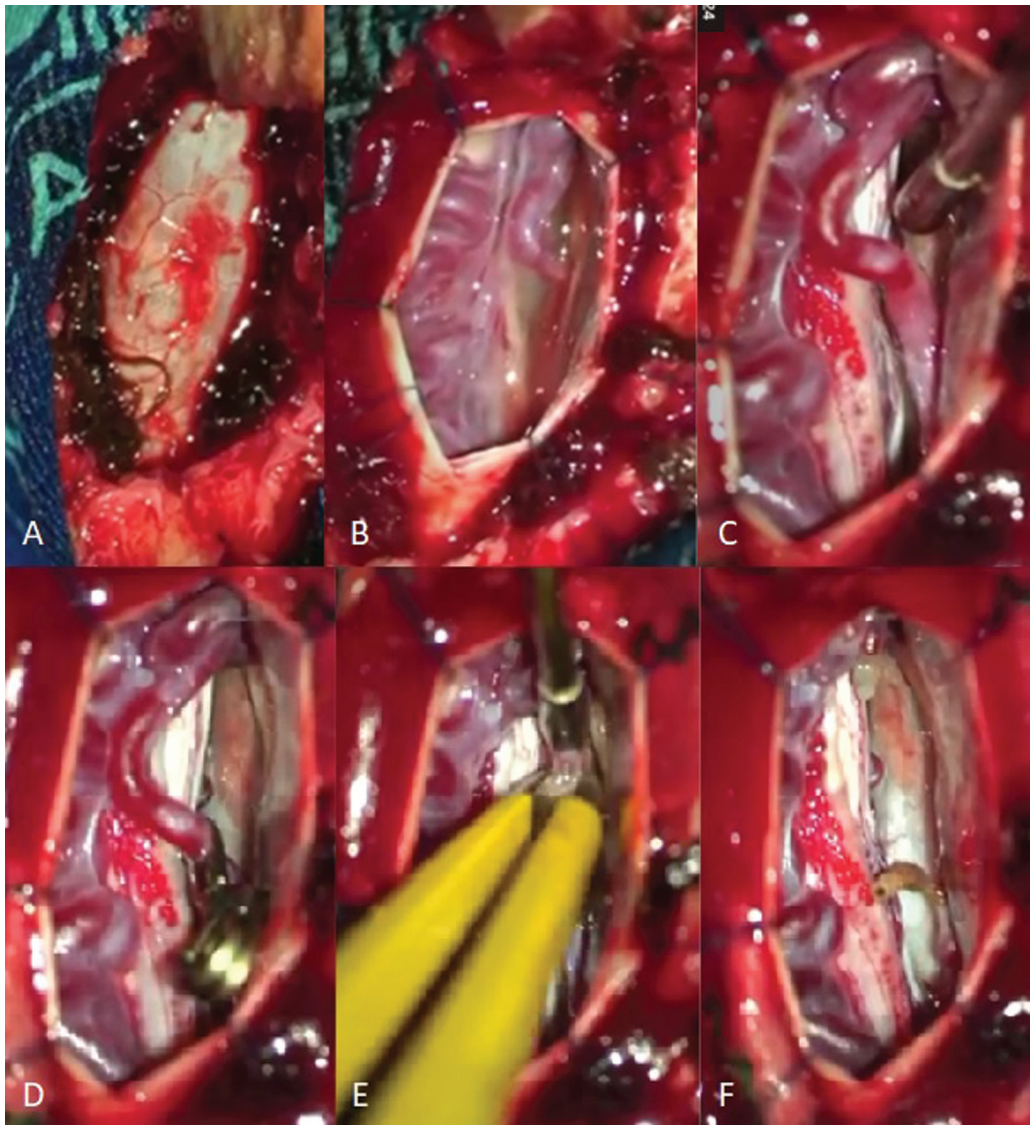

Fig. 3 Surgical approach to SDAVF. (**A**) Standard T9 laminoplasty was performed, and the dura mater was exposed. (**B**) After dural opening, visualization of tortuous, congested posterior venous system. (**C**) After finding the right T9 nerve route, the location of the arterialized draining vein was confirmed, and the vessel, dissected. (**D**) Clipping of the draining vein to prevent bleeding during coagulation. (**E**) Coagulation of the draining vessel with bipolar cautery. (**F**) Final aspect of the procedure. The dura mater was closed watertight, and the T9 vertebra was fixed with miniplates.

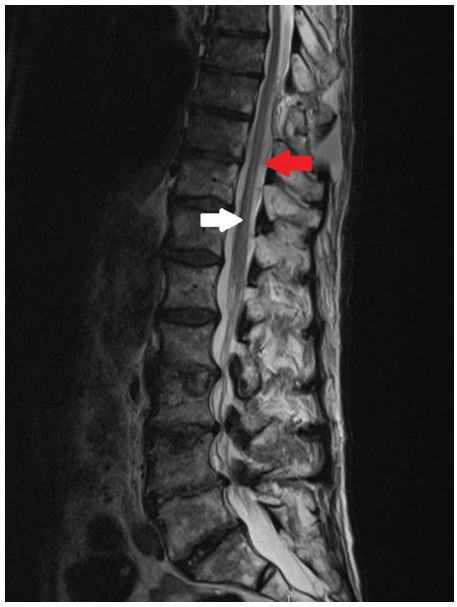

Fig. 4 Postoperative MRI scan acquired on the fourth postoperative day, showing decrease in the centromeddulary hypersignal (**white arrow**) on T2-weighted imaging, indicating improvement in cord edema, and disappearance of posterior flow voids (**red arrow**).

intradural arteriovenous fistulas, which can be further subdivided into subtype A or B, single or multiple arterial feeders respectively.[2,11] The SDAVFs are composed of a radicular artery shunting into a medullary vein through a complex network of arteriovenous microfistulas within the dural nerve root sleeve, with no capillary system interposed.[3,11] This connection raises venous blood pressure, generating venous congestion and cord edema, ultimately leading to myelopathic symptoms.[2] Their etiology is not yet clear.[2,12] Epidemiologically, they more commonly affect men aged approximately 55 to 60 years, with a male:female ratio of up to 6:1.[2,11]

The symptoms manifest as those of progressive myelopathy. A large retrospective study[4] including 326 affected patients reported that the most common findings at the initial presentation were lower-limb motor deficits and sensorial alterations (pain, dysesthesia, paresthesia, and/or hypoesthesia), present in 71.8% and 70.2% of the patients, respectively. With disease progression, weakness and sensory disturbance not only worsened, but were more frequently accompanied by sphincter abnormalities (especially urinary incontinence), shown by 52.5% of patients by the time of diagnosis.[4] Other less prevalent symptoms included headache and vomit—associated with subarachnoid hemorrhage

Table 1 Di Chiro's[7] classification of spinal vascular malformations (modified by Rosenblum et al.[8])

Type	Description/name
I	Dural arteriovenous fistula
II	Intramedullary glomus arteriovenous fistula
III	Intramedullary juvenile arteriovenous fistula
IV	Intradural direct arteriovenous fistula

Table 2 Spetzler et al.'s[9] SDAVF classification

Type	Description/name
AVF	Extradural arteriovenous fistula
	Intradural ventral arteriovenous fistula
	Intradural dorsal arteriovenous fistula
AVM	Extradural-intradural arteriovenous malformation
	Intradural compact arteriovenous malformation
	Intradural diffuse arteriovenous malformation
	Intradural conus arteriovenous malformation

Abbreviations: AVF, arteriovenous fistula; AVM, arteriovenous malformation; SDAVF, spinal dural arteriovenous fistula.

—dizziness, upper-limb weakness, and thoracic pain.[4] Similarly, other studies[12–14] have reported gait disturbances, lower-extremity motor impairment, sensory alterations, sphincter dysfunctions, and pain as the most common initial symptoms. In accordance with the literature, the patient herein described was affected by lower-limb weakness, urinary and fecal incontinence, and pain.

The unspecific nature of the clinical presentation makes SDAVFs a diagnostic challenge, which may lead to diagnostic delay and/or misdiagnosis, favoring unnecessary interventions.[4,13,15–17] Donghai et al.[4] found that most affected patients (81.3%) had at least 1 previous misdiagnosis before reaching the reference center—with the most common ones being degenerative disc disease, myelitis, prostatic hyperplasia, and intramedullary tumors. A substantial amount (19%) remained with a mistaken diagnosis even after evaluation at the reference hospitals—69.3% of whom were submitted to surgical treatment for alternative etiologies.[4] The prognosis worsened after unnecessary medical therapies or procedures.[4] Other studies[13,16,17] have confirmed this tendency towards misdiagnosis and unnecessary invasive treatments. The average diagnostic delay ranges from 12 to 24.6 months,[4,12,14,17–19] but it can reach more than 20 years[4,14] and may be associated with misdiagnosis and worsened prognosis.[13,15,18] Though presenting classical signs of the disease, the patient herein described remained undiagnosed for one and a half years since the first symptom (urinary incontinence). Moreover, the initial diagnostic hypotheses ranged from degenerative diseases to infectious and metabolic disturbances, but SDAVF was not suspected until neuroimaging scans were acquired. The clinical picture was worsening, but, fortunately, no major procedure was performed prior to proper elucidation. Syphilis and vitamin deficiency still have high prevalence rates, especially in low- and middle-income countries,[20,21] and may present as differential diagnoses of myelopathy, both also considered hypotheses for our patient. Providers should have SDAVF in mind as a differential for these diagnoses and rule it out when appropriate.

After the initial suspicion raised on clinical grounds, MRI is usually the first imaging modality performed to guide the diagnosis.[22] The classic signs according to Fox et al.[18] are depicted in ►**Fig. 1**. The definitive diagnosis is ideally based

on DSA, but even this gold standard is not 100% sensitive, and repeating the exam may be necessary if clinical suspicion is high.[4] The DSA is also useful for planning fistula occlusion, especially in terms of precisely locating the fistula level.[2] Noninvasive imaging techniques (such as MRI or computed tomography angiography) may be useful to narrow down the vertebral segments requiring DSA.[4] Segments T6 to L2 are the most affected, being responsible for around 80% of SDAVFs,[2,11,23] as represented by the T9 fistula in the case herein reported.

The treatment is curative, and to date surgery is still considered the gold-standard therapy. Details on the technique used on the patient herein described are available in ►**Fig. 3**. The surgical procedure is relatively simple and safe,[24] and, despite the advances in endovascular embolization, it results in higher occlusion and lower recurrence rates, with better neurological outcomes without adding significant complications or morbidity.[15,24–26] The main pitfall in the surgical treatment is the adequate location of the level of the draining vein,[24] highlighting the importance of careful evaluation of preoperative DSA. After successful obliteration, strength and gait tend to improve at higher rates than urinary function.[13,15,26] Shorter times between onset and adequate treatment are associated with better prognosis[13,15]

In sum, we have described a patient with a typical presentation of an SDAVF, illustrating misdiagnosis, diagnostic delay, worsening of symptoms, and the final resolution with surgical obliteration of the fistula. We hope that, by sharing this case, we will assist in reducing the time until the correct diagnosis and institution of treatment, contributing to minimize the neurological impairments caused by the disease.

References

1 Day AL, Turkmani AH, Chen PR. Spinal arteriovenous fistulae: surgical management [Internet]. 1st ed. Vol. 143;Handbook of Clinical Neurology Elsevier B.V.; 2017:189–198. Available from: http://dx.doi.org/10.1016/B978-0-444-63640-9.00018-7

2 Maimon S, Luckman Y, Strauss I. Spinal Dural Arteriovenous Fistula: A Review. Adv Tech Stand Neurosurg 2016;43(43):111–137

3 McCutcheon IE, Doppman JL, Oldfield EH. Microvascular anatomy of dural arteriovenous abnormalities of the spine: a microangiographic study. J Neurosurg 1996;84(02):215–220

4 Donghai W, Ning Y, Peng Z, et al. The diagnosis of spinal dural arteriovenous fistulas. Spine 2013;38(09):E546–E553

5 Ehresman J, Catapano JS, Baranoski JF, Jadhav AP, Ducruet AF, Albuquerque FC. Treatment of Spinal Arteriovenous Malformation and Fistula. Neurosurg Clin N Am 2022;33(02):193–206. Doi: 10.1016/j.nec.2021.11.005 [Internet]

6 Takai K. Spinal arteriovenous shunts: Angioarchitecture and historical changes in classification. Neurol Med Chir (Tokyo) 2017;57(07):356–365

7 Di Chiro G. Angiography of obstructive vascular disease of the spinal cord. Radiology 1971;100(03):607–614

8 Rosenblum B, Oldfield EH, Doppman JL, Di Chiro G. Spinal arteriovenous malformations: a comparison of dural arteriovenous fistulas and intradural AVM's in 81 patients. J Neurosurg 1987;67(06):795–802

9 Spetzler RF, Detwiler PW, Riina HA, Porter RW. Modified classification of spinal cord vascular lesions. J Neurosurg 2002;96;(2, Suppl)145–156

10 Rodesch G, Hurth M, Alvarez H, et al. Classification of spinal cord arteriovenous shunts: proposal for a reappraisal—the Bicêtre experience with 155 consecutive patients treated between 1981 and 1999. Neurosurgery 2002;51(02):374–379

11 Kona MP, Buch K, Singh J, Rohatgi S. Spinal vascular shunts: A patterned approach. AJNR Am J Neuroradiol 2021;42(12): 2110–2118

12 Jellema K, Tijssen CC, van Gijn J. Spinal dural arteriovenous fistulas: a congestive myelopathy that initially mimics a peripheral nerve disorder. Brain 2006;129(Pt 12):3150–3164

13 Ronald AA, Yao B, Winkelman RD, Piraino D, Masaryk TJ, Krishnaney AA. Spinal Dural Arteriovenous Fistula: Diagnosis, Outcomes, and Prognostic Factors. World Neurosurg 2020;144: e306–e315. Doi: 10.1016/j.wneu.2020.08.126 [Internet]

14 Muralidharan R, Saladino A, Lanzino G, Atkinson JL, Rabinstein AA. The clinical and radiological presentation of spinal dural arteriovenous fistula. Spine 2011;36(25):E1641–E1647

15 Takai K, Endo T, Yasuhara T, et al. Neurosurgical versus endovascular treatment of spinal dural arteriovenous fistulas: a multicenter study of 195 patients. J Neurosurg Spine 2020;34(03):514–521

16 Saladino A, Atkinson JLD, Rabinstein AA, et al. Surgical treatment of spinal dural arteriovenous fistulae: a consecutive series of 154 patients. Neurosurgery 2010;67(05):1350–1357, discussion 1357–1358

17 Yang B, Lu T, He X, Li H. Spinal dural arteriovenous fistula: A rare but treatable disease that should not be missed by orthopedic surgeons. Front Neurol 2022;13:938342

18 Fox S, Hnenny L, Ahmed U, Meguro K, Kelly ME. Spinal dural arteriovenous fistula: a case series and review of imaging findings. Spinal Cord Ser Cases 2017;3(01):17024

19 Gemmete JJ, Chaudhary N, Elias AE, et al. Spinal dural arteriovenous fistulas: clinical experience with endovascular treatment as a primary therapy at 2 academic referral centers. AJNR Am J Neuroradiol 2013;34(10):1974–1979

20 Dowd EC, Frank MJ, Collins A, Goldd JM, Barch DM. Kuschner. HHS Public Access. Physiol Behav 2017;176(03):139–148. Available from https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3856065/pdf/ijms-14-22274.pdf%0Ahttp://www.mdpi.com/1422-0067/14/11/22274 [Internet]

21 Jiang J, Trundle P, Ren J, et al. We are IntechOpen, the world ' s leading publisher of Open Access books Built by scientists, for scientists TOP 1%. Intech [Internet] 2010;34(08):57–67. Doi: 10.1007/s12559-021-09926-6%0A. https://www.intechopen.com/books/advanced-biometric-technologies/liveness-detection-in-biometrics%0Ahttp://dx.doi.org/10.1016/j.compmedimag.2010.07.003

22 Condette-Auliac S, Boulin A, Roccatagliata L, et al. MRI and MRA of spinal cord arteriovenous shunts. J Magn Reson Imaging 2014;40 (06):1253–1266

23 Durnford AJ, Hempenstall J, Sadek AR, et al. Degree and Duration of Functional Improvement on Long-Term Follow-Up of Spinal Dural Arteriovenous Fistulae Occluded by Endovascular and Surgical Treatment. World Neurosurg 2017;107:488–494. Doi: 10.1016/j.wneu.2017.07.140 [Internet]

24 Bretonnier M, Hénaux PL, Gaberel T, et al. Spinal Dural Arteriovenous Fistulas: Clinical Outcome After Surgery Versus Embolization: A Retrospective Study. World Neurosurg 2019;127: e943–e949

25 Goyal A, Cesare J, Lu VM, et al. Outcomes following surgical versus endovascular treatment of spinal dural arteriovenous fistula: a systematic review and meta-analysis. J Neurol Neurosurg Psychiatry 2019;90(10):1139–1146

26 Steinmetz MP, Chow MM, Krishnaney AA, et al. Outcome after the treatment of spinal dural arteriovenous fistulae: a contemporary single-institution series and meta-analysis. Neurosurgery 2004; 55(01):77–87, discussion 87–88

