*CASE REPORT*

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# A rare case of spinal dural arteriovenous fistula in a 22-year-old farmer

**Sara Kierońska-Siwak[1,2,A-B,D,F], Natalia Rulewska[3,C], Jan Klimowicz[3,A], Grzegorz Meder[4,E], Milena Świtońska[5,E], Paweł Sokal[1,2,F]**

[1] *Department of Neurosurgery, Functional and Stereotactic Neurosurgery , Collegium Medicum, Nicolaus Copernicus University, Bydgoszcz, Poland*
[2] *Department of Neurosurgery and Neurology, Jan Biziel University Hospital No 2, Collegium Medicum, Nicolaus Copernicus University, Bydgoszcz, Poland*
[3] *Students' Scientific Circle, Department of Neurosurgery and Neurology, Jan Biziel University Hospital No 2, Collegium Medicum, Nicolaus Copernicus University, Bydgoszcz, Poland*
[4] *Department of Radiology, Jan Biziel University Hospital No. 2, Collegium Medicum Nicolaus Copernicus University, Bydgoszcz, Poland*
[5] *Department of Neurology and Clinical Neurophysiology, Collegium Medicum, Nicolaus Copernicus University, Bydgoszcz, Poland*
*A – Research concept and design, B – Collection and/or assembly of data, C – Data analysis and interpretation, D – Writing the article, E – Critical revision of the article, F – Final approval of the article*

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## Abstract

Spinal Dural Arteriovenous Fistulas (DAVFs) are rare vascular malformations characterized by abnormal connections between a spinal artery and venous plexus. Though uncommon, DAVFs can be misdiagnosed due to their presentation with non-specific symptoms, leading to potential treatment delays or incorrect procedures. This case report presents a 22-year-old male farmer admitted with gradual lower limb muscle weakness and urinary retention. Despite no trauma history, his recent exposure to chemicals raised concerns. Neurological examination revealed significant mobility impairment, particularly in the left leg, alongside sensory deficits. MRI findings demonstrated spinal cord oedema and venous dilatation, suggesting DAVF. The patient underwent successful endovascular embolization, with a post-operative course free of complications. This case highlights the challenges in diagnosing DAVF and underscores the importance of timely intervention.

## Key words

spinal fistula, sDAVF, embolisation

## INTRODUCTION

Spinal Dural Arteriovenous Fistulas (DAVFs) occur when an abnormal connection forms between a spinal artery and a venous plexus without capillaries in between. Foix and Alajouanine were the first to report on the presence of spinal DAVF in 1926. Nevertheless, there remains disagreement regarding how to categorize and name the spinal arteriovenous lesions [1]. Different terms have been used to refer to spinal vascular abnormalities, which have been categorized based on clinical characteristics, anatomical location, and their blood vessel structure [2]. The first classification of spinal AV shunts was described in 1971 by Di Chiro et al., and distinguished type I (single coiled vessel), type II (glomus), and type III (juvenile). The classification was changed several times until in 1987 Rosenblum introduced the most commonly accepted categorization, including type I (dural AVF), type II (intradural intramedullary glomus AVM), type III (intradural intramedullary juvenile AVM), and type IV (intradural direct AVF). Takai recently introduced an updated edition of the earlier categorizations [3]. Types of spinal AVF was presented in Table 1.

**Table 1.** Classification of spinal AV shunts by Takai

| Type | Academic name | Subtype | Feeder, AVF, and venous drainage |
|---|---|---|---|
| I | Dural AVF | | |
| II | Intramedullary glomus AVM | | |
| III | Intramedullary juvenile AVM | | |
| IV | Perimedullary AVF | IVa | A single feeder and small AVF |
| | | IVb | Multiple feeders and medium AVFs |
| | | IVc | Multiple feeders and a giant AVF |
| V | Extradural AVF | Va | w intradural venous drainage |
| | | Vb | w/o intradural venous drainage |

Spinal AVF are the most prevalent type of spinal cord vascular malformation, comprising 50% – 85% of all spinal vascular lesions. The incidence of DAVFs is estimated to be 0.5 – 1 per 100,000 individuals per year. Despite that, they remain rare and often misdiagnosed as medullary tumour, acute or chronic inflammatory demyelinating polyneuropathy, sensory polyneuropathy and degenerative disc disease, with as many as 22% of patients receiving avoidable laminectomy or other procedure, with a deterioration in symptoms1 The lesions typically occur in the thoracolumbar region of the spine as a single lesion emerging from vertebral segmental arteries at the level of T6-L2 (>80% cases of DVAFs)4. According to available data, it is believed that Spinal DAVFs are acquired lesions and tend to impact middle-

✉ Address for correspondence: Sara Kierońska-Siwak, Department of Neurosurgery, Functional and Stereotactic Neurosurgery Collegium Medicum, Nicolaus Copernicus University, 85-168 Bydgoszcz, Poland
E-mail: sara.kieronska@gmail.com

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aged men. In most series, at least 80% of the patients are male [5]. The age distribution of SDAVF typically occurs in the fifth or sixth decade, with below 1% cases occurring below 30 years old [9]. Specific etiological factors of spinal Dural arteriovenous fistula remains uncertain, trauma is not regarded as significant underlying cause. SDAVFs commonly have slow and progressive onset with non-specific symptom, including gait disturbances, paresthesia, sensory loss and lower extremity weakness (32–81% of patients), radicular pain affecting one or both lower extremities, lower back pain without radicular distribution, with roughly 5% individuals presenting acute nature of symptoms [2, 6, 7]. Severe neurogenic bladder, erectile dysfunction, as well as bowel and bladder incontinence, have been linked to later stages of the disease, and there are concerns of reduced amelioration of symptoms after treatment. Moreover, due to character of symptoms it takes an estimated 15 – 18 months or more from the onset to diagnosis [8–10].

CT angiography is considered an effective way of localizing the lesion [9], while MRI continues to be the primary method for assessing patients with such symptoms as motor and sensory disturbances in the lower limbs and myelopathy. Hyperintensity on T-2 weighted MRI images corresponding with spinal cord oedema is almost completely sensitive for SDAVF, but these findings are non-specific. T2-weighted imaging can also reveal flow voids corresponding to dilated intradural veins which are more accurate determinations of Dural AFV [11].

Spinal catheter angiography remains the optimal method of diagnosis thanks to more accuracy compared to MR angiography. It is worth noting that cases of SDAVF presenting cord oedema distant from the location of AV shunt lesion localization may be particularly arduous and complex7. Treatment options for Dural AVF includes surgical disconnection, with one of the potential complications being leakage of cerebrospinal fluid. In addition to surgical disconnection, endovascular embolization is another commonly used method. In instances of relapse following embolization, a combination of treatment modalities may be used [12].

## CASE REPORT

A 22-year-old male farmer working on the harvesting of grain and seasonal fruits was admitted to the Department of Neurosurgery, University Hospital No. 2 in Bydgoszcz, Poland, due to gradual muscle weakness of the lower limbs with urinary retention lasting 24 hours. The patient denied the occurrence of the injury. However, for the past 2 months, he has been working with chemicals, fertilizing crops. On admission, the neurological examination showed muscle bulk appearing to be symmetrical – there was no atrophy and fasciculations in the lower limbs. Deep reflex examination revealed weakness of the deep tendon knee jerk in both legs. By assessing the patient's muscle strength according to the Lovett scale, pronounced mobility impairment was demonstrated, and the patient was unable to overcome gravity while moving in the left leg (⅖ Lovett points). Examination of the right limb also revealed that the patient had slightly impaired mobility and overcoming gravity while moving (⅗ Lovett points). Flaccid paresis was especially expressed in the quadriceps and biceps femoris muscles, more pronounced in the left limb. In addition, the disturbances of superficial and deep sensation below the level of the iliac spines was found in both legs. No other neurologic deficits were identified.

Laboratory tests revealed leukocytosis (WBC 13.63 G/l) with left shift (PMNs→ NEUT 73.6%) and monocytosis (MON 1.6G/l.) Other results: electrolyte panel, clotting times, and inflammatory markers were within normal limits.

The lumbosacral MRI was performed in T1, T2-weighted sequences and the STIR in sagittal, coronal, transverse projections and in the SPACE option with a secondary 3D reconstruction. The contrast was given. At the level of Th12-L1, the dilated spinal cord was visible. Areas of increased signal corresponding to oedema and ischemic changes within it. In the spinal canal section from Th12 – L5, features of circulatory stasis, the presence of dilated and tortuous venous vessels partially penetrating into the medulla, were observed (Fig 1). The fistula is most probably supplied from left intercostal arteries at the level of L3-L4. The height and the signal of vertebral bodies were normal on the examination.

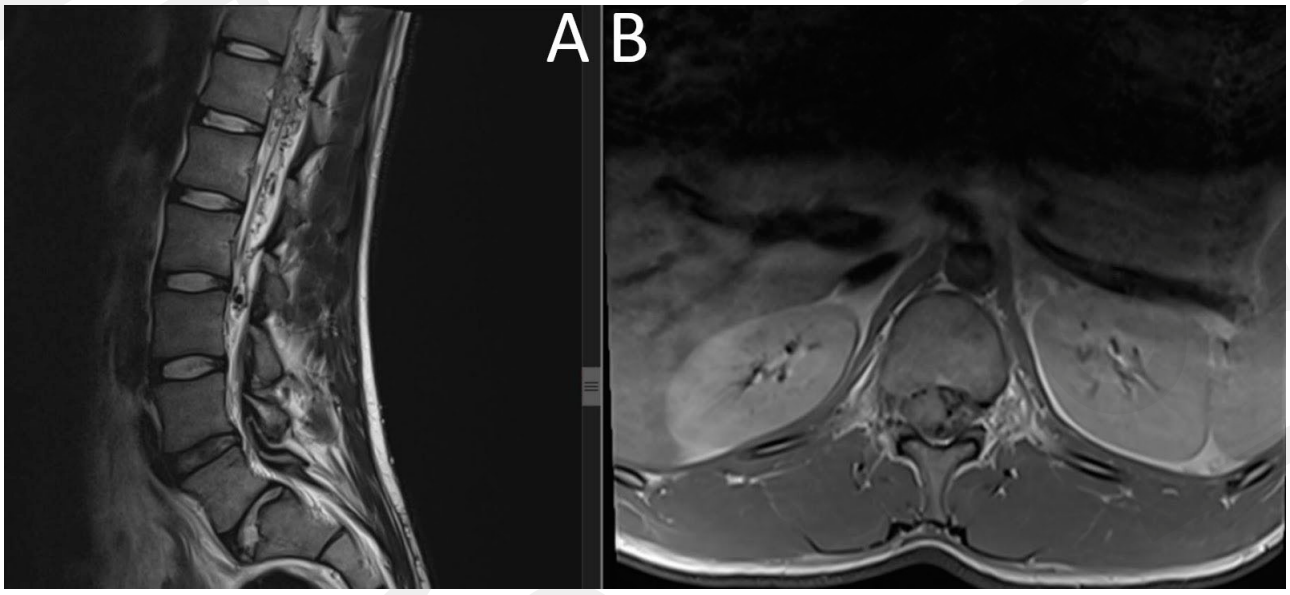


**Figure 1.** MRI of the Th and L/S spine on admission, T2 sequence with image of dAVF in sagittal plane (A) and axial plane (B)

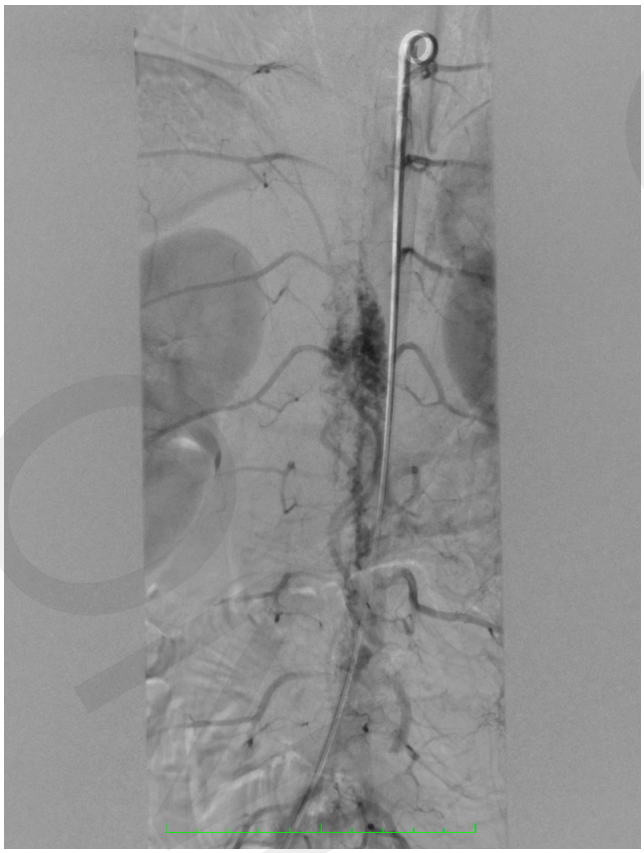

**Figure 2.** General aortography revealing high-flow spinal dAVF in lumbar region.

The patient was qualified for immediate endovascular embolisation of the fistula and nt signed informed consent for the procedure. The procedure of embolization was performed from the femoral approach and under general anesthesia. At the beginning, non-selective aortography was performed which revealed typical dAVF features, with arterial feeders located in the lumbar area. In the next step, selective angiography of the lumbar arteries was performed and a single arterial feeder was located at the level of the left 3rd lumbar artery. The final step consisted of super selective catheterization of the feeding artery, close to the fistula with the microcathether (Apollo – Medtronic Minneapolis, USA) and injection of ethylene vinyl alcohol (EVOH) copolymer – (ONYX – Medtronic Minneapolis, USA) to occlude: arterial feeder, fistula and proximal venous outflow (Fig. 2 – 5).

The post-operative course was uncomplicated. In the postoperative period until discharge from the hospital, intravenous hydration, analgesics, anticoagulants, anti-oedemas, as well as a proton pump inhibitor, were administered.

**Follow up.** During hospitalization, the patient's neurological condition improved, during which a significant improvement in the motor functions of the lower limbs was observed. On the 7th day after the procedure, MRI of TH and L/S was repeated (Fig 6), neurological superficial and deep sensory disturbances were reduced, mainly on the anterior surfaces of the thighs on both sides. A reduction in lower limb paresis was also observed.

The patient was discharged on the 7th day after embolization to the rehabilitation department with paresis

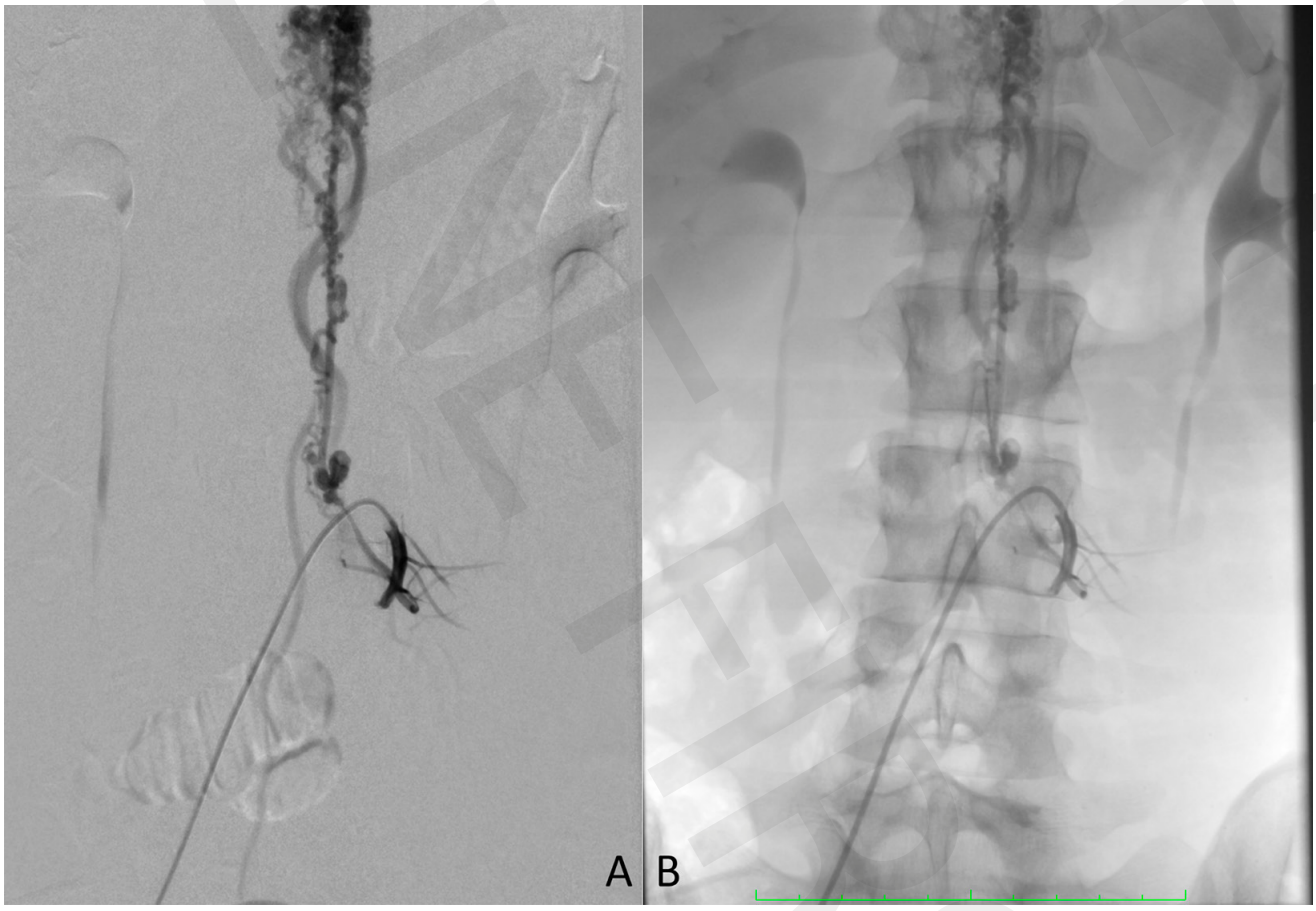


**Figure 3.** Selective angiography from left 3rd lumbar artery confirming origin of the fistula (A unsubtracted and B 3 subtracted image)

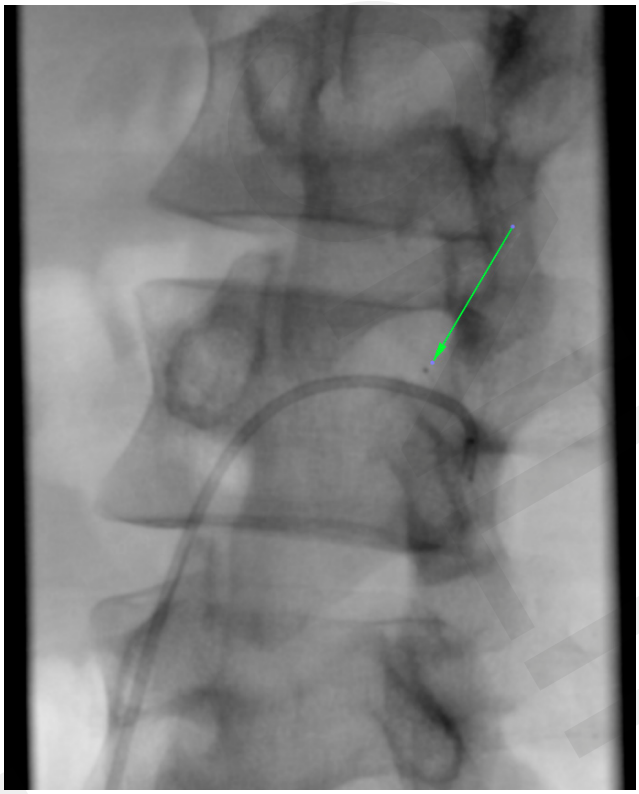

**Figure 4.** Microcathaterer tip (green arrow) placed in the fistula arterial feeder

of the left lower limb – ⅗ Lovett points and the right lower limb – ⅘ Lovett points, with the following recommendations: physical rehabilitation for at least 3 months, MRI of the Th and L/S spine in 6 months.

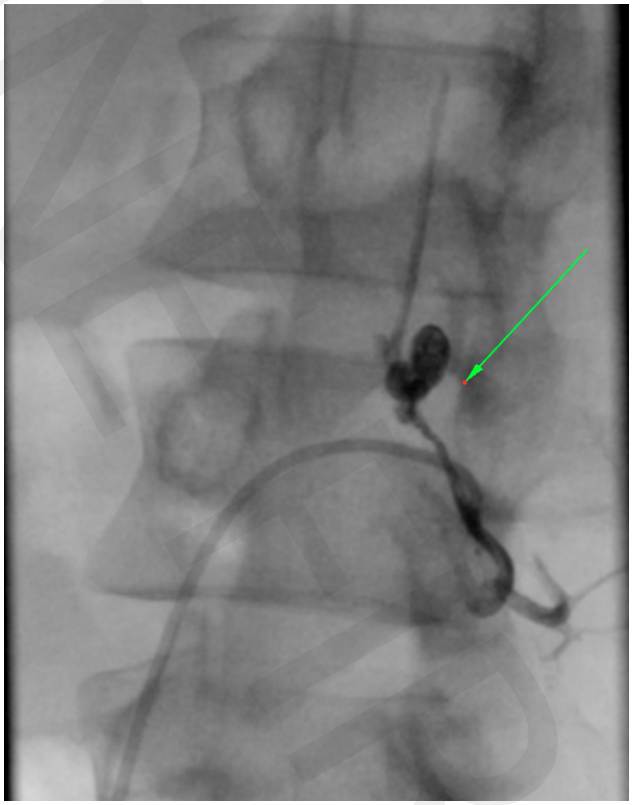

**Figure 5.** Cast of Onyx in the arterial feeder, fistula and proximal part of the draining vein

## DISCUSSION

SDAVFs are the most frequent vascular malformation of the spine, account for 50% – 85% of all spinal vascular lesions. However, the disease seems to be under-diagnosed. The characteristic features of SDAVF are: progression of the area of sensory disturbances cranially to the sacral area and asymmetric distribution of symptoms and rare involvement of the upper limbs, possible when the fistula is present in the cervical spine, but location in the cervical spine is considered exceedingly rare and has been reported most often in the context of subarachnoid haemorrhage and myelopathy1. The other clinical manifestations of the SDAVF in the early stage often include paresthesia, weakness, numbness, additional bowel problems, gait disturbance, and diffuse or patchy sensory loss, which gradually progress in the course of months to years [13].

The symptoms are not specific to this disease and are very similar to the clinical manifestations of polyneuropathy, degenerative spine diseases or proliferative disease; thus the median time to diagnosis ranges from 12 – 44 months [14]. The reason for this delay may be a frequently overlooked diagnosis of SDAVF because of the rareness of the disease [8].

In the discussed patient, efficient and quick diagnostics enabled early determination of the etiology of the disease and implementation of appropriate treatment, which prevented further progression of symptoms. MRI is the first step in the imaging test that helps detect SDAVF [15, 16]. The diagnosis is confirmed by a triad of findings on routine MRI that is present in > 95% of cases: 1) spinal cord oedema (hyperintense signal on T2-weighted images); 2) enlarged pial flow voids around the spinal cord (flow voids on T2-weighted images); 3) disruption of the blood-brain barrier (cord enhancement on T1-weighted images after contrast injection) [17, 18]. The gold standard for the diagnosis is selective spinal angiography because it allows to locate the lesion well, but also can guide its obliteration and elimination [8].

Implementation of appropriate treatment, in the form of surgical disconnection or endovascular occlusion of the meningeal fistula, is the only option to stop progression of the disease, additionally relieving the symptoms in most patients. The choice of fistula closure method, between surgical and endovascular, depends mainly on local anatomical conditions and the individual experience of the treatment centre, because DSA is an invasive procedure requiring a high level of expertise [11].

The traditional treatment of sDAVF involves a haemilaminectomy, opening the dura and coagulation or clipping of the vein [5, 14, 17]. A recent review of the literature published by Zanin et al. describes the failure rate as being significantly higher for endovascular therapy (20%) compared to surgery (4%). Neurological complications were generally rare, with similar rates among the 2 groups (endovascular 2.9%; surgery 2.6%). Endovascular treatment showed a statistically significantly higher rate of persistent neurological complications than surgical treatment (2.9% versus 0.2%; $p < 0.01$). There was no significant difference in the effectiveness of both treatments in enhancing clinical outcomes [5, 14, 17].

Traditional treatment, being an open surgery, is associated with several procedural complications including epidural haematoma, CSF leak or infection. However, surgical treatment is still the preferred method in patients with arterial dissection or severe atherosclerosis [18].

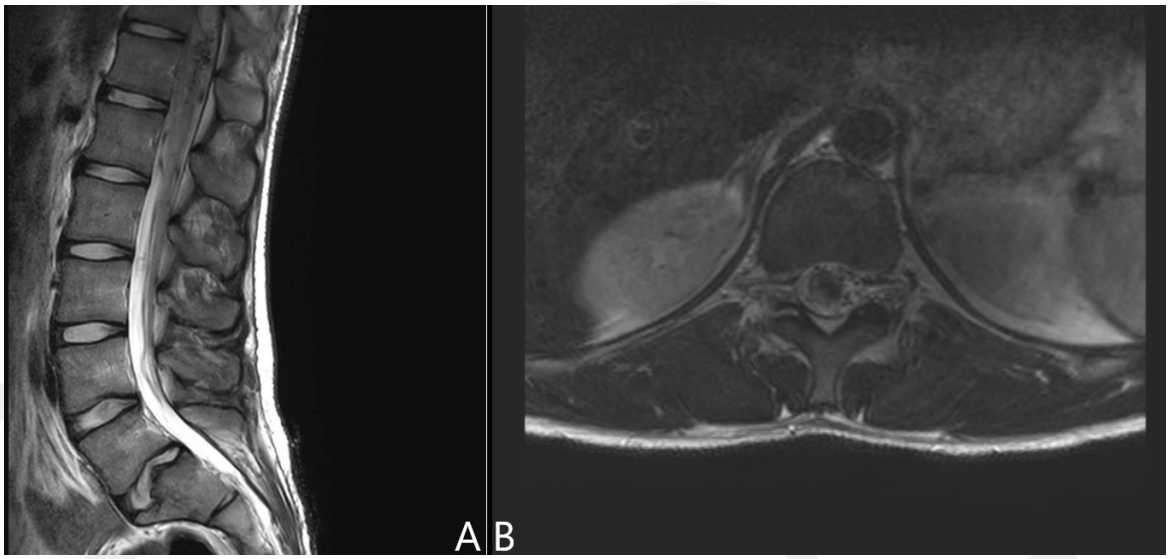


**Figure 6.** MRI of the Th and L/S spine in 7 days after embolisation in sagittal plane (A) and axial plane (B)

An alternative therapeutic strategy for patients with SDAVF is minimally invasive endovascular embolisation. For embolization, N-butyl 2-cyanoacrylate (NBCA) can be used, which is commonly referred to as as glue or Ethylene vinyl alcohol (Onyx). This is a liquid embolic material dissolved in dimethyl sulfoxide that polymerizes soon after contact with the bloodstream [14, 17].

Szmygin et al. showed that embolization using Onyx is a minimally invasive procedure, and its high effectiveness combined with a low rate of procedural complications is similar to that in surgical studies [19].

Bretonnier et al., in multicentre study comparing 2 techniques – endovascular and surgical – did not find significant differences in overall clinical outcomes, but concluded that patients undergoing embolisation have a higher risk of late recurrence (endovascular 21.4 %; surgery 9.1 %; P = 0.28), and the initial occlusion rate was in favour of surgery, with 91.3 % versus 70 % for endovascular treatment (P = 0.050) [10].

## CONCLUSIONS

Spinal dural arteriovenous fistula is an important spinal vascular pathology but frequently under-diagnosed since the clinical presentations resemble other more common causes of myelopathy. Even though there are no precise guidelines for treatment, endovascular embolization and surgery have been proven to be effective. Timely identification and intervention play a crucial role in determining the outcome for the patients.

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