Spinal Dural Arteriovenous Fistulae – A Diagnostic Challenge

Nirit Lev MD[1], Shimon Maimon MD[2], Zvi H. Rappaport MD[3] and Eldad Melamed MD[1]

Departments of [1]Neurology, [2]Invasive Radiology and [3]Neurosurgery, Rabin Medical Center (Beilinson Campus), Petah Tiqva, Israel

Key words: spinal cord, dura mater, arteriovenous fistula, vascular malformation, myelopathy, angiography

Abstract

Background: Spinal dural arteriovenous fistulae comprise the majority of spinal vascular malformations. The most common clinical presentation is that of progressive myeloradiculopathy, probably related to venous hypertension, which may lead to permanent disability and even death.

Objective: To report our clinical experience with spinal dural arteriovenous fistulae.

Methods: Nine patients with spinal dural AVF were managed at our center during a one year period (1998–1999). The patients, eight men and one woman ranging in age from 46 to 75 years, presented with initially fluctuating and eventually permanent and progressive paraparesis, sensory disturbances and sphincter dysfunction. The neurological signs generally began symmetrically and progressed from the distal to proximal limb regions. The duration of symptoms before diagnosis ranged from 6 to 36 months during which the patients underwent an extensive but fruitless work-up and even unnecessary operations due to misdiagnosis. All patients finally underwent magnetic resonance imaging and spinal angiography, which demonstrated the pathological vascular fistula. Interruption of the AVF was achieved by embolization or by surgical resection.

Results: Following treatment, six patients experienced improvement of gait and sphincter control, and the severe neurological deficits stabilized in the other three patients with long duration of illness. There was no further deterioration in any of the treated patients.

Conclusions: The history, neurological findings and radiological changes on MRI scan should alert clinicians to the possibility of spinal dural AVF, leading to diagnostic spinal angiography. Early diagnosis and treatment may significantly improve outcome and prevent permanent disability and even mortality.

IMAJ 2001;3:492–496

Spinal cord arteriovenous malformations represent a heterogeneous group of vascular anomalies. These lesions have been classified into four types: Type I – spinal dural arteriovenous fistulae, type II – intramedullary glomus malformations, type III – extensive juvenile malformations, and type IV – intradural perimedullary arteriovenous fistulae [1]. The distinction between these arteriovenous malformations is based on the location of the malformation and the vessels involved. Thus for example, type I lesions are fistulae between a segmental dural artery and an intradural vein, while types II and IV are usually fed by the anterior spinal artery. Type II lesions are intramedullary and located usually in the cervical cord, while type IV are intradural extramedullary and are most commonly located in the conus medullaris or cauda equina. Type III lesions are complex lesions and compromise intramedullary, extramedullary and even extraspinal components.

Spinal dural AVFs (type I) are the most common spinal arteriovenous malformation [2]. Typically, these lesions are located in the dura mater around the sensory ganglion of the proximal nerve root. These fistulae represent an abnormal connection between the segmental dural arterial supply of the nerve root sleeve and the underlying medullary vein [3]. The venous drainage of spinal dural AVFs is usually retrograde toward the spinal cord via the medullary vein. These low flow, high pressure lesions cause venous hypertension and spinal cord congestion [1], resulting in progressive myelopathy. In these patients, if the fistulae are interrupted and venous hypertension is relieved before an irreversible injury, reversal of myelopathy is possible [4–6].

Due to lack of awareness among clinicians these lesions are under-diagnosed, leading to severe disability and even death. With the emergence of better diagnostic techniques this disorder can be identified and treated efficiently.

Nine cases of spinal dural AVFs were managed at our center. Three of them are discussed here in detail and six are briefly presented in a table, in order to familiarize physicians and surgeons with this elusive, yet treatable neurological entity.

Material and Methods

Nine patients with spinal dural AVFs were managed at our center during a one year period (1998–1999). The clinical and angiographic data are outlined in Table 1. All the patients, eight men and one woman with an age range of 46–75 years, presented with progressive myelopathy resulting in different degrees of disability. They had progressive paraparesis, sensory disturbances, and eventually bladder and bowel dysfunction. Symptoms were initially fluctuating and later became permanent. The neurological signs generally began symmetrically and

AVF = arteriovenous fistulae

Table 1. Summary of the clinical data

No.	Age	Gender	Presentation	Duration	Location	Treatment	Complications	Follow-up	Outcome
1	63	F	Paraplegia, L1 sensory level, incontinence	30 months	Rt. D9	Surgery	UTI	11 months	Stabilized
2	46	M	Paraparesis, sensory disturbances	24 months	Lt. D10	Surgery	Radicular pain, dysesthesias	12 months	Improved
3	75	M	Paraparesis, L1 sensory level, urinary retention	5 months	Rt. L2-3	Surgery	Low back pain	9 months	Improved
4	53	M	Severe paraparesis, D12 sensory level, loss of sphincter control	9 months	? Low thoracic	Surgery	None	8 months	Stabilized
5	62	M	Paraparesis, sensory disturbances, loss of sphincter control	6 months	Lt. D6	Embolization surgery	None	8 months	Improved
6	63	M	Paraparesis, L2 sensory level, urinary disturbances*	36 months	Lt. D7	Embolization	None	4 months	Improved
7	52	M	Paraparesis, D11 sensory level, loss of sphincter control	6 months	Rt. D9	Embolization	DVT, uncontrolled diabetes	10 months	Improved
8	64	M	Paraparesis, sensory disturbances	8 months	Lt. L1	Surgery	UTI	12 months	Improved
9	60	M	Paraparesis, D12 sensory level	24 months	Lt. D12	Embolization	None	2 months	Mild improvement

* See Figure 2.
DVT = deep vein thrombosis, UTI = urinary tract infection

progressed from the distal to proximal limb regions. Presenting symptoms predated diagnosis by 6–36 months, during which the patients underwent an extensive work-up that included several neurological, orthopedic and urological examinations, imaging by computed tomography and MRI and even unnecessary operations due to misdiagnosis. A high index of suspicion and accumulating experience led us to suspect and pursue the diagnosis of this disorder in these patients in view of the clinical and radiological findings. Suggestive findings on MRI included increased spinal cord signal on T2-weighted images and decreased signal on T1-weighted images, swelling of the cord, cord enhancement, and prominent intradural vessels with flow voids [Figure 1A and B].

Selective spinal angiography was conducted in all the patients as part of their diagnostic work-up [Figure 2]. Four patients underwent embolic interruption of the fistulae, one of whom required subsequent surgical treatment due to recurrence. Five patients were treated by direct surgery through a posterior laminectomy. Pathological examination of the surgically resected material demonstrated blood vessels. There were none or

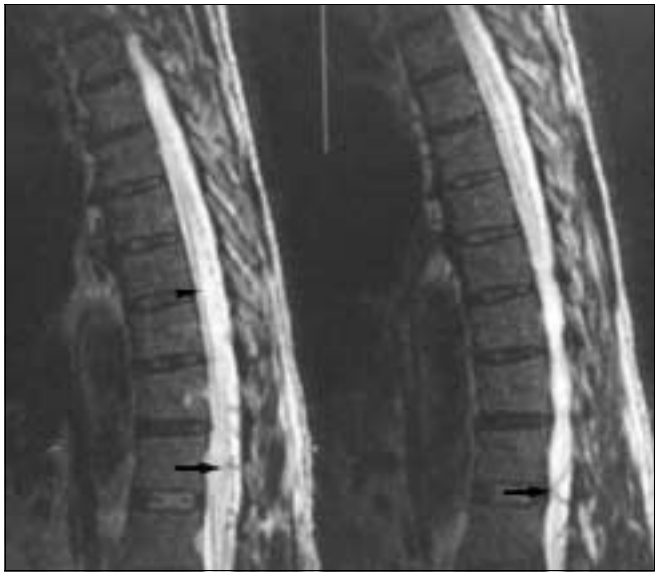

Figure 1A. MRI of the thoracic spine, T2-weighted sagital view showing high intensity of the thoracic spinal cord (arrowhead), with multiple flow voids (arrows) that represent the abnormal blood vessels.

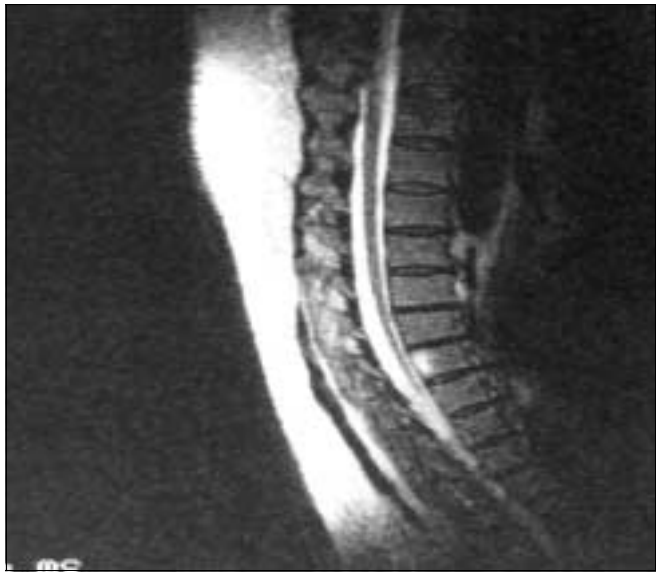

Figure 1B. MRI of a normal thoracic spine, T2-weighted sagittal view. Comparing the normal spinal image to the pathologic one, no increased signal of the spinal cord nor flow voids are seen.

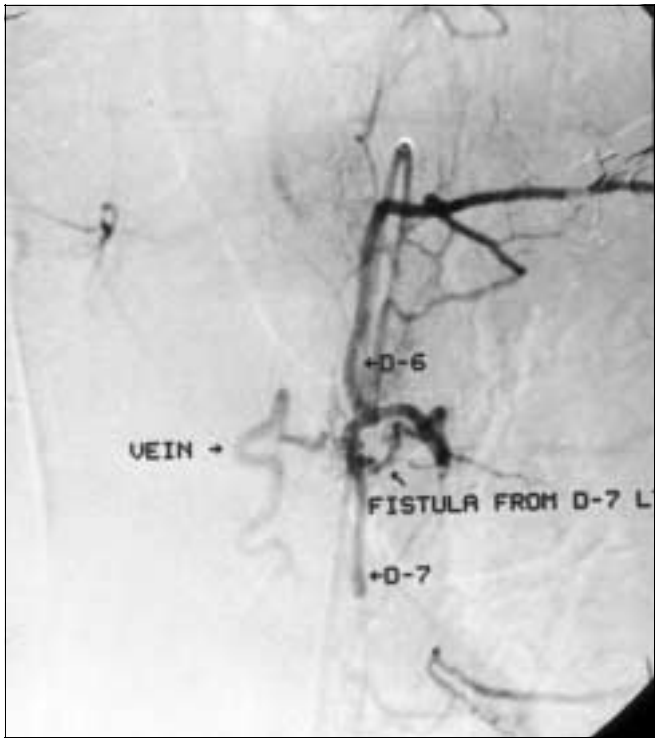

Figure 2. Spinal angiography. Injection of contrast to D6 intercostal artery filling also D7. A dural branch arising from the D7 intercostal artery supplies a network of the arteriovenous fistula and a dilated medullary draining vein.

only minor surgical complications, including low back pain, wound-healing problems and urinary tract infections. There were no complications related to the spinal angiography or endovascular embolization, except for a deep vein thrombosis that was related to the paraparesis. With regard to outcome, after a follow-up of 2–12 months six patients showed improvement after interruption of the arteriovenous fistulae, and three patients demonstrated stabilization or only minor improvement of their neurological deficits. There was no further deterioration in any of the patients.

Patient Descriptions

Patient 1

A 63 year old woman presented with progressive sensory and motor deficits in her lower limbs, and urinary and bowel dysfunction. Two and a half years before her admission to our hospital she started to feel numbness in her distal lower limbs and gradually developed gait problems. Lumbar MRI revealed sacral lipoma to which the symptoms were related. Despite surgery and resection of the lipoma, her condition continued to deteriorate. She had progressive motor and sensory impairment in her lower limbs and later began to suffer from impaired bowel and urinary functions. She subsequently developed weakness of the upper limbs. Neurological examination at admission to our department revealed flaccid paraplegia, slight weakness of her upper limbs, urinary and bowel incontinence with decreased anal tone, and L1 sensory level. MRI of the spinal cord demonstrated remnants of the sacral lipoma, tethered cord, hyperintensity, and swelling of the lower thoracic spinal cord and the conus medullaris on T2-weighted images [Figure 1A]. There was a high intensity intramedullary focus at the level of D10, and flow voids in the subarachnoid space. Selective spinal angiography demonstrated spinal dural AVF fed by the right D9 intercostal artery. Surgery was performed and the AVF was obliterated. The postoperative course was uneventful except for a urinary tract infection. She regained normal function of the upper limbs, but remained paraplegic and incontinent (follow-up to date is 11 months).

Patient 2

This 46 year old man presented with progressive numbness in his lower limbs, gait difficulties, and urinary and bowel dysfunction. His symptoms began 2 years before his admission, with numbness in his feet that progressed gradually up to his lower abdomen. He underwent lumbar L4-5 laminectomy without improvement. Two months before his admission he noticed heaviness of his legs while walking, and difficulties in urination and defecation. On examination, he had mild spastic paraparesis, with brisk reflexes and Babinsky signs in his lower limbs, sensory level at D10-11, and a normal anal sphincter tone. MRI of the thoracic spinal cord demonstrated increased cord signal on T2-weighted images and multiple flow voids compatible with spinal dural AVF. Spinal angiography revealed dural AVF fed by left D10 intercostal artery. He underwent D9-11 laminectomy with resection of the spinal dural AVF. There was a radicular pain in the distribution of D9-10 after the operation and dysesthesias in his feet. His condition improved and he regained normal function. The follow-up to date is 12 months.

Patient 3

This 75 year old patient presented with a 5 month history of progressive paraparesis and urinary retention. He also complained of sensory disturbances in his lower limbs that included numbness and pain. His neurological complaints were initially fluctuating but later became permanent and progressed proximally. At admission he was unable to walk and had an indwelling urinary catheter. His past medical history included ischemic heart disease. On examination, he had severe spastic paraparesis, L1 sensory level and urinary retention. MRI demonstrated hyperintensity in the lower spinal cord on T2-weighted images and multiple flow voids. Spinal angiography demonstrated spinal dural AVF fed by right L2-3 lumbar arteries. He underwent L2-3 laminectomy with excision of the fistula. After the operation he suffered from low back pain related to his pre-existing spondylosis. The patient's neurological condition improved, with residual weakness in his right lower limb. Nine months after interruption of the fistula he is able to walk long distances unassisted and has regained bladder control.

Discussion

Spinal dural AVF is a rare and elusive pathology that tends to be under-diagnosed. It affects males more commonly than females, typically between the fifth and eight decades [1], as was also found in our series. Our patients presented with a progressive myeloradiculopathy. Initially, the sensory and motor neurological symptoms were transient and usually began in the feet distally, but gradually they became permanent and progressed proximally. Later in the course of illness progression, the patients lost sphincter control.

The mechanism of the spinal cord damage was not well understood until recent years. It is now believed that these arteriovenous fistulae, which are low flow high pressure lesions, cause venous hypertension [1]. Dural AVF drains intradurally into the coronal venous plexus, which becomes tortuous and elongated with time [3]. The anatomical absence of valves between this plexus and the intramedullary radial veins facilitates the transmission of high venous pressure to the cord tissue. Venous congestive myelopathy produces gradually progressive clinical deterioration. Initially there are transient neurological symptoms related to deterioration of the spinal cord capacity to adjust to venous congestion (compensation/decompensation phenomena). Since the spinal cord edema progresses rostrally and the damage to neurons increases, without treatment the patients will remain permanently impaired or even die, usually because of respiratory complications. Venous hypertension and congestion predispose the veins of the spinal cord to vascular thrombosis resulting in necrotic myelopathy, as described by Foix and Alajouanine (''Foix-Alajouanine syndrome'') in the early 1900s [6,7]. When thrombosis occurs, the damage is no longer reversible. In these patients, if the fistula is interrupted and venous hypertension is relieved before the occurrence of irreversible injury by ischemia or venous thrombosis and cord infarction, reversal of myelopathy is possible [4–6]. Thus, rapid diagnosis and treatment are imperative.

In a microangiographic study, the microvascular anatomy of dural arteriovenous abnormalities was found to be direct arteriovenous fistulae that link the dural branch of the radiculo-medullary-dural artery to the intradural medullary vein [8]. Microscopy showed that the epidural artery feeding the lesion is histologically normal but the draining veins are dilated, fibrotic and irregularly thickened, probably because of their adjustment to arterial pressure [9]. In our patients there was no history that could point to an acquired etiology, e.g., trauma.

Diagnosis is based on the history, clinical findings, and imaging that includes MRI and magnetic resonance angiography. When there are complaints of whole-limb sensory disturbances, physicians should suspect thoracic spine pathology and not lumbar. If there is suspicion of thoracic pathology other than the spinal column, MRI should be performed despite its high cost. MRI findings include regions of increased cord signal on T2-weighted imaging, with a corresponding hypointense signal on T1-weighted images, swelling of the cord, cord enhancement, and prominent intradural vessels with flow voids [10]. These intradural vessels and flow voids are the congested blood vessels draining the fistula, since normal blood vessels are not usually seen on thoracic MRI. Reportedly, MRA can be of assistance in identifying these lesions [5]. MRI findings are not specific for this diagnosis, but if the clinical course and characteristic MRI findings suggest the possibility of spinal dural AVF, selective spinal angiography is indicated since it is the definitive diagnostic procedure [10].

The goal of treatment is to achieve complete occlusion of the fistula without damaging the spinal cord blood supply. This can be carried out by endovascular embolization of the fistula or by its surgical occlusion. Endovascular embolization is a relatively simple and non-invasive procedure. Previously, embolization using polyvinyl-alcohol was employed, but the recurrence rate was very high (around 60%) [11]. Occlusion of the fistulae by embolization with acrylic glues yielded good results and a low recurrence rate. In a series of 49 cases treated by embolization an initial satisfactory result was achieved in 80%, but after the introduction of special microcatheters it increased to 87%, and in over 70% of the cases there was no recurrence of the fistula [4]. In our patients, there was a recurrence of the fistula in one patient (of the four who were treated by initial embolization).

Microsurgical closure of the arteriovenous fistula provides long-lasting obliteration of the fistula with almost no risk of recurrence [5,11,12]. Only a few minor complications have been reported, such as accumulation of cerebrospinal fluid, wound-healing impairment, epidural hematoma and venous thrombosis [12]. Surgery is indicated if a spinal cord artery shares the same pedicle as the feeder of the spinal dural AVF, if endovascular treatment is difficult or dangerous (as in a severe atherosclerotic disease), and after an unsuccessful embolization [4]. Six of our patients underwent surgical closure of their fistulae. There were none or only minor complications, including a temporary wound-healing problem, low back pain and urinary tract infection.

Most of our patients experienced improvement of gait and sphincter control following closure of the fistulae. Some sensory disturbances usually remained. Most had suffered from their symptoms for a long time prior to therapeutic intervention. In three patients their neurological deficits stabilized or showed only mild improvement. One of them had progressive symptoms for 30 months, reaching flaccid paraplegia, complete loss of sphincter control and the start of arm weakness. After interruption of the AVF she regained strength in her arms but remained paraplegic and incontinent. Prolonged duration of symptoms and poor functional status at diagnosis and treatment were associated with poor prognosis. Age was not found to influence outcome; in fact the oldest of our patients had an impressive improvement – he regained the ability to walk unaided and to control his bladder. Prompt diagnosis and treatment, even when neurological damage is severe, can result

MRA = magnetic resonance angiography

in clinical improvement. Since the success of therapy and prognosis depend on rapid occlusion of the fistula, a high index of suspicion and a rapid diagnosis and treatment are imperative in such cases.

References

1. Barrow DL, Colohan AR, Dawson R. Intradural perimedullary arteriovenous fistulas (type IV spinal cord arteriovenous malformations). *J Neurosurg* 1994;81:221–9.
2. Pirouzmand F, Wallace MC, Willinsky R. Spinal epidural arteriovenous fistula with intramedullary reflux – case report. *J Neurosurg* 1997;87:633–5.
3. Merland JS, Riche MC, Chiras J. Intraspinal extramedullary arteriovenous fistulae draining into the medullary veins. *J Neuroradiol* 1980;7:271–320.
4. Niimi Y, Berenstein A, Setton A, Neoohytides A. Embolization of spinal dural arteriovenous fistulae: results and follow-up. *Neurosurgery* 1997;40:675–83.
5. Lee TT, Gromelski EB, Bowen BC, Green BA. Diagnostic and surgical management of spinal dural arteriovenous fistulas. *Neurosurgery* 1998;43:242–7.
6. Van der Meulen MFG, Rinkel GJE, Witkamp TD, Van Gijn J. A man with progressive weakness in his legs. *Lancet* 1999;354:830.
7. Rendowden SA, Molyneux AJ. Case report: spontaneous thrombosis of a spinal dural AVM (Foix-Alajouanine syndrome) – magnetic resonance appearance. *Clin Radiol* 1993;47:134–6.
8. McCutcheon IE, Doppman JL, Oldfield EH. Microvascular anatomy of dural arteriovenous abnormalities of the spine: a microangiographic study. *J Neurosurg* 1996;84:215–20.
9. Benhaiem N, Poirier J, Hurth M. Arteriovenous fistulae of the meninges draining into the spinal veins. A histological study of 28 cases. *Acta Neuropathol Berl* 1983;62:103–11.
10. Jones BV, Ernst RJ, Tomsick TA, Tew J Jr. Spinal dural arteriovenous fistulas: recognizing the spectrum of magnetic resonance imaging findings. *J Spinal Cord Med* 1997;20:43–8.
11. Hall WA, Oldfield EH, Doppman JL. Recanalization of spinal arteriovenous malformations following embolization. *J Neurosurg* 1989;70:714–20.
12. Huffmann BC, Gilsbach JM, Thron A. Spinal dural arteriovenous fistulas: a plea for neurosurgical treatment. *Acta Neurochir Wien* 1995;135:44–51.

Correspondence: Dr. N. Lev, Dept. of Neurology, Rabin Medical Center (Beilinson Campus), Petah Tiqva 49100, Israel. Phone: (972-3) 9376358, Fax: (972-3) 922-3352, email: niril@hotmail.com