Chapter 23
Treatment of Spinal Vascular Malformations:

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INTRODUCTION

Vascular malformations in the spinal cord represent a heterogeneous and challenging group of entities. They can manifest a wide clinical spectrum of symptoms that range from progressive neurological deficits during the course of years to an insidious presentation from acute hemorrhage. Many systems based on diverse criteria have been used to classify these lesions.

This review summarizes the senior author’s (RFS) experience (38) (Table 23.1) and presents a classification system for these vascular malformations on the basis of broad categories, such as the anatomic location (intradural or extradural, anterior or posterior) of the malformation or its angiographic appearance (arteriovenous fistula, AVF; or arteriovenous malformation, AVM). Neoplastic lesions that behave similar to vascular malformations in terms of their high vascularity and propensity to hemorrhage are also included in this classification.

NEOPLASTIC LESIONS

Cavernous Malformations

Including cavernous malformations in the same category as neoplasms is controversial. We do so, however, because these lesions can grow, have a chromosomal abnormality, and may be familial or acquired (11). The familial form is associated with a dominant trait (37) located on chromosome 7q (33). They also may be induced after radiation of the spine (28). From a histological perspective, spinal cavernous malformations are identical to those that occur in the brain (4).

Cavernous malformations become symptomatic acutely from a new hemorrhagic event or progressively from neurological deterioration related to spinal cord compression. Symptoms are based on the location of the lesion within the spinal cord. T2-weighted (Fig. 23.1) magnetic resonance imaging (MRI) often shows evidence of blood at different stages. The lesions are well demarcated, with mixed signal intensity. The core is surrounded by a rim of low intensity that corresponds to hemosiderin. The clinical history of patients with spinal cord cavernous malformations is acute neurological decline that improves but without returning to baseline status. After a few hemorrhages, impairment is significant (35).

Symptomatic patients usually worsen, and only surgical resection halts their deterioration. Furthermore, patients treated soon after symptoms develop have a better chance of improving (76%) within 3 years of treatment compared with patients whose treatment is delayed (52%, \( P < 0.02 \)) (41). The primary prognostic factor is preoperative status (11).

Patients with acute hemorrhage or progressive neurological deficits should undergo surgical resection. In patients who are improving or in those with minor symptoms, the risks of surgery must be weighed against the risk of
irreversible damage from acute hemorrhage. Dorsal or exophytic lesions are easier to treat than ventral or intramedullary lesions.

During surgery, the cavernous malformation is identified as an area of blue discoloration within the spinal cord (Fig. 23.2). During resection, the glial rim is preserved as a dissection plane around the lesion. Electrocoagulation should be avoided. Resection must be complete because residual lesions can regrow, with the potential for catastrophic consequences (40). During surgery, venous anomalies are often encountered. Because they drain normal spinal cord tissue, they must be left intact to avoid the potentially devastating consequences of a stroke.

In the senior author’s (RFS) experience with more than 45 spinal cavernous malformations, Frankel functional outcome scores remained the same or improved in 91% of the patients. Five (11%) patients developed new episodes of bleeding related to incomplete resection but improved after a second operation (RF Spetzler, unpublished data). If symptoms recur after surgery in the absence of residual cavernous malformation, a tethered cord should be considered (40). Because cavernous malformations are angiographically occult, endovascular therapy has no role in their treatment, nor does radiation therapy.

Hemangioblastomas

Hemangioblastomas can occur sporadically or have a familial presentation associated with von Hippel-Lindau disease (10). Hemangioblastomas are neoplastic lesions consisting of three cell types: pericytes, stromal cells, and endothelial cells. These tumors can become vascular enough to resemble AVMs. Even macroscopically, they can have large venous channels (26). During angiography, it is sometimes possible to see a vascular shunt within the lesion. These tumors are identical to those that occur in the brain (16, 25).

MRI (Fig. 23.3) shows a cystic lesion with an enhancing nodule with or without a syrinx. When feeders are clearly visible during angiography (Fig. 23.4), embolization can facilitate surgical resection. The definitive treatment is surgical resection (Fig. 23.5).

AVMs AND AVFs

AVMs are divided into extradural–intradural and intradural. The latter is further subdivided into intramedullary and conus AVMs. AVFs can be intradural or extradural. Intradural lesions are located either ventrally or dorsally within the spinal cord.

Malformations

Intradural–Extradural AVMs

Intradural–extradural AVMs are also known as Type III, metameric, or juvenile AVMs. They usually involve the entire metamere compromising the skin, muscle, bone, and spinal cord (Fig. 23.6A and B). This manifestation is known as Cobb’s syndrome. They can involve several levels, maintaining the same characteristics of wide and diffuse compromise. These lesions are highly vascular, and a multidisciplinary and multistaged treatment is recommended. Preoperative embolization is important. Treatment, however, is typically palliative because complete obliteration of
these extensive lesions is rarely obtained.

Intradural AVMs

Intradural AVMs are also known as Type II, glomus, or classic AVMs. Intradural AVMs have a nidus within the spinal cord, resemble brain AVMs, and are challenging to treat. These lesions usually become symptomatic with hemorrhage within the spinal cord or with progressive neurological deterioration related to vascular steal. Acute nonhemorrhagic deterioration may be related to spontaneous venous thrombosis (12). Because these lesions often compromise the anterior spinal artery, a definitive cure is usually obtained with surgery (13).

On the basis of the angioarchitecture of the nidus, intradural AVMs can be subcategorized as compact or diffuse (Fig. 23.7A and B). Compact niduses are most amenable to surgical resection (Fig. 23.7C). They can be approached through a standard laminotomy. A small myelotomy is performed following the posterior median sulcus, and the spinal cord is split between the two posterior columns. If the lesion is closest to the lateral surface of the spinal cord, a posterolateral myelotomy is performed through the dorsal root entry zone between two or more nerve roots. The posterior midline approach is especially useful for compact lesions.

Superselective catheterization can help identify associated aneurysms, which often disappear after the AVM has been treated. Embolizing major feeders is helpful during surgery (5, 8), and preoperative embolization of major arterial feeders has been described.

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Conus AVMs

This special category of malformations is characterized by the simultaneous presence of an anterior and dorsal intradural AVF (Fig. 23.8) with multiple feeders and an intramedullary AVM. Invariably, conus AVMs are located at the conus medullaris. Consequently, an abnormality during neurulation has been proposed to underlie their development (24). They have also been associated with a tethered cord. Patients with a conus AVM usually present with myelopathy or radicular symptoms (Fig. 23.8B). Early bowel and bladder compromise reflects their typical location at the conus. Their extensive nature and multiple arterial feeders make it difficult to treat conus AVMs by embolization alone. An initial embolization followed by surgical resection is ideal.

Fistulas

Extradural AVF

Extradural AVFs involve direct communication between an extradural artery and the epidural venous system. These high-flow fistulas cause engorgement (Fig. 23.9A and B) of the epidural veins, which compresses the spinal cord. The shunt of arterial blood into the venous system can steal flow from the spinal cord, causing symptoms related to ischemia. Extradural AVFs can be treated effectively with endovascular techniques by embolizing the arterial feeder (9). They rarely require surgery. Occasionally, extradural AVFs can cause spontaneous epidural hematomas. If the patient’s clinical condition permits the necessary time, we recommend spinal angiography during the evaluation of
Intradural Dorsal AVFs

Intradural dorsal AVFs are the most common vascular malformations affecting the spinal cord (23, 39). They are also known as Type I malformations, angioma racemosum, angioma racemosum venosum, long dorsal AVFs, and dorsal extramedullary AVFs. For unknown reasons, they occur in the thoracic and lumbar regions, usually in men during the fourth decade of life. The communication between a radiculomedullary artery and a radicular vein that drains into the coronal venous plexus is located (Fig. 23.10A and B) intradurally at the dural root sleeve, causing intradural venous hypertension (17). The absence of valves between the coronal and radicular veins encourages venous congestion (36).

Venous hypertension decreases spinal cord perfusion pressure. Therefore, ischemia is a primary contributor to symptoms. Symptoms tend to be exacerbated by Valsalva maneuvers, which further increase venous pressure and decrease blood flow in the spinal cord. During surgery, pressure measurements in the epidural space do not change after the fistula is occluded. Pressure decreases significantly in the intradural space but never to the level of normal central venous pressure. These findings indicate the presence of the fistula at the intradural compartment and some obstruction of venous drainage (Figs. 23.3, 23.4, and 23.7) (7, 20).

Intradural dorsal AVFs are subclassified into two subtypes: Type A has a single feeder and Type B has multiple feeders (2, 3). Clinically, patients become symptomatic with early fecal and urinary incontinence and progressive and incapacitating myelopathy (39). Within 6 months, 19% of patients are disabled and within 3 years, 50% are severely disabled (1).

Surgery provides the most definitive treatment, although good results have been obtained with endovascular treatment using liquid acrylic rather than alcohol or particles, which are associated with a higher incidence of recurrence (9, 17). Endovascular therapy offers the possibility of making the diagnosis and treating the fistula during the same angiographic procedure, but it is associated with a higher incidence of recurrence than surgery (17). Surgery is associated with its own intrinsic complications, but provides a definitive cure for most patients. We recommend ligation and section at the site of the fistula, without stripping the dilated veins that drain the normal spinal cord. Doing so may precipitate clinical deterioration (32).

Intradural Ventral AVFs

Intradural ventral AVFs were initially described by Djindjian et al. (14). As their name implies, they usually occur anterior to the spinal cord (Fig. 23.11A–C) and cause symptoms related to compression or hemorrhage (spinal subarachnoid hemorrhage). They usually occur at the conus medullaris or filum terminale. They can present as part of Rendu-Osler-Weber or Cobb's syndromes (19). These high-flow lesions are also known as Type IV or perimedullary fistulas (21). They often involve the anterior spinal artery, and the fistula occurs within the spinal canal but outside the spinal cord (18). The fistulous communication occurs between the anterior spinal artery and perimedullary veins (30). In contrast, dorsal AVFs are fed by segmental arteries and located dorsal to the spinal cord.

Intradural ventral AVFs can be subdivided as Type 1, 2, or 3 on the basis of their size (18). Good outcomes usually
follow endovascular treatment. If endovascular treatment is unsuccessful, the fistula can be interrupted surgically (6). The absence of a nidus involving the spinal cord (30) is an advantage favoring surgical treatment. Typically, an endovascular approach is used as an adjunct to surgery for large fistulas (9). Precise identification of the angioarchitecture of the fistula, especially the contribution from the anterior spinal artery, is important to avoid compromising blood flow distal to the fistula.

ANEURYSMS

Spinal aneurysms are rare. Only a few cases have been reported in the literature. In 1993, Rengachary et al. reviewed 10 cases of isolated spinal aneurysms in the literature (34). We have treated four patients with spinal subarachnoid hemorrhage related to ruptured aneurysms.

Spinal aneurysms can manifest as compressive lesions but typically do so with rupture. Spinal aneurysms are a rare cause of intracranial subarachnoid hemorrhage, but the diagnosis should be considered when no other sources of bleeding are found or when the subarachnoid hemorrhage is limited to the spine. On MRI, they appear as round, well-localized flow voids within the spinal canal (Fig. 23.12A). Spinal aneurysms differ from intracranial aneurysms in ways that affect their management. They seldom occur at branching points; rather, they develop along the course of an artery. The caliber of the spinal arteries is much smaller than that of intracranial arteries, and they tend to be less affected by atherosclerosis (27). Spinal aneurysms lack a clear neck and usually appear as fusiform dilations (34). Partial thrombosis of spinal aneurysms, a frequent finding during surgery, probably accounts for their becoming symptomatic (15, 22).

The fusiform nature of these lesions makes clipping difficult and favors sacrifice of the parent vessel. During surgery, if the aneurysm is found along the course of the artery with evidence of distal flow, surgical reconstruction with a termino–terminal anastomosis can be performed.

If the aneurysm is distal on the circulation or thrombosed, the parent vessel can be occluded and the aneurysm can be resected. If there is evidence of flow distal to the aneurysm, the lesion can be wrapped (Fig. 23.12B) in the hope that a fibrotic scar will surround the aneurysm and protect it from further bleeding (40).

The position of the aneurysm with respect to the spinal cord and the choice of surgical approach are important determinants of the procedure of choice. We used a traditional laminoplasty in three patients with an aneurysm posterior to the spinal cord (Fig. 23.12C). In one case, an aneurysm anterior to the spinal cord in the lower thoracic region was exposed through an anterior (transthoracic) approach, which provides a narrow and challenging corridor.

Endovascular techniques can be considered, but the diagnosis of spinal cord aneurysm must be definitive before intervention. We have only been able to make the definitive diagnosis at surgery. Distal aneurysms can be addressed with thinner and more flexible catheters only if the diagnosis is certain.

Table 23.1. Classification of spinal cord vascular malformations
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(Modified with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96(2)145–156, 2002 [38]).
Extradural–intradural

Intradural intramedullary

Conus malformations

AVF

Extradural

Intradural dorsal

Intradural ventral

Spinal aneurysms

(Modified with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96(2)145–156, 2002 [38]).

References


18. Gueguen B, Merland JJ, Riche MC, Rey A: Vascular malformations of the spinal cord: Intrathecal perimedullary


Fig. 23.1 Sagittal T2-weighted MRI of a thoracic intramedullary cavernous malformation. The cavernous malformation exhibits a heterogeneous signal (from previous hemorrhagic episodes) surrounded by a low-intensity rim that corresponds to hemosiderin (with permission from, Riina HA, Lemole GM Jr, Kim LJ, Spetzler RF: Spinal arteriovenous malformations, in Mohr JP, Choi D, Grotta J, Wolf P (eds): Stroke: Pathophysiology, Diagnosis, Management. Churchill Livingston, 2004, pp 1417–1422 [35]).

Fig. 23.2 Intraoperative photograph from a different patient from Figure 28.1 that shows the presence of a cavernous malformation within the spinal cord and associated venous anomaly. During surgery, it is very important to preserve this venous channel because it drains the normal spinal cord tissue (with permission from, Porter RW, Detwiler PW, Spetzler RF, et al.: Cavernous malformations of the brainstem: Experience with 100 patients. J Neurosurg 90:50–58, 1999).

Fig. 23.3 Sagittal T1-weighted image with gadolinium that shows an homogeneous enhancement within the spinal cord that corresponds to a hemangioblastoma (with permission from, Riina HA, Lemole GM Jr, Kim LJ, Spetzler RF: Spinal arteriovenous malformations, in Mohr JP, Choi D, Grotta J, Wolf P (eds): Stroke: Pathophysiology, Diagnosis, Management. Churchill Livingston, 2004, pp 1417–1422 [35]).

Fig. 23.4 Digital subtraction angiography that demonstrates the significant vascularity of these tumors (with permission from, Riina HA, Lemole GM Jr, Kim LJ, Spetzler RF: Spinal arteriovenous malformations, in Mohr JP, Choi D, Grotta J, Wolf P (eds): Stroke: Pathophysiology, Diagnosis, Management. Churchill Livingston, 2004, pp 1417–1422 [35]).

Fig. 23.5 Intraoperative photograph showing an intramedullary hemangioblastoma (with permission from Thieme International).

Fig. 23.6 A, illustration showing a complex AVM that has a metameric involvement. Note the compromise of the vertebral body, extradural compartment, and intradural compartment (with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96:145–156, 2002 [38]). B, axial T1-weighted MRI with contrast that shows flow voids within the vertebral body, the extradural space, and within the intradural compartment. Because of this diffuse extension, surgical resection is difficult to
accomplish.

Fig. 23.7 Illustration showing the presence of an AVM. A, the compact nidus is more amenable for surgical resection than B, the diffuse nidus, which extends over multiple levels. C, vertebral injection showing the presence of an intradural compact AVM (with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96:145–156, 2002 [38]).

Fig. 23.8 A, illustration showing the conus type of AVM demonstrating the simultaneous presence of a dorsal and a ventral fistula with an intradural AVM. These abnormalities occur for unknown reasons exclusively at the conus medullaris. B, intraoperative picture showing the presence of multiple fistulas that occupy the canal anterior and posterior. Note the extensive involvement of roots and spinal cord at the conus medullaris (with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96:145–156, 2002 [38]).

Fig. 23.9 A, illustration showing the presence of an extradural AVF. Also note the compressive effect that the engorged veins cause on the spinal cord. B, anteroposterior digital angiogram showing the presence of multiple and diffuse serpiginous fistula in the epidural space, causing significant cord compression (with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96:145–156, 2002 [38]).

Fig. 23.10 A, illustration showing a dorsal AVF. Note the arterial feeder arising from the radiculomedullary artery and communicating directly into the coronal plexus, causing venous engorgement. B, selective angiogram showing the fistula between a radiculomedullary artery and the coronal venous plexus. The fistula was coagulated and transected (with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96:145–156, 2002 [38]).

Fig. 23.11 A, illustration showing an AVF fed mainly by the anterior spinal artery (with permission from, Spetzler RF, Detwiler PW, Riina HA, Porter RW: Modified classification of spinal cord vascular lesions. J Neurosurg Spine 96:145–156, 2002 [38]). B, T1-weighted MRI showing the presence of flow voids located anterior to the spinal cord (with permission from Barrow Neurological Institute).

Fig. 23.12 A, sagittal T2-weighted MRI showing a flow void (arrow) in the spinal canal in a patient with subarachnoid hemorrhage limited to the spinal cord. B, selective angiogram showing the presence of a nondistal aneurysm that