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REVIEW

Spinal vascular malformations: treatment strategies and outcome

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Abstract Spinal vascular malformations (SVMs) are a heterogeneous group that can cause acute, subacute, or chronic spinal cord dysfunction. The majority of the patients present to neurosurgical attention after a protracted course with severe neurological dysfunction. Spinal vascular lesions comprise approximately 3–4 % of all intradural spinal lesions. They are pathologically similar to their intracranial counterparts, but their clinical impact is often comparatively worse. Early, correct recognition of the pathology is mandatory to halt the progression of the disease and minimize permanent spinal cord injury. The first clinical observation of a SVM was published in 1890, but it was not until 1914 that the first successful surgical treatment of a spinal vascular malformation was reported. Intervention—either by microsurgical or endovascular means—aims to halt or reverse the progressive neurological deterioration by eliminating flow through the abnormal fistulous or nidal connections, and restoring normal spinal cord perfusion and intravascular pressures. In fact, complex spinal arteriovenous malformations (AVMs) and arteriovenous fistulas (AVFs) frequently require a multimodality approach that utilizes both microsurgery and endovascular embolization effectively. The goal of this review is to describe the various types of vascular malformations of the spine, their pathophysiology, clinical presentation, treatment strategies, and outcome. For purposes of discussion on the current manuscript, vascular malformations of the spine were divided into arteriovenous fistulas (AVFs) and arteriovenous malformations (AVMs). Spinal cord aneurysms are extremely rare, and the majority of the lesions that come to the neurosurgeon's attention are concomitant to a spinal AVM.

Keywords Spinal arteriovenous malformation · Spinal arteriovenous fistula · Spinal dural arteriovenous fistula · Treatment strategies · Outcome · Review

Introduction

Spinal vascular lesions comprise approximately 5–9 % of all vascular malformations of the central nervous system, [8] or 3–4 % of all intradural spinal lesions [45]. Despite their pathological similarities with their intracranial counterparts, their clinical impact is often comparatively worse. Spinal vascular malformations (SVMs) are a heterogeneous group that can cause acute, subacute or chronic spinal cord dysfunction. The majority of the patients present to neurosurgical attention after a protracted course with severe neurological dysfunction. In 1974, Aminoff and Logue showed that up to 48 % of patients with untreated arteriovenous malformations of the spinal cord (AVMs) were confined to bed or wheelchair within 3 years of symptom onset, and complications of chronic paraplegia were directly responsible for a mortality rate of 15 % [2, 3]. Early, correct recognition of the pathology is mandatory to halt the progression of the disease and minimize permanent spinal cord injury.

The goal of this review is to describe the various types of vascular malformations of the spine, their pathophysiology, clinical presentation, treatment strategies, and outcome. A prelude on the historical background, vascular anatomy and radiographic findings is presented. For purposes of discussion on the current manuscript, vascular malformations of the spine were divided into arteriovenous fistulas (AVFs) and

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arteriovenous malformations (AVMs). Spinal cord aneurysms are extremely rare, and the majority of the lesions that come to the neurosurgeon's attention are concomitant to a spinal AVM.

Historical perspective

The first clinical observation of a SVM was published in Germany in 1890. Berenbruch operated on a patient with a spinal abnormality, subsequently recognized as a vascular malformation at autopsy [5]. In 1910, Fedor Krause was the first to recognize a spinal lesion observed at laminectomy as a vascular abnormality [5.] In 1912, Charles Elsberg pioneered the first successful surgical intervention for a spinal cord vascular lesion, presumably a perimedullary AVF [1, 5]. Recovery of the operation was uneventful, but without clinical improvement. In 1915, Cobb reported several cases of SVMs and outlined their variable clinical features. He was the first to report the combination of a SVM and vascular anomalies of the overlying skin, now known as Cobb syndrome [1] The classic subacute necrotic myelopathy was described by Foix and Alajouanine in 1926, but its pathophysiology was not further elaborated until the landmark studies of Wyburn-Mason [1, 61].

Before the advent of modern anesthesia and microsurgical techniques, the few case reports on the surgical treatment of SVMs had overall discouraging results. Several of the performed operations focused on elevation and coagulation of the abnormal blood vessels from the dorsal aspect of the spinal cord, mainly due to the lack of an adequate comprehension of the pathophysiology of the disease. The so called modern era in the treatment of SVMs began in 1969 with Krayenbuhl and Yasargil and the publication of their microsurgical techniques, based heavily on the use of the operating microscope and bipolar cautery [28, 62]. In 1977, Kendall and Logue demonstrated that lesions on the surface of the spinal cord, formerly thought to be venous angiomas, were actually arterialized veins dilated by communication through a dural AVF [5, 25]. The current understanding of the different spinal vascular malformations is a result of combined efforts from what Black described as the American/English/French (ABF) connection [5]. Their contributions led to the development of the widely used classification of spinal arteriovenous malformations (types I–III), with a fourth type added later by Heros et al. [22]. Over the last decade, other classifications have been introduced by different groups [26, 46, 52].

Epidemiology

Dural AVF is the most common vascular malformation; it accounts for 50 to 85 % of all lesions [13, 36, 39, 48, 49, 58]. Men are affected five times more often than women, and the mean age at the time of diagnosis is 50–60 years [29, 30, 41]. Patients younger than 30 years of age constitute less than 1 % of patients with a DAVF. Most lesions are centered at the thoracolumbar spine, with up to 90 % of those located between T4 and L3 [39, 49]. True intradural, perimedullary AVFs are significantly rarer, have no sex predilection, and tend to occur at the thoracolumbar region [4, 39]. Most patients present at a relatively young age, typically within the second or third decades [39, 49].

Spinal cord AVMs usually present in the 3rd decade of life, but they can be diagnosed in the pediatric population [13, 18, 39, 46, 49, 55]. In a recent meta-analysis analyzing spinal glomus AVMs, Gross and Du reported no sex predilection. The majority of AVMs were thoracic (51 %) and cervical (29 %), and 29 % had an associated aneurysm [18].

Anatomy and pathophysiology

The anterior two thirds of the spinal cord are supplied by a single anterior spinal artery (ASA). It originates from the spinal branches of the vertebral arteries and is additionally supplied at multiple levels by spinal radicular branches of the segmental arteries. The majority of the radicular arteries regress during development, with an average number of six still present in adult life in an unpredictable pattern [36]. In the cervical region, the artery of the cervical enlargement is the largest feeder, often encountered between C4 and C8 [23]. In the thoracolumbar region, those radicular arteries arise from the dorsal branches of either an intercostal or a lumbar artery. They follow the ventral nerve root through the intervertebral foramen until anastomosing with the ASA. After a short initial ascending course, the radicular artery follows a characteristic hairpin configuration at its junction with the ASA, with smaller cephalad and larger caudad branches arising from its apex. This classic configuration is very useful for angiographic identification of the anterior spinal artery. The most prominent radicular feeder in the thoracolumbar region is the artery of the lumbar enlargement or artery of Adamkiewicz. It arises most commonly between T9 and T12, typically on the left side, seldom from the lumbar region or higher between T6 and T8. In the conus, the anterior spinal artery terminates by anastomosing with the posterior spinal arteries, forming a basket-like configuration (rami cruciantes) [23].

The posterior third of the spinal cord is supplied by an extensive plexus formed by the duplicated posterior spinal arteries (PSAs). They originate from the vertebral artery or the posterior inferior cerebellar artery. The pial plexus surrounds the surface of the cord and connects the anterior and posterior vessels. Numerous posterior radicular feeders arise from the extraspinal arteries and anastomose with the PSAs. As seen on the anterior spinal circulation, the posterior

radicular arteries also assume a classic hairpin configuration as they join the PSAs [36].

The variability of the venous system is much more pronounced. The intrinsic radial veins drain in a centrifugal manner towards the venous plexus of the pia mater, which in turn drains towards the anterior and posterior median spinal veins. They communicate via medullary and radicular veins with the epidural venous plexus. The valveless radicular veins pierce the dura to follow the nerve roots. At this level, there is an anatomic narrowing that some consider a functional antireflux mechanism [36]. The epidural venous plexus, in turn, drains into the paravertebral veins, such as the vertebral vein in the neck, the azygous and hemiazygous veins in the thorax, the ascending lumbar vein, and the internal iliac vein [23].

Kendall and Logue were the first to correctly recognize a spinal cord arteriovenous fistulous shunt site as dural, related to the nerve root sleeve [25]. The arterialization of the coronal venous plexus caused by the fistulous connection resulted in venous hypertension and spinal cord ischemia and myelopathy. In fact, venous hypertension has been considered the major factor causing spinal cord ischemia in several types of SVMs. Direct intraoperative measurements have documented mean venous pressures as high as 74 % of the systemic arterial pressure in patients with spinal dural AVFs [20, 21].

Three other physiological mechanisms have been proposed to explain the neurological deterioration in patients with SVMs: hemorrhage, vascular steal, and mass effect. Hemorrhage is most commonly seen with AVMs and can be subarachnoid, intraparenchymal, or both. Vascular steal was first recognized in the late 1960s and 1970s [1]. It was then associated with high-flow, low-pressure AVMs and with large perimedullary AVFs fed by the ASA [1, 23, 26]. Mass effect can occur with large AVFs with massively dilated venous structures and feeding vessel aneurysms.

Imaging

The development of spinal angiography in the 1960s revolutionized the understanding of the spinal vascular malformations. Spinal aortography was introduced by Rene Djindjian and associates in France at the Lariboisiere Hospital in 1962. Contemporary to that group, Doppman and DiChiro demonstrated the importance of subtraction angiography and selective catheterization techniques on their early studies on the theme at the National Institutes of Health (NIH) in the USA [1].

Before the advent of modern spinal imaging techniques, the descriptions of SVMs were largely derived from post-mortem pathological studies. It was not until the advent of Lipiodol myelography in the 1920s that clinicians were able to identify spinal vascular lesions in the living patient [1]. Myelography rapidly became a viable diagnostic tool but was limited by its inability to directly visualize the vascular anatomy itself. By the 1950s and 1960s, cerebral angiography became the gold standard for diagnosing and analyzing spinal vascular lesions. However, the continuing advances on CT and MRI imaging may ultimately rival spinal angiography in regards to diagnosis and characterization of these lesions.

MRI

MRI is frequently the first imaging study ordered in the work-up of patients with SVMs. The classic clinical presentation of progressive thoracic myelopathy associated with venous congestion seen on patients with spinal dural AVFs correlate with MRI findings of hyperintense T2 cord signal and cord edema over multiple spinal levels. The edematous cord may demonstrate enhancement on post-gadolinium sequences. In advanced disease, cord atrophy may be present. On T2-weighted sequences dilated, serpiginous perimedullary vessels can be seen as flow voids lining the dorsal or ventral surface of the cord [36]. The T2-hyperintensity involves the conus in up to 90 % of cases, and lack of T2 cord signal in the presence of an AVF is extremely rare.

Spinal AVMs share some imaging features on MRI with intracranial AVMs. Typically, they form a mass of dilated peri- and intramedullary vessels visualized as flow voids on T2-weighted sequences. As with dural AVF, venous congestion may be present with hyperintense T2 cord signal and swelling. In AVMs with fistulous components, serpiginous flow voids extending through several levels are common. AVMs that hemorrhage may demonstrate varying cord signal intensities consistent with acute of subacute blood products or subarachnoid hemorrhage.

MRA

The MR radiographic findings of T2-cord hyperintensity, enhancement, or flow voids are not predictive of the level of the lesion. Recent advances in MR angiography (MRA) have improved the ability to confirm the diagnosis of spinal AVF or AVM and, in many instances, localize the lesion to a specific segment or spinal level. Traditionally, the resolution of spinal MRA was hampered by the trade-off between obtaining a large field of view to encompass the thoracolumbar spine while maintaining high spatial resolution. New protocols utilizing fast contrast-enhanced MRA allow for more precise imaging of dilated perimedullary and radicular veins in dural or perimedullary AVFs. Correct identification of the level of fistulous connection can be done in up to 81 % of cases [33]. The majority of the patients often require further spinal angiography. Nevertheless, CE-MRA may at minimum allow for a focused angiogram of the involved segments, potentially cutting down on procedure time, contrast load, time of fluoroscopic radiation exposure, and procedural complications.

CTA

CTA and CT myelography remain viable options in the evaluation of spinal vascular disease. A small series comparing CT spinal angiography to CE-MRA and spinal angiography found a 75 % rate of detection of spinal vascular malformation, which was comparable to CE-MRA [50]. CTA may suffer from impaired contrast resolution in the obese patient and the potential negative impact of iodinated contrast use and ionizing radiation exposure.

Spinal angiography

Spinal angiography remains the gold standard for diagnosis and characterization of spinal vascular lesions, particularly spinal dural AVFs and AVMs. It is still superior to MR and CT imaging for characterization of spinal vascular lesions because it allows a precise determination of the involved vessels. In many circumstances it can pinpoint the exact fistulous component. Specific protocols in the workup of spinal dural AVFs include identifying the level of the artery of Adamkiewicz and any venous stasis suggestive of a fistulous connection, followed by selective thoracic and lumbar intercostal injections. If these are unrevealing, further workup involves injecting the lateral sacral arteries, aorta, and subsequently the arterial supply to the cervical cord and posterior fossa [59]. At our institution, and as preconized by other authors, [32] selective catheterization of bilateral internal iliac arteries is routinely performed during the angiographic evaluation of patients with suspected dural arteriovenous fistula where a more cephalad fistulous component could not be identified. The addition of 3D rotational spinal angiography has further improved the imaging quality of spinal vascular lesions [43]. Despite these advances, conventional spinal angiography can require extended procedure times and multiple studies to define the offending pathology (particularly with dural AVFs). They may involve high iodinated contrast loads and radiation doses to the patient and continue to carry a small risk of procedural complication, including spinal cord ischemia and paraparesis.

Classification

The modern classification of the spinal vascular malformations was first proposed by DiChiro, Doppman, and Ommaya in 1969 [9]. They divided the SVMs into three types based on their landmark studies on spinal angiography. In 1986, Heros et al. reported a patient with an intradural perimedullary AVF and proposed this SVM be classified as a distinct fourth type [22]. The resultant classification—also known as the American/British/French connection (ABF) classification [5]—has had widespread acceptance and use in the neurosurgical literature since then (Table 1).

Table 1 ABF classification of spinal vascular malformations

Type I	Spinal dural arteriovenous fistula (previous angioma racemosum venosum): located at the dural sleeve of a spinal root, associated with a single-coiled vessel on the dorsal pial surface of the spinal cord
Type II	Glomus AVM (previous angioma racemosum arteriovenosum): characterized by a true intramedullary nidus and with the arteriovenous shunting occurring deep into the pia
Type III	Metameric or juvenile AVM (previous Cobb Syndrome): involvement of one or more metameres (and consequently of portions of the neural tissue, dura, bone, muscle and skin)
Type IV	Direct or perimedullary AVF: direct arteriovenous fistula, usually supplied by the anterior spinal artery, and drainage through the pial venous network, resulting in aneurysmal dilation of the draining veins

ABF American/British/French, *AVM* arteriovenous malformation, *AVF* arteriovenous fistula

In 2002, Spetzler et al. proposed a modified classification system for spinal vascular malformations based on anatomical and pathophysiological factors [52]. It incorporated to the ABF classification distinct categories for spinal cord aneurysms and neoplastic vascular lesions. The authors also proposed a separate category called conus medullaris arteriovenous malformations, characterized by their exclusive involvement of the conus medullaris and filum terminale, multiple feeding arteries, multiple extramedullary and pial niduses, and complex venous drainage (Table 2).

In cases of spinal dural AVF, the fistulous connections are typically located in the dura between the radiculomeningeal artery and the radicular vein as it exits the dural sleeve. In rare instances, though, the fistula occurs extradurally between branches of the radicular artery and the epidural venous plexus. Those cases are denominated as spinal epidural or extradural arteriovenous fistula. They are rarely symptomatic but may present with symptoms secondary to a compressive mass effect on adjacent nerve roots or congestive myelopathy [26, 44]. The presence of secondary intradural drainage can explain why some of those lesions can be mistakenly

Table 2 Spetzler classification of spinal vascular malformations

Neoplastic vascular lesions	Hemangioblastoma
	Cavernous malformation
Spinal cord aneurysms	
Arteriovenous fistulas	Extradural
	Intradural
	Ventral (type IV AVM)
	Dorsal (type I AVM)
Arteriovenous malformations	Extradural-intradural (type III AVM)
	Intradural
	Intramedullary (type II AVM)
	Conus medullaris

AVM arteriovenous malformation

classified as a type I SDAVF; in fact, epidural AVFs may account for some of the recurrences seen after surgical disconnection. In these cases, obliteration of the intradural vein may be associated with early recurrence as the epidural shunt recruits new intradural veins adjacent to the disconnected level. A recent literature review on the topic identified only 45 ventral epidural AVFs in 20 published papers. The mean age was 63.9 years. The lumbar spine was the most frequently involved segment, and progressive myelopathy signs were five times more common than radiculopathy. The arterial feeders tended to be multiple and originating from segmental arteries such as intercostal artery, lumbar artery, lateral sacral artery, ascending/deep cervical arteries, and vertebral arteries [27].

Clinical presentation

The clinical presentation of SVMs can be subdivided in two different categories: acute presentation or protracted, progressive neurological decline. Acute presentation (associated with hematomyelia or subarachnoid hemorrhage) is usually seen in patients with spinal cord aneurysms, and intradural/intramedullary AVMs. Classic examples of a protracted course (secondary to venous hypertension, cord ischemia, or mass effect) include AVFs (extradural or intradural) and conus medullaris and juvenile AVMs. By the time of diagnosis, the majority of patients already have certain degree of motor and sensory deficits [49]. Aminoff and Logue defined the course of the SVMs as one of progressive neurological decline and functional disability [3]. In that study, one fifth of the 60 patients required crutches or were nonambulatory by 6 months after the onset of symptoms other than pain. Half of all patients were confined to a wheelchair or bed within 3 years of the onset of gait impairment, and 91 % had restricted activity within 3 years of the onset of symptoms [2, 3, 39].

Common to all patients are symptoms of myelopathy, such as lower extremity weakness, loss of pain and temperature sensation, and bladder and bowel incontinence. Patients with spinal dural AVF often suffer from neurogenic claudication, with symptoms exacerbated by physical activities such as walking and standing and relieved by sitting. Exercise or posture-induced symptoms are uncommon with AVMs. Subarachnoid hemorrhage is the presenting event in about one third of patients with AVMs of the spinal cord, but it is exceptionally rare with AVFs [58]. The presence of an associated nidal or feeding artery aneurysm has been reported in 16–48 % of AVMs, and is often cited as a risk factor for hemorrhage [14, 18, 19, 49]. A spinal bruit may be heard on high-flow, juvenile AVMs. In a recent institutional review of 110 treated AVFs and AVMs, the most common presentation was paresis/paralysis (75.5 %), paresthesias (60 %), pain (51.8 %), and bowel/bladder dysfunction (41.8 %) [45]. The frequencies of presenting signs and symptoms were similar between the two separate groups, except for a higher incidence of subarachnoid hemorrhage with AVMs (37.9 %).

Despite the similarities, spinal dural AVFs are distinguished from intradural SVMs by several clinical features. They have a strong male predilection (>80 %) and present later in life (80 % after the age of 40) [39, 49]. The majority of those lesions are located on the thoracolumbar region, which helps explain why upper extremities involvement is so unlikely. The typical patient with a spinal dural AVF is an older (>40 years) male with gradual onset of progressive lower extremity symptoms exacerbated by walking or standing. Differential diagnosis involves spinal stenosis, demyelinating disease, spinal cord tumors or, more rarely, conditions such as Guillain-Barre syndrome, amyotrophic lateral sclerosis, or peripheral vascular disease [17]. Conversely, the patient with an AVM tends to be younger (<30 years), men (if included the pediatric population), has a higher chance of abrupt onset, and have more frequently upper extremity symptoms, depending on the lesion location [58]. A distinct sensory level is present in most patients, and generally reflects the location of the vascular nidus along the spinal axis [49].

Foix-Alajouanine syndrome is a classic but frequently misunderstood syndrome associated with spinal cord vascular malformations. Traditionally described as an acute or subacute myelopathy, it is attributed to spinal cord venous thrombosis related to an arteriovenous malformation, resulting in venous infarction and necrosis. Spinal dural AVFs had not yet been described at the time of the original report in 1926 [16]. However, in retrospect, it has been speculated that the patients in the original report by Foix and Alajouanine had type I AVFs. Pathological analysis of these initial cases did not show evidence of thrombosis, and symptoms may have been attributable to venous hypertension [16, 17].

In spinal cord AVMs that present with subarachnoid hemorrhage, the massive hemorrhage may often result in sudden, rapid development of excruciating back pain, with or without neurological deficit (*le coup de poignard rachidien*, or *Coup de poignard of Michon*). It can be seen as the corollary of the thunderclap headache of intracranial SAH. Michon gave the first description of spinal SAH in 1928, which he likened to being stabbed in the spine (poignard = French for dagger) [40].

Unlike other SVMs, conus medullaris AVMs frequently produce radiculopathy and myelopathy at the same time, and the radicular deficits are often prominent [52]. Wilson et al. described myeloradiculopathy as the initial presentation for 63 % of patients. More than half of the study population had bladder or bowel dysfunction, and 75 % of the patients were ambulatory at presentation [60]. Overall, 31 % of their patients had a history of spinal hemorrhage.

In the pediatric population, AVMs have been associated with inherited disorders such as hereditary hemorrhagic teleangiectasia, familial cerebral cavernous hemangiomas, pulmonary AVMs, Klippel-Trenauway-Weber syndrome,

and Rendo-Osler-Weber syndrome [14, 24]. Most of the patients present with acute onset of symptoms, with spinal cord AVMs (44.4 %), perimedullary AVFs (23.6 %), and Cobb syndrome (13.9 %) reported as the most frequently diagnosed subtypes [14]. Spinal dural AVFs appear to be extremely rare in the pediatric population, findings that argue in favor of an acquired etiology for those lesions.

Treatment and outcomes

Indications

There is no data to support a standardized approach to the treatment of spinal vascular malformations. Most studies are based upon retrospective series that include less than 50 patients [10, 53]. Spinal dural AVFs tend to produce a slowly progressive motor and sensory myelopathy over the course of months to years. They rarely present with acute hemorrhage. Intervention—either by microsurgical or endovascular means—aims to halt or reverse this progression by eliminating flow through the abnormal fistulous connection and restoring normal spinal cord perfusion and intravascular pressures. Spinal AVMs, as well as perimedullary fistulae, however, are more likely to present with an acute neurological deficit secondary to intramedullary or subarachnoid hemorrhage. The goals of treatment in those lesions include prevention of future hemorrhagic events, evacuation of acute hemorrhage products or selective obliteration of parts of the malformation that are felt to be symptomatic (i.e., feeding artery aneurysms).

Microvascular treatment

Dural AVFs

The first successful surgical treatment of a spinal vascular malformation involved a thoracic laminectomy performed by Charles Enberg at Mount Sinai Hospital in 1914. He reportedly identified enlarged blood vessels adjacent to a thoracic nerve root, excising several centimeters of the abnormality where it penetrated the dura, and the patient recovered almost completely [1]. Later efforts were initially less successful as they often involved stripping the entire venous complex off the surface of the spinal cord, presumably incurring cord ischemia or worsening pre-existing venous hypertension. It was not until Kendall and Logue identified the critical pathology of the dural AVF, later corroborated by Symon and Oldfield, that microsurgery became an almost uniformly successful treatment modality for these lesions.

Dural AVFs (Fig. 1a, b) involve one or several fistulous connections between a dural branch of a radicular artery and radicular vein located along the inner surface of the dura and laterally at the nerve root sleeve, most commonly in the thoracic or lumbar spine. Once the level and side of the lesion is identified, the exposure of the lesion typically is relatively straightforward. For classic lesions located at the nerve root sleeve, a laminectomy or laminoplasty is completed ipsilateral to the lesion. The laminectomy may extend a level above or below the lesion to allow adequate access and ability to open the dura rostral and caudal to the pathology and may extend laterally to the level of the pedicle above the involved neural foramen. The draining vein or veins are often abnormal, enlarged, and arterialized. The fistulous connection is identified and either a microsurgical clip is placed at the point of connection between the artery, and the vein or the fistula is coagulated and subsequently cut [37]. The use of intraoperative ICG angiography has proven helpful in identifying the pathology and confirming obliteration of the fistula [45, 57]. Electrophysiological monitoring of MEPs and SSEPs may be a useful adjunct during surgery to minimize the risk of cord injury and vessel sacrifice as changes in MEPs or SSEPs after

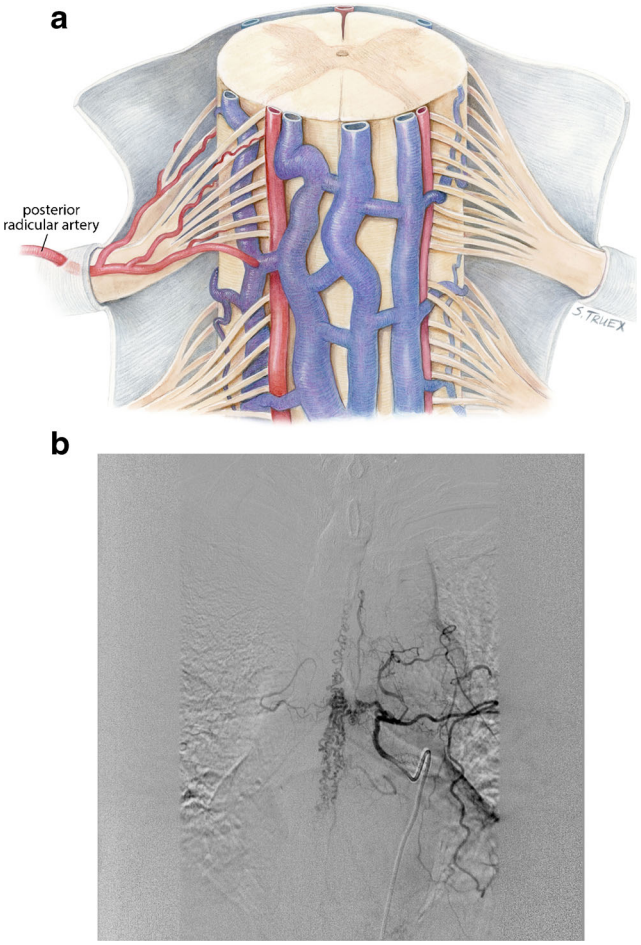

Fig. 1 **a** Schematic drawing of an intradural dorsal arteriovenous fistula. Notice the fistulous connections between a dural branch of the posterior radicular artery and radicular vein, and the resultant engorgement and dilatation of the venous plexus. The fistula is usually located along the inner surface of the dura and laterally at the nerve root sleeve. **b** Selective catheter spinal angiography demonstrating a similar lesion with striking dilatation of the medullary venous plexus network

temporary vessel occlusion with microclips may potentially be reversible by clip removal [4, 38]. Once the fistula is obliterated, one may see the involved arterialized red draining vein develop stasis and a purple hue. Definitive confirmation of AVF resection requires a postoperative spinal angiogram. A standard watertight dural closure is then completed to prevent cerebrospinal fluid leak and pseudomeningocele formation. Instrumented arthrodesis may be required in cases where significant bone removal of the ipsilateral facet complex and pedicle was necessary for accessibility [53].

Spinal AVMs and perimedullary AVF

Spinal AVMs are less well-defined vascular lesions than dural AVFs, and as such, their surgical treatment is less well-characterized. From a microsurgical standpoint, the anatomy of the individual lesion often dictates the goals of surgery and surgical approach. The location of the AVM within the spinal canal determines the extent of bony removal and whether a posterior, posterolateral, or anterior approach is warranted. Most lesions are accessible through a posterior laminectomy and partial facetectomy. Intradural AVMs that are ventral or ventrolateral to the spinal cord may warrant a generous arachnoid dissection and dentate ligament resection to facilitate mobilization of the spinal cord medially. Nerve roots—particularly, thoracic—may need to be sacrificed intradurally for further exposure. Perimedullary AVFs (Fig. 2) are fed by radiculomedullary arteries which drain to superficial perimedullary veins, in contrast to dural AVFs. Their surgical treatment, like dural AVFs, involves disconnecting the fistulous sites. Glomus AVMs (Fig. 3a, b) contain a nidus that resembles that of a brain AVM and tend to be intramedullary. Depending on their location within the spinal cord they may not be amenable to surgical resection or may carry an obligatory risk of postoperative neurological deficit. Resection of the nidus may require a myelotomy, which traditionally can be dorsal midline, dorsal root entry zone, lateral, or anterior midline. Pathological features such as feeding artery aneurysms or varices may also be targeted for resection in a focused manner in an attempt to minimize the risk of postoperative deficit from resection and to prevent further lesional hemorrhages or edema. For glomus AVMs with a significant intramedullary component, some authors have recently advocated subtotal resection of the extramedullary component of the lesion to minimize postoperative morbidity [55]. In this so-called pial resection technique, feeding arteries and draining veins along the surface of the spinal cord are coagulated and divided while minimizing subpial dissection. Myelotomies may then be reserved for intramedullary hematoma evacuation and fenestration of associated intramedullary syrinx. Complex spinal AVMs and AVFs require a multimodality approach that utilizes both microsurgery and endovascular embolization effectively.

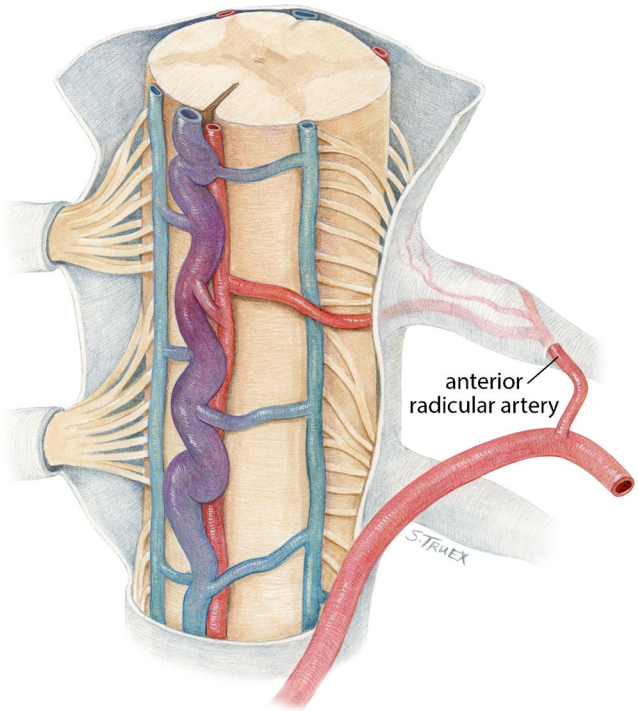

Fig. 2 Schematic drawing of an intradural ventral arteriovenous fistula (i.e., type IV AVM or perimedullary AVF). Notice the ventral fistulous connection between the anterior spinal artery and the venous plexus network

Endovascular treatment

Endovascular treatment of SVMs was initially described by Doppman et al. in 1968 [1]. Since then, the adjunct of modern spinal angiography, better microcatheter navigability and the development of liquid embolic agents such as n-butyl cyanoacrylate (nBCA) and Onyx have vastly expanded the role of embolization in the treatment of the various types of spinal vascular malformations. For some of those lesions, surgery remains the treatment of choice, particularly when the malformation vascular supply is in intimal association with the ASA, PSA, or artery of Adamkiewicz; in those cases, the risk of spinal cord ischemia and worse neurological function with curative embolization may be prohibitive. Recent reports have shown high rates of complete angiographic obliteration, and similar results on long-term neurological outcome with minimal morbidity [10, 12, 41, 47, 53].

Except for a few cases where preoperative embolization is the treatment goal, the use of particle embolization (polyvinyl alcohol (PVA), embospheres, gelfoam) is not indicated and has been largely abandoned, due to its high recanalization rates [15, 41, 47, 48]. The utility of endovascular treatment as monotherapy for spinal vascular malformations is directly dependent on the lesion subtype, its angioarchitecture, and embolic agent selection.

The higher rates of angiographic obliteration after endovascular treatment are described for spinal dural AVFs.

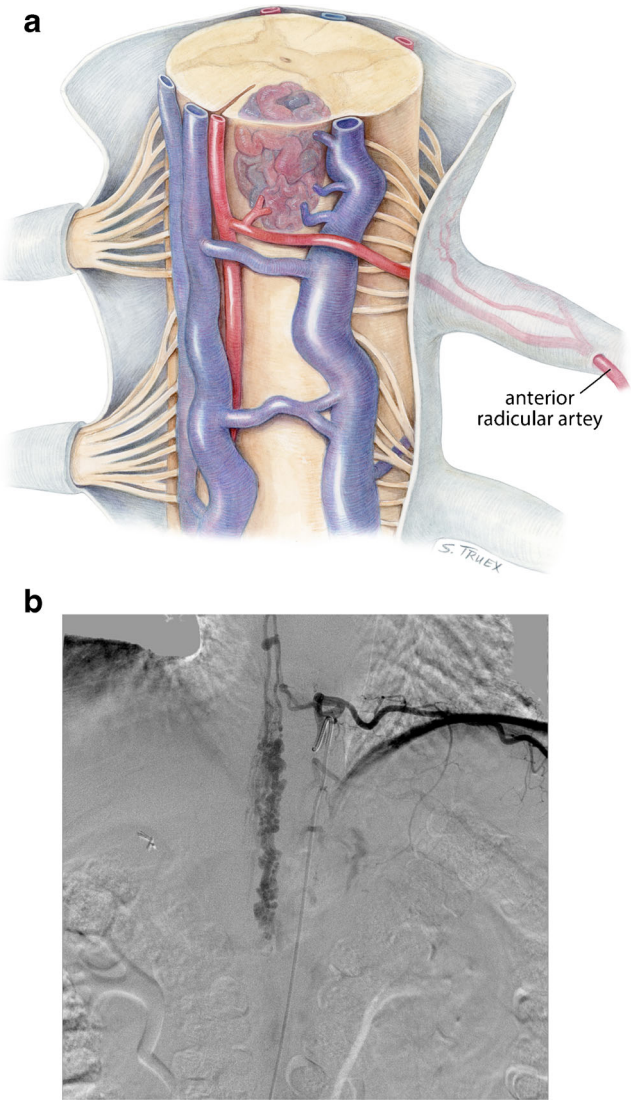

Fig. 3 **a** Schematic drawing of an intradural intramedullary arteriovenous malformation (i.e., glomus AVM). The AVM nidus can be compact—as shown here—or diffuse, but it is primarily parenchymal. **b** Selective catheter spinal angiography showing a large intradural intramedullary arteriovenous malformation with diffuse nidus. Notice the enlarged anterior spinal artery and artery of Adamkiewicz with an associated flow-related aneurysm

In fact, several authors preconize embolization as the treatment of choice [10, 13, 17, 41, 46, 53]. The goal of embolization is obliteration of the fistulous connection as well as the proximal aspect of the arterialized draining vein. Collateral supply must be ruled out at the time of treatment by injections at the correspondent levels on the contralateral side, as well as adjacent segmental arteries above and below the fistula. The initial obliteration rates vary from 25 to 100 % (depending on the embolic agent used), with up to 76 % recurrence rates on the early PVA series [15]. Recurrence is much less frequent in the cases treated with nBCA or Onyx (0–25 %). The results for intradural AVFs are more heterogeneous, in part due to the significant variability in the nomenclature. The lesions with progressively larger shunts and marked dilated venous network appear to be the ones with better results, with initial obliteration rates of 67–100 % [15].

The role of embolization for the treatment of spinal cord AVMs has been studied by several authors [7, 10, 31, 56]. In several institutions, it has become the treatment of choice [12, 13, 31, 47, 56]. Similar to what occurs with ruptured cerebral AVMs, after a spinal AVM presents with hemorrhage, most authors would agree with a delay in treatment to promote hematoma reabsorption and some improvement in neurological function to recover. However, contrary to its intracranial equivalent, partial treatment or obliteration of spinal AVMs may be sufficient to dramatically improve prognosis, especially in those where complete resection or embolization would incur in neurological deficits [31]. In unruptured spinal AVMs that have become symptomatic with venous congestion rather than hemorrhage, a reasonable goal of treatment would be to reduce the shunting volume. A great example of the application of this treatment paradigm would be on the management of metameric or juvenile AVMs (Fig. 4a–c), where surgical resection or complete endovascular obliteration is virtually unrealistic. In the less complex AVMs, though, the reported obliteration rates with liquid embolic agents varies from 33–100 %, depending on location and nidus size [7, 12, 15, 18, 31, 47]. Corkill et al. published in 2007 their single-center experience with embolization of intramedullary AVMs with Onyx [12]. Seventy percent of those patients had some history of spinal hemorrhage at the time of presentation. After an average of 1.23 sessions per patient, total or subtotal obliteration was achieved in 68.75 % of patients. Despite a relatively low rate of complete obliteration (37.5 %), improvement in neurological and/or functional status was seen in 82 % of treated patients, with a permanent complication rate of 4.3 % [12]. Partial obliteration of spinal AVMs may be acceptable also in patients with high-risk features, such as associated nidal or prenidal aneurysms or large venous varices. Differently from what is seen in the intracranial literature, there appears to be a protective effect against hemorrhage even with partial obliteration of a spinal AVM. This has been studied on a recent pooled analysis of literature cases of glomus (type II) AVMs [18]. In this study, the overall annual hemorrhage rate was 4 %, increasing to 10 % in AVMs with previous hemorrhage. Despite a rate of complete endovascular obliteration of 33 %, no postembolization AVM hemorrhages were reported over a total of 240.7 patient-years. The reduction in the annual hemorrhage risk was statistically significant even in the subgroup of partially embolized AVMs.

Despite the improvement on the obliteration rates with endovascular techniques, the modern treatment of spinal vascular malformations relies heavily on a multidisciplinary approach. Even in high volume surgical centers, almost half of the SVMs are preoperatively embolized or treated with embolization alone [4, 24, 26, 39, 45, 52, 55, 58, 60]. The use of

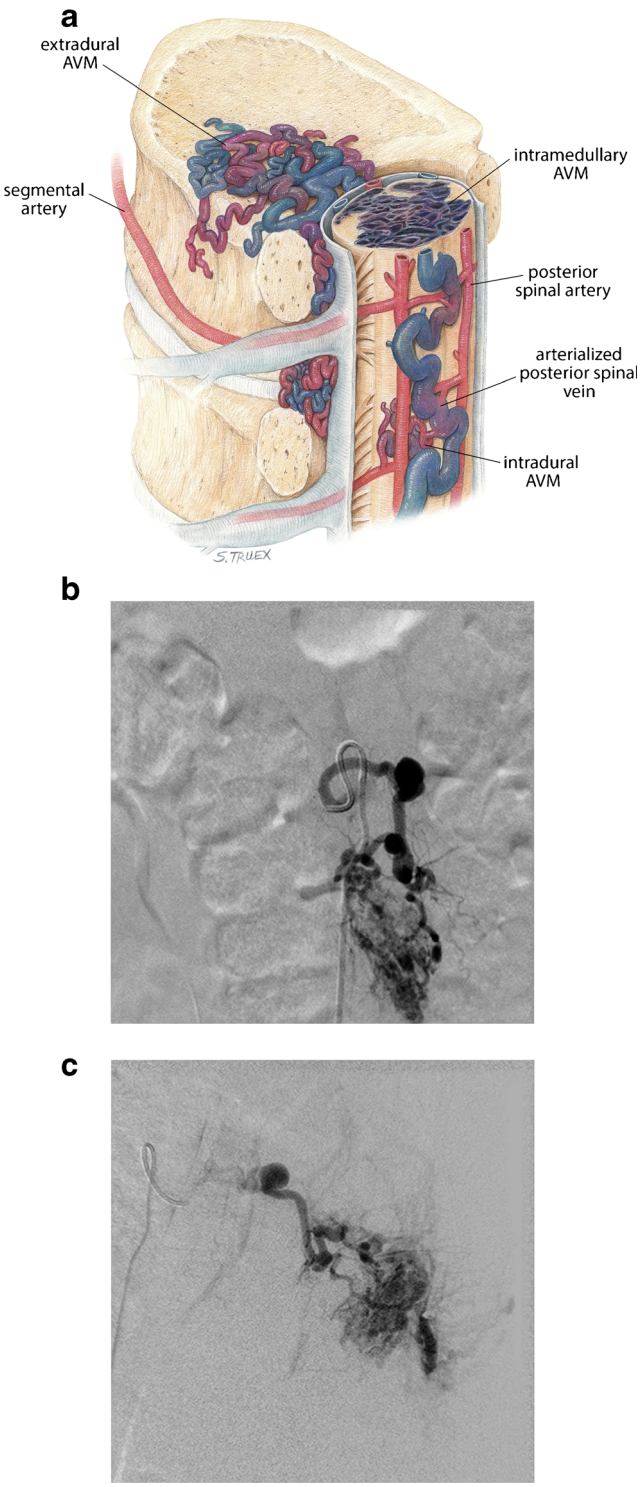

Fig. 4 **a** Schematic drawing of an extradural-intradural arteriovenous malformation (i.e., metameric AVM). Notice the two distinct nidus components—intramedullary and extradural involving the vertebral bodies and ventral epidural space. AP (**b**) and lateral (**c**) projections of a selective catheter spinal angiography, demonstrating a large metameric arteriovenous malformation. Notice the enlarged radicular artery with flow-related and intranidal aneurysms

endovascular techniques to exclude high-risk features or obliterate deep arterial feeders otherwise not easily approached by microsurgery alone is of paramount importance to decrease the perioperative blood loss, minimize the spinal cord dissection injury and the incidence of postoperative new or worse neurological deficits.

Radiosurgery

Radiosurgical treatment of SVMs has not been extensively studied and, thus, is not recommended. Over the last decade, two reports have described the use of multisession CyberKnife radiosurgery for treatment of intramedullary spinal cord AVMs [42, 51]. Overall, these results suggest a potential benefit of radiosurgery on hemorrhage risk; however, its effect on angiographic obliteration and long-term treatment results are yet to be determined.

Outcome

The treatment paradigm of spinal vascular malformations has significantly changed over the last three decades. The lower treatment morbidity has been coupled with improvements in long-term obliteration rates, making conservative management a suboptimal treatment choice. Several SVMs can be safely treated with a multimodality approach that involves preoperative embolization and surgical resection. The treatment success rates still depend directly on the lesion subtype and mode of presentation. The diversity of lesions and their rarity may help explain the paucity of adequate studies on the natural history of SVMs. Most of the treatment recommendations and outcomes published are based on case series or anecdotal experiences, and any generalization of clinical practice into guidelines is set to failure.

Of all the vascular lesion subtypes, spinal dural arteriovenous fistulas represent the most widely studied group. Their surgical obliteration rates approach 100 %, and long-term functional improvement of 50 % or greater is consistently reported on the case series [10, 34, 35, 37, 39, 41, 45–47, 49, 54, 58]. The degree of preoperative neurological function correlates strongly with the extent of postoperative recovery, independently on the treatment modality used 58]. Motor symptoms tend to respond better to treatment (approximately 66 % overall improvement), while sensory symptoms such as numbness, dysesthesias or burning pain tend to improve less frequently (12–45 % of patients) 17]. Recovery of sphincteric dysfunction tends to be disappointing, with persisting symptoms in up to 73 % of patients. Nevertheless, clinical recovery is possible even for patients with severe deficits, including paraplegia. Treatment should not be withheld from patients who are severely affected, since surgery may still be beneficial [37, 54]. Although mild transient worsening of symptoms after surgery or embolization is common, it does not influence the short- or long-term outcome. Because many patients progress over a considerable period of time before a diagnosis is

Table 3 Case series of microsurgical treatment of spinal vascular malformations

Authors	Patients (*n*)	Mean age (years old)	Lesion type	Treatment modality (%)	Obliteration (%)	Mean follow-up	Outcome	Recurrence (%)	Complication (%)	Mortality (%)
Rangel-Castilla et al. (2014)[45]	110	42.3	AVF	S ± E (86.4), E (12.7)	95.5	30.5 months	Improved 71.4 %, stable 26.3 %	13.6	15.4	0
			AVM		75.5		Improved 43.6 %, stable 42.8 %	15		
Gross & Du (2014)*[19]	51	15	Juvenile AVM	E (44), E + S (24), S (9)	32	2.6 years	Improved 73 %, stable 10 %	–	–	–
Cho et al. (2013)[11]	64	59	SDAVF	E (40), S (19), E + S (4)	94	20 months	Improved 50 %, stable 41 %	0	23.4	0
		32	PMAVF		68	42 months	Improved 58 %, stable 37 %			
		24	AVM		50	56 months	Improved 33 %, stable 25 %			
Gross & Du (2013)*[18]	293	29.1	Glomus AVM	S (68.2)	78	774.8/patient-years	Improved 57 %, stable 31 %	0.9/patient-year	–	–
				E (31.7)	33		Improved 66 %, stable 21 %	11/patient-year		
Velat et al. (2012)[55]	20	30	Glomus AVM	S ± E (100)	75	45.4months	Improved 55 %, stable 45 %	0	5	0
Wilson et al. (2012)[60]	16	34	Conus AVM	S ± E (100)	88	70 months	Improved 43 %, stable 43 %	19	7	0
Bostrom et al. (2009)[6]	20	–	Glomus AVM	S (65), E + S (35)	78.5	55 months	Improved 20 %, stable 75 %	15	15	0
Du et al. (2009)[16]	72	9	AVF, AVM, CM	S (14), E (54), E + S (28)	64	–	Improved 40 %, stable 51.7 %	–	–	0
Zozulya et al. (2006)[63]	91	42.9	AVF	S (76.9), E (14.3), E + S (8.8)	100	–	Improved 82.4 %, stable 11 %	0	–	0
			AVM							
Steinmetz et al. (2004)**[53]	19	60	SDAVF	S (100)	100	35 months	Improved 55 %, stable 34 %	0	5	0
Connolly et al. (1998)[11]	15	28	Glomus AVM	S ± E (100)	94	8.5 years	Improved 40 %, stable 53 %	20	7	0
Rosenblum et al. (1987)[47]	81	49	AVF	S (85)	100	3.7 years	Improved 72 %, stable 28 %	0	–	1.2
		27	AVM		59		Improved 33 %, stable 51 %			

AVM arteriovenous malformation, *AVF* arteriovenous fistula, *SDAVF* spinal dural arteriovenous fistula, *PMAVF* perimedullary arteriovenous fistula, *S* surgery, *E* embolization

*Pooled analysis; **Single-institution series and meta-analysis

made, it can be argued that the delay in diagnosis rather than the degree of neurologic impairment is the major reason for incomplete recovery.

Outcomes of microsurgery for dural AVF (Table 3) vary based on the lesion complexity, the surgeon's experience, and perhaps most importantly, the preoperative neurological status of the patient. Obliteration rates on postoperative angiography in several modern series are typically in the range of 94–100 % with recurrence rates typically less than 15 % [10, 45, 53]. The vast majority of patients are either clinically improved or stable postoperatively. Finally, complication rates have been reported to range from 5–15 % in the modern era and typically include pseudomeningocele, spinal instability, or worsened neurological deficit.

The high initial recanalization rates with endovascular treatment using particles in the 1980s and 1990s have been overcome with the advent of nBCA and Onyx, and in many centers, embolization has become the treatment of choice [13, 15, 41, 47, 53]. However, surgical intervention historically has higher rates of obliteration, with lower rates of recurrence and comparable morbidity to endovascular treatment, and is the treatment of choice of several authors [26, 37, 39, 45, 53]. Radiological findings on MRI do not appear to be a reliable predictor of outcome, as neither the extent of preoperative nor the change in postoperative T2 signal abnormality correlates with postoperative clinical disability [17].

Distinct from spinal dural AVFs, perimedullary or intradural ventral AVFs are rare; most of the case series with long-term follow-up and treatment results are relatively new. The preferred treatment modality differs by subtype (which takes into consideration the number and location of feeders and size of the fistulous component). There is general consensus that the smaller lesions with single or few arterial feeders (types A and B) are better treated with surgery, while the larger lesions (type C) are usually managed with endovascular techniques. Using a multidisciplinary approach, Cho et al. reported successful obliteration of 70 % of perimedullary AVFs, with 95 % of favorable outcomes in long-term follow-up [10]. The majority of lesions with complete obliteration were types A and B and were treated with surgery. Similar results have been recently published by other authors [45].

Due to the heterogeneity and the complexity of the lesions, spinal AVMs represent the subgroup with the worst obliteration rates (32–94 %), even in multimodality groups. Nevertheless, treatment is justifiable if one takes into consideration the high annual hemorrhage risk and their characteristic stepwise deterioration. On a pooled analysis of the literature on glomus AVMs, Gross and Du estimated an overall hemorrhage risk of 4 %, increasing to

Table 4 Clinical series of endovascular treatment of spinal arteriovenous malformations

Classification	Authors	Year	Patients (*n*)	Endovascular treatment	Embolizate	Initial obliteration (%)	Recurrence (%)	Outcome
Extradural	Rangel-Castilla et al.	2011	7	6	Onyx, nBCA	100	0	57 % excellent recovery, 43 % persistent symptoms
Intradural dorsal	Morgan et al.	1989	17	14	PVA	88	76	88 % improved
	Merland et al.	1990	63	36	iBCA	NR	NR	80 % improved
	Lundqvist et al.	1990	11	11	iBCA, PVA	NR	9	55 % improved, 45 % unchanged
	Niimi et al.	1997	49	49	iBCA, nBCA,	80	16	98 % initial improvement
	Van Dijk et al.	2002	48	44	nBCA	25	0	100 % improvement
	Narvid et al.	2008	63	39	nBCA	69	25	65 % improved gait
	Nogueira et al.	2008	3	3	Onyx	100	0	100 % improvement
Intradural ventral	Rodesch et al.	2005	32	18	nBCA	Macro 67/ Micro 75	NR	100 % improvement
	Cho et al.	2005	19	11	nBCA, PVA	45	NR	45 % improvement
Type A	Oran et al.	2005	5	5	nBCA	80	0	100 % improvement
Type B	Lundqvist et al.	1990	2	2	PVA	100	NR	NR
Type C	Casasco et al.	2012	6	6	nBCA, coils	100	0	100 % improvement
	Guegen et al.	1987	4	4	Balloons, gelfoam	50	NR	50 % improved, 50 % stable
	Ricolfi et al.	1997	12	12	Balloon, gelatin	66	NR	50 % good
Intramedullary	Biondi et al.	1990	35	35	PVA	3	80	63 % improved, 20 % worsened
	Corkhill et al.	2007	17	17	Onyx	37	6	82 % improved
Conus medullaris	Wilson et al.	2012	16	8	Onyx, nBCA	88	13	43 % improved, 43 % stable, 14 % worsened

SAVM spinal arteriovenous malformation, *iBCA* isobutyl 2-cyanoacrylate, *nBCA* n-butyl cyanoacrylate, *PVA* polyvinyl alcohol, *NR* not reported

Modified from: Ducruet AF, Crowley RW, McDougall CG, Albuquerque FC. J Neurointervent Surg 2013; 5: 605–611. Used with permission

10 % for AVMs with previous hemorrhage [18]. Even considering only the metameric or juvenile AVMs, historically thought to have lower hemorrhage risk than glomus AVMs, the same authors reported in a similar subsequent study an annual hemorrhage risk of 2.1 %/year [19]. The treating neurosurgeon might also keep in mind the substantial difference in treatment goals between spinal AVMs and their intracranial correspondents. As it has been noted by several authors, significant clinical recovery and functional improvement do not necessarily correlate with completeness of angiographic obliteration [10, 12, 45, 55]. For example, targeted embolization of specific AVM angioarchitecture features (such as nidal aneurysms) may protect against future devastating events, such as intramedullary hemorrhage [12, 47, 48].

Tables 3 and 4 represent a contemporary analysis of the published series on microsurgical and endovascular treatment of spinal vascular malformations.

Conclusions

Spinal vascular lesions are rare but represent a formidable challenge for the treating neurosurgeon. Despite their pathological similarities with their intracranial counterparts, their clinical impact is often comparatively worse. A thorough understanding of their complex spinal vascular anatomy and pertinent radiological findings is of paramount importance for the correct diagnosis and prompt treatment intervention. The clinical presentation may have an acute or more protracted presentation with progressive neurological decline. In both situations, early treatment is associated with better long-term neurological outcome. The treatment success rates still depend directly on the lesion subtype and mode of presentation. A modern, multimodality approach involving endovascular embolization and microsurgical resection has been shown to provide high obliteration and low recanalization rates, with better overall results in spinal arteriovenous fistulas, when compared to arteriovenous malformations. The advent of modern liquid embolic agents has revolutionized the endovascular treatment of those lesions, but microsurgical resection remains a viable option with excellent long-term results.

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Compliance with ethical standards

Conflict of interest The authors declare that there are no conflicts of interest.

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Comments

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The authors provide a comprehensive summary on spinal vascular malformations. We like to make a few comments on the type 1 spinal dural arteriovenous fistulas (SDAVFs), which represent the most common spinal vascular malformation. Unfortunately, despite advances and widespread utilization of noninvasive neuroimaging, the diagnosis continues to be delayed [1, 7]. When we compared the time interval from symptom onset to final diagnosis between those patients treated from 1986 to 1999 and those treated between 2000 and 2008, we were surprised to find that the median delay in diagnosis had not changed over the time interval being 12 months in both periods [7]. At times, delays in diagnosis are related to the difficulty of performing a complete spinal angiography in some of these patients who are often elderly with advanced atherosclerosis. We routinely use magnetic resonance angiography (MRA) as a screening tool helpful in narrowing down the segments most likely bearing the fistula [3]. In this manner, catheter angiography can be started on the segments in question. In the exceptional cases where catheter angiography cannot localize the fistula, advanced MRI techniques such as time-resolved MRA and PC-Fiesta imaging can be helpful in identifying the site of the fistula [6].

As noted by the authors, type 1 DAVFs have a striking male predominance (usually in the 7th and 8th decade of life). The presence of clinical symptoms and MRI findings suspicious for a type I DAVFs in a young patient or in a woman, should raise the suspicion of an epidural fistula or a paraspinal AVM with secondary retrograde intradural venous drainage [2]. With increased awareness of this entity and better imagine techniques, epidural AVFs now account for about 30 % of the spinal vascular malformations that we see. Differentiation between an epidural AVF and a type 1 AVF is an important one, as epidural fistulas are amenable to successful and permanent obliteration with endovascular trans-arterial embolization [5]. Moreover, epidural AVFs are often more difficult to obliterate surgically then the classic type I SDAF because the AV shunt is often located in the ventral epidural venous plexus.

After complete obliteration of a type 1 DAVF, the degree of clinical improvement is highly variable. The vast majority of patients experience some degree of improvement of motor function. As noted by the authors of this review, improvement of sensory function and sphincter control is less dramatic. Interestingly, there is no correlation between the degree and pattern of improvement of signal changes on MRI and the degree of clinical improvement [4]. Resolution of the flow voids on MRI and of serpiginous vessels on MRA is an excellent predictor of complete fistula obliteration [4], and in straightforward cases, we rarely perform a postoperative catheter angiography after surgical obliteration.

After successful treatment of a type I DAVF, it is not uncommon in patients who had presented with myelopathy to complain of delayed subjective recurrence of symptoms in association with an intercurrent systemic illness. This is usually related to the loss of the ability to compensate for the lost function in the presence of an intercurrent systemic illness and not to the recurrence of the fistula. Similarly, patients can report a subjective sense of increasing weakness and fatigability 6 to 18 months after successful treatment. This is often related to increased muscular tone which occur as a result of some degree of "spinal cord healing." These symptoms usually improve with pharmacological treatment of the spasticity. It is important to warn patients about these possible "setbacks" and their significance as to avoid excessive worrying and unnecessary expensive imaging studies.

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Flores et al. are to be congratulated for reporting a comprehensive update on the modern classification systems, treatment and management strategies, and outcomes data for spinal vascular malformations. They note that endovascular embolization with Onyx or nBCA is the treatment of choice for dAVF (with the exception of perimedullary or intradural ventral dAVF with single or small feeders) and often can result in cure. Even for spinal AVM, unlike its cranial counterpart, complete embolization can result in a durable cure and even partial embolization can improve neurological function. Before a proper treatment strategy can be developed, however, comprehensive spinal angiography must be performed. The authors dutifully note the obligation of the angiographer to image all of the thoracic intercostal and lumbar radicular arteries not only to look for any fistulous connection, but also to identify the artery of Adamkiewicz. Critical to comprehensive angiography, however, is knowing what to image when the thoracic and lumbar segmental arteries fail to show any fistulous connection. The authors mention multiple strategies including injecting the lateral sacral arteries, arterial supply to the cervical cord and posterior fossa, as well as the bilateral internal iliac arteries. These are very useful pearls to the article readership and are much appreciated.