

Spinal arteriovenous malformations

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Abstract

Pathologies that affect the spinal cord are diverse. In addition to trauma, common etiologies of myelopathy include autoimmune, infectious, neoplastic, vascular, and hereditary-degenerative diseases. Physicians should be aware of the possibility of a spinal arteriovenous malformation (sAVMs) when facing a challenging case. Performing an adequate clinical history and neurological examination is the key element in the proper approach to these pathologies. However, a better determination of the problem can be obtained with the help of neuroimaging studies. Given the rare nature of sAVMs, the purpose of this article is to review the spinal vascular anatomy, the pathogenesis and pathology of arteriovenous malformation, discuss key points about their classification, and summarize the imaging findings and treatment approaches available.

Key words: Arteriovenous malformations. Spine. Review.

Malformaciones arteriovenosas de la médula espinal

Resumen

Las patologías que afectan la médula espinal son diversas. Además del trauma, las etiologías comunes de la mielopatía incluyen enfermedades autoinmunes, infecciosas, neoplasias, vasculares y hereditarias degenerativas. Los médicos deben mantener en mente la posibilidad de una malformación arteriovenosa espinal cuando se enfrentan a un caso difícil. La historia clínica y la exploración neurológica adecuada siguen siendo pieza clave en el adecuado abordaje de estas patologías, sin embargo, puede obtenerse una mejor determinación del problema con ayuda de los estudios de neuroimagen. Dada la naturaleza rara de las malformaciones arteriovenosas espinales, el propósito de este artículo es revisar la anatomía vascular espinal, la patogenia y la patología de las malformaciones arteriovenosas, discutir puntos clave sobre su clasificación y resumir los hallazgos por imagen y los tratamientos disponibles.

Palabras clave: Malformaciones arteriovenosas de la médula espinal. Lesiones vasculares. Médula espinal. Epidural. Revisión.

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Introduction

Arteriovenous malformations (AVMs) are rare lesions, which are often diagnosed late because clinicians do not think about them. The best estimates for the detection of an AVM are 1/100,000 population per year (about 3000 new cases detected each year in the U.S.), with prevalence about 10/100,000¹. According to Moore et al., they represent 2.5% of cases of spastic paraparesis or quadripareisis compared with cervical spondylotic myelopathy (24%), tumors (16%), multiple sclerosis (18%), and motor neuron disease (4%)².

In this article, we briefly summarize the anatomy, clinical presentation, classification, and treatment of vascular spinal cord lesions. Given the unique nature of spinal AVM (sAVM), our purpose was to construct a bibliographic review about the spinal vascular anatomy, the pathogenesis and pathology of AVMs, discuss key points about their classification, and summarize the imaging findings and treatment approaches available. In this regard, an electronic search strategy of the literature available was conducted (January 2020) using PubMed, Access Medicine, Embase, and UpToDate to retrieve publications regarding the key points mentioned previously. The terms used in the literature search were our keywords as subject headings from the Medical Subject Headings thesaurus. Publications in both English and Spanish were included in the study. The reference sections of the included publications were also screened for potentially relevant references. In addition to the electronic search strategy, other sources were examined for potentially relevant publications. These included different books on neurology and spinal cord disorders.

Vascular anatomy of the spine

Spinal vascular anatomy incorporates not only the vascular supply to the cord but also that to the adjacent structures, including the nerve roots, dura, and paraspinal musculature³.

Arterial supply

Anterolateral supply

Superficially, the anterior two-thirds of the spinal cord is supplied by the anterior spinal artery (ASA), which originates from a medial branch of the intracranial vertebral artery on both sides, which join together in the midcervical spine (C2-C4) and travel along the anterior

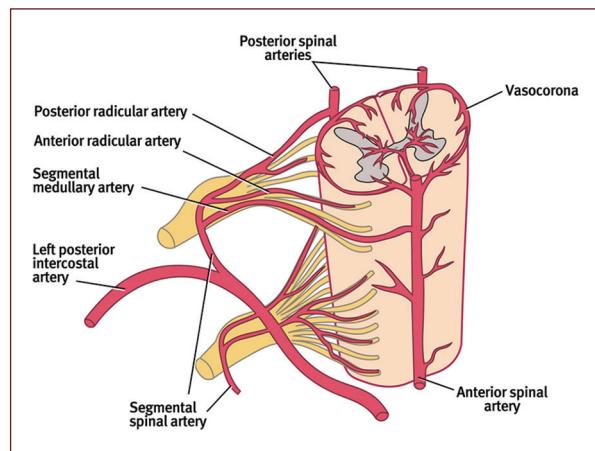


Figure 1. Blood supply of the spinal cord.

medial fissure (Fig. 1). The dominant contributors to this artery are six to eight segmental radiculomedullary arteries, which typically include:

- An artery at C3 (also referred to as the artery of the cervical enlargement, from the vertebral artery),
- C6 (from the deep cervical artery),
- C8 (from the costocervical trunk),
- T4/T5
- Moreover, the anterior great radiculomedullary artery (artery of Adamkiewicz), which usually originates on the left side between the T8 and L2 level^{3,4}.

This anterior blood supply is also referred to as the “sulcocommissural system” or centrifugal system and gives rise to 200-400 sulcocommissural arteries within the ventral sulcus to supply the anterior commissure, dorsal nucleus of Clarke, corticospinal tract, spinothalamic tract, and the anterior portions of the cuneate and gracile fasciculi⁵.

Posterior supply

Superficially, the posterior one-third of the spinal cord is supplied by a pair of posterior spinal arteries (PSA), which originate at the level of the foramen magnum from:

- Branches of the intradural portion of the vertebral artery or
- The posterior inferior cerebellar artery, with
- Contributions from 6 to 11 posterior radiculomedullary arteries as they descend caudally in a discontinuous manner³.

This posterior system is also referred to as the dorso-lateral pial supply or centripetal system (Fig. 1). This

network gives off radial arteries that circumferentially run along the spinal cord, reaching anastomosis with the ventral system (forming a pial plexus called the vasocorona), and giving off axially oriented perforating branches into the white matter⁵.

Inferiorly on the spinal cord, the ASA and PSA systems build a reticulating network around the conus in a structure commonly referred to as the arterial basket of the conus medullaris^{6,7}.

ASA

The ASA descends over the central sulcus of the anterior cord all the way to the conus medullaris. En route, it acquires multiple feeders: in the cervical spine, the most prominent feeder is the artery of the cervical enlargement; in the thoracolumbar spine, the most prominent feeder are multiple anterior radiculomedullary feeders from segmental branches directly from the aorta, and in the lumbar spine, there usually are four pairs of lumbar segmental arteries from L1-L4 and lower lumbar segmental arteries arising from the common iliac arteries and the median sacral artery^{4,6}.

Venous drainage

The venous system of the spinal cord is not wholly analogous to the arterial anatomy. Venous drainage uses to be more regional with a central and peripheral venous system.

Peripheral system

The peripheral, or radial, veins originate in the capillaries at the gray-white junction and are directed centrifugally.

Central system

The central, or sulcal, veins drain from the medial aspects of both halves of the spinal cord, specifically from the anterior horns, anterior commissure, and the white matter in the anterior funiculus⁸. Intrinsic spinal cord veins and small superficial pial veins lead into the superficial longitudinal median spinal cord veins. Eventually, these drain into a set of veins that mirror the arterial system, but have many anastomoses creating a vast interconnected network⁹. The venous blood reaches the epidural plexus and the extraspinal veins and plexus, with a reflux-impeding mechanism within the dura mater⁸. Ultimately, the extraspinal plexus joins the

cava system, mainly the innominate veins at the cervical level, the azygos vein at the thoracic level, and the ascending lumbar vein at lumbar level⁶.

Pathogenesis and pathology of sAVMs

sAVMs represent an abnormal, often tiny, arteriovenous shunt located within the dura of the spinal cord. Afferent supply to the lesion is through a radicular artery, usually at a level with no associated arterial supply to the spinal cord¹⁰. Blood flow through the fistula runs through a radicular vein in a retrograde manner to the coronal and pial venous plexus, which becomes dilated and tortuous. Outflow impairment to the epidural drainage system or blockage of venous return in the cava system explains this venous dilatation¹¹. Since there is no involvement of arterial supply to the spinal cord, no possibility of arterial steal may be invoked to explain the neurologic dysfunction in these patients¹⁰. Venous congestion is now recognized as a primary source of neurologic disability with sAVMs. Recent studies have validated the elevated pressure in the draining vein as a causative agent in myelopathy, and shown pathologic correlates, including hyalinized small blood vessels, perivascular/intraparenchymal lymphocytic infiltration, glial cell proliferation, and neuronal degeneration¹².

Although impairment of venous drainage from the spinal cord parenchyma is most significant at the level of the shunt, the spread of venous hypertension in the cranial and caudal directions causes damage to the cord over a long distance¹¹. It is the caudal end of the spinal cord commonly first affected by congestive edema and ultimately infarction, regardless of the level of the fistula¹².

Hemorrhage can also precipitate neurologic decline: 25% of sAVMs patients will present with a ruptured lesion, as evidenced by subarachnoid hemorrhage (SAH), intraparenchymal hemorrhage, or a combination. Hemorrhage can injure neurologic structures either directly through disruption of fiber tracts or indirectly with mass effect. Venous engorgement, venous varices, and aneurysmal dilatations can further contribute to mass effect and compress the spinal cord or nerve roots¹².

Classification

The first classification system originated from Virchow in 1858, who observed lesions at autopsy that he referred as neoplasms or vascular tumors, and that was either considered angioma cavernosum if there was no parenchyma between the blood vessels, or angioma racemosum (further subdivided into telangiectasias and

angioma racemosum arteriale sive venosum [i.e., sAVM]^{13,14}. In 1916, Elsberg proposed an updated scheme to include aneurysms of spinal vessels; angiomas and dilated posterior spinal veins or hemorrhoids of the spinal cord¹⁵. Since then, many classification systems have been reported and have changed over time in the literature. Until now, a detailed and widely accepted systematization of spinal vascular malformations has not been proposed.

During the 1960s, there was enormous progress in understanding and treating sAVMs. Three major centers (including the National Institutes of Health in Bethesda, the National Hospital for Neurology and Neurosurgery in London, and Hôpital Lariboisière in Paris), generated a combined effort of independent and collaborative investigation that resulted in what is known as the American/English/French classification system. This system separates sAVMs into three types. Type I lesions (single coiled vessel) are dural sAVMs and consist of a radicular artery draining into an engorged spinal vein on the dorsal aspect of the dural sheath of a nerve root, and comprises 80% of sAVMs. Type II lesions (spinal glomus sAVMs) consist of a medullary artery feeding into a nidus, and then into a venous drainage system; they comprise 15-20% of sAVMs and cause damage via hemorrhage, mass effect, vascular steal, and venous hypertension. Type III lesions (juvenile spinal sAVMs) are intramedullary lesions supplied by pial, dural, and paraspinal arteries with an expansive and diffuse nidus that occupies the entire cross-sectional area of the spinal cord and invade the nearby vertebral body; the particularly distinct and unique attribute within this Type III category is the intradural and extradural extension of the lesion¹². A Type IV lesion was later described by both Djindjian et al. (1977)¹⁶ and Heros et al. (1986)¹⁷, consisting of a direct fistula between a spinal and a dilated perimedullary venous plexus without an intervening nidus; these lesions are intradural and extra/perimedullary. In 1980, Merland et al. further described three subtypes of Type IV lesions: in the first subtype, there is a single arterial feeder from the ASA, mild venous dilatation, and a single, small, and slow fistula. The second and third subtypes have multiple arterial feeders, the first having multiple sAVMs of medium size, whereas the latter has a single but giant sAVM and giant venous ectasia^{13,18}.

As we mentioned before, different authors have tried to find a clear systematization of spinal vascular lesions and several major classification systems appeared on the literature, including a description by Di Chiro et al. of sAVMs diagnosed and classified using spinal angiography in 1971¹⁹; a classification system by Mourier

and Merland based on a case series of intradural direct perimedullary sAVM treated with endovascular intervention in 1993²⁰; two classification systems based on a case series of sAVMs treated by microsurgery by Spetzler²¹ or endovascular interventions in 2002 by Rodesh and Lasjaunias²²; and a classification system based on a case series of extradural SAVMs treated by microsurgery and endovascular interventions in 2011 by Rangel-Castilla et al.^{23,24}. Most remarkably, in 2002, Spetzler et al. expanded upon the prior American/English/French system, offering new classifications of spinal vascular pathological entities based on anatomical, pathophysiological or angioarchitectural features, and in 2011 Patsalides et al. offered a classification based on hemodynamic criteria^{8,25}.

In addition to the many classification systems reported, the Bicêtre group classified spinal cord AVMs into three main groups:

1. Genetic hereditary lesions caused by a genetic disorder affecting vascular germinal cells, such as malformations associated with hereditary hemorrhagic telangiectasia.
2. Genetic non-hereditary lesions such as the Cobb syndrome (or spinal arteriovenous metameric syndrome), Klippel-Trenaunay, and Parkes-Weber syndromes, and
3. Single lesions that may reflect the incomplete expression of one of the situations above.

These include spinal cord, nerve root, and filum terminale arteriovenous lesions. Most of the spinal vascular malformations with pial and dural arteriovenous shunts are included in this group^{26,27}.

Epidemiology

Little has been published regarding the epidemiology of these lesions²⁸. sAVMs make up the most common vascular anomaly of the spine, with an estimated proportion of 60-80%³³. sAVMs account for 3-4% of all intradural spinal cord mass lesions²⁹, and dural sAVMs are the most common vascular malformation, accounting for 50-85% of all lesions³⁰⁻³².

Lad et al. analyzed National Inpatient Sample data from 1995 to 2006, screening admissions with a primary diagnostic code for spinal vascular malformation. In these 11 years, 3291 patients were admitted with a diagnosis of spinal vascular malformation, with an average of 299 annual admissions with a new diagnosis of sAVM; the majority of admissions corresponded to male patients (57%) in the 45-64-year age range (36% of admissions)³⁰.

Jellema et al.³³ analyzed all reported series with more than five patients affected by direct sAVMs, detecting there were 968 men against 210 women (ratio almost 5:1). The mean age at the time of diagnosis was found to be 55-60 years; patients under the age of 30 were rarely reported (14 patients under age 30 were found in the 1178 patients or 1%). The youngest patients reported were 22 years at the time of diagnosis. In their study was concluded that no patient under the age of 20 has ever been reported.

Clinical presentation

sAVM most commonly affects the upper thoracic and cervical spine. The sAVMs can be classified in those diagnosed by their clinical presentation and those present when making a diagnostic approach. Initial symptoms are often non-specific and include symmetrical or asymmetrical sensory symptoms such as paresthesia in one or both feet, and diffuse or patchy sensory loss (17-72%), gait difficulties and motor disturbances (50-81%), pain including back or radicular pain (13-64%), and micturition difficulties (4-75%). Other but less frequent symptoms include defecation problems and sexual dysfunction³³⁻³⁴. In most patients (40-63%), progression lasts for 1-3 years before the diagnosis is made, but a protracted course with a duration of > 3 years occurs in 10-34%. A gradually progressive course with stepwise deterioration is recorded in approximately 11-32% of patients^{34,35}. An acute onset is reported in 5-18% of patients. If symptoms develop within minutes to hours, they can mimic an ASA syndrome^{33,36}. The sudden episodes mostly occur after exercise, prolonged standing, and even singing and may disappear after rest³³.

At the time of diagnosis, there are often considerable neurological deficits and much of the clinical presentation depends on the type of the sAVM.

- Type I (dural arteriovenous fistulas [DAVFs]): the typical presentation is radiculomyelopathy followed by progressive neurological deterioration. SAH is very uncommon, and acute deterioration in neurological function is unlikely. Most are solitary lesions found in the thoracolumbar region. The fistula is located inside the dura, where a radiculomeningeal artery enters the corresponding radicular vein, close to the spinal nerve root^{8,33,37}.
- Type II (intramedullary glomus AVMs): the clinical course of these lesions is marked by progressive and fluctuating myelopathy, paraplegia and pain, overlaid by periods of acute neurological deterioration secondary to hemorrhage within the AVM. Sudden-onset

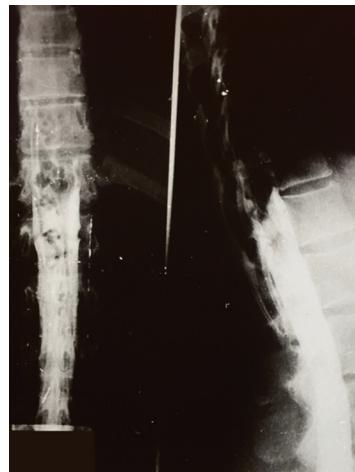


Figure 2. Myelography.

presentation (often with profound neurological impairment and possible transverse myelopathy) is frequent. Subarachnoid and intramedullary hemorrhage often occurs in these lesions. The mortality related to type-II malformations has been reported to be 17.6%. After initial hemorrhage, the prevalence of re-bleed is 10% within the 1st month and 40% within the 1st year^{8,33,37}.

- Type III (Juvenile AVMs): their clinical presentation is similar to that seen with Type II sAVMs. Acute onset of symptoms can occur secondary to SAH, whereas progressive motor and sensory deterioration, as well as sphincter disturbance, usually result from vascular steal, venous hypertension, or mass effect on the spinal cord and/or nerve roots from the dilated veins^{8,33,37}.
- Type IV (perimedullary AVFs): presentation occurs in the third-to-sixth decade. SAH is possible, with subsequent acute neurological deterioration. A gradual but progressive neurological deterioration is common^{8,33,37}.

Diagnosis

Imaging is an essential component in the diagnosis, management, and follow-up of patients with AVMs in the brain and spine. A wide variety of imaging modalities are available for its use, such as computed tomography (with its permutations: non-contrast, contrast enhanced, angiography perfusion, and myelography (Fig. 1) and magnetic resonance imaging (with its permutations: non-contrast, contrast enhanced, angiography, and myelography (Fig. 2). Although advances in

Table 1. Comparison between common causes of myelopathy

Disorder	Typical age of presentation	Clinical findings	Course
Infarction	Older adults	Profound weakness with decreased pain and temperature sensation below the lesion. Intact JPS, vibration	Acute
Motor neuron disease	Adults	Upper (increased tone, hyperreflexia) and/or lower (atrophy, fasciculations) motor signs. Normal sensory findings	Progressive, rate of progression may vary
Neoplastic disease	Older adults	Weakness below the level of the lesion, sensory loss	Subacute-chronic with acute worsening
Radiation myelopathy	Any age	Weakness, spasticity	Delayed months after radiation May be slowly progressive
sAVMs	Typically adults	Focal or radicular pain, weakness, sensory symptoms Neurogenic claudication	Acute, may be step-wise or slowly progressive
Spinal epidural abscess	Any age	back pain, focal weakness, sensory symptoms, fever	Subacute with acute worsening
Spinal epidural hematoma	Any age	Focal or radicular pain, weakness	Acute
Spondylotic myelopathy	Older adults	Gait disturbance, spasticity, atrophy, and weakness of extremities	Progressive, step-wise decline
Subacute combined degeneration	Any age	Numbness, paresthesia, weakness, gait disturbance. Decreased JPS and vibration Sensory ataxia	Slowly progressive
Transverse myelitis	Young adults and children	Focal sensory and motor symptoms Sensory level at/below the lesion	Acute or subacute

this noninvasive imaging, digital subtraction angiography continues to be the gold standard for diagnosing and characterizing the detailed anatomic localization, arteriovenous transit, and venous drainage patterns of sAVMs^{29,33,38}.

In sAVM, spinal angiography demonstrates a tangle of vessels at the level of the spinal cord parenchyma (Fig. 3). After delineation of the lesion, the neighboring vessels should be studied to identify potential feeders and to evaluate the effect of the lesion on the spinal cord. Anteroposterior angiography should be performed to identify feeding vessels and their origin from the anterior or posterior spinal artery (if any). Mass effect can distort the midline structures on frontal projection angiography and in this situation, the lateral projection angiography may help to delineate angioarchitecture. Three-dimensional angiography may help to define the relationship between the vascular malformations, bony structures, and the spinal cord. During the selective injection, contrast reflux into the lesion helps to identify the contributing feeders immediately above and below and is critical for planning endovascular treatment^{39,40}.

**Figure 3.** Angiography.

A careful diagnostic workup helps realign clinical reasoning that might otherwise lead these patients to

misdiagnosis and late specialized care, further delaying optimal management. Every patient with lower limb motor/sensory disturbances, micturition disorders, or reflex abolition not well explained by common findings (spinal stenosis, lumbar disk herniation, and spondylolisthesis) should have a focused clinical examination of the thoracic region and/or magnetic resonance imaging of the dorsal spine⁴¹. Table 1 summarizes the key clinical aspects of AVMs compared with their main differential diagnosis.

Management

All patients must have a thorough preoperative evaluation, including neurological, radiographic, and angiographic evaluations, to fully understand the location and anatomy of the malformations, and the strategy necessary to treat it. The decision to treat vascular malformations of the spine should be made after discussion between the various members of the team consisting of neurovascular surgeons, interventionalists, and radiation therapists, and with the patient or caregivers. Endovascular embolization should be considered as a standalone procedure or as an adjunct to microsurgical resection for spinal vascular malformations when appropriate³⁹⁻⁴².

Conclusion

Spinal DAVFs are rare but can cause serious gait and micturition disturbances. Delays in diagnosis and treatment result in poor clinical outcomes. When a common diagnosis fails to explain the symptoms such as thoracic myelopathy, epiconus syndrome, and conus medullaris syndrome, the possibility of sAVMs should be considered. This is particularly true after spinal trauma in young subjects or spinal stenosis or lumbar disk herniation in older individuals. A thorough exploration of abdominal reflexes or of the sensitivity of the abdominal/thoracic region, particularly when the topography of the deficit suggests a lesion of the spinal cord, should be performed.

Conflicts of interest

None.

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Ethical disclosures

Protection of human and animal subjects: The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data: The authors declare that no patient data appear in this article.

Right to privacy and informed consent: The authors declare that no patient data appear in this article.

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