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1. Introduction

Spinal cord vascular lesions can be broadly classified into one of the 3 following groups: neoplasm (hemangioblastomas and cavernous malformations), aneurysms, and arteriovenous malformations (AVM).[1],[2] Spinal AVMs account for 1% to 2% of vascular neurologic pathologies and 3%-12% of space-occupying pathologies of the spinal cord.[3]

Several classifications have been proposed for spinal AVMs.[2],[4]-[6] They can be divided into intramedullary AVMs and arteriovenous fistulas (AVFs). Based on anatomical characteristics, spinal AVFs are classified as extradural AVFs and intradural lesions, including spinal dural AVFs (SDAVFs) and perimedullary spinal AVFs (PMAVFs).[7],[8] Rangel-Castilla et al. classified extradural AVFs as type A or type B based on the availability of intradural venous drainage. In Type A spinal extradural AVFs, there is intradural venous drainage and arteriovenous shunting develops in the epidural space. Type B can be further classified as B1 and B2, both of which are limited to the epidural space without any intradural draining veins. The only difference between these two types is the presence or absence of compression on the spinal cord and nerve roots.[9] There is a paucity of literature about spinal AVFs and due to the very small incidence of these lesion, almost all available studies are small cases series. However, spinal AVFs can manifest with severe neurologic symptoms, leading to permanent neurologic deficit. Therefore, in this chapter we aim to review the available literature about spinal AVFs.

2. Literature search

A literature search of Medline through PubMed was performed using following search terms: ("Arteriovenous Malformations"[Mesh] OR "Arteriovenous Fistula"[Mesh]) AND "Spinal
"Cord." Then we limited our search to English-language articles and studies on human subjects. We expanded our literature search using the “related citation” option in PubMed. We also searched Google Scholar using the following keywords: spinal, arteriovenous fistula, extradural arteriovenous fistula, spinal dural arteriovenous fistula, and perimedullary spinal arteriovenous fistula. We also manually searched the reference lists of important papers to identify those that we missed during our primary electronic search. Data on epidemiology, classification, etiology, clinical manifestations, diagnosis, management, prognostic factors, and outcome of SDAVFs were extracted from selected articles.

3. SDAVFs

Although SDAVFs are rare, they are the most common spinal vascular malformation and are responsible for 70% of spinal cord arteriovenous shunts,[10] with an annual incidence of 5-10 cases per million.[11] It has been suggested that SDAVFs are acquired lesions, though their exact etiology remains unknown. SDAVFs are seen more frequently in elderly males and tend to affect the thoracolumbar segment more frequently than other spinal segments.[10]

SDAVFs are low-flow lesions and cause venous hypertension and chronic spinal cord hypoxia. Patients present with progressive myelopathy. Paraparesis is often the initial presentation, which is followed by root and/or back pain, sensory impairment, and sphincter dysfunction. [7] Due to non-specific presentations, SDAVFs are often diagnosed late. The mean time interval between onset of symptoms and diagnosis of SDAVF has been reported as high as 22.9 months (range 12 to 44 months).[12]-[14] Some authors suggest that there is a correlation between time until diagnosis of SDAVF and severity of symptoms.[5],[15] As mentioned earlier, the majority of authors consider SDAVFs to be acquired lesions and that they should be differentiated from congenital intradural PMAVFs.[16]

Digital subtraction angiography is the gold standard for diagnosis of spinal vascular malformations, including SDAVF[17]; however, magnetic resonance imaging (MRI) has been suggested as an accurate and reliable tool for diagnosis of SDAVF.[7] Intramedullary high-intensity changes on T2-weighted MRI are seen when SDAVF is present.[18] After treatment of SDAVF, intramedullary high-intensity changes start to reduce in the majority of cases between 1 and 4 months after treatment and these changes disappear in most cases between 2 weeks and 23 months after treatment. Although there is a possible correlation between the severity of these changes and preoperative neurologic deficits, there is not necessarily an association between reduction of intramedullary changes and clinical improvement.[18] One of the radiologic findings correlating with severity of functional status of patients with SDAVF is craniocaudal extension of the enlarged intrathecal draining veins.[19] For diagnosis of residual or recurrent flow in peri- or intramedullary vessels after treatment of SDAVFs, magnetic resonance angiography (MRA) may be more sensitive than MRI.[20]

SDAVFs can be treated surgically, endovascularly embolized, or a combination of surgery and endovascular techniques may be used.[21],[22] Traditionally, surgery is considered the treatment of choice for SDAVFs. If the lesion can be localized using imaging studies preoper-
atively, surgery can be performed with a low complication rate.[22] In comparison, endovascular approaches are less invasive. However, surgery has an advantage over endovascular techniques when there are multiple SDAVFs because surgery provides direct visibility of all feeding vessels. Since the first case of surgical treatment of SDAVF in 1916,[23] advances in treatment have led to simplified techniques and good outcomes. Nowadays, the typical surgical approach consists of a posterior approach with a laminectomy or hemilaminotomy. The dura is opened, the arterialized vein is identified, and either cauterization or microscissor interruption of the SDAVF is performed.[13],[24],[25] Compared to clipping the draining vein alone, excision or coagulation of the nidus and disconnection of the draining vein may be associated with more favorable long-term results.[26] Complications following surgery are rare; however, instability after laminectomy and pseudomeningocele are two potential complications that can be minimized by limited facet removal (<50%) and meticulous closure of the dura, muscle, and skin.[24]

Endovascular techniques are less invasive and preserve spinal cord tissue and its function; in some cases, access to the feeding vessels of SDAVFs is not possible endovascularly.[22] The first case of SDAVF embolization using metal pellets was described in 1968 by Doppman et al. [27] Since then, new agents including polyvinyl alcohol (PVA), isobutyl-2-cyanoacrylate (IBCA), and N-buty1 cyanoacrylate (NBCA) have been introduced and used for endovascular embolization of various vascular lesions including SDAVF.[13] Although the rate of recanalization following use of PVA was high (up to 93%),[28] the success rate of SDAVF embolization increased to 70%-90% using NCBA.[13] Ethylene vinyl alcohol (Onyx, EV3) liquid embolic system has also been used for embolization of SDAVFs. Onyx was approved by the Food and Drug Administration to be used for embolization of intracranial AVMs but has been used for treatment of spinal AVFs in recent years.[29]-[31] Onyx, which is a nonadhesive liquid agent, carries the advantages of lower likelihood of adhesion of the catheter to the vessels, which facilitates the injection of a larger amount of the agent.[30] In a small series of 3 SDAVFs, Nogueira et al. reported successful management of SDAVF without evidence of residual or recurrent AVF confirmed clinically and radiologically (MRI and MRA) during follow up of more than 7 months.[29] In another small series of 6 patients with SDAVF, Carlson et al. reported complete occlusion of AVF in 5 of the patients during 2 to 4 months of follow up using the Onyx embolization system.[32] It seems that in the future, Onyx will be the preferred embolization method for management of SDAVFs.

In a 2004 meta-analysis, Steinmetz et al. suggested that for treatment of SDAVF, surgery might be superior to embolization. Surgery is usually successful and recurrence and complications are rare. The authors also suggested that endovascular intervention might be a reasonable initial option; however, this technique is associated with a relatively high rate of recurrence. [24] Nowadays, in spite of significant improvements in endovascular embolization and the introduction of new embolization agents, surgery still seems to be the treatment of choice.[13] After treatment, the majority of patients experience improvement; however, symptoms may remain unchanged or deterioration might occur in a few cases.[24],[33] Overall, if timely treatment occurs, patient outcomes for motor abilities and gait disability scores will be particularly good.[34] However, if urinary dysfunction occurs, it less likely responds to
treatment. It seems that time from onset of symptoms to treatment is the largest determinant of outcome in patients with SDAVF.

4. PMAVFs

In 1977, Djindjian and colleagues described PMAVFs for the first time. They described PMAVF as abnormal direct connections between the anterior spinal artery and/or posterior spinal artery and the medullary veins without any intervening nidus. PMAVF is located on the pial surface of the spinal cord. Later, Heros et al. considered PMAVFs to be type IV spinal vascular malformations. PMAVFs can be classified into type I, type II, or type III according to the size of the fistula, the number of feeding arteries, and the severity of venous hypertension.

Type I is a small and single AVF which is fed by the terminal segment of a thin anterior spinal artery. The anatomic location of the AVF is against the anterior surface of the conus medullaris or the filum terminale. Draining perimedullary veins are slightly dilated. Type I AVFs are hemodynamically similar to SDAVFs.

Type II is an intermediate-sized AVF and is fed by multiple arteries. The anatomic location of the AVF is against the conus medullaris. The shunt may be found more frequently in a posterolateral position and less frequently in the anterior position. In the posterolateral position, the feeding artery originates from ipsilateral posterior spinal artery. In anteriorly-located AVFs, the feeding artery originates from the anterior spinal artery. The AVF drains directly to a dilated and tortuous venous system containing a relatively high flow of arterialized blood. However, venous drainage in type II AVFs is slow.

Type III AVFs are large and single and located at the cervical or thoracic spinal cord. The AVF is fed by multiple arteries originating from the anterior and posterior spinal arteries. High-flow AVF drains to a very dilated and tortuous venous system.

The etiology of PMAVF is not clear. Although it is believed these lesions are congenital, few cases of traumatic PMAVF have been reported in the literature. PMAVF is usually diagnosed between the third and sixth decades of life and are very rare, particularly in children.

The exact prevalence of PMAVFs is unknown but it is estimated that they constitute 4%-40% of spinal AVMs. Although the thoracolumbar spinal cord is the most common site of PMAVFs, these lesions can be seen at cervical and thoracic levels as well.

PMAVFs are high-flow lesions leading to venous hypertension. PMAVFs manifest with progressive myelopathy in the majority of patients and can result in complete transverse myelopathy if treatment does not occur. Subarachnoid hemorrhage has also been reported in PMAVF cases. Due to non-specific presentation, the time from onset of symptoms to diagnosis may vary from 2 to 25 years. Angiography is still the gold standard for diagnosis of PMAVF and its benefits include possible simultaneous treatment. However, angiography is invasive and time-consuming, and is only available in specialized centers.
Recent studies indicate that MRI has high sensitivity for diagnosis of spinal AVFs.[7] It is recommended that MRI be ordered as the initial imaging study, followed by MRA if necessary.

PMAVFs can be treated surgically, with endovascularly embolized, or combination of techniques can be performed.[47]-[50] Early diagnosis and treatment may be associated with better results and may prevent irreversible spinal cord injury. Removal of shunt vessels while the spinal cord perfusion is preserved is the purpose of surgery; however, sometimes complex vasculature of the shunt and risk of spinal cord perfusion impairment make surgery complicated. Therefore, various intraoperative diagnostic tools should be used to appropriately identify shunt vessels to avoid damaging spinal cord perfusion. Spinal cord angiography, Doppler ultrasound, and videoangiography using indigo carmine and indocyanine green are some of these modalities.[51]-[53]

Due to the paucity of literature on PMAVF, which results from the very low incidence and prevalence of these lesions, it is not possible to standardize surgical approaches for these lesions; therefore, a variety of surgical approaches have been used for management of them. For treatment of giant PMAVFs, a combination of surgery and endovascular embolization is recommended.[57],[58] Embolization of PMAVF can be performed using glue, coils, and balloons.[47],[50],[57] Overall, treatment of PMAVFs is more difficult than treatment of SDAVFs.[38] Because the majority of PMAVFs are high-flow shunts, endovascular embolization should be considered as the initial therapeutic modality in pediatric patients.[47] In summary, surgery is successful in the treatment of PMAVFs, even in cases with involvement of the anterior spinal artery. In high-flow PMAVFs, endovascular embolization is an appropriate adjunct to surgery.[59]

5. Extradural AVFs

Extradural AVFs are the least frequent type of spinal AVFs. In spinal extradural AVFs, a direct connection exists between the artery or arteries and the extradural venous plexus, which is not available in normal individuals. This abnormal connection is located within the spinal canal and/or intervertebral foramen. Extradural AVFs can cause venous hypertension, mass effect, and vascular steal leading to myelopathy.[8]

Type A extradural spinal AVFs usually manifest as congestive myelopathy or cauda equina syndrome based on the location of the AVF; however, type B1 extradural spinal AVFs present with spinal cord or nerve root compression. Type B2 extradural AVFs are asymptomatic.[30] MRI findings in Type A spinal extradural AVFs are spinal cord edema and perimedullary flow voids. On T2-weighted images, spinal cord edema (hyperintensity) is seen over multiple segments with a hypointense rim, most likely indicating deoxygenated blood.[8] In contrast, type B2 spinal extradural AVFs are difficult to diagnose with MRI and spinal angiography is the imaging modality of choice for diagnosis of these lesions.

Treatment consists of complete obliteration of the extradural AVF by either embolization or surgical excision.[8] It has been reported that after partial obliteration spinal extradural AVFs
will recruit new blood supplies and make treatment more difficult. For management of Type A spinal extradural AVF it is not mandatory to occlude the intradural draining vein. However, it is necessary to inject embolic material into the entire malformation to achieve complete obliteration.[30]

Improvement of endovascular techniques and agents that are using for embolization of lesions may yield better results in the treatment of extradural AVFs. Recently, Rangel-Castilla used Onyx for the embolization of 7 extradural AVFs. Four patients had excellent recovery at 6-24 months and 3 patients with type A extradural AVF experienced good motor recovery without improvement of bladder/bowel problem.[9]

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