A Review of Vascular Abnormalities of the Spine

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Abstract

Patients with spinal vascular lesions present with unique symptoms and have important anatomical and physiologic changes that must be considered prior to treatment. In this mini-review, we provide an overview of normal spinal vascular anatomy and discuss several key spinal vascular lesions. We provide an overview of cavernous malformations, intradural arteriovenous malformations, perimedullary arteriovenous fistulas, and dural arteriovenous fistulas. Important considerations are addressed in terms of pathologic characterization, specific imaging findings, and treatment approaches.

BACKGROUND

Classification of spinal vascular lesions is important for determining the appropriate diagnosis and treatment approach. Krings proposed a system for spinal vascular shunting lesions based on the location of the feeding vessel (dural vs. pial). Dural venous fistulas extend from either radiculomedullary or radiculopial arteries whereas pial lesions can be plexiform or nidus-type arteriovenous malformations (AVMs) with connections between arteries and veins. Once the location has been determined, the type of shunt can be further broken down into macro or micro fistulas based on volume of flow [1]. Non-shunting lesions such as cavernomas are also important to consider in the differential diagnosis for spinal vascular lesions. Below we expand upon this original classification by describing key features of spinal vascular lesions based on anatomic considerations, symptoms, and appropriate treatment approaches (Table 1).

SPINAL VASCULAR ANATOMY

A thorough understanding of the normal spinal vascular anatomy is necessary to fully appreciate the pathophysiology of spinal vascular lesions. The spinal cord is supplied by the anterior and posterior arterial systems. The anterior spinal artery extends along the anterior median fissure and comprises the anterior arterial system. Sulcal arteries arise from the anterior spinal artery and supply the anterior horns corticospinal tracts, and spinothalamic tracts. The posterior arterial system is a network of plexiform collaterals between the two posterior spinal arteries. It supplies the posterior third of the spinal cord including the dorsal columns.

In adults, 6-10 medullary arteries feed into the anterior and posterior spinal arteries. In the cervical region, the medullary arteries are derived from the vertebral arteries and branches of the thyrocervical trunk. In the thoracic and lumbar region, these medullary arteries are derived from the intercostal arteries, which arise from the aorta and iliac arteries. The artery of Adamkiewicz is the largest of these medullary arteries and typically originates on the left side to supply the spinal cord between T8 and L2 (Figure 1).

The spinal cord venous system is comprised of the sulcal and radial veins, which drain into the coronal venous plexus on the
cord surface. Medullary veins drain the pial venous plexus into the epidural (Batson’s) venous plexus. The medullary veins are unique in that they are valveless.

CAVERNOUS MALFORMATIONS

Cavernous malformations (CMs) do not involve a shunt. These are often small lesions, with low blood flow, and are supplied by thin-walled sinusoidal vessels (Figure 2). A rim of hemosiderin and gliosis often surrounds CMs (Figure 3). They can be best diagnosed using magnetic resonance imaging [2]. These lesions are much more common in the brain than in the spinal cord but still comprise up to 12% of spinal vascular abnormalities [3]. Patients often present in the third to sixth decades of life with signs of myelopathy, although the course of the disease can vary in both acuity and severity [4].

Conservative management with serial surveillance imaging is a viable option for patients with asymptomatic cavernous angiomas or for patients with minor, non progressive symptoms [5]. Surgical intervention is indicated in patients with progressive neurological deficits. Ventrally located lesions are associated with poor outcomes after surgery compared to dorsal lesions due to the more extensive myelotomy required for exposure leading to greater risk of neurological injury during surgery [6]. Recent reports have shown that fiber optic CO₂ laser guided treatment may be a viable option with reduced morbidity [7]. The goal of surgery should be complete excision or obliteration of the lesion because residual cavernomas may re-bleed, leading to recurrent myelopathy [8].

INTRADURAL ARTERIOVENOUS MALFORMATIONS

Intradural arteriovenous malformations (Figure 4) are congenital lesions that affect men and women equally. Patients typically present by their third decade of life. The nidus of these high-flow lesions may be completely intramedullary or partially intramedullary with an extramedullary component (Figure 5). Spinal magnetic resonance angiography with contrast injection has been shown to provide the best resolution for detecting the location of the nidus [9].

Figure 2 Cavernous malformations are composed of back-to-back, thin-walled vessels with minimal intervening tissue and are surrounded by gliosis with associated hemosiderin pigment deposition.

Figure 3 Axial T1WI showing heterogenous areas of low signal from presence of prior hemorrhage characteristic of cavernous malformations.

Figure 4 Arteriovenous malformations are characterized by abnormal vessels of varying caliber with intervening gliotic neural tissue.

Figure 5 Sagittal T2WI showing intramedullary mass with mulberry-like areas of hypointensity consistent with hemosiderin staining consistent with prior hemorrhage.
Juvenile type AVMs are high flow lesions that commonly consist of multiple enlarged medullary arteries draining into anterior and posterior spinal arteries that supply the nidus. The nidus is often extensive and may fill the thecal sac. These lesions have a relatively uniform distribution along the spinal cord [10]. As in intracranial AVMs, there is often neural tissue within nidus.

The nidus of glomus type AVMs are often more focal and usually confined to the anterior half of the spinal cord. These lesions are typically supplied by the anterior spinal artery, which is supplied by one or two medullary arteries.

Subarachnoid or intramedullary hemorrhage leading to acute onset back pain, acute neurological decline, or meningismus is a common presentation in patients with spinal AVMs. However, patients may also present with a progressive myelopathy that is thought to be due to a vascular steal phenomenon [11]. The goal of treatment for intradural vascular malformations should be complete and permanent obliteration of the AVM while maintaining the blood supply to the spinal cord. Preservation of neurological function and avoidance of iatrogenic disability may prohibit definitive treatment of lesions that are large in size, involve the ventral half of the spinal cord, have a blood supply associated with extension to the ventral spinal cord surface, and multiple feeding vessels from the anterior spinal artery [12].

Surgery should be considered only after careful evaluation of the spinal arteriography and MRL. Glomus types AVMs are usually compact, lack intervening neural tissue, and tend to have a single arterial feeding vessel thus facilitating safe complete excision without neurological deficits. These glomus type lesions often benefit from endovascular embolization prior to surgical resection [10]. Most juvenile-type AVMs (Spetzler grade 3) however, are high-flow lesions that contain intervening neural tissue. They get their blood supply from multiple medullary arteries, therefore making these inoperable lesions [13]. Furthermore, intramedullary AVMs in the thoracic and lumbar region have many small feedings vessels, thus posing an unacceptably high operative risk [14]. Gamma knife is now being investigated as a potential approach for these hard to treat lesions, but reported results are limited thus far [15].

Surgical treatment of these lesions begins with operative exposure extending one level above and below the nidus of the AVM. A midline durotomy is performed and the arachnoid is preserved to avoid injuring any of the adherent vessels underlying the dural opening. The preoperative angiogram should be reviewed to confirm the major feeding and draining vessels. After the AVM is gently separated from the surrounding arachnoid, the pia at the edge of the nidus is incised to allow for exposure of the AVM. The feeding vessels are sacrificed after coagulation with bipolar cautery. Next, a giotic plan is developed between the AVM and the adjacent neural tissue. Bipolar coagulation is used to shrink the AVM and reduce its turgidity. Any vessels supplying or draining the lesion that are encountered during this dissection are sacrificed. Of note, at least one major draining vein is preserved until dissection around the periphery of the malformation has been completed and all feeding vessels have been sacrificed [16].

The literature is sparse with regards to information regarding the long-term outcomes for patients that underwent surgical treatment of spinal dural AVFs. The reported studies are limited to isolated case reports or small surgical series. The reported clinical results indicate that patients who receive treatment before the onset of neurological sequelae have superior outcomes.

There is no reported long-term data regarding the incidence of clinical relapse or progression because of incomplete surgical excision [10]. Although embolic occlusion is associated with a high incidence of recanalization, repeated arteriographic embolization via endoscopic techniques may be a viable treatment option in certain lesions that are otherwise deemed inoperable [14].

**PERIMEDULLARY ARTERIOVENOUS FISTULAS**

Perimedullary arteriovenous fistulas (AVF) are often located in the lower thoracic or lumbar regions (Figure 6). Typically, the anterior spinal artery drains directly into the coronal venous plexus. The vein just distal to the fistula is often a venous varix. These lesions occur equally in men and women and patients typically present by their fifth decade of life [17]. Zhou and colleagues showed that these lesions can best be visualized by dynamic contrast-enhanced magnetic resonance angiography [18].

Merland and colleagues stratified these lesions into 3 groups based on size and complexity of the nidus [19]. Type I lesions are low flow shunts in the conus or filum terminale that consist of a small fistula supplied by a single small anterior spinal artery. Type II perimedullary AVF are moderate or high flow fistulas. Their nidus is small or medium-size and consists of multiple discrete shunts that are supplied by several enlarged anterior or posterior spinal arteries (Figure 7). Type III shunts are high flow shunts that consists of multiple dilated arteries feeding into a single large fistula.

Patients with perimedullary AVFs most commonly present with a slowly progressive myelopathy. However, patients with type II and III perimedullary AVF may present with subarachnoid hemorrhage [20]. The type of perimedullary fistula dictates the appropriate treatment modality for perimedullary

![Figure 6 Sagittal CTA Perimedullary AVF showing prominent early filling of perimedullary vein.](image-url)
AVFs. Surgical intervention is usually safer than embolization for the treatment of type I perimedullary fistulas [21]. Pre-operative embolization may be effective in reducing blood supply to type II AVFs, but is not a viable curative option due to the multiple feeding vessels [22] and surgical interruption of these AVFs has been performed successfully by various approaches [23]. Selective balloon occlusion, coil embolization, onyx embolization, and guidance with indocyanine green for surgical interruption have all been reported as successful treatments of type III AVFs in the literature [24,25].

DURAL ARTERIOVENOUS FISTULAS

Spinal dural arteriovenous fistulas (AVFs) are thought to be acquired lesions, and were previously referred to as type I AVMs. They are low flow shunts that are supplied by the dura branch of the intervertebral artery. The nidus is composed of a network of separate vessels that converge into the medullary vein and is located in both the dural root sleeve and the adjacent spinal dura [26]. A medullary vein carries the shunted blood retrograde to the coronal venous plexus. Due to the lack of an alternative route for venous drainage in these patients, the coronal venous plexus carries arterialized blood rostrally into the cranial venous system. The high-pressure system can cause vessels of the coronal venous plexus to dilate, elongate, and become tortuous. The best imaging modality to diagnose these lesions is first-pass contrast-enhanced magnetic resonance angiography [27].

Dural AVFs are thought to be acquired lesions [28]. Due to the valveless nature of the intrathecal venous system, increased pressure within the medullary veins subsequently transmits pressure onto the radial and sulcal veins. This venous hypertension leads to venous congestion, which produces ischemic injury to the spinal cord and progressive myelopathy [29].

Patients typically present with a gradual, slowly progressive neurological decline. Spinal dAVFs most commonly affect men in their 5th decade of life. Because these lesions are typically present in the thoracic and lumbar region, patients most often report an insidious onset of paraphrases or sphincter dysfunction [30]. Foix-Alajouanine syndrome (Figure 8) describes a more rapid decline in neurological function and occurs in up to 15% of patients. This likely represents significant venous congestion, which results in venous thrombosis and irreversible spinal cord injury unless treated emergently [31]. Acute hemorrhage is a rarely reported presentation in patients with spinal dural AVFs.

The goal of treatment for spinal dural AVFs is to eliminate the venous congestion either by interruption of the venous drainage from the fistula between the dura and the dilated coronal venous plexus or by elimination of the nidus. Success has been shown with endovascular embolization prior to surgical intervention [32].

Patients with Foix-Alajouanine syndrome exhibit rapid decline in neurological function from venous hypertension leading to venous thrombosis and spinal cord infarction. Embolization is indicated in these patients to transiently provide a rapid reduction in venous congestion until definitive surgical treatment can be provided [32]. The role of embolization for definitive treatment of dural AVFs is contentious. Particular embolization has been reported to provide only transient relief as the embolized dural AVF recanalizes and myelopathy returns in most cases [33]. N-butyl-2-cyanoacrylate (NBCA) is a liquid polymerizing agent that has been used as the primary treatment in patients with dural AVFs. However, the frequency of recanalization and need for additional therapy in these patients questions the utility of embolization alone as a definitive treatment approach thereby necessitating further surgical intervention [34]. Additionally, there is a high rate of inadequate occlusion of the fistula site and draining vein in patients treated with liquid embolic agents [35]. Finally, there have been reported cases of hemorrhage and delayed paraplegia after embolization of spinal dural AVFs [36].

Onyx (ethylene vinyl alcohol copolymer) is a new liquid embolic agent that offers additional advantages: it can be delivered in a slower and more controlled manner, has a lower risk of retaining the catheter, and carries a lower risk for premature venous occlusion [37]. However, there is insufficient long-term data regarding its efficacy in the treatment of spinal dural AVFs.

Surgical interruption of spinal dural AVFs provides immediate relief of venous hypertension without risking occlusion of the

Figure 7 Spinal DSA showing perimedullary AVF supplied by multiple arterial feeders.

Figure 8 Dural arteriovenous fistulas are complicated by necrotic/vacuolated spinal cord tissue with associated vascular congestion.
Table 1: Characteristic findings of each subtype.

<table>
<thead>
<tr>
<th>Type of Spinal Vascular Lesion</th>
<th>Key Features</th>
<th>Diagnostic Modality and Symptoms</th>
<th>Treatments of Choice</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cavernous Malformation</td>
<td>* Do not involve a shunt</td>
<td>* Diagnose: magnetic resonance imaging</td>
<td>* Conservative management</td>
</tr>
<tr>
<td></td>
<td>* Are typically small</td>
<td>* Symptoms: onset in 6th decade often with myelopathy</td>
<td>* CO2 laser guided treatment</td>
</tr>
<tr>
<td></td>
<td>* Supplied by thin-walled vessels</td>
<td></td>
<td>* Surgical intervention</td>
</tr>
<tr>
<td>Intradural Arteriovenous Malformations</td>
<td>* Congenital lesions with equal male to female ratio</td>
<td>* Diagnose: spinal magnetic resonance angiography</td>
<td>* Endovascular embolization followed by surgical obliteration</td>
</tr>
<tr>
<td></td>
<td>* Nidus typically intra-medullary</td>
<td>* Symptoms: subarachnoid hemorrhage with back pain, meningitis, or myelopathy</td>
<td>* Gamma knife for hard to treat lesions</td>
</tr>
<tr>
<td>Perimedullary Arteriovenous Fistulas</td>
<td>* Located in lower lumbar regions</td>
<td>* Diagnose: contrast-enhanced magnetic resonance angiography</td>
<td>* Oxy or coil presurgical embolization</td>
</tr>
<tr>
<td></td>
<td>* Occur equally between males and females</td>
<td>* Symptoms: slower progressive myelopathy or subarachnoid hemorrhage</td>
<td>* Selective balloon occlusion</td>
</tr>
<tr>
<td></td>
<td>* Presents in 5th decade of life</td>
<td></td>
<td>* Surgical interruption</td>
</tr>
<tr>
<td>Dural Arteriovenous Fistulas</td>
<td>* Acquired lesions</td>
<td>* Diagnose: contrast-enhanced magnetic resonance angiography</td>
<td>* Endovascular embolization prior to surgical intervention</td>
</tr>
<tr>
<td></td>
<td>* Low flow shunt</td>
<td>* Symptoms: ischemic injury to spinal cord with progressive myelopathy</td>
<td>* Omyx embolization has been shown to be superior to other methods</td>
</tr>
<tr>
<td></td>
<td>* Coronal venous plexus dilatation</td>
<td></td>
<td>* Surgical interruption</td>
</tr>
</tbody>
</table>

normal vessels that supply the spinal cord. Operative treatment of dural AVFs begins with localization of the dural nidus and draining intramedullary veins. Following subperiosteal dissection and laminctomy for exposure, a midline dural opening is performed and the arterialized medullary vein is identified. This vessel is confirmed to be the draining vein by comparing its anatomy to the preoperative arteriogram and following it as it crosses the subarachnoid space and joins the coronal venous plexus. After confirmation, the medullary vein is coagulated with bipolar forceps and sharply divided as it penetrates the inner layer of the dura between the dura and the coronal venous plexus [38].

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