Vascular malformations of the spinal cord occur infrequently, yet are a significant cause of morbidity. They form a complex set of disorders that can present with a wide spectrum of signs and symptoms, which may contribute to delays in their diagnosis and treatment. Spinal vascular malformations can be further classified according to multiple features, including anatomical location, angioarchitecture, or flow dynamics.

Spinal DAVFs are the most commonly occurring spinal vascular malformations, accounting for 60–80% of all such lesions.3,21,34,44,52 Although recent advances in neuroimaging, microneurosurgical, and endovascular techniques have significantly improved the diagnosis and treatment of this complicated spectrum of pathology, a complete understanding of the pathophysiology of this disease has yet to be achieved. In any case, multidisciplinary treatment has made these lesions curable causes of myelopathy in a large proportion of patients with spinal DAVFs.

In this report, we review the existing literature regarding the clinical characteristics, classification, and endovascular management of spinal DAVFs. As previous attention has primarily focused on the open surgical management of these lesions, we will discuss the literature and our strategy for endovascular embolization of
TABLE 1: Modified classification of spinal cord vascular malformations*

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>AVMs</td>
<td>Extramural and intramedullary lesions</td>
</tr>
<tr>
<td></td>
<td>Ventral (small/medium/large shunt)</td>
</tr>
<tr>
<td></td>
<td>Dorsal (single or multiple feeders)</td>
</tr>
</tbody>
</table>

* From Anson and Spetzler, 1992.

Intradural location. Spinal DA VFs comprise the most common subtype of the intradural group (see below). Spinal DA VFs can lead to the development of spinal cord ischemia, this phenomenon can occur via varying mechanisms in each case. In smaller spinal DA VFs, such as Type I-A lesions, congestion of the radial veins draining the spinal cord results from disproportionate arterial inflow and venous outflow. As a result, segmental spinal cord edema develops that may progress to congestive ischemia and necrotizing myelopathy. In contrast, the high flow observed in lesions such as Type IV-C spinal DA VFs can create a vascular steal phenomenon leading to adjacent spinal cord ischemia. Additionally, Type IV lesions can also develop aneurysms and varices that can rupture, causing hemorrhage, or can exert a mass effect on the spinal cord. In comparison, Type IV-A spinal DA VFs rarely hemorrhage spontaneously.

Clinical Presentation

Although the arrival of MR imaging and selective angiography has significantly improved the ability to characterize spinal DA VFs, these lesions remain inefficiently diagnosed. The time between the onset of symptoms and diagnosis has been reported to be between 12 and 44 months, with a mean duration of 22.9 months. This delay in diagnosis is likely (in part) due to a frequently nonspecific clinical presentation. Presenting symptoms of motor weakness, gait disturbances, and paresthesias commonly lead clinicians to consider and rule out many other disorders before considering spinal DA VFs. Common misdiagnoses include degenerative disc disease, spinal cord tumors, peripheral vascular disease, neuromuscular diseases, or neuropathy. The long-term clinical significance of the delay in diagnosis has yet to be fully elucidated. It is likely that many patients would benefit from more prompt diagnosis and intervention.

The peak age of a patient at the time of diagnosis of a spinal DAVF is in the sixth or seventh decade of life. For reasons yet to be fully determined, there is a preponderance of this diagnosis in males. Although a gradual progression in clinical symptoms has been reported in the majority of patients with spinal
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DAVFs, a more punctuated, stepwise deterioration has been reported in some patients. At the time of diagnosis, many patients have already experienced a considerable progression in the severity of symptoms, and approximately half of all patients report motor disturbances, including weakness and gait abnormalities. One study reported that more than one-third of patients diagnosed with a spinal DAVF are confined to a wheelchair at the time of diagnosis. Other initial complaints include sensory disturbances (paresthesias, lower extremity sensory loss, back/radicular pain) and urinary symptoms. A significant proportion of patients present with bowel and bladder deficits or sexual dysfunction.

Diagnosis

Once sufficient clinical suspicion has been reached, the diagnosis of a spinal DAVF is typically made using MR imaging, MR angiography, or CT myelography. However, the gold standard modality for diagnosing and characterizing flow patterns of these lesions is selective angiography.

Magnetic resonance imaging is sensitive enough to diagnose spinal DAVFs in the majority of patients. Frequent MR imaging findings associated with spinal DAVFs include increased T2 cord signal, Gd enhancement, mass effect, and flow voids. In particular, almost all cases of spinal DAVFs demonstrated increased signal intensity along the center of the spinal cord on T2-weighted MR sequences. Spinal cord hyperintensity typically spans 5 to 7 vertebral levels, with a reported range of 1–11 levels. The majority of spinal DAVFs occur in the thoracolumbar regions, with < 6% of spinal DAVFs occurring in the cervical or sacral regions. Multiple spinal DAVFs are encountered in approximately 4% of patients. Another common finding on MR imaging is the presence of flow voids in 35% of patients, which are believed to represent blood flow within dilated medullary veins. More recently, MR angiography has been used as a diagnostic modality for spinal DAVFs, demonstrating abnormal intradural vessels in 100% of patients according to 1 study. Magnetic resonance angiography has also been reported as a sensitive technique in the assessment of residual blood flow following treatment for spinal DAVFs.

Standard catheter angiography remains the gold standard in the diagnosis of spinal DAVFs. When attempting...
to locate a feeder vessel to a suspected AVF, the intercostal, lumbar, sacral, deep cervical, and ascending cervical arteries should all be visualized. In addition, the internal iliac arteries should be imaged for lumbosacral spinal DA VFs because 12.5% of lesions are fed primarily by these vessels. The vasculature associated with the spinal DA VF should be examined systematically and completely. The presence of arterial feeders that contribute to the ASA, in particular the segmental medullary artery, should be ruled out. Embolization of such feeding branches is contraindicated due to the high risk of spinal cord ischemia and infarction.

**Illustrative Case**

This 85-year-old male presented with progressive gait instability and a history of frequent falls for 3 years. Additional symptoms included numbness and dysesthesias in the bilateral lower extremities, as well as increased lower extremity fatigue and urinary incontinence. Magnetic resonance imaging and MR angiography of the spine demonstrated diffuse cord edema in the lumbar spine, with multiple abnormal blood vessels surrounding the spinal cord. These findings prompted a referral of the patient to the University of Southern California for further evaluation and treatment of a suspected spinal AVM. Upon admission, a neurological examination revealed bilateral lower-extremity motor weakness and sensory deficits, with the left side more severely affected. There was no motor or sensory deficit in the upper extremities.

Spinal angiography was performed. Selective injection of the left L-2 intercostal artery showed filling of an ascending branch along the vertebral body to the pedicle of the left L-1 vertebra, where it filled a fistula arising from the region of the nerve root sleeve (Figs. 1 and 2). No other contributions to the spinal DA VF were identified. The ASA was found to arise from the left T-11 intercostal artery. At this time, the spinal DA VF and feeding branch from the left L-2 intercostal artery were embolized using NBCA. (Fig. 3)

Postembolization angiography demonstrated obliteration of the spinal DA VF without compromise to the left L-2 intercostal artery. One day following his embolization procedure, his gait began to markedly improve and he was discharged. He continued to improve

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**TABLE 2: Reported series of spinal DAVFs treated with endovascular embolization**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Embolization Technique</th>
<th>Successful Embolization (%)</th>
<th>Surgery Required (%)</th>
<th>Repeat Embolization (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Criscuolo et al., 1989</td>
<td>1</td>
<td>PVA</td>
<td>0 (100)</td>
<td>1 (100)</td>
<td>0</td>
</tr>
<tr>
<td>Hall et al., 1989</td>
<td>3</td>
<td>PVA</td>
<td>1 (33)</td>
<td>2 (67)</td>
<td>0</td>
</tr>
<tr>
<td>Hasuo et al., 1996</td>
<td>2</td>
<td>PVA</td>
<td>2 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Morgan &amp; Marsh, 1989</td>
<td>14</td>
<td>PVA</td>
<td>2 (14)</td>
<td>5 (36)</td>
<td>NA</td>
</tr>
<tr>
<td>Nichols et al., 1992</td>
<td>14</td>
<td>PVA</td>
<td>3 (21)</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Schaaf et al., 2002</td>
<td>1</td>
<td>PVA/Tornado coil</td>
<td>1 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Mourier et al., 1993</td>
<td>22</td>
<td>latex detachable balloons</td>
<td>15 (68)</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Rodiek, 2002</td>
<td>1</td>
<td>TGMs</td>
<td>1 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Sugiu et al., 2001</td>
<td>1</td>
<td>CAP</td>
<td>1 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Warakaule et al., 2003</td>
<td>2</td>
<td>Onyx</td>
<td>1 (50)</td>
<td>1?</td>
<td>0</td>
</tr>
<tr>
<td>Jellema et al., 2005</td>
<td>24</td>
<td>histoacryl + lipiodol</td>
<td>12 (50)</td>
<td>4 (17)</td>
<td>5 (21)</td>
</tr>
<tr>
<td>Eskandar et al., 2002</td>
<td>21</td>
<td>liquid acrylic</td>
<td>9 (43)</td>
<td>9 (43)</td>
<td>0</td>
</tr>
<tr>
<td>Cenzato et al., 2004</td>
<td>10</td>
<td>cyanoacrylic glue</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Birchall et al., 2000</td>
<td>1</td>
<td>NBCA</td>
<td>0 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Cognard et al., 1996</td>
<td>7</td>
<td>NBCA</td>
<td>6 (86)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Guillemin et al., 2005</td>
<td>26</td>
<td>NBCA</td>
<td>21 (81)</td>
<td>5 (19)</td>
<td>0</td>
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<tr>
<td>Mascacchi et al., 2001</td>
<td>18</td>
<td>NBCA</td>
<td>11 (61)</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Matsubara et al., 2008</td>
<td>2</td>
<td>NBCA</td>
<td>2 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Rodesch et al., 2005</td>
<td>18</td>
<td>NBCA</td>
<td>13 (72)</td>
<td>3 (17)</td>
<td>0</td>
</tr>
<tr>
<td>Song et al., 2001</td>
<td>20</td>
<td>NBCA</td>
<td>14 (70)</td>
<td>5 (25)</td>
<td>0</td>
</tr>
<tr>
<td>Ushikoshi et al., 1999</td>
<td>6</td>
<td>NBCA</td>
<td>4 (67)</td>
<td>2 (33)</td>
<td>0</td>
</tr>
<tr>
<td>Van Dijk et al., 2002</td>
<td>44</td>
<td>NBCA</td>
<td>11 (25)</td>
<td>31 (70)</td>
<td>0</td>
</tr>
<tr>
<td>Lundqvist et al., 1990</td>
<td>10</td>
<td>NBCA or PVA</td>
<td>9 (90)</td>
<td>1 (10)</td>
<td>0</td>
</tr>
<tr>
<td>Narvid et al., 2008</td>
<td>39</td>
<td>NBCA or PVA</td>
<td>27 (69)</td>
<td>12 (31)</td>
<td>0</td>
</tr>
<tr>
<td>Niimi et al., 1997</td>
<td>47</td>
<td>NBCA or IBCA</td>
<td>39 (83)</td>
<td>0</td>
<td>NA</td>
</tr>
<tr>
<td>Westphal and Koch, 1999</td>
<td>35</td>
<td>NBCA, embospheres, PVA, fiber coils</td>
<td>13 (37)</td>
<td>20 (57)</td>
<td>2 (6)</td>
</tr>
</tbody>
</table>

* CAP = cellulose acetate polymer; NA = not available; TGMs = trisacryl gelatin microspheres.
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and was almost back to his baseline neurological and ambulatory status at the 6-month follow-up.

Endovascular Treatment of Spinal AVFs

Treatment of spinal DAVFs consists primarily of surgical ligation, endovascular obliteration, or both. Although open surgical treatment of spinal DAVFs has been the primary intervention for several decades, recent advances in neurointerventional techniques have proven that endovascular treatment options are safe and effective alternatives. Typically, open surgical ligation consists of a posterior approach with a laminectomy or laminotomy, identification of the arterialized vein, dissection along the dural root sleeve, and cautery or microscissor interruption of the fistula. In addition, intraoperative or postoperative angiography has been frequently used to confirm complete surgical obliteration of spinal DAVFs.

According to some studies, direct surgical obliteration of spinal DAVFs has been reported to provide improved disability scores and lower recurrence rates as compared with embolization procedures, and is therefore generally considered to be the gold standard for management of these lesions by many authors. Surgical management of spinal DAVFs is also required when these lesions are not amenable to endovascular treatment, and if failed embolization or repeated recanalization has occurred. Furthermore, if a major feeding artery to the spinal DAVF also contributes to perfusion of the spinal cord, the risk of spinal cord ischemia may be too high to attempt embolization. This situation is typically encountered in fistulas that are fed by the segmental medullary arteries, which contribute to the ASA.

Over the last several decades, improvements in endovascular technique and embolic agents have greatly improved the ability to definitively treat a majority of spinal DAVFs. This improvement has been associated with shorter hospital stays, minimal procedural morbidity, and earlier initiation of rehabilitation for patients undergoing embolization for these lesions. Rates of definitive embolization have ranged between 25 and 100%, depending in part on the embolic agent used and the use of variable stiffness microcatheters (Table 2). This practice of many institutions, including our own, has therefore been to primarily attempt minimally invasive endovascular embolization and reserve surgical intervention for refractory cases or those not amenable to embolization.

The success of endovascular treatment is believed to be highly dependent on complete occlusion of the proximal radiculomedullary draining vein and the site of the fistula itself. Various radiographic outcome measures have been used in the past to define endovascular occlusion of a spinal DAVF, depending on whether the draining vein is obliterated or the fistula itself is filled with a liquid embolic agent. Complete occlusion of the fistula usually requires that the microcatheter be positioned as close to the site of the fistula as possible. If embolization is attempted too proximally in the radicular feeding artery, liquid embolic agents may occlude proximal to the fistula, allowing collateral feeders to develop distally and reconstitute the fistula. Spinal DAVFs are believed to be fed by 1 or more radicular arteries draining into a single radiculomedullary vein. Recurrence, possibly due to recruitment of collateral vessels, can form despite occlusion of the main feeder artery if the fistula and the draining veins are not effectively embolized or if PVA is chosen as the embolic agent.

The origin of endovascular treatment of spinal DAVFs dates back to 1968, when Doppman and colleagues performed the first embolization of a spinal DAVF using metal pellets. Since that time, multiple embolic agents have been used to attempt permanent closure of the draining vein and fistula. Prior to the introduction of modern embolic agents such as IBCA and NBCA, PVA was used in standard practice. However, the rates of recanalization of the draining vein were found to be exceedingly high using PVA. In a series of 14 patients, Morgan and Marsh reported that 13 (93%) showed angiographic evidence of recanalization within 9 months, and 8 (57%) required surgery as definitive management. This result has partially attributed to PVA occluding the proximal area of the feeder without reaching the actual site of the fistula, resulting in recanalization or formation of collateral vessels to the fistula.

Rates of successful embolization have significantly improved with the transition to liquid glue embolic agents such as IBCA or NBCA, which are believed to provide improved penetration of distal draining vessels and therefore more permanent occlusion of the fistula. The majority of studies reporting outcomes following embolization of spinal DAVFs using NBCA have reported success rates of 70–90%. Even more recently, ethylene vinyl alcohol (Onyx, EV3) has been used in the treatment of spinal DAVFs. Although few reports exist discussing the use of Onyx in the treatment of spinal DAVFs, the success of Onyx in intracranial DAVFs has generated significant interest in its potential use in their spinal counterparts. The potential benefits of Onyx include obviating the need for rapid microcatheter withdrawal and improved distal draining vein penetration based on the relatively lower viscosity of this compound. This may be particularly beneficial when microcatheter access cannot be achieved directly at the site of the fistula, precluding the ability to use NBCA, which tends to polymerize near the catheter tip in these slow-flow fistulas. As a result the operator has the opportunity to inject embolic material more slowly—that is, over a period of minutes rather than seconds—potentially leading to a more accurate embolization. Whereas isolated case studies have shown successful spinal DAVF occlusion at 7 months, the recanalization rates of spinal DAVFs treated with Onyx remain to be determined and more long-term follow-up will be required before Onyx can be fully recommended in standard practice.

Clinical Outcomes

Because the primary concern of treatment is symptomatic and functional improvement, in particular for gait and urinary dysfunction, the Aminoff-Logue disability scale was introduced to facilitate long-term follow-up trends (Table 3). Retrospective studies with significant
TABLE 3: Modified Aminoff-Logue scale of disability*

<table>
<thead>
<tr>
<th>Grade</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>gait disturbance</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>leg weakness or abnormal gait, no restricted activity</td>
</tr>
<tr>
<td>2</td>
<td>Grade 1 with restricted activity</td>
</tr>
<tr>
<td>3</td>
<td>requiring 1 stick for walking</td>
</tr>
<tr>
<td>4</td>
<td>requiring 2 sticks for walking (crutches/walker)</td>
</tr>
<tr>
<td>5</td>
<td>unable to stand, confined to bed/wheelchair</td>
</tr>
<tr>
<td>micturition</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>hesitancy, frequency, urgency</td>
</tr>
<tr>
<td>2</td>
<td>occasional urinary incontinence or retention</td>
</tr>
<tr>
<td>3</td>
<td>total urinary incontinence or retention</td>
</tr>
</tbody>
</table>

* From Aminoff and Logue, 1974.

long-term follow-up data following endovascular embolization are only recently emerging in the literature. Narvid et al.48 reported a mean follow-up of 49 months in a large series of patients over a 20-year period at a single institution. In general, previous studies that presented data on both endovascular and surgical treatment have highlighted a 1-grade mean reduction for gait on the Aminoff-Logue scale.14,57,63 In particular, gait disturbances were more likely to improve following either treatment, whereas micturition disturbances were less likely to improve.37 The majority of studies reporting outcomes following endovascular treatment, albeit with limited follow-up times, have reported gait improvement in 40–100% of patients.8,14,23,50,57 Niimi et al.50 reported that 25 (71%) of 35 patients who underwent embolization using either IBCA or NBCA and had adequate follow-up with angiographic testing were free of symptomatic recurrence after 12 months. In another study of 44 patients, who were primarily treated with endovascular therapy, Jallema et al.33 found an improvement in motor function in 56% of patients and gait improvement in 64% of patients 6 years after their procedure. Other symptoms such as pain and urinary symptoms were less likely to significantly improve following treatment.23,57

Conclusions

Recent advances in neuroimaging, endovascular, and surgical techniques have made spinal DAVFs a curable cause of myelopathy. Although open neurosurgical management provides definitive treatment and is currently the preferred treatment modality at most institutions, recent advances in embolic agents and techniques in endovascular neurosurgery have allowed embolization to serve as a less invasive and increasingly effective treatment alternative. The majority of recent studies reporting outcomes following embolization of spinal DAVFs using NBCA have reported success rates of 70–90%. Newer embolic agents such as Onyx provide significant promise for definitive initial management of these lesions, yet require longer clinical follow-up analysis. An interdisciplinary approach to the management of spinal DAVFs is recommended.

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Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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