Spinal dural arteriovenous fistulas: outcome and prognostic factors

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The aim of this study is to review the clinical outcome of patients treated for spinal dural arteriovenous malformations and investigate the presence of pretreatment indicators of outcome after short- and midterm follow-up. The authors retrospectively reviewed the records of 65 consecutive patients treated either surgically or endovascularly in 3 neurosurgery departments between 1989 and 2009. After treatment, 80% of patients reported improvement of at least 1 symptom. Motor symptoms improved more than sensory disorders, pain, or sphincter impairment. Spinal dural arteriovenous fistulas at the thoracic level, and in particular at the lower level, responded better than those in other spinal areas. Spinal dural arteriovenous fistula is a rare pathology with a malignant course that should be treated aggressively. This study appears to confirm that neurological status before treatment, anatomical location of the fistula, and symptoms at presentation are all factors that can predict outcome. Early diagnosis of spinal dural arteriovenous malformations in the lower thoracic area in patients with an Aminoff scale score < 3 was associated with the most favorable outcome.

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Key Words • embolization • outcome • spinal dural arteriovenous fistula • treatment • venous congestion

Spinal dural arteriovenous fistulas, also known as Type I spinal arteriovenous malformations, are a rare pathology with an expected incidence of only 5–10 new cases per million inhabitants per year. They constitute approximately 80% of all spinal vascular malformations and usually become symptomatic in middle-aged patients. Most SDAVF s are located in the thoracolumbar region of the spine, whereas sacral and multiple fistulas are even more rare and occur in approximately 4% and 2% of patients, respectively.

Hebold and Gaupp are credited with the first descriptions of isolated spinal cord vascular malformation in 1885 and 1888, respectively. They portrayed the latter as “a large varix of veins arising from the pia which pressed upon and flattened the spinal cord.” Over the following decades, many surgeons reported their experience with exploratory laminectomies in patients with progressive paraparesis who were subsequently found to have vascular malformations.

Only recently, however, did Kendall and Logue report the first case of an SDAVF as a distinct entity. Later, Merland et al. illustrated the anatomical and pathological structure and location of an SDVAF in the dura mater and the intradural site of a radicular pouch. Only with the recent work of McCutcheon et al., however, have the anatomical characteristics of SDAVF s been completely understood.

Diagnosis of SDAVF s is very difficult in clinical practice. In fact, symptoms are generally nonspecific and include problems in climbing stairs and gait disturbance, and more often sensory symptoms such as paresthesia and diffuse or patchy sensory loss, but also radicular pain and lower back pain. Bowel and bladder incontinence, erectile dysfunction, and urinary retention are more often observed in the latter course of the disease.

Neurological symptoms of SDAVF s are related to the high venous pressure that causes venous engorgement and subsequently induces progressive ischemic damage to the spinal cord. Symptoms are slowly progressive, sometimes interrupted by intermediate remission, and for these reasons several months of misdiagnosis and incorrect evaluations often precede correct diagnosis.

Because presenting clinical symptoms are unspecific,
the neuroradiologist is often the first clinician to consider a diagnosis of SDAVF, which initially relies primarily on MRI. Magnetic resonance imaging is fundamental for identifying alterations of the spinal cord and the vascular pattern characteristic of SDAVs. For a thorough understanding of the disease and for planning the therapeutic strategy, however, selective spinal digital subtraction angiography is necessary. Treatment of SDAVs is based on the occlusion of the shunting zone, which can be obtained through either a surgical approach or endovascular treatment, although the optimal management strategy remains unclear.

Despite improvements in neuroradiological tools, which permit prompt diagnosis and safe surgical or endovascular intervention, it remains difficult to predict whether patients will obtain clinical improvement or whether the treatment will simply halt progression of symptoms. Moreover, it is difficult to predict the impact on quality of life that the patient may expect. An unfavorable outcome could be the consequence of several factors as noted by many authors. It would appear that clinical status before treatment, modality of onset of symptoms, patient age, and anatomical location of the fistula all play a predictive role in determining clinical outcome after treatment. In this paper we present an extension of a previous analysis of pretreatment indicators of outcome in 65 consecutive SDAVs over a period of 20 years.

Methods

The study analyzed the records of 65 consecutive patients ranging in age from 24 to 82 years who were treated for SDAVs in the Department of Neurosurgery of San Raffaele Hospital (between 1989 and 2005), the Department of Neurosurgery of Spedali Civili Hospital in Brescia (between 2005 and 2007), and the Department of Neurosurgery of Niguarda Ca’ Granda Hospital in Milan, Italy (between 2007 and 2009). Most patients were between 55 and 70 years of age (mean 61 years, median 64 years).

Clinical and Radiological Evaluation

All patients underwent pretreatment MRI and digital subtraction angiography with selective catheterization of each radicular artery. These examinations permit a correct diagnosis to be made, identifying the specific characteristics of each fistula and demonstrating spinal cord damage and its extension. Digital subtraction angiography was repeated shortly after treatment to confirm occlusion. Between 3 months and 2 years (mean 6 months) after treatment of an SDAVF, all patients returned for an angiographic control study to confirm occlusion; 60% of the patients were also reexamed using MRI. We reviewed radiological images to identify any feature that might be related to outcome. In the angiographic examination, we evaluated the diameter of drainage veins, transit time at shunt level, and length of pathological drainage veins in terms of metameric extension. Magnetic resonance images were examined to evaluate the extension of the myelopathy and diameter of the spinal cord.

All patients underwent complete neurological examination before treatment, and at 6 months and 3 years after intervention. Follow-up visits included clinical evaluation of motor deficit, sphincter disturbances, sensorial deficiency, and the presence of pain. For evaluation of motor deficit, the disability scale of Aminoff and Logue was used; changes in this score were used for statistical analyses.

Surgical Intervention

A team of neurosurgeons and neuroradiologists selected the optimal treatment for each patient. Surgical intervention was considered the first choice in all cases, whereas endovascular therapy was preferred for patients with unfavorable general medical conditions. A total of 55 patients underwent microsurgical interruption of the drainage vein immediately distal to the shunt (Fig. 1), and 10 patients underwent endovascular exclusion with cyanoacrylic glue as the sole treatment, mostly in the earlier cases. Surgical procedures were performed through a bilateral laminectomy, while in the later years the procedure became less invasive using a hemiinterlaminectomy.
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tomy centered at the level of the corresponding radicle.\textsuperscript{29} Statistical analysis was performed using a chi-square test with the StatView program for Macintosh (SAS Institute).

Neuroradiological Examination

All patients underwent MRI of the spine that demonstrated spinal cord damage together with characteristic vascular images due to the presence of an arteriovenous fistula. Both the anatomical site of cord damage and metameric extension were recorded for all patients. Subsequently, spinal angiography was necessary for correct diagnosis and to localize the presence of single or multiple arteriovenous shunts between a dural artery and a pial vein.\textsuperscript{6,7} The incidence of SDA VF was higher at the midthoracic (above T-9; n = 26, 40.0%) and lower thoracic levels (T9–12; n = 20, 31.0%) compared with the lumbar area (n = 18, 27.7%). One case of cervical SDAVF was observed (1.5%).

Symptomatology

The average duration of clinical symptoms before diagnosis was 19 months (range 1 day [patient with acute paraplegia] to 8 years). However, 51% of patients were diagnosed within 1 year after the appearance of symptoms, and the median duration of symptoms was 13 months. The duration of disease has been postulated to influence the severity of spinal cord impairment.

As summarized in Table 1, there were 7 patients with a high Aminoff score (4–5) that was not correlated with duration of disease before treatment (< 13 months, 21%; ≥ 13 months, 23%). Several differences were noted between clinical symptoms at onset and the time of diagnosis (Table 2). At onset, more than 37% of patients presented with various grades of motor deficits and a low percentage of other disturbances. In contrast, at the time of diagnosis the majority of patients had motor deficits (91%), sensorial deficiency (82%), micturition disturbances (69%), and pain (51%). The onset of symptoms was gradual and slowly progressive in 58 patients and acute in 7 cases.

Results

Treatment

Ten patients underwent endovascular embolization without operative complications, whereas follow-up angiography demonstrated occlusion of the SDAVF in 7 patients (70%). Three patients (30%) developed recurrence due to an insufficient occlusion of the SDAVF vein and underwent surgical revision. The majority of our cohort (55 patients, 85%) underwent surgical intervention, with no deaths and no significant postoperative complications.

<table>
<thead>
<tr>
<th>TABLE 1: Severity of motor impairment (Aminoff score) and disease duration before treatment*</th>
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</thead>
<tbody>
<tr>
<td>Disease Duration (mos)</td>
</tr>
<tr>
<td>&lt;13</td>
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<tr>
<td>≥13</td>
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</tbody>
</table>

* Data given as number of patients (%).

Radiological follow-up demonstrated complete anatomical occlusion of the fistula in all patients who had undergone surgical occlusion.

Pretreatment Indicators of Outcome

We observed improvement of at least 1 symptom in a high percentage of patients (80%). The greatest percentage of symptom improvement after treatment (63%) was observed in motor deficits, whereas sensory and sphincter disturbances showed less improvement (40% and 44%, respectively; Table 3). A similar situation was observed at the 3-year follow-up, although slow progressive increases in improvement were observed.

Improvement of different symptoms was related to the anatomical location of the fistula, and in particular, only 70% of patients with a fistula above T-9, a region with lesser vascularization, showed improvement in at least 1 symptom. This increased to 78% when the fistula involved the lumbar nerve roots and to 95% when the fistula was anatomically located in the lower thoracic region. A statistically significant difference (p = 0.004) was observed between the motor component of symptomatology and location of the fistula after treatment, which decreased somewhat (p = 0.02) at the 3-year follow-up (Table 3; Fig. 2). The lumbar site of the fistula also appeared to play a role in spinal pain improvement after treatment (86%) and at the 3-year follow-up (86%).

The Aminoff score scale of motor disability was used to rate outcomes. After treatment, 44% of patients were able to walk without help (Aminoff scale score 0 or 1), 22% had reduced tolerance, 19% walked with a cane, 12% required crutches, and 3% were confined to a wheelchair (Table 4; Fig. 3). Interestingly, we observed only a slight difference between clinical outcome and duration of the onset-to-treatment interval at the 6-month and 3-year follow-up evaluations (Table 5). Younger patients (< 40 years old) showed more improvement compared with older patients (> 60 years old), but with a slight difference in motor deficits (Table 6). Motor outcomes are undoubtedly related to pretreatment deficiency: generally none of the patients (< 60 years old) with severe motor deficits returned to normal, although treatment was associated with improvement of 1–2 points on the Aminoff scale. Finally, we analyzed angiographic and MRI findings, but none of the morphological features appeared to be related to treatment outcome.

Discussion

What is clear about the natural history of SDAVF's...
is that without treatment, clinical symptoms unavoidably worsen until the patient becomes severely disabled. In fact, it is estimated that 50% of untreated patients will become severely disabled within 3 years of the onset of leg weakness and that progressive myelopathy caused by venous hypertension will be unrelenting, with almost no possibility of spontaneous improvement. We consider SDA VF a rare pathology with a malignant course that should be treated aggressively. Despite the malignant course of SDA VFs, early detection remains a challenge. Based on some reports, the average duration of clinical symptoms before diagnosis of SDAVF varies from 12 to 22 months. Neurological symptoms have a generally gradual and slow progressive onset; they are usually non-localizing and subtle and are often attributed to aging, stress, or exercise.

Surgical treatment of SDA VFs was the first technique used to cure the disease, as described by the pioneering procedure by Hebold and Gaupp at the end of 19th century. At present, treatment of SDAVFs has improved greatly due to the development of microsurgical techniques. Surgical disconnection between the point of fistula within the dural leaves and the draining vein emerging intradurally at the level of the neural foramen, along with its corresponding nerve, is a technically simple and highly effective procedure, with a 98% success rate (Fig. 1). However, with advances in endovascular techniques, embolization has been proposed as an alternative to surgical intervention by a number of centers despite some recurrence.

Surgical and endovascular treatment can both be considered safe procedures, and only 1 recurrence of a fistula after embolization was observed in our case series that was successfully retreated with surgery. No significant surgical complications were observed. Although we report on 2 very different treatment groups based on sample size, there were no significant differences in complications or outcomes between the surgical and endovascular groups. Surgical or endovascular disconnection between the drainage vein and artery of the dural leaves is effective, as shown by the fact that 80% of patients showed improvement after treatment. Nevertheless, dural fistulas remain highly disabling, and treatment is not fully curative.

**Variables That May Affect Outcome**

**Onset of Symptomatology.** A limited number of patients (n = 7, 11%) showed an acute onset of symptoms that ranged from 1 to 7 days. The majority (n = 58, 89%), however, showed slow and gradual progression of neurological deterioration. Interestingly, clinical neurological improvement after treatment of the fistula was lower in patients with acute onset of the symptomatology (70%) compared with those with gradual onset (81%). As previously assumed, medical treatment is ineffective for the

**TABLE 3: Improvement of different symptoms (when present) in relation to location of fistula***

<table>
<thead>
<tr>
<th>Variable</th>
<th>Above T-9</th>
<th>After Tx</th>
<th>3-yr FU</th>
<th>T9–12</th>
<th>After Tx</th>
<th>3-yr FU</th>
<th>Below T-12</th>
<th>After Tx</th>
<th>3-yr FU</th>
<th>Total</th>
<th>After Tx</th>
<th>3-yr FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>motor deficit (Aminoff</td>
<td>12/24 (50)</td>
<td>14/24 (58)</td>
<td>17/19 (89)†</td>
<td>17/19 (89)‡</td>
<td>8/16 (50)</td>
<td>10/16 (62)</td>
<td>37/59 (63)</td>
<td>41/59 (69)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>scale)</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>sensory disturbance</td>
<td>7/19 (37)</td>
<td>8/19 (42)</td>
<td>8/17 (47)</td>
<td>8/17 (47)</td>
<td>5/14 (35)</td>
<td>6/14 (43)</td>
<td>20/50 (40)</td>
<td>22/50 (44)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>pain</td>
<td>6/10 (60)</td>
<td>6/10 (60)</td>
<td>4/9 (44)</td>
<td>6/9 (66)</td>
<td>6/7 (66)</td>
<td>6/7 (66)</td>
<td>16/26 (61)</td>
<td>18/26 (69)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>sphincter disturbance</td>
<td>7/15 (46)</td>
<td>8/15 (53)</td>
<td>9/16 (56)</td>
<td>10/16 (62)</td>
<td>2/10 (20)</td>
<td>2/10 (20)</td>
<td>18/41 (44)</td>
<td>20/41 (49)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>at least 1 symptom</td>
<td>19/27 (70)</td>
<td>19/27 (70)</td>
<td>19/20 (95)</td>
<td>19/20 (95)</td>
<td>14/18 (78)</td>
<td>14/18 (78)</td>
<td>52/65 (80)</td>
<td>52/65 (80)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* All data given as number of patients improved (%). Abbreviations: FU = follow-up; Tx = treatment.
† p = 0.004.
‡ p = 0.02.

Fig. 2. Percentage of improvement in Aminoff score at the 3-year follow-up evaluation according to location of the fistula. Patients with lower thoracic SDAVFs (T9–12) showed the greatest improvement after operative treatment.
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Duration and Severity of Symptomatology. Consistent with results from our previous report and those of other authors, we found no statistically significant correlation between duration of pretreatment clinical symptoms and posttreatment extent of improvement. It is widely reported that patients with more serious symptoms generally have a longer duration of disease. In our case series, there was a small difference in the Aminoff score between patients with a long-term pretreatment history (≥13 months) and those with a short pretreatment history (<13 months; Table 1). After treatment and at 3-year follow-up, only a few patients (n = 3, 5%) who received early treatment (<13 months) had an Aminoff score less than 3, compared with a higher number of patients (n = 6, 10%) who received late treatment.

Based on these data, it appears clear that patients with a longer clinical history generally had a more severe clinical picture, and consequently a less favorable long-term outcome; however, the extent of improvement in the 2 groups did not differ greatly. As previously reported, duration of symptoms does not appear to limit the chances of improvement, and outcome is mainly dependent on the clinical picture before treatment.

Location of Fistula. Spinal dural arteriovenous fistulas may be located at any spinal level, although most are commonly observed at the middle and lower thoracic areas. In our series, 46 patients (71%) had spinal fistulas at those levels. Previously, we described a correlation between clinical outcome and anatomical location of the fistula, showing a significant statistical association (p = 0.04) for the first time. However, the small sample size and a limited long-term clinical follow-up were the limitations of that study. The analysis of a larger series of 65 patients after treatment and at 3 years of follow-up allowed us to confirm our previous results (Table 3; Fig. 2).

Patients with SDAVFs located in the lower thoracic area improved more than those with a fistula elsewhere independent of follow-up, and this difference reached statistical significance for motor outcome. We have previously analyzed the possible reasons for this relationship, although no clear conclusions have been made. The fact that the lower thoracic section has a better vascular supply

### TABLE 4: Aminoff score of motor disability before and after treatment

<table>
<thead>
<tr>
<th>Score</th>
<th>Before Tx</th>
<th>After Tx</th>
<th>3-yr FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>0 (no deficit)</td>
<td>4 (6)</td>
<td>4 (6)</td>
<td>6 (9)</td>
</tr>
<tr>
<td>1 (hyposthenia)</td>
<td>3 (5)</td>
<td>25 (38)</td>
<td>24 (37)</td>
</tr>
<tr>
<td>2 (reduced tolerance)</td>
<td>16 (25)</td>
<td>14 (22)</td>
<td>14 (22)</td>
</tr>
<tr>
<td>3 (need for a cane)</td>
<td>15 (23)</td>
<td>12 (18)</td>
<td>11 (17)</td>
</tr>
<tr>
<td>4 (need for crutches)</td>
<td>17 (26)</td>
<td>8 (12)</td>
<td>8 (12)</td>
</tr>
<tr>
<td>5 (patient in wheelchair)</td>
<td>10 (15)</td>
<td>2 (3)</td>
<td>2 (3)</td>
</tr>
</tbody>
</table>

* All data given as number of patients (%).

### TABLE 5: Outcome in relation to the duration of onset-to-treatment interval

<table>
<thead>
<tr>
<th>Onset-To-Treatment Interval</th>
<th>Aminoff Score Improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>After Tx</td>
</tr>
<tr>
<td>&lt;13 mos</td>
<td>20/32 (63)</td>
</tr>
<tr>
<td>≥13 mos</td>
<td>17/27 (63)</td>
</tr>
<tr>
<td>total</td>
<td>37/59 (63)</td>
</tr>
</tbody>
</table>

* All data given as number of patients (%).
may help to explain this difference, considering the role of ischemia in the genesis of spinal cord damage in these patients, as previously noted.10,11 The poor vascularization of the thoracic spinal cord may make it impossible to maintain a sufficiently steep arteriovenous gradient to perfuse the spinal cord, whereas at the thoracolumbar passage, the entrance of the Adamkiewicz artery may allow for slightly better perfusion.

Nature of Symptomatology and Patient Age. Motor symptoms were associated with more improvement after treatment and at long-term follow-up, and the majority of patients gained 1 or 2 points on the Aminoff scale. Similarly, spinal pain improved in a significant percentage of patients after treatment, with a slight increase after 3 years of follow-up. Sensitivity and sphincter disturbances were among the symptoms showing less improvement, and in fact remained impaired in a high percentage of patients after treatment. Nagata et al.16 reported a relationship between patient age and clinical outcome after treatment of SDAVs, reporting that younger patients had a better chance for good outcome. We note that all patients < 40 years old showed improvement in at least 1 symptom, while a lower percentage (68%) of older patients (> 60 years old) showed improvement, even though this difference was not statistically significant.

Conclusions

Surgical obliteration is a safe and effective procedure and is currently considered first-line therapy for SDAVs. Our study appears to confirm that neurological status before treatment, anatomical location of the fistula, and presenting symptoms are all predictive factors for outcome. However, no patient gained more than 2 points on the Aminoff scale after treatment. Early diagnosis of lower thoracic area SDAVs in patients and an Aminoff scale score < 3 were associated with the best outcome.

Disclosure

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

Author contributions to the study and manuscript preparation include the following. Conception and design: Cenzato, Debernardi, Versari. Acquisition of data: Stefini, D’Aliberti, Coppini. Analysis and interpretation of data: Cenzato, Debernardi, Stefini, Piparo, Talamonti. Drafting the article: Cenzato, Debernardi, Stefini, D’Aliberti, Piparo, Talamonti, Versari. Critically revising the article: Cenzato, Debernardi, Stefini, D’Aliberti, Piparo, Talamonti, Versari. Reviewed submitted version of manuscript: Cenzato, Debernardi, Stefini, D’Aliberti, Piparo, Talamonti, Versari. Approved the final version of the manuscript on behalf of all authors: Cenzato. Statistical analysis: Debernardi.

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