Curable Cause of Paraplegia
Spinal Dural Arteriovenous Fistulae

Nozar Aghakhani, MD; Fabrice Parker, MD, PhD; Philippe David, MD; Pierre Lasjaunias, MD, PhD

Background and Purpose—The rarity of spinal dural arteriovenous fistulae makes physicians often overlook this potential diagnosis in patients with progressive gait disturbance and paraparesis. Consequently, patients with spinal dural arteriovenous fistulae can gradually become completely paraplegic if the final diagnosis is delayed considerably. The objective of the current study is to demonstrate that, particularly in patients with paraplegia, surgical treatment of fistula is necessary and often has a favorable outcome.

Methods—Of 42 patients with spinal dural arteriovenous fistulae treated in our institution (surgery or endovascular treatment), 6 were paraplegic preoperatively (Grade IV on the McCormick scale and Grade V on the Aminoff scale, Grade 5 of modified Rankin Scale with motor ASIA between 0 and 10 for both lower limbs). Their clinical history revealed that paraplegia appeared progressively within a period of <3 months. All patients were clinically evaluated at 6 weeks, 6 months, and then annually during an average follow-up of 3 years. Patients received at least one spinal angiography and MRI test during the follow-up period.

Results—Total exclusion of the fistula was performed surgically in all cases and was confirmed by spinal angiography. No surgical complications were recorded. All patients improved postoperatively. Three patients showed almost normal walking (Grade I on the McCormick scale, I on the Aminoff scale, Grade 1 of modified Rankin Scale) and 3 were able to walk with a cane (Grade II on McCormick, Grade III on Aminoff scale, Grade 2 of modified Rankin Scale). MRI tests were normal in all patients.

Conclusions—Our results indicate that treatment of fistula is a necessary intervention, even in patients with complete paraplegia. (Stroke. 2008;39:2756-2759.)

Key Words: paraplegia ■ spinal dural arteriovenous fistulae ■ surgery ■ treatment

Spinal dural arteriovenous fistulae (SDAVF) are the most common type of spinal vascular malformations.1 Although considered a well-known pathology in the neurological and neurosurgical milieu, SDAVF is frequently ignored and misdiagnosed by other physicians. Early diagnosis is difficult due to the rarity of SDAVF and the nonspecific symptoms (progressive myelopathy). In a large series reported by Jellema et al,2 the median delay of diagnosis was 15 months by which time 62 of 80 patients had severe weakness of the legs and 15 had become wheelchair-bound.

There are several reports on the clinical presentation, treatment, and outcome of patients with SDAVF in the literature. Steinmetz1 analyzed data collected from 20 papers and 532 patients. The typical clinical presentation of SDAVF is progressive myelopathy with neurological deterioration such as paraparesis, sensory disturbance, and sphincter dysfunction. If the SDAVF diagnosis is missed on initial evaluation, the neurological deterioration can worsen over time. As reported by Aminoff et al3 within 3 years after the onset of symptoms, over 90% of patients are unable to walk independently.

The appropriate treatment option in these deteriorated patients is a matter of controversy. To contribute to this debate, we analyzed data from 6 patients with paraplegia with SDAVF treated in our institution. Clinical and radiological data, patient outcomes, and our pathophysiological hypothesis are presented and discussed.

Patients and Methods
We reviewed all patients treated for SDAVF in our institution (through endovascular or surgical intervention) between December 1992 and December 2004. During this period, 42 patients were treated (22 by endovascular procedure and 20 by surgery). We analyzed 6 patients who were already paraplegic at the time of the treatment. Clinical symptoms were evaluated with regard to motor deficit (using the Aminoff scale,3 McCormick scale,4 modified...
Table 1. McCormick Classification

<table>
<thead>
<tr>
<th>Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Neurologically normal; mild focal deficit not significantly affecting function of involved limb; mild spasticity or reflex abnormality; normal gait</td>
</tr>
<tr>
<td>II</td>
<td>Presence of sensorimotor deficit affecting function of involved limb; mild to moderate gait difficulty, severe pain or dysesthesia syndrome impairing patient’s quality of life; still functions and ambulates independently discomfort in professional activity</td>
</tr>
<tr>
<td>III</td>
<td>More severe neurological deficit; requires cane/brace for ambulation or significant bilateral upper extremity impairment; may or may not function independently</td>
</tr>
<tr>
<td>IV</td>
<td>Severe deficit, requires wheelchair or cane/brace with bilateral upper extremity impairment, usually not independent</td>
</tr>
</tbody>
</table>

Table 2. Aminoff-Logue Disability Scales for Gait and Micturition

<table>
<thead>
<tr>
<th>Gait</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leg weakness, no restriction in gait</td>
<td>1</td>
</tr>
<tr>
<td>Reduced tolerance to exercise</td>
<td>2</td>
</tr>
<tr>
<td>Need for a cane to walk</td>
<td>3</td>
</tr>
<tr>
<td>Need for 2 canes or crutches to walk</td>
<td>4</td>
</tr>
<tr>
<td>Inability to stand, patient in wheelchair or in bed</td>
<td>5</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Micturition</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>0</td>
</tr>
<tr>
<td>Hesitancy, urgency, frequency, altered sensation, but continent</td>
<td>1</td>
</tr>
<tr>
<td>Occasional urinary incontinence or retention</td>
<td>2</td>
</tr>
<tr>
<td>Total incontinence or persistent retention</td>
<td>3</td>
</tr>
</tbody>
</table>

Rankin Scale®, and ASIA classification®; Tables 1 and 2), sphincter disturbances (Aminoff-Logue scale®), and pain.

The optimal treatment for each patient was selected after discussion between neurosurgical and neuroradiological staff. Fistula occlusion was checked by postoperative angiography before discharge. Patients were evaluated at 6 weeks and 6 months after treatment. They also underwent at least one MRI during the follow-up period. In cases exhibiting persistence of intramedullary hyperintensity on T2-weighted MRI and vascular images posterior to the spinal cord, another angiography was performed.

Results

Table 3 shows the clinical presentation, the localization of the fistulae, and follow-up results in our patients.

The mean duration of symptoms before surgery was 39 months (range, 11 to 144 months). The mean duration of paraplegia before surgery was 1.6 months (range, 24 hours to 3 months). At the time of treatment, all patients had paraplegia (Grade IV on the McCormick scale and Grade V on the Aminoff scale, Grade 5 on modified Rankin Scale) with a motor ASIA score between 0 and 10 for both legs. Five patients had a gradual and progressive degradation. One patient (No 1) had acute paraplegia after spinal angiography. Pain typically presented as a radicular pain in the legs and was present in 4 (66%) of our patients. Sphincter disturbance was present in 5 cases.

T2-weighted sagittal MRI sequences showed in all patients perimedullary flow voids on the dorsal aspect of the cord and a diffuse intramedullary hyperintensity that extended to the conus medullaris. The hyperintensity extended up to the lower thoracic region in 2 cases, to the cervical region in 2 cases, and were limited on the conus region in 2 others.

Surgery was used in all cases. Endovascular treatment was considered unsuitable because, in 3 cases, the fistula originated at the same level as the Adamkiewicz artery (Case Nos 1, 5, and 6), and in the remaining 3 cases, the feeding arteries were tortuous or narrow. Surgery was performed under general anesthesia in the prone position through a one-level laminectomy. After a dural opening, the drainage vein was microsurgically interrupted at the level of the shunt with coagulation of the inner and outer layers of the dura. There were no surgical complications. After discharge, all patients were transferred to a rehabilitation center for at least 2 months. Functional improvement started immediately during the postoperative period and continued over 3 to 6 months. Six months after the operation, all patients were able to walk; 3 had a normal gait (Grade I on the McCormick scale, Grade I on the Aminoff scale, Grade 1 on modified Rankin Scale) and 3 needed a cane to walk (Grade II on the McCormick, Grade III on Aminoff scale, Grade 2 of modified Rankin Scale). In all cases, the motor ASIA scores for lower limbs were improved and ranged between 40 and 45 (mean, 42.8). These results remained totally stable over the follow-up period (mean, 3 years). There was no patient with secondary clinical degradation in our experience.

Of 4 patients experiencing pain, 3 patients (75%) reported that their pain was lessened after surgery and the fourth reported it was unchanged. Only 2 of 5 patients (40%) experiencing sphincter disturbance before surgery reported an improvement.

The extension of the intramedullary T2-weighted hyperintensity on pretreatment MRI and its evolution on posttreatment MRI was not correlated with the outcome.

Considering all 36 other patients, long-term follow-up showed that 78% of patients with pretreatment gait disturbance improved, 10% worsened, and 10% remained stable. No patient becomes paraplegic after treatment.

Discussion

Although several studies have analyzed SDAVF s, none have focused particularly on patients with paraplegia. Steinmetz, Cenzato, and Mourier® reported on patients with Grades of IV and V on the Aminoff scale. Among patients confined to bed, Mourier et al reported that 60% were able to walk with crutches, but only one could return to work. Among the 18 patients reported by Steinmetz, 4 were Grade V on the Aminoff scale. After treatment, one patient reached Grade 0, another reached Grade II, and 2 reached Grade III on the Aminoff scale. In these 2 series, no information concerning the duration of the paraplegia was available. Jellema et al reported in 2004 a series of 44 treated patients and mentioned 13 patients who were wheelchair-bound at the time of
treatment. All we know about the outcome of these patients is that at the time of follow-up, this number (wheelchair-bound patients) was 5, including 3 of the original patients. All Grade V patients of the series published by Song10 remained in the same grade after treatment. Afshar11 reported over a series of 19 patients, 6 who were Grade V. Three of these patients remained in the same grade after treatment; however, 3 improved (2 went to Grade IV and one Grade III). Using the qualitative Aminoff scale alone to classify these patients is a debatable decision, because Grade V of this classification system includes patients who are unable to stand and are confined to a wheelchair or bed, although it is unclear whether these patients are completely paraplegic. Atkinson et al12 published a series of 94 dural arteriovenous fistulas (spinal and cranial fistulas) and proposed a modified Aminoff-Logue scale dividing the Grade V into 2 subgroups: Grade 5T (patients with ability to aid in transfer with some movement of legs but confined to bed or wheelchair and unable to stand) and Grade 5P (those who are paraplegic with no ability to aid in transfer nor functional lower extremity movement and who are also confined to bed or a wheelchair). They reported 7 patients with paraplegia (Grade 5P patients of their modified Aminoff-Logue scale). One patient was lost for long-term follow-up, 3 remained paraplegic (Grade 5T), and 2 could ambulate with aids (Grade IV). In these cases, the duration of paraplegia was not precise.

We used the ASIA score to precisely quantify the motor deficit. All of our patients had a motor ASIA score of <10. They had no voluntary movement of legs and the duration of their paraplegia before treatment is clearly noted. Paraplegia in our patients aged from 24 hours to 3 months, and we admit that recovery is more hazardous in long-standing paraplegia.

SDAVFs are an abnormal communication between a radicular artery and a vein within the dura matter near a nerve root. This direct communication provokes venous hyperpressure at the level of the valveless spinal cord veins. Using intraoperative pressure recording, Hassler et al13,14 clearly demonstrated an increase in intrinsic venous pressure (the venous pressure in dural arteriovenous fistula was approximately 70% of the systemic arterial pressure) and the fact that this venous pressure increases concomitantly with blood pressure. This leads to a reduction in the intramedullary arteriovenous pressure gradient and thus in blood stagnation at the spinal cord level. A decrease in tissue perfusion and progressive hypoxia of neural tissue may follow. This provokes histological changes, including a thickening of intramedullary and pial vessel walls with hyalinization and often with thrombus within the vessels. There can also be neuronal depopulation, gliosis, and lipid-laden macrophages. Necrosis of the white or gray matter may follow.15

All these modifications explain the progression of the neurological deficit. Given the gradual and progressive installation of these phenomena, it is possible that treatment of the venous hyperpressure could arrest the progression of symptoms. If there are no definitive histological lesions, it is possible that all signs of disease would decline once the fistula is treated.

The value of MRI changes as a predicting factor of outcome remains controversial. Willinsky et al16 considered that there was some correlation between MRI and clinical outcome but at the same time they pointed out that MRI evaluation could not distinguish those who had improved from those who stabilized. However, the majority of authors7,10,12 admit that no correlation is found between clinical outcome and pre- or posttreatment MRI changes. There is especially no correlation between the extent of abnormal T2-weighted hyperintensity on pretreatment MRI and the clinical outcome.10 Our results support this feature.

**Conclusion**

Our results demonstrate that endovascular or surgical treatment is beneficial for completely paraplegic SDAVF patients. All of our patients were able to walk independently within 6 months after surgery. Thus, in light of these encouraging results, we believe that paraplegia due to SDAVF is typically a reversible symptom, and therefore treatment should be performed even in patients with paraplegia.

**Disclosures**

None.
Table 3. Continued

<table>
<thead>
<tr>
<th>First ASIA</th>
<th>First Aminoff Scale</th>
<th>First McCormick Scale</th>
<th>Last ASIA</th>
<th>Last Aminoff Scale</th>
<th>Last McCormick Scale</th>
<th>MRS</th>
<th>Pain</th>
<th>Sphincter*</th>
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<tr>
<td>8</td>
<td>5</td>
<td>IV</td>
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<td>1</td>
<td>I</td>
<td>1</td>
<td>Improved</td>
<td>2</td>
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Stroke. 2008;39:2756-2759; originally published online July 17, 2008;
doi: 10.1161/STROKEAHA.108.517037
Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

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