A variety of classifications of spinal vascular malformations have been proposed.1,2 The Bicêtre group suggested a classification based on genetic and hereditary perspectives that divided conditions into 3 main groups: genetic hereditary lesions, genetic nonhereditary lesions, and single lesions that may reflect an incomplete expression of a genetic condition.3,4 The vast majority of spinal vascular malformations fall into this third group.

Spinal cord arteriovenous malformations comprise ≈20% to 30% of all spinal vascular malformations.5,6 The angioarchitecture at the transition from artery into vein can be either through a network (the nidus) or direct (ie, fistulous).2,3 Fistulous AVMs are located superficially and only rarely possess intramedullary compartments and are therefore usually called perimedullary arteriovenous fistulas.7 Their cranial counterpart would be the pial arteriovenous fistulas that are typically also superficial lesions present in the subpial space. Nidus-type AVMs, however, may be considered as the counterpart of brain AVMs. The nidus is located in the spinal cord parenchyma; however, a superficial compartment can also be exposed to the subpial space.

Clinical presentations of spinal cord AVMs can involve either progressive myelopathy or acute hemorrhage (subarachnoid or intramedullary).8–11 Nonhemorrhagic acute neurological deficits can be present because of acute thrombosis of venous pouches with mass effect and disturbance of venous outflow. Fistulous AVMs have been reported to present more frequently with progressive myelopathy, whereas the nidus types have been associated with an increased risk of hemorrhage.12,13 However, given the rarity of these diseases, there are few larger patient series that evaluated clinical findings, treatment modalities, and long-term outcome of these lesions. Therefore, in this article, we report our single-center experience with treatment of spinal cord AVMs and describe the

**Background and Purpose**—As a result of the rarity of spinal cord arteriovenous malformations (AVM), there are only a few series available that describe clinical features, outcome after treatment, and natural history of these lesions. In this article, we aim to describe our experience with both nidus- and fistulous-type spinal cord AVMs.

**Methods**—Forty-four consecutive patients with spinal cord AVMs were retrospectively reviewed. There were 26 patients with a nidus-type and 18 patients with a fistulous-type AVM. Treatments were performed with embolization (n=23), surgery (n=13), combined embolization–surgery (n=3), or conservative management (n=5). Clinical features, radiological findings, treatment results, and clinical outcomes were assessed.

**Results**—Patients with nidus-type AVMs were younger at presentation and more often presented with hemorrhage, with a higher proportion of hematomyelia than fistulous-type AVMs (P<0.05). Progression of clinical presentation from hemorrhage to congestive myelopathy during follow-up was noted in 5 patients, all of which had AVMs of the nidus type. Complete obliteration could be achieved more often in the fistulous type (72%) than in the nidus type (27%). Improved or stable clinical status at last follow-up was noted in 100% of fistulous-type and 77% of nidus-type patients. Long-term clinical deterioration was noted in 6 of 26 patients with nidus-type (23%) AVMs and was related to recurrent bleeding (n=3) or progressive venous congestion (n=3). Overall rebleed rate after presentation with hemorrhage was 7 in 145.5 patient-years (4.8%/y) if the lesion was not treated, 3 in 102 patient-years (2.9%/y) after partial treatment, and 0 in 47.5 patient-years (0%) after complete treatment.

**Conclusions**—Nidus and fistulous spinal cord AVMs have different clinical features and obliteration rates, which may affect their long-term prognosis. (Stroke. 2014;45:2606-2612.)

**Key Words:** arteriovenous malformations ◼ embolization ◼ spinal cord ◼ surgery ◼ treatment outcome

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relationship of the types of angioarchitecture (nidus versus fistulous type) with their respective clinical features and long-term outcomes.

Methods

Patients

We retrospectively reviewed 44 consecutive patients with spinal cord AVMs who were managed at the Toronto Western Hospital between January 2000 and October 2013 and who were entered prospectively into our institutional database. The study was approved by our institutional review board. The following inclusion and exclusion criteria were used for this study: purely intradural, nonradicular, nonmetameric spinal vascular malformations, that is, we excluded patients with paravertebral, epidural, dural, or radicular AVMs and cases that involved both the intra- and extradural spaces (eg, juvenile AVMs or Cobb syndrome). The study group consisted of 16 men and 28 women with a mean age at first presentation of 32.0 years (range, 8–77 years). The clinical follow-up was performed by neurosurgical and interventional neuroradiology staffs and fellows during our weekly AVM outpatient clinic, whereas angiographic imaging follow-up was performed by endovascular staff; neither clinical nor imaging follow-up was necessarily done by the treating physician. Mean clinical follow-up from the onset was 9.6 years (range, 1–36 years; median 5.0 years).

Radiological Evaluation

Pretreatment spinal MRI was available in 43 patients, and contrast-enhanced spinal MR angiography was available in 33 of these patients. MRI was evaluated with respect to presence of congestive edema or hemorrhage, as well as type of hemorrhage. Congestive myelopathy was suspected based on clinical examination and MRI demonstrating enlargement of the spinal cord with central confluence, focal, or extensive T2 hyperintensity over multiple segments, with or without patchy contrast enhancement.14 Spinal catheter angiography with complete evaluation of all possible feeding arteries was performed in all patients, and a confirmative diagnosis was made on the basis of spinal angiograms. Angioarchitecture, locations, and types of the vascular malformation were evaluated by spinal angiography. Regarding the type of lesions, spinal cord vascular malformations were divided into nidus and fistulous types according to the initial opinion about existence of intervening nidus on spinal angiogram made by the interventional neuroradiologists (K.G.T. and T.K.) and reread by the first author who was blinded to the initial evaluation. The angioarchitecture was evaluated regarding arterial feeder multiplicity, venous drainage (anterior spinal vein, posterior perimedullary veins, and radicular veins), and existence of focal area of weakness such as intramedullary aneurysms or venous outpouchings. The lesion locations were classified into cervical cord (from T1 to T7), lower thoracic cord (from T8 to L1), and filum terminale (below the conus medullaris).

Treatment

Spinal cord AVMs were treated with embolization in 23, surgery in 13, combined embolization–surgery in 3, or conservative management in 5 patients. In all cases of patients treated by an endovascular approach, glue (N-butyl cyanoacrylate) was used as the sole embolizing agent after superselective microcatheterization using, in all cases, a Magic 0.12 microcatheter (Balt, Montmorency, France) aided by a 0.07 microguidewire (Mirage, Microvention, Hybrid, Balt). The microcatheter was introduced coaxially through a 5F Cobra-2 catheter (Cook Medical, Bloomington, IN) placed in the segmental artery that supplied the AVM under roadmap conditions. Working projections were chosen in recent years after 3-dimensional rotational angiography. Superselective series were obtained during slow advancement of the catheter to the nidus. Embolizations were performed only when we can reach the feeder far enough to avoid accidental embolization of anterior spinal artery. Surgery was indicated when the embolization was not feasible, failed, or was perceived to be too dangerous.

The surgical approach and the intraoperative technique used varied according to lesion location, type, and clinical status. In general, after laminectomy at the predetermined level, surgical resection of AVM was performed with a microsurgical technique under the guidance of microscope. Although dorsal deep lesions were typically approached through the dorsal nerve root entry zone, ventral lesions were approached after cutting of the dentate ligaments and gentle rotation of the cord. In selected cases, monitoring of evoked potential, intraoperative conventional angiography, and, more recently, intraoperative indocyanine green video angiography were used. Conservative management was proposed when embolization or surgery was deemed too dangerous while symptoms were benign (ie, incidental finding or mild congestion only) and stable. We classified the results of treatment as complete obliteration or partial obliteration according to the angiographic results immediately after surgery or at 2 to 4 months after embolization. The latter date was chosen because determining the obliteration rate at the end of embolization session may lead to an overestimation of true obliteration: Directly after an embolization, there may be local vasospasm that can obscure residual flow into an AVM. In addition, if glue has not penetrated through the nidus into the proximal feeding vein (so-called ligation embolization), belated reopening of the AVM through local collaterals may occur and therefore also lead to false-positive occlusions. Complete obliteration was defined as an angiographically negative status, regardless of treatment modalities. Partial obliteration was defined if any residual arteriovenous shunting remained visible on the angiogram. In the present series, follow-up angiography later than the 2 to 4 months control was only performed if the patient’s clinical status deteriorated.

Clinical Evaluation

Pretreatment and further clinical status were evaluated according to the Aminoff and Logue scale collected based on the medical record: (0) normal, (I) leg weakness/abnormal gait but no restriction of activity, (II) restrictive activity, (III) requiring 1 stick to walk, (IV) requiring 2 sticks/crutches to walk, and (V) wheelchair-bound.6 Severe neurological impairment at onset was defined by an Aminoff and Logue scale of IV or V, whereas a scale value of 0, I, II, or III was not considered to be severe. Clinical outcomes were evaluated at short-term (within 1 month after treatment) and at last available clinical follow-up. Short-term and long-term outcomes were rated according to Aminoff and Logue scale as improved, stable, or deteriorated.

Statistical Analysis

A Student t test was used for the mean age comparison, and a Fisher exact test was used to compare incidences among the 2 groups. x² goodness-of-fit test was used for sex ratio. Statistical significance was defined as a value of P<0.05.

Results

The pretreatment clinical data and angiographic findings are summarized in Table 1. Nidus-type AVMs were found in 26 patients, whereas fistulous-type AVMs were found in 18 patients, including 7 patients with filum terminale arteriovenous fistulas. Patients with nidus-type AVM were younger at presentation compared with patients with fistulous-type AVM (24.2 versus 43.2 years; P=0.0002). A slight but nonsignificant predominance in female patients was observed in both nidus (65%) and fistulous types (61%). First clinical presentation was hemorrhage in 25 patients (57%) and myelopathy in 16 patients (36%), whereas in 3 patients the AVMs were found incidentally (7%). Hemorrhagic presentation including subarachnoid hemorrhage (SAH) or hematomyelia was more frequent in nidus-type AVMs than fistulous-type AVMs (81% versus 22%; P=0.0002), whereas progressive myelopathic presentation was more frequent in the fistulous-type

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Among the patients with hemorrhagic presentation, hematomyelia (n=19) was found more often in nidus-type patients (18 versus 1; \( P = 0.03 \)) than fistulous-type patients (1; \( P = 0.03 \)), whereas SAH (n=6) was similar in both types (3 versus 3; Table 2).

Severe neurological impairment at onset was found in 6 of 19 patients with hematomyelia (32%) and in 1 of 6 patients with SAH (17%). Recurrent hemorrhage before treatment was noted in 7 of 25 patients with hemorrhagic presentation (28%). Recurrent hemorrhage with hematomyelia occurred in 4 patients with nidus-type AVM (who bled, respectively, after 3, 4, 5, and 16 years after the first hematomyelia). Recurrent hemorrhage with SAH occurred in 2 patients with nidus-type AVM (who bled 11 and 16 years after first SAH, respectively) and in 1 patient with fistulous-type AVM (who bled 1 month after first SAH). Relatively long conservative management periods before rebleeding in 6 of these 7 patients were related to belated referral or good clinical recovery with clinically stable conditions.

Although patients with myelopathic presentation did not experience hemorrhage during follow-up, progression toward congestive myelopathy during follow-up was found in 5 of 25 patients with initial hemorrhagic presentation (20%). These 5 patients had nidus-type AVMs in the lower thoracic region and had initially presented with hematomyelia. The progression occurred during conservative management before active

### Table 1. Clinical Features and Angiographic Findings

<table>
<thead>
<tr>
<th></th>
<th>All (n=44)</th>
<th>Nidus Type (n=26)</th>
<th>Fistulous Type (n=18)</th>
<th>( P ) Value*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age (y, SD)</td>
<td>32.0±17.8</td>
<td>24.2±12.1</td>
<td>43.2±18.9</td>
<td>0.0002</td>
</tr>
<tr>
<td>Sex (men/women)</td>
<td>16/28</td>
<td>9/17</td>
<td>7/11</td>
<td>NS</td>
</tr>
<tr>
<td>Mean F/U (y, SD)</td>
<td>9.6±9.7</td>
<td>13.8±10.2</td>
<td>3.2±3.6</td>
<td></td>
</tr>
<tr>
<td>Clinical presentation</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hemorrhage</td>
<td>25 (57%)</td>
<td>21 (81%)</td>
<td>4 (22%)</td>
<td>0.0002</td>
</tr>
<tr>
<td>Myelopathy</td>
<td>16 (36%)</td>
<td>5 (19%)</td>
<td>11 (61%)</td>
<td>0.01</td>
</tr>
<tr>
<td>Others†</td>
<td>3 (7%)</td>
<td>0</td>
<td>3 (17%)</td>
<td>NS</td>
</tr>
<tr>
<td>Angioarchitecture</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nidus</td>
<td>26 (59%)</td>
<td>26 (100%)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Multiple arterial feeder</td>
<td>33 (75%)</td>
<td>22 (85%)</td>
<td>11 (61%)</td>
<td>NS</td>
</tr>
<tr>
<td>Perimedullary venous drainage</td>
<td>30 (68%)</td>
<td>16 (62%)</td>
<td>14 (78%)</td>
<td>NS</td>
</tr>
<tr>
<td>Radicular venous drainage</td>
<td>13 (30%)</td>
<td>10 (38%)</td>
<td>3 (17%)</td>
<td>NS</td>
</tr>
<tr>
<td>Anterior spinal venous drainage</td>
<td>34 (77%)</td>
<td>19 (73%)</td>
<td>15 (83%)</td>
<td>NS</td>
</tr>
<tr>
<td>Area of weakness</td>
<td>14 (32%)</td>
<td>9 (35%)</td>
<td>5 (28%)</td>
<td>NS</td>
</tr>
<tr>
<td>Location</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cervical</td>
<td>10 (23%)</td>
<td>7 (27%)</td>
<td>3 (17%)</td>
<td>NS</td>
</tr>
<tr>
<td>Upper thoracic</td>
<td>5 (11%)</td>
<td>5 (19%)</td>
<td>0</td>
<td>NS</td>
</tr>
<tr>
<td>Lower thoracic</td>
<td>22 (50%)</td>
<td>14 (54%)</td>
<td>8 (44%)</td>
<td>NS</td>
</tr>
<tr>
<td>Filum terminale</td>
<td>7 (16%)</td>
<td>0</td>
<td>7 (39%)</td>
<td>NS</td>
</tr>
<tr>
<td>Clinical status at onset‡</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nonsevere deficit</td>
<td>35 (80%)</td>
<td>19 (73%)</td>
<td>16 (89%)</td>
<td>NS</td>
</tr>
<tr>
<td>Severe deficit</td>
<td>9 (20%)</td>
<td>7 (27%)</td>
<td>2 (11%)</td>
<td>NS</td>
</tr>
</tbody>
</table>

F/U indicates follow-up; and NS, not significant.

*Comparison between nidus and fistulous type.
†Incidental finding.
‡Severe deficit at onset was defined by an Aminoff and Logue scale of IV or V, whereas a scale value of 0, I, II, or III was considered to be nonsevere deficit.

### Table 2. Comparison Between Hematomyelia and SAH Regarding Types, Recurrent Hemorrhage, Myelopathic Progression, and Clinical Outcomes

<table>
<thead>
<tr>
<th></th>
<th>All (n=25)</th>
<th>Hematomyelia (n=19)</th>
<th>SAH (n=6)</th>
<th>( P ) Value*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nidus type:fistulous type</td>
<td>21:4</td>
<td>18:1</td>
<td>3:3</td>
<td>0.03</td>
</tr>
<tr>
<td>Severe neurological impairment at first presentation</td>
<td>7 (28%)</td>
<td>6 (32%)</td>
<td>1 (17%)</td>
<td>NS</td>
</tr>
<tr>
<td>Recurrent hemorrhage before treatment</td>
<td>7 (28%)</td>
<td>4 (21%)</td>
<td>3 (50%)</td>
<td>NS</td>
</tr>
<tr>
<td>Progression to cord congestion</td>
<td>5 (20%)</td>
<td>5 (26%)</td>
<td>0</td>
<td>NS</td>
</tr>
<tr>
<td>Recurrent hemorrhage after treatment</td>
<td>3 (12%)</td>
<td>1 (5%)</td>
<td>2 (33%)</td>
<td>NS</td>
</tr>
<tr>
<td>Long-term outcomes (improved/stable/deteriorated)</td>
<td>8/13/4 (32%/52%/16%)</td>
<td>5/12/2 (26%/63%/11%)</td>
<td>3/1/2 (50%/17%/33%)</td>
<td>NS</td>
</tr>
</tbody>
</table>

NS indicates not significant; and SAH, subarachnoid hemorrhage.

*Comparison between hematomyelia and SAH.
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treatment in 3 patients and occurred after partial embolization in 2 patients. The range of time before progression was 4 to 34 years (mean, 12 years; median, 5 years). In 2 patients, spontaneous thrombosis of the draining vein was suggested on MRI during conservative management before treatment was initiated; both patients improved after partial embolization or complete surgical removal (Figure 1).

Multiple feeders were found in 22 of 26 nidus-type A VMs (85%) and 11 of 18 fistulous-type A VMs (61%). Absence of regional radicular venous outflow was noted in 16 of 25 patients with hemorrhagic presentation (64%) and 15 of 16 patients with myelopathic presentation (94%). The single patient who presented with myelopathy despite the presence of radicular venous outflow harbored a high-flow shunt. Focal areas of weakness were found in 11 of 25 patients with hemorrhagic presentation (44%) and 3 of 19 patients with nonhemorrhagic presentation (16%).

Treatment Results
Of the 26 patients with nidus-type A VMs, 18 patients underwent embolization, 4 surgery, 2 combined embolization–surgery, or 2 conservative management (with 26 patient-year follow-up; Table 3). Of the 18 patients with fistulous-type A VMs, 5 patients underwent embolization, 9 surgery, 1 combined embolization–surgery, or 3 conservative management. The 3 patients who were managed conservatively had a total follow-up of 7 patient-years. Complete angiographic obliteration could be achieved more often in the fistulous type (72%, 13 of 18) than in the nidus type (27%; 7 of 26; Table 3). Complete obliteration could be achieved with embolization in 3 of 18 nidus-type A VMs (17%; Figure 2) and in 5 of 5 fistulous-type A VMs (100%). Complete obliteration after surgery (including combined embolization–surgery) was noted in 4 of 6 nidus-type A VMs (67%) and 8 of 10 fistulous-type A VMs (80%).

When a focal area of weakness (aneurysm) was deemed to be the cause of hemorrhage (n=11), this component could be excluded by embolization in 8 patients or by surgery in 3 patients. Of these 11 patients who underwent partial (targeted) treatment for a presumed area of weakness, recurrent hemorrhage occurred in 2 patients (after 8 and 9 years, respectively).

Overall rebleed rate after presentation with hemorrhage was 7 in 145.5 patient-years (4.8%/y) if the lesion was not treated. This rebleed rate was 3 in 102 patient-years (2.9%/y) after partial treatment and 0 in 47.5 patient-years (0%) after complete treatment.

In 2 patients, belated reconstitution of shunting after initial angiographic obliteration was noted. One occurred in a nidus-type patient with myelopathic presentation 1 year after embolization. The other occurred in a fistulous-type patient with myelopathic presentation 2 years after surgery. At the last follow-up, the former had clinically deteriorated, whereas the latter was stable.

In 5 patients (4 nidus-type and 1 fistulous-type AVM), the attempt to embolize failed, because a safe position of the microcatheter tip in the anterior spinal artery branch supplying the lesion could not be obtained. Three of them (2 nidus-type and 1 fistulous-type AVMs) underwent surgical resection, whereas the other 2 patients with nidus-type AVM did not undergo surgery because of the perceived surgical risk.

Table 3. Long-Term Clinical Outcomes According to the Treatment Options

<table>
<thead>
<tr>
<th>Long-Term Outcomes</th>
<th>No. of Patients</th>
<th>Nidus Type</th>
<th>Fistulous Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>All</td>
<td>44 (16/22/6)</td>
<td>26 (8/12/6)</td>
<td>18 (8/10/0)</td>
</tr>
<tr>
<td>Embolization</td>
<td>23 (6/11/6)</td>
<td>18 (4/8/6)</td>
<td>5 (2/3/0)</td>
</tr>
<tr>
<td>Partial obliteration</td>
<td>15 (3/7/5)</td>
<td>15 (3/7/5)</td>
<td>0</td>
</tr>
<tr>
<td>Complete obliteration</td>
<td>8 (3/4/1)</td>
<td>3 (1/1/1)</td>
<td>5 (2/3/0)</td>
</tr>
<tr>
<td>Surgery</td>
<td>13 (6/7/0)</td>
<td>4 (1/3/0)</td>
<td>9 (5/4/0)</td>
</tr>
<tr>
<td>Partial obliteration</td>
<td>4 (2/2/0)</td>
<td>2 (1/1/0)</td>
<td>2 (1/1/0)</td>
</tr>
<tr>
<td>Complete obliteration</td>
<td>9 (4/5/0)</td>
<td>2 (0/2/0)</td>
<td>7 (4/3/0)</td>
</tr>
<tr>
<td>Combined embolization-surgery*</td>
<td>3 (3/0/0)</td>
<td>2 (2/0/0)</td>
<td>1 (1/0/0)</td>
</tr>
<tr>
<td>Conservative management</td>
<td>5 (1/4/0)</td>
<td>2 (1/1/0)</td>
<td>3 (0/3/0)</td>
</tr>
</tbody>
</table>

*Partial embolization followed by complete surgery.
no demonstrable area of weakness on angiography, and their benign clinical course. Except for these 2 patients who underwent no further treatment despite embolization failure, all partial treatments were effective in achieving treatment goals (ie, resolution of the current clinical presentation by elimination of areas of weakness or reduction of venous congestion).

**Clinical Outcome**

Short-term outcome results showed that 2 patients deteriorated in the immediate post-treatment period because of postprocedural complications. One was a patient with nidus-type AVM, who presented with hematomyelia. Although no focal area of weakness was identified on the spinal angiogram, partial embolization was performed and an SAH occurred several hours after the treatment. This was presumed to be related to flow redirection toward an area of weakness not demonstrated at initial angiography because of the presence of the hematoma. This patient made a significant clinical improvement 1 month after treatment. The other patient who deteriorated after treatment had a fistulous AVM and presented with

![Figure 2](image1.png)

**Figure 2.** Complete obliteration of a nidus-type arteriovenous malformation (AVM) with embolization in a 17-year-old woman. Intramedullary hematoma (arrow) with associated edema is noted at upper thoracic cord on preoperative T2-weighted image (A). Spinal angiogram from right supreme intercostal artery shows a nidus-type AVM (arrow) supplied by the radiculopial artery arising from the segmental artery T3 (B). Superselective angiogram with microcatheter tip (arrow) positioned at right radiculopial artery shows precise angioarchitecture of the nidus (C). Follow-up spinal angiogram taken 4 months after embolization shows complete obliteration of the AVM (D). This patient recovered fully from initial paraplegic impairment on 2 years after embolization.

![Figure 3](image2.png)

**Figure 3.** Venous ischemia after embolization in a 52-year-old woman with fistulous-type arteriovenous malformation. Dilated perimedullary veins (arrow) without remarkable cord signal change are noted on preoperative T2-weighted image (A). Spinal angiogram from the right costocervical trunk shows a radiculopial artery (arrow) supplying the shunt and dilated draining veins (B). One month after the embolization, new T2 hyperintensity in the cervical cord (arrow) is noted and draining veins are remarkably shrunken (C). A small hyperintensity on T1-weighted image is suggestive of draining venous thrombosis (D).
myelopathic signs. One week after complete embolization, the cord deteriorated and MR showed new T2 hyperintensity in the spinal cord, which was presumably related to progressive thrombosis of the draining vein (Figure 3). This patient became stable with partial clinical recovery 6 months after treatment.

Long-term outcome (mean, 13.8 years; median, 12.5 years) of patients with nidus-type AVMs showed improved clinical status in 8 patients (31%), clinical stability in 12 patients (46%), and worsening in 6 patients (23%). Long-term clinical deterioration was related to recurrent hemorrhage after treatment in 3, recurrent myelopathy after treatment in 2, and progression from hemorrhage to myelopathy in 1 patient.

Long-term outcome (mean, 3.2 years; median, 2 years) of patients with fistulous-type AVMs showed improved clinical status in 8 patients (44%) and stability in 10 patients (56%).

Discussion

Clinical manifestations and age at first presentation differed according to the type of spinal cord vascular malformation. Patients with nidus-type AVMs were younger at first presentation and more often presented with hemorrhage, in particular hematomyelia, compared with patients with fistulous-type AVMs. Nidus-type AVMs had lower complete obliteration rate that might lead to delayed rebleeding, clinical progression from hemorrhage to myelopathy, and tendency to long-term deterioration. Lesion location or clinical status at onset did not show significant difference between the nidus and fistulous types. The retrospective design and the sample volume in our study limit the evaluation of superiority of a certain treatment modality, which would require a randomized prospective design. In our opinion, treatment has to be individualized in relation to the AVM angioarchitecture, the clinical status, and the expertise of the local treatment team.

The most frequent clinical presentation was different between the 2 types of vascular malformations in this study. Fistulous types were more often associated with congestive myelopathy, whereas nidus types were more commonly associated with hemorrhage. One may argue that the larger arteriovenous shunt present in the fistulous malformations should induce more profound arterialization resulting in higher venous pressure, leading to increased cord congestion, reduced tissue perfusion, and therefore venous cord ischemia. However, the nidus-type AVM might be more fragile, which could lead to more frequent rupture.

The difference in the mean age at first presentation may be related to the preferential clinical manifestations because progressive myelopathy manifests itself if there is significant radicular venous outflow obstruction that occurs later in life.17 The mean age at first presentation was younger in nidus-type patients than in fistulous-type patients, which is not consistent with results from Lv et al10 and Cho et al,12 which indicated that the mean age at presentation was younger in the fistulous type. However, the age of patients with nidus-type AVMs in our study (mean age, 24.2 years) is comparable with a recent meta-analysis of 293 nidus-type AVMs where they reported a mean age at presentation of 29.1 years.13

A higher rate of complete obliteration could be achieved with the fistulous-type AVMs. This may explain the difference in the mean follow-up period between the 2 types because the completely obliterated lesions did not undergo further follow-up. Treatment options for fistulous types are determined by the size of the vessels.18 Microsurgery may be preferred if the feeding artery is too small for microcatheter navigation although the ventrally located fistulas are still challenging. Embolization is the first line of treatment in our institution in most other cases. For nidus-type AVMs, with conditions such as multiple feeders or a high risk of anterior spinal arterial compromise, treatment may be challenging or result in incomplete obliteration. However, treatment does have role in these patients if one can demonstrate reduction in bleeding rate after partial embolization. In a recent meta-analysis,13 a hemorrhage risk of nidus-type AVMs before treatment was 4%/y in unruptured AVMs and 10% for AVMs with previous hemorrhage. In this meta-analysis, it was stated that the hemorrhage rate decreased significantly even after partial obliteration. Therefore, we propose that the goal of treatment of nidus-type AVMs presenting with hemorrhage should be the identification and elimination of focal areas of weakness such as aneurysms. In patients presenting with hemorrhage, we found a probable cause for the hemorrhage in 11 patients, which could be eliminated by embolization in all cases. The hemorrhagic rate in partially treated patients was 2.9%/y (ie, 3 hemorrhages in 102 patient-years of follow-up), which is significantly lower than the reported annual rupture rate of previously ruptured AVMs according to the aforementioned meta-analysis.13 In those patients who rebled several years after treatment, we found a change in the angioarchitecture. Because belated recurrences occurred in 2 of our patients after initial angiographic complete obliteration, we are now advocating to perform the belated angiography at 12 months rather than 2 to 4 months to determine the success of the embolization.

Regardless of the AVM type, patients who presented with congestive myelopathy did not experience hemorrhage during follow-up. However, clinical presentation that progressed from hemorrhage to congestive myelopathy was noted in 5 patients. These results may suggest that the long-term prognosis of patients with congestive myelopathic presentation may differ from the patients who present with hemorrhage. The patients who experienced progression from hemorrhage to congestive myelopathy were all nidus-type patients who initially presented with intramedullary hemorrhage.

In conclusion, clinical features and long-term prognosis of actively treated spinal cord AVMs differ according to their types, nidus versus fistulous. This in turn may be related to a lower complete obliteration rate after treatment in nidus-type AVMs as well as different inherent hemodynamics between the 2 types.

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Disclosures
None.

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Clinical Features and Outcomes of Spinal Cord Arteriovenous Malformations: Comparison Between Nidus and Fistulous Types
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