Surgical Treatment of Spinal Arteriovenous Malformations: Vascular Anatomy and Surgical Outcome

Po-An Tai, Yong-Kwang Tu, and Hon-Man Liu

Background and purpose: Spinal arteriovenous malformations (AVMs) are rare but debilitating lesions of the central nervous system. This study evaluated the outcome in patients with spinal AVMs treated surgically, and the relationship between outcome and the vascular anatomy of the lesions.

Methods: These AVMs were classified into four types: dural AVM (type I), glomus AVM (type II), juvenile AVM (type III), and intradural direct arteriovenous fistula (type IV). Either interruption of the feeding vessels or excision of the AVMs was performed in all patients.

Results: Intradural AVMs manifested as subarachnoid or intramedullary hemorrhages, whereas dural AVM manifested as epidural hemorrhage in two patients, and as an episode of subarachnoid hemorrhage in one patient. The nidus in five of the six dural AVMs was below the mid-thoracic level. In six of the seven patients with intradural AVMs, the nidus was located in the cervical region. The prognosis of patients with dural AVMs was generally good, but in patients with intradural AVMs, motor recovery was worse and resection was more difficult.

Conclusions: Correct diagnosis and classification of spinal AVMs are the key prerequisites for successful treatment. The surgical outcome is closely related to the vascular anatomy of the lesion. For dural AVMs, only surgical interruption of the arteriovenous shunting at the dural sleeve of the nerve is required and good surgical results are often obtained. Intradural direct arteriovenous fistula responded well to surgery, whereas combined endovascular and microsurgical techniques can minimize the chance of rebleeding in intramedullary AVMs.

Spinal arteriovenous malformations (AVMs) are rare lesions of the central nervous system that can cause serious neurologic deficits and can mimic a variety of neurologic conditions. If unrecognized and left untreated, they may result in major disability within a few years after the onset of the initial symptoms [1, 2]. There are two major types of spinal AVMs, defined by radiographic appearance at selective spinal angiography: small AVMs of the radicular arteries on the dural sleeve of the nerve roots, referred to as spinal dural AVM due to their dural arterial origin; and intradural AVMs perfused by the medullary arteries and involving the pia or parenchyma of the cord, which are further classified into three types (juvenile AVM, glomus AVM, and intradural arteriovenous fistula) [3, 4]. Despite the similar gross appearance of these two types of spinal AVMs, their clinical presentation and response to treatment are often entirely different [3, 5]. Unlike intradural AVMs, dural AVMs have a strong male predilection and 80% have symptom onset after age 40. Dural AVMs are predominantly located in the lower thoracic and lumbar regions, and usually present as a gradual onset of paraparesis and/or sphincter dysfunction [3]. This study evaluated the
relationship between outcome and vascular anatomy in patients with spinal AVMs treated by surgical management.

**Patients and Methods**

Medical records from 10 male and three female patients with spinal AVMs who were treated at the National Taiwan University Hospital during the period from 1990 through 1997 were reviewed. All patients underwent definite treatment such as surgery or embolization and had complete records. Radiologic studies, including computerized tomography (CT), magnetic resonance (MR) imaging, and selective spinal angiography, were reviewed. The age of the patients ranged from 1 to 69 years (mean, 22.4 yr). All patients underwent selective spinal angiography for preoperative diagnosis and postoperative follow-up.

Dural AVM, also called dural arteriovenous fistula (AVF) or type I spinal AVM, is defined as a vascular lesion in which the nidus of the arteriovenous shunt is embedded in the dural sleeve of the dorsal nerve root and the adjacent spinal dura is inside the intervertebral foramen. Dural branches of the radicular arteries supply these AVMs. Intramedullary AVMs are defined as a vascular lesion in which the nidus is in the spinal cord or on the pia of the spinal cord and which receives its blood supply from medullary arteries. Intramedullary AVMs are subclassified into intramedullary AVMs, of which the nidus is within the parenchyma of the spinal cord, and intramedullary direct AVF (type IV spinal AVM), in which the transition from artery to vein occurs without an intervening glomus of abnormal vessels. Intramedullary AVMs are further divided into glomus type (type II spinal AVM), composed of a tightly packed localized nidus of abnormal vessels within the spinal cord, and juvenile type (type III spinal AVM), which occupy the entire spinal canal at longer segments, and are supplied by numerous, frequently large arteries [3].

Of the 13 patients, six had dural AVMs (type I) and seven had intradural AVMs. Of the intradural AVMs, five patients had intramedullary AVMs and two patients had intradural direct AVFs (type IV). Among the five intramedullary AVMs, three were of the glomus type (type II) and two were of the juvenile type (type III).

All patients with dural AVMs or intramedullary AVMs underwent operative procedures to interrupt the AVFs or excise the AVMs microsurgically. Preoperative embolization with 30% histoacryl mixed with 70% lipiodol was performed in one case. Data on clinical presentation, vascular anatomy, therapy, and outcome were analysed.

**Results**

There was male predominance among the six patients with dural lesions and seven with intradural lesions. The age of patients with symptoms from dural AVMs ranged from 11 to 69 years (mean, 31.5 yr). In contrast, all patients except one with symptoms from intradural AVMs were less than 24 years old (1–45 yr; mean, 15.3 yr) ($p < 0.05$; between patients with the two types of spinal AVMs).

Presenting symptoms caused by dural and intradural AVMs included motor weakness, sensory abnormalities, sphincter disturbances, and pain (Table 1). All 13 patients presented with motor weakness, either paraparesis or hemiparesis. The majority of patients with dural AVMs presented with progressive motor, sensory, and sphincter symptoms. Three patients with dural AVMs had hemorrhages (1 subarachnoid hemorrhage and 2 epidural hemorrhages). Patients with dural AVMs had equal frequency of gradual onset or acute onset of symptoms.

In the present series, dural AVMs differed from intradural AVMs with respect to the level of involvement along the longitudinal spinal axis, arterial supply, and site of the nidus. The nidus of the dural AVMs originated in the thoracic or lumbar region in four of the six patients and in the sacral region in one patient (Table 2). In only one patient with dural AVM was there a cervical nidus. Six of the seven patients with intradural AVMs had a cervical location of the nidus. The intradural spinal AVM of the patient in whom the nidus was located at the thoracic level was, in fact, a direct AVF.

Other associated conditions included coexisting cerebellar AVM in a patient with dural AVM and a cavernous hemangioma within the bony part of the C4 transverse process in a patient with intradural AVM (Table 3). Two patients had spinal intradural lipomas. One of these lipomas was a conus lipoma with a tethered cord in a patient with a dural AVM supplied by bilateral S1 radicular arteries. The other was a...
Table 2. Location of the vascular nidus in patients with different types of spinal arteriovenous malformations (AVMs)

<table>
<thead>
<tr>
<th>Location of the nidus</th>
<th>Dural AVMs (n = 6)</th>
<th>Intradural AVMs (n = 7)</th>
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</thead>
<tbody>
<tr>
<td>C2–4</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>C5–7</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>T1–3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>T4–6</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>T7–9</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>T10–12</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>L1–3</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>L4–S1</td>
<td>1</td>
<td>0</td>
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medullocerebellar lipoma in a patient with a type III spinal AVM at C1–3 levels. Both lipomas came into contact with the AVMs and contributed to the physical manifestation. The two pediatric patients with intradural direct AVFs were noted to have prominently enlarged venous aneurysms.

All patients underwent microsurgery to interrupt feeding vessels or excise AVMs. Two patients with dural AVMs presented with spontaneous epidural bleeding, which required emergency removal of epidural hematomas. These patients both underwent subsequent elective excision of the AVM (Fig. 1). The other five patients with dural AVMs underwent simple division of the feeding radicular artery and electrocoagulation of the small nidus of the AVM on the dura of the root sleeve (Fig. 2). One patient had a concurrent cerebellar AVM and underwent further excision of the intracranial vascular malformation at another operation (Fig. 1). Subsequent selective spinal angiography showed only one patient with a residual dural AVM, which was later successfully treated by transarterial embolization. All patients with dural AVMs showed great improvement in motor and sensory function in the postoperative course. Residual numbness and paresthesia were noted in two patients. The prognosis of patients with dural AVMs in the present series was generally good (Table 4).

Table 3. Associated conditions of the 13 patients with different types of spinal arteriovenous malformations (AVMs)

<table>
<thead>
<tr>
<th>Associated condition</th>
<th>Dural AVMs (n = 6)</th>
<th>Intradural AVMs (n = 7)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intracranial AVMs</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Hemangioma</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Spinal lipoma</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Venous aneurysm</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Spina bifida</td>
<td>1</td>
<td>0</td>
</tr>
</tbody>
</table>

Although all patients with intradural AVMs underwent surgery to resect the AVM, their prognosis varied among different types of lesions. Among patients with intradural direct AVF (type IV), one underwent direct surgery and the other underwent preoperative embolization followed by surgery (Fig. 3). Both regained normal neurologic function. All three patients with glomus AVMs (type II) had normal neurologic function after several months of follow-up. Two lesions were obliterated totally as shown on postoperative spinal angiography; one patient with a small residual AVM refused further treatment and did not bleed again during 39 months of follow-up (Fig. 4). In patients with juvenile AVMs (type III), the lesions were difficult to treat. They were large, intermingled with normal parenchyma, and shared common blood supply with the spinal cord. Surgery was carried out with the aims of feeder division and electrocauterization on the nidus surface to shrink the nidus. Of the two patients with juvenile AVMs, one patient underwent one operation and the other had two operations 4 months apart. Neither of these two lesions could be obliterated totally; however, both patients showed marked reduction in the flow and size of their AVM on postoperative angiography. One patient had a total recovery of her motor function with mild sensory impairment; the other had persistent hemiparesis and marked spasticity postoperatively. No recurrent hemorrhage was noted during the follow-up period.

Discussion

Since the introduction of selective spinal arteriography in the 1960s, a classification of spinal AVMs based on the vascular anatomy seen during arteriography has been employed to categorize spinal AVMs into four types [3, 4]. Screening studies with MR imaging or myelography can detect or raise the suspicion of a vascular abnormality. Myelography always reveals abnormally engorged draining veins in patients with spinal AVMs. However, non-invasive MR imaging is usually first employed in patients suspected to have spinal AVMs. T1- and T2-weighted MR imaging most often show a serpentine pattern of low signal in the subarachnoid space [6]. Selective spinal arteriography is then used to confirm the diagnosis, to precisely localize the lesion, and to define the vascular anatomy of the malformation and the blood supply of the spinal cord, thus facilitating treatment.

In the present study, we did not perform myelography in any of the 13 patients. MR imaging revealed serpentine signal void lesions inside or outside the
spinal cord in all patients. Selective angiography invariably disclosed feeding vessels and dilated draining veins. Interestingly, angiographic and operative findings were discrepant in two cases because selective angiography failed to demonstrate bilateral feeding vessels from vertebral arteries or thyrocervical trunks. These abnormalities were identified at operation in one patient with intramedullary AVM and another with intradural direct AVF.

Clinical and demographic features of the patients clearly distinguish intradural AVMs from dural AVMs. Rosenblum et al compared dural and intradural AVMs in 81 patients and found several dissimilarities in clinical and radiologic findings in these two subgroups [3]. The age at onset of symptoms in patients with intramedullary AVMs averaged 27 years compared to 49 years for patients with dural AVMs. The most common initial symptom associated with dural AVMs was steadily progressive paresis, whereas hemorrhage was the most common presenting symptom in cases of intramedullary lesions. The nidus of dural AVMs was in the low thoracic or lumbar regions in 96% of cases, and only 15% of these shared a common medullary artery with the spinal cord; most intradural AVMs (84%) were in the cervical or thoracic segments of the spinal cord and all were supplied by medullary arteries.

**Fig. 1.** An 11-year-old boy presented with spontaneous epidural hematoma at C3 to T8 levels. A) and B) Magnetic resonance imaging reveals signal void serpentine lesions in the epidural space anterior to the dural sac (white arrows). C) After removal of the epidural hematoma, angiography shows a dural arteriovenous malformation (AVM) supplied by the left T6 spinal artery with a large draining vein. He also had a cerebellar AVM that was totally excised at another operation.
In the present study, patients in both dural and intradural AVMs groups were younger than in the previous series, yet there was a distinct difference in mean age. Five of the six cases of dural AVMs had a vascular nidus below the mid-thoracic segment, while the nidus in six of the seven intradural AVMs was located in the cervical region.

Dural AVMs of the spine, previously called long dorsal spinal AVMs, typically affect men in the later half of their lives, and are most often located in the low thoracic or lumbar segments, and only rarely in the cervical segment. The dural branch of the intervertebral artery supplies the dural AVM. The small arteriovenous nidus occurs at the dural covering of the dorsal nerve root and is drained by a medullary vein, the solitary venous outflow, which carries blood under high pressure retrograde to the coronal venous plexus. This becomes dilated and elongated as a result of excess blood flow under high pressure. Because there are no valves between the coronal venous plexus and radial veins, venous hypertension is directly transmitted to the spinal cord and causes gradual myelopathy [7]. Dural AVMs presenting with acute cord dysfunction are thought to indicate an advanced stage of venous congestion, leading to ischemia but not bleeding [7].

In the present series, although all patients with dural AVMs presented with paresis, half had gradual onset as a consequence of congestion of the circulation of the spinal cord. In particular, two patients had an epidural hematoma as their first presenting symptom, which has only been reported in one previous series [8]. These two patients had both intradural and extradural venous drainage. Pure spinal epidural AVMs are even rarer [9–11], and were not noted in our series. One patient with dural AVM had the unusual condition of subarachnoid hemorrhage.

Before the use of spinal angiography enabled the delineation of the nidus of the dural AVM, dural AVM was treated by total excision of the vascular malformation including stripping of the tortuously engorged coronal venous plexus, which is now thought to be normally transformed after venous hypertension [12]. However, when the engorged vessels extend between many spinal segments, it is very difficult to excise the AVM completely without extended laminectomy. In addition, extensive removal of these vessels impairs the venous drainage of the spinal cord and increases the chance of cord trauma. Prior to the advent of superselective spinal angiography, we also treated many patients with dural AVMs by stripping the engorged long dorsal vein; however, the result was poor and patients usually deteriorated after surgery. Improved knowledge of the vascular anatomy and pathophysiology of the dural AVM led us to interrupt the arteriovenous shunting at the dural covering of the nerve root. After such procedures, the dilated, arterialized, reddish coronal venous plexus flattened and changed to a blue color.

A higher cure rate for spinal dural AVMs might be obtained by adequate surgery than by embolization.
Niimi et al reported that, despite adequate initial embolization, about 25% of patients underwent re-embolization due to late collateralization or development of new fistulae [14]. However, embolization is safe, avoids the need for surgery, and allows earlier rehabilitation. If embolization fails, surgery can always be performed and the surgical procedure is not complicated by the initial embolization. Furthermore, embolic material may serve as a marker to localize the site of the fistula during operation. If there is a spinal artery arising from the same pedicle as the feeder or if embolization fails, surgical treatment should be performed without delay.

In the present study, owing to the young age of this group of patients and their clinical presentation, degenerative spinal disease was suspected in most patients with dural AVM on their first visit, which is often the case in general practice. This leads to delay in correct diagnosis and poorer surgical results. Therefore, physicians should be alert to the possibility of spinal dural AVMs, which respond well to early surgery.

Patients with intradural AVMs are usually younger than those with dural AVMs, generally less than 40 years old (6 of our 7 patients were less than 24 years old). Intradural AVMs affect males and females with equal frequency. They are more likely to present with an acute onset, and about 50% present with subarachnoid hemorrhage. The increased risk of hemorrhage is assumed to be partly due to the fact that about half of intradural AVMs have associated arterial or venous aneurysms [3, 15].

Intramedullary AVMs can be successfully treated only by permanently occluding the nidus while preserving the blood supply to the spinal cord. Nevertheless, because intramedullary AVMs intermingle within the spinal cord and share a common arterial supply, it is almost impossible to eradicate them either by surgery or by embolization without causing severe neurologic sequelae. If the AVM is located in the dorsal aspect of the cord and fed by posterior spinal arteries only, total excision without damaging the normal cord blood perfusion may be possible. This involves meticulous separation of the AVM from the adjacent cord tissue and careful hemostasis under microsurgery. However, when the malformation lies within the ventral half of the spinal cord supplied by the anterior spinal artery,
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Fig. 4. An 11-year-old boy underwent coagulation of the glomus arteriovenous malformation (AVM) at C2–4, which was supplied by branches from bilateral vertebral arteries. Three years later, the angiogram shows a residual AVM perfused by the anterior spinal artery. To avoid jeopardizing the blood supply of the normal spinal cord, no further operation was performed. No neurologic deterioration was noted within 39 months of follow-up.

surgical excision is contraindicated because of inevitable cord damage. Transarterial embolization may offer an alternative treatment, but late recanalization remains a problem [16–18]. For example, Biondi et al reported 35 cases of intramedullary AVMs treated with particulate materials [16]. After embolization, 20 patients showed clinical improvement. However, spinal angiography revealed re-canalization in 28 patients at 6 months to 15 years after embolization. Thus, intramedullary AVMs may be treated surgically and/or endovascularly, but the outcome is usually disappointing. In the present series, we could only interrupt the feeding arteries and electrocoagulate the engorged glomus to shrink it as much as possible at surgery; total excision of the AVM could not be performed. However, the possibility of further hemorrhage was greatly decreased due to the reduced blood flow achieved by surgery. No recurrent bleeding was noted in this series.

Intradural direct AVFs are very rare vascular malformations of the spinal cord; only sporadic cases have been reported, first in Europe in 1977 [19] and in America in 1986 [20]. The anatomy and pathophysiology of these lesions are different from those of intramedullary AVMs. These extramedullary AVFs involve direct connection between spinal arteries, anterior or posterior, and medullary veins on the anterior or posterior surface of the cord. With the advent of high-resolution and rapid-sequence imaging of digital subtraction arteriography, the nidus of AVFs can be easily demonstrated. These lesions are often accompanied by arterial or venous aneurysms. Surgery or embolization is used to interrupt the abnormal communication between artery and vein. Optimal treatment depends on identification of three further subtypes as follows: small simple fistulae with a long, thin anterior spinal artery and only moderate venous enlargement, in which embolic occlusion is contraindicated because of the danger of occluding the anterior spinal artery, whereas surgery offers a good alternative; fistulae fed by an enlarged anterior spinal artery with a dysplastic venous aneurysm at the site of shunt, in which a combined operation and embolization may be considered; and large multipediced fistulae with a high blood flow and grossly dilated draining veins, in which surgery is impossible, but embolization with detachable balloon can provide effective treatment [21–23].

Both of our patients with intradural direct AVFs belonged to the second subtype. They had markedly dilated medullary arteries from radicular shunting and drained directly to the dorsal veins. There were also venous aneurysms at the T5–6 level in one patient and at the C4–5 level in the other. The neurologic status of the patients returned to normal after microsurgical ligation of the fistula and electrocauterization of the venous aneurysm to completely obliterate the lesion in one and combined embolization and microsurgery in the other. Results were satisfactory in both patients, which is compatible with the previous literature [21–23].

In conclusion, the surgical outcome of spinal AVMs is closely related to their vascular anatomy. For dural AVMs, only surgical interruption of the arteriovenous shunting at the dural sleeve of the nerve is required and good surgical results are often obtained. However, misdiagnosis is frequent due to the similarity of clinical presentations to degenerative diseases. Intradural AVFs have better results from surgery than intradural direct AVMs, which may be treated by reducing blood flow as much as possible, combined endovascular management, and the use of microsurgical techniques to minimize the chance of rebleeding.
References