Spinal arteriovenous malformations: a comparison of dural arteriovenous fistulas and intradural AVM’s in 81 patients

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The medical records and arteriograms of 81 patients with spinal arteriovenous malformations (AVM’s) were reviewed, and the vascular lesions were classified as dural arteriovenous (AV) fistulas or intradural AVM’s. Intradural AVM’s were further classified as intramedullary AVM’s (juvenile and glomus types) and direct AV fistulas, which were extramedullary or intramedullary in location. Dural AV fistulas were defined as being supplied by a dural artery and draining into spinal veins via an AV shunt in the intervertebral foramen. Intramedullary AVM’s were defined as having the AV shunt contained at least partially within the cord or pia and receiving arterial supply by medullary arteries.

Of the 81 patients, 27 (33%) had dural AV fistulas and 54 (67%) had intradural AVM’s. Several dissimilarities in clinical and radiographic findings of the two subgroups were evident. The patients with intramedullary AVM’s were younger; the age at onset of symptoms averaged 27 years compared to 49 years for dural AV fistulas. The most common initial symptom associated with dural AV fistulas was steadily progressive paresis, whereas hemorrhage was the most common presenting symptom in cases of intramedullary lesions. No patients with dural AV fistulas had subarachnoid hemorrhage. Activity exacerbated symptoms more frequently in patients with dural lesions. Associated vascular anomalies occurred only in cases of intradural AVM’s. In 96% of the dural lesions the AV nidus was in the low thoracic or lumbar region; in only 15% did the intercostal or lumbar arteries supplying the AVM also provide a medullary artery which supplied the spinal cord. In contrast, most intradural AVM’s (84%) were in the cervical or thoracic segments of the spinal cord and all of them were supplied by medullary arteries. Transit of contrast medium through the intradural AVM’s was rapid in 80% of cases, suggesting high-flow lesions. Forty-four percent of the patients with AVM’s of the spinal cord had associated saccular arterial or venous spinal aneurysms. No dural AV fistulas displayed these characteristics. A good outcome occurred in 88% of patients with dural AV fistulas after nidus obliteration, while 49% of patients with intramedullary AVM’s did well after surgery or embolization.

These findings suggest that dural and intradural AVM’s differ in etiology (acquired vs. congenital) and that they have different pathophysiology, radiographic findings, clinical presentation, and response to treatment.

KEY WORDS • spinal arteriovenous malformation • dural arteriovenous fistula • intradural arteriovenous malformation

Although spinal arteriovenous malformations (AVM’s) are relatively rare, they are important clinical entities because they produce considerable morbidity. Early classification of spinal AVM’s relied upon descriptive pathological analyses.Later classification was based on the radiographic appearance at selective spinal angiography, although the location of the nidus of dural spinal AVM’s was not immediately recognized. Kendall and Logue in 1977 first distinguished two arteriographically distinct types of AVM of the spinal cord: dural and intradural. Clinical descriptions of intradural AVM’s have been reported previously. Although the anatomy, pathology, pathophysiology, and response to surgery or embolic occlusion of the dural lesions were emphasized in the original paper by Kendall and Logue, as well as in subsequent reports, there has been no study comparing the features of dural
arteriovenous (AV) fistulas with those of intradural spinal AVM's.

We reviewed the case histories and arteriograms of 81 patients with spinal AVM's. The purposes of this study were: 1) to examine clinical and radiographic differences that may suggest a congenital versus an acquired etiology of dural AV fistulas and intradural AVM's; 2) to correlate the anatomical and blood flow patterns seen radiographically with the postulated pathophysiology of the different types of spinal AVM's; and 3) to compare the clinical features and the response to treatment of the different types of spinal AVM's. Our results indicate that dural AV fistulas and intradural AVM's differ in etiology, clinical presentation, mechanism of cord injury, and response to treatment.

Clinical Material and Methods

The clinical records and radiological studies of 81 patients with spinal AVM's who were admitted to the Clinical Center, National Institutes of Health (NIH), between 1964 and 1985 were reviewed. Only patients who underwent spinal arteriography, and for whom complete records and arteriograms were available, were included. Selective spinal arteriography has been performed since 1964. These data were analyzed with regard to epidemiology, clinical presentation, vascular anatomy, and flow patterns, based on arteriography, therapy, and outcome.

Dural AV fistulas are defined as spinal vascular lesions in which the vascular nidus of the AV shunt is embedded in the dura covering the proximal nerve root and in the adjacent spinal dura (Figs. 1 and 2). Dural branches of the intercostal or lumbar arteries supply these AVM's. Intradural AVM's are defined as spinal vascular lesions in which the nidus of the AVM is in the cord or pia and which, together with the spinal cord, receive their blood supply from medullary arteries. Intradural AVM's are subclassified into intramedullary AVM's, in which the nidus of the AVM is within the parenchyma of the spinal cord or pia, and direct AV fistulas, which are intramedullary or paramedullary in location, and in which the transition from artery to vein occurs without an intervening glomus of abnormal vessels (Figs. 3 to 5). The intramedullary AVM's are further subclassified as glomus AVM's, composed of a tightly packed localized nidus of abnormal blood vessels within the spinal cord, and juvenile AVM's, which occupy the entire spinal canal at the involved level and which are supplied by numerous, frequently large, feeding arteries.
Dural and intradural spinal AVM’s

Results

Demographic Factors

Of the 81 patients, 27 (33%) had dural AV fistulas and 54 (67%) had intradural AVM’s. The 54 patients with intradural AVM’s were subclassified as having intramedullary AVM’s (43 patients) or direct AV fistulas (11 patients). The intramedullary AVM’s were further divided into glomus (14 patients) and juvenile AVM’s (29 patients).

There was male predominance associated with both dural and intradural lesions. Men comprised 85% of dural and 70% of intradural AVM’s. The age of patients with symptoms from dural AV fistulas ranged from 22 to 72 years (49 ± 11.4 years, mean ± standard deviation) (Fig. 6). Only one patient was under 25 years of age. In contrast, patients with symptoms from AVM’s of the spinal cord were younger, aged 4 to 58 years (27 ± 12.1 years). Sixty-five percent were younger than 25 years of age (Fig. 6).

Clinical Features

Presenting symptoms caused by dural and intradural AVM’s included paresis, sensory abnormalities, sphincter disturbances, and pain (Table 1). The initial symptom in patients with dural AV fistulas was most frequently paresis, which occurred in 12 (44%) of the 27 patients. There was a striking difference in the incidence of subarachnoid hemorrhage (SAH) between the two major types of AVM. Subarachnoid hemorrhage was responsible for the initial symptoms in 17 (31%) patients with intradural AVM’s (Table 1). It occurred
TABLE 1

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Initial Symptoms</th>
<th>Symptoms at Diagnosis†</th>
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<tr>
<td></td>
<td>Dural AVM's</td>
<td>Intradural AVM's</td>
</tr>
<tr>
<td></td>
<td>Intradural AVM's</td>
<td>Dural AVM's</td>
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<tr>
<td></td>
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<td>total cases</td>
<td>27</td>
<td>54</td>
</tr>
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</table>

* AVM = arteriovenous malformation.
† The values listed indicate the number of patients with these symptoms when the diagnosis of spinal AVM was established. Most patients had several of the symptoms.

Fig. 6. Onset of symptoms in patients with intramedullary arteriovenous malformations (AVM's) was usually in childhood and early adulthood. Dural AVM's presented predominantly in and after middle age.

most commonly with glomus AVM's (10 of 14 patients), while weakness was the most common initial complaint in patients with juvenile-type AVM's and direct AV fistulas of the spinal cord. By the time a diagnosis of spinal AVM was established, the majority of patients with dural AV fistulas and intradural AVM's had motor and sensory deficits (Tables 1 and 2). In both groups of patients, spastic paresis of the lower extremities (Table 3) and loss of the sensations of pain and temperature were the most common patterns. A distinct sensory level was present in most patients; the level generally reflected the location of the vascular nidus of the AVM along the spinal axis. Functionally, the majority of patients were in a good category (Aminoff and Logue Grade 1 or 2, Table 4).

No dural AV fistula caused SAH, whereas by the time the diagnosis of spinal AVM was established 52% of the patients with intradural AVM's had experienced SAH (80% of which were confirmed by lumbar puncture) with one to five hemorrhages per patient (mean of two) (Table 1). Most patients with glomus lesions suffered SAH (12 of 14 patients), while this complication was less common with juvenile-type AVM's (13 of 29 patients) and direct AV fistulas (three of 11 patients). A spinal bruit was heard in three patients. All had intradural AVM's with rapid flow. Most patients with dural and intradural lesions experienced a gradual onset and progressive deterioration of neurological function (Table 5). An acute onset of initial symptoms occurred in half of the 54 patients with intradural AVM's. This was even more frequent in the subgroup of patients with glomus lesions (11 of 14 cases), and reflected the higher incidence of SAH in these patients (Table 5). Bending, standing, or activity elicited exacerbation of symptoms in a few patients with intradural AVM's; activity exacerbated symptoms in 19 of the 27 patients with dural AV fistulas (Table 6). The neurological condition of one patient with a dural AV fistula and three with intradural AVM's deteriorated during pregnancy (Table 6).

Radiographic Anatomy and Pattern of Blood Flow

Eight patients with intradural AVM's (15%) showed
Dural and intradural spinal AVM’s

TABLE 4
Preoperative function and outcome after surgery*

<table>
<thead>
<tr>
<th>Preoperative Functional Grade</th>
<th>Postoperative Functional Grade</th>
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<tr>
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<td>Dural AVM’s</td>
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<td></td>
<td>1 2 3 4 5</td>
</tr>
<tr>
<td>motor function†</td>
<td>1 0 0 0 0</td>
</tr>
<tr>
<td>functional grade‡</td>
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</table>

* AVM = arteriovenous malformation. Surgery was performed on 26 patients with dural AVM and 43 with intramedullary AVM.
† For a description of grades of motor function see Table 2.
‡ Functional grades: Grade 1 = disturbance of gait, leg weakness, or abnormal stance, no restriction of activity; Grade 2 = restricted activity; Grade 3 = requires a cane for walking; Grade 4 = requires crutches for walking; Grade 5 = unable to stand and confined to bed or wheelchair. Grading scales adapted from Aminoff and Logue.

TABLE 5
Course of illness in each patient group*

<table>
<thead>
<tr>
<th>Course of Illness</th>
<th>Dural AVM’s</th>
<th>Intramedullary AVM’s</th>
</tr>
</thead>
<tbody>
<tr>
<td>gradual</td>
<td>23</td>
<td>27</td>
</tr>
<tr>
<td>acute</td>
<td>4</td>
<td>27</td>
</tr>
<tr>
<td>total cases</td>
<td>27</td>
<td>54</td>
</tr>
</tbody>
</table>

* AVM = arteriovenous malformation.

increased interpedicular distance on spine x-ray films. Six of these eight patients had high-flow lesions (disappearance of arterial flow within 6 seconds during arteriography). There was no instance of a widened spinal canal in patients with dural AV fistulas. Three patients with dural AV fistulas and one patient with an intramedullary AVM were reported to have normal myelograms. None of the patients who underwent myelography at the NIH had a normal study. Associated vascular anomalies at other sites (that is, additional vascular malformations and extraspinal aneurysms) occurred in 19% of the patients with intradural malformations and in none of the patients with dural AV fistulas (Table 7).

Dural AV fistulas differed from intradural AVM’s with respect to the level of involvement along the longitudinal spinal axis, arterial supply, rate of blood flow, site of AV nidus, and route of venous drainage of the AVM. The feeding vessels to the dural AV fistulas originated in the lower thoracic or the lumbar region in 26 of 27 patients and in the sacral region in one patient (Fig. 7). Only three of the 27 dural AV fistulas had more than one feeding artery (maximum of two). In four instances the feeding arteries shared a common origin with the artery of Adamkiewicz or a posterolateral spinal artery. Rapid blood flow (angiographic arterial phase lasting 6 seconds or less) did not occur with dural AV fistulas, and no patient with this type of lesion had an associated arterial or venous aneurysm. The vascular nidus was lateral to the spinal cord in all cases of dural AV fistulas. The direction of predominant venous drainage of most dural AV fistulas was rostral. The dilated tortuous veins were always evident on the dorsal cord surface, but they were also present anterior to the cord in 15% of cases. Epidural and paravertebral outflow from the dural AV fistula was visualized in only one patient.

Intradural AVM’s occurred more diffusely along the longitudinal spinal axis than dural AV fistulas (Fig. 7). The AV nidus occurred within the spinal cord in 80% of cases, on the dorsal surface of the cord in 9%, and on the ventral surface in 11%. All cervical AVM’s, except one juvenile malformation, were intramedullary glomus-type AVM’s. Multiple feeding arteries were recognized in 72% (range two to six feeding vessels), and the flow was rapid in 80% of the intradural AVM’s. Associated arterial or venous aneurysms of the vessels that supplied or drained the spinal AVM were demonstrated arteriographically in 24 of the 54 patients with intradural AVM’s (17 patients had single aneurysms, three patients had two, four patients had more than two). Most patients (54%) with intradural AVM’s and associated spinal aneurysms experienced SAH, but this complication also occurred in 50% of patients.

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AV fistulas and 43 (80%) of the 54 patients with intradural AVM's that had been treated.

Treatment and Prognosis

Treatment was delayed longer after the onset of symptoms in patients with intradural AVM's than in patients with dural lesions; the duration from onset of symptoms to initial treatment averaged 2.7 years (range 3 months to 9 years) with dural AV fistulas and 4.2 years (range 1 week to 21 years) with intradural AVM's. Therapy was undertaken on eight of the 27 dural AV fistulas and on 24 of the 54 intradural AVM's prior to initial treatment averaged 2.7 years (range 3 months to 9 years) with dural AV fistulas and 4.2 years (range 1 week to 21 years) with intradural AVM's.

At the NIH, 26 (96%) of the 27 patients with dural AV fistulas and 43 (80%) of the 54 patients with intradural AVM's underwent surgery. The presence or degree of preoperative sensory loss and the rate of progression of neurological deficits did not correlate with therapeutic outcome with dural AV fistulas or intradural AVM's. Similarly, age at onset of symptoms did not correlate with outcome. There was, however, a direct relationship between preoperative strength and function and postoperative strength and function for both dural and intradural AVM's (Table 4).

Of the 26 patients with surgery for dural AV fistulas, 19 (72%) improved postoperatively and seven (28%) remained stable. Following surgery on the 43 operative patients with intramedullary AVM's, the neurological status was unchanged in 22 (51%), improved in 14 (33%), and worsened in six (14%). One patient died during a circulatory bypass procedure after excision of an AVM and an anterior spinal artery aneurysm.

Following surgery, 11 (42%) of the 26 patients with dural AV fistulas were in Aminoff and Logue's functional Grade 1 (follow-up period 1 month to 17 years; mean 3.7 years), as were 10 (24%) of the 43 patients with intramedullary AVM's (follow-up period 1 month to 3.9 years; mean 3.1 years). All but one of the patients with dural AV fistulas and 32 of the 43 surgical patients with intramedullary AVM's received postoperative arteriography. None of the postoperative angiograms of dural AV fistulas demonstrated residual evidence of abnormality, whereas persistent AVM was seen in 13 of the 32 patients with intramedullary AVM's.

Discussion

The two major types of spinal AVM's are distinguished by the location of the vascular nidus, whether dural or intradural. Intradural AVM's are further classified into three distinct subtypes; juvenile AVM's and glomus AVM's, in which the nidus lies completely or partially within the cord parenchyma, and direct AV fistulas, which may lie in the cord tissue or on the cord surface, and in which the transition from artery to vein occurs without an intervening glomus of abnormal vessels. Several studies indicate that the incidence of dural AV fistulas is higher than that of intradural AVM's. In the period reported here, patients often underwent surgical stripping of the dilated veins on the posterior cord surface at laminectomy and consequently were not referred. It is likely that a referral bias in favor of intradural AVM's accounts for the relative incidence of dural and intradural AVM's in this series.

The nidus of dural AV fistulas is imbedded in the dura covering the proximal nerve root and in the adjacent spinal dura (Fig. 2). Arteriography demonstrates an AV nidus of fine vessels in the lateral aspect of the spinal canal and within an intervertebral foramen. Contrast medium flows through the fistula and intradurally, into a tightly coiled, continuous vessel on the cord surface (Fig. 2). It is the dural branch of the spinal ramus of the intercostal or lumbar artery that supplies the AV fistula (Fig. 2). Blood flowing through the fistula is carried through the medullary vein in a retrograde...
Dural and intradural spinal AVM’s

manner to the coronal venous plexus, which becomes dilated, tortuous, and elongated. The absence of valves between the coronal venous plexus and the radial veins facilitates the transmission of high venous pressure to the cord tissue, which causes congestive myelopathy.

In the intramedullary AVM’s of the spinal cord the site of the AV fistula is within the parenchyma of the spinal cord (Figs. 3 and 4). One of the feeding vessels of these AVM’s is almost always an enlarged medullary artery that also supplies the spinal cord via the anterior spinal artery. Most juvenile-type spinal AVM’s fill a segment of the spinal canal and contain cord tissue within the interstices of the AVM (Fig. 4). The glomus AVM is a localized congeries of smaller blood vessels confined to a short segment of the spinal cord and is usually fed by a single feeding vessel, almost always a medullary artery (Fig. 3).

Distinguishing clinical features between dural AV fistulas and intradural spinal AVM’s may help in the differential diagnosis. Our findings indicate that intradural AVM’s should be suspected when a patient is less than 30 years old, with an acute onset of symptoms and the presence of SAH or a spinal bruit, and if the symptoms affect the arms. In contrast, patients with dural AVM’s are usually older than 40 years of age, have a gradual onset and progressive worsening of symptoms, and experience exacerbation of symptoms by activity. The dural lesions are always in the lower half of the spinal column and produce symptoms that affect the legs, but not the arms.

Our findings support an acquired etiology for dural AV fistulas and a congenital origin for intramedullary spinal AVM’s. Dural AV fistulas first become symptomatic in later adult life, whereas medullary AVM’s affect children and young adults. Associated congenital vascular malformations are found only with intradural AVM’s. If spinal AVM’s were congenital malformations of vascular origin, a uniform distribution along the longitudinal axis of the spine would be expected. Such a distribution was demonstrated in the patients with medullary AVM’s, but not in the patients with dural AV fistulas. The abnormalities in the latter group were all located in the lower half of the spine, a distribution that is compatible with an acquired etiology dependent on an upright posture. An embryonic origin of an AV fistula would likely have an associated increased development of the regional draining veins to accommodate the excess blood flow. The intramedullary AVM’s were drained by veins flowing both rostrally and caudally. However, with dural lesions, inferiorly directed venous drainage rarely occurred; almost all patients had superiorly directed venous efflux (that is, a drainage against a greater hydrostatic pressure in the upright position than caudal drainage). In addition, although dural AV fistulas were restricted to the lower half of the spine, there was frequent rostral venous filling of the distended coronal venous plexus all the way to the cranium. Thus, in dural AVM’s venous drainage via normal medullary veins appeared to be deficient. This indicates that diminished, and not increased, venous drainage might accompany, or be associated with the etiology of these lesions, as has been suggested by Merland, et al.19 Intramedullary AVM’s are intrinsic vascular anomalies of the spinal cord, fed by medullary arteries and drained by the coronal venous plexus, often via the medullary veins into the paravertebral venous systems. These observations suggest that dural AV fistulas are probably acquired while medullary AVM’s are congenital in nature.

The pathophysiology of the neurological deterioration may well be different in patients with dural and those with intradural spinal AVM’s, with three mechanisms (venous hypertension, arterial steal, and SAH) playing different roles in the two groups of lesions. Based on pathological examination of the spinal cord, the neurological deterioration of patients with dural AV fistulas has been attributed to increased venous pressure.1 Venous hypertension arises when arterial blood passes through the dural AV fistula into a medullary vein (the solitary venous outflow of the dural AV fistula) and reaches the valveless coronal venous plexus and the radial veins (Fig. 2). Some angiographic features of dural AV fistulas, such as sluggish retrograde drainage of the fistula via enlarged medullary veins, indicate that vascular steal is unlikely and support the relative importance of venous hypertension as the pathogenesis of the neurological findings in patients with dural AV fistulas. Furthermore, the medullary arterial supply for the spinal cord and the vessel feeding the dural AV fistula arose from a common vessel in only four of our 27 patients. Since the aorta was the only vessel that contributed blood to the supply of the dural AV fistula and to the spinal cord, and since the rate of blood flow is so slow through the dural AV nidus, vascular steal cannot be the cause of myelopathy in these patients.

On the other hand, in patients with intradural AVM’s, rapid blood flow appears to be the dominant pathogenetic mechanism. Compatible with a high-flow lesion, these spinal AVM’s are associated with interpedicular widening, the acute onset of symptoms associated with SAH, a spinal bruit, multiple feeding vessels, and the presence of spinal aneurysms of the feeding or draining vessels. Venous and arterial aneurysms were seen only with intradural AVM’s and are probably caused by high-pressure, high-volume, turbulent blood flow through the intradural nidus and its feeding and draining channels. The combination of high flow through the AVM and the fact that the feeding vessels of the AVM are always medullary arteries creates the physiological and anatomical basis for arterial steal and the ensuing myelopathy. Sufficient quantities of blood may be diverted away from cord parenchyma and into the AV shunt flow to cause ischemia. As in spinal dural AV fistulas, the distended draining veins of intramedullary AVM’s may also cause ischemia by increased venous pressure and consequent reduction in tissue perfusion pressure. However, the presence of bidirectional flow in the venous drainage of these lesions and
access to the extradural and paravertebral veins suggest that the venous drainage of the excess blood flow is often adequate.

These results suggest that dural AV fistulas are consistently amenable to treatment, whereas some patients with AVM's of the spinal cord have lesions that cannot be completely excised without unacceptable neurological risks. Postoperative angiography uniformly indicated complete obliteration of the AV fistula in patients with dural lesions, whereas in many patients with intradural AVM's arteriography showed residual portions of the AVM glomus. Recent advances in embolotherapy suggest that intradural AVM's, both intra- and extra-medullary, may be effectively palliated by microparticulate (polyvinyl alcohol, 150 to 500 µm) embolization of glomus lesions and by occlusion of the feeding vessels to the intradural AV fistulous types.6'9'11'13'15'22 However, the permanence of this therapy has not yet been demonstrated.

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References


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