Spinal arteriovenous malformations: new classification and surgical treatment

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Object. Spinal vascular malformations represent rare and insufficiently studied pathological entities characterized by considerable variation. Insufficient study of this disease is connected with the complexity of its diagnosis, which restricts the development of surgical treatments that are differentiated according to the type of malformation. Great difficulties are caused by the lack of a clear structural–hemodynamic classification of spinal arteriovenous malformations (AVMs). At present the classification created between 1991 and 1998 by the combined efforts of different authors is the most widely used one. According to this classification, four categories are distinguishable: Type I, dural arteriovenous fistulas (AVFs); Type II, intramedullary glomus AVMs; Type III, juvenile or combined AVMs; and Type IV, intradural perimedullary AVFs. Vascular tumors are also classified, as follows: hemangiomas, hemangioblastomas, angiosarcomas, hemangiopericytomas, angiofibromas, angiolipomas, and hemangioendotheliomas, as well as cavernous malformations.

Methods. In this study the authors analyze the diagnostic data and results of treatment in 91 patients with AVMs and AVFs who were treated at the Institute of Neurosurgery between 1995 and 2005. The patients’ ages ranged from 9 to 83 years; the mean age was 42.9 years. For spinal vascular malformations we devised a classification that took into account the aforementioned features of AVMs: the anatomical characteristics of a malformation and its angiostructural and hemodynamic features. In all patients the neuroimaging modalities used in the investigation of their lesions included magnetic resonance (MR) imaging and selective spinal angiography. Three-dimensional computerized tomography angiography studies were obtained in 14 patients, and MR angiography was used in 17.

Conclusions. For successful surgical treatment of spinal AVMs it is necessary to obtain data about their localization, vascular structure, and hemodynamics that are as complete as possible. This information will promote the use of optimum surgical procedures and the latest methods of microsurgical and endovascular interventions, with treatments differentiated according to the type of malformation. One should try to use the least invasive endovascular approach in these cases, where possible, to occlude the AVM or reduce the intensity of blood flow by means of embolization. To perform an AVM resection or occlusion, one should use a direct approach to the malformation, blocking only the vessels supplying blood to the malformation and preserving the vessels feeding the spinal cord.

KEY WORDS • spinal arteriovenous malformation • structural–hemodynamic classification • surgical treatment

S P I N A L vascular malformations represent a rare and insufficiently studied pathological entity characterized by considerable variation.7,5,7,12,14,20,22 These lesions have been insufficiently studied because of the complexity of their diagnosis, which restricts the development of surgical treatments that are differentiated according to the type of malformation.9,13 Great difficulties are caused by lack of a clear structural–hemodynamic classification of spinal AVM. At present, the classification created between 1991 and 1998 by the combined efforts of different authors is the one most widely used. According to this classification, the following four categories are distinguishable: Type I, dural AVFs; Type II, intramedullary glomus AVMs; Type III, juvenile or combined AVMs; and Type IV, intradural perimedullary AVFs.23 Vascular tumors are also identified: hemangiomas, hemangioblastomas, angiosarcomas, hemangiopericytomas, angiofibromas, angiolipomas, and hemangioendotheliomas, as are cavernous malformations.

In 2002 Spetzler, et al.,19 offered a new classification of spinal vascular pathological entities. Nevertheless, this classification does not cover the whole spectrum of spinal malformation types. Therefore, we consider a detailed systematization of spinal vascular malformations to be necessary; such a scheme can facilitate the development of tactics for surgical interventions that are differentiated with regard to the localization, angiostructural type, and hemodynamic peculiarities of malformations and will allow optimization of the results of treatment.

Clinical Material and Methods

In this study we analyzed the diagnostic data and results of treatment in 91 patients with AVMs and AVFs who were treated at the Institute of Neurosurgery between
1995 and 2005. The patients’ ages ranged from 9 to 83 years; the mean age was 42.9 years. For spinal vascular malformations we devised a classification that took into account the aforementioned features of malformations: anatomical characteristics of a lesion and its angiostuctural and hemodynamic features (Table 1). Based on their anatomical features, AVMs are divided into intramedullary, perimedullary, dural, epidural, intraventricular, and combined (covering some adjacent areas). The feeding and draining vessels were designated because many malformations had essential differences depending on the supply vessels. Aneurysms of spinal vessels were observed in our studies only in combination with AVMs. Hemodynamic differences and differences in vascular structure in many cases required different surgical tactics. We did not include spinal vascular tumors and cavernous malformations in this study because they represent separate pathological entities.

In all patients the evaluations included MR imaging and selective spinal angiography. Three-dimensional computed tomography angiography studies were obtained in 14 patients and MR angiography in 17.

Surgical intervention was performed in all 91 patients: in 13 of them endovascular interventions were applied, in 70 we performed microsurgery, and in eight we performed combined operations with application of endovascular and microsurgical techniques. During open operations, dorsal and dorsolateral approaches were chiefly used. Ventral and ventrolateral approaches were used in eight patients, and in 11 patients operative interventions were completed by stabilization of the spine.

Statistical analysis (digital neurological scales, t-criterion, and comparison of these data before and after the operation) was conducted using the program Microsoft Excel 2002 and its statistics supplement Analysis ToolPak. Correlative, cluster, and linear regression analyses were performed using the STATISTICA program (version 6; StatSoft, Inc., Tulsa, OK).

Results

Types of Malformations

Intramedullary AVMs. For intramedullary AVMs of all kinds, clustering zones of low-intensity MR signals in the spinal cord were typical. The spinal cord seemed expanded in the region of the malformation; sometimes expanded perimedullary draining veins were discovered around the cord on MR images. Changes of the spinal cord density were observed around the vascular nidus. The size and density of arrangement of the malformation’s vessels varied essentially. The vessels were densely packed in glomus AVMs and scattered in the spinal cord in diffuse AVMs in which the spinal cord matter was observed among the vessels. The selective spinal angiography studies revealed a vascular conglomerate consisting of vessels that either adjoined each other tightly (glomus type) or were scattered in the spinal cord matter (diffuse type). Direct AVM feeding vessels passed from the ventral or dorsal spinal arteries, and sometimes from the radiculopinal arteries. The AVM was drained by the perimedullary veins. According to the MR imaging and surgical findings, intramedullary AVMs were limited by spinal cord or the conglomeration of vessels spread on the surface of the spinal cord (Figs. 1–6).

Intradural or Perimedullary AVMs. These lesions were visualized on MR imaging as conglomerations of vessels in the form of low-intensity zones around the spinal cord on MR images. These vessels could be localized on the ventral as well as on the dorsal or lateral spinal cord surface. The spinal cord was not expanded; in most cases its compression and displacement by the malformation was observed. Spinal cord edema was rare with this type of malformation. The selective spinal angiography studies demonstrated feeding vessels from the ventral or dorsal radiculomедullary arteries. Comparing the MR imaging and selective spinal angiography data, we often found thrombosis in AVM vessels. These vessels drained into the ventral or dorsal perimedullary veins (Figs. 7 and 8).

Intradural or Perimedullary AVFs. In perimedullary AVFs, MR imaging demonstrated a serpentine pattern of vessels around the spinal cord, although in contrast to AVMs, there were no vascular conglomerates, signs of compression, or displacement of the spinal cord. In the patients with AVF in whom insignificant blood shunting occurred, the perimedullary veins on the spinal cord surface were hardly noticeable. As distinct from the perimedullary AVMs, spinal cord edema was marked in the AVFs. The typical finding on selective spinal angiography was an AVF in the form of a direct passage of the feeding vessel (the ventral or dorsal spinal artery) into the draining ventral or dorsal perimedullary veins.

As a rule, with ventral feeding vessels, an AVF drained into ventral perimedullary veins and vice versa, whereas with dorsal feeding vessels an AVF drained into the dorsal perimedullary veins. The draining veins were twisted and could be traced for a long distance along the spinal cord. In AVFs, the feeding vessels more often were the ventral or the dorsal spinal arteries, whereas in AVMs they were the ventral or dorsal radiculomедullary arteries. In vascular malformations arising in the conus medullaris, the feeding vessels coursed with the cauda equina roots. Based on the amount of blood shunting nd the size of the lesion, perimedullary AVFs and AVMs were divided into lesions with the following characteristics: A) insignificant blood shunting; B) moderate blood shunting; or C) considerable blood shunting (Figs. 9–13).

Dural AVMs and AVFs With Retrograde Drainage Into Perimedullary Veins. In dural AVMs and AVFs with retrograde drainage into perimedullary veins, expansion of these veins was found on MR imaging studies. Dural AVMs or AVFs of the thoracic region were more often drained into the dorsal perimedullary veins. Rare dural AVFs of the cervical spine could be drained into the ventral perimedullary veins. In patients with an insignificant amount of blood shunting, the perimedullary vein expansion looked like serpiginous flow voids on the dorsal surface of the spinal cord, most often in the middle and lower thoracic spine. In case of moderate blood shunting, expanded veins were found on the T₁- and T₂-weighted sequences in the form of a serpentine vascular pattern in the subarachnoid space. In all cases, vast spinal cord edema and thickening were typical. Angiographic studies revealed expanded radiculomедullary arteries, which directly (in AVFs) or through a vascular conglomerate in
the region of the intervertebral neural foramen (in AVMs), shunted into the expanded perimedullary veins. Most often, a malformation had only one tributary and was characterized by slow blood flow; several tributaries were rare. The blood flow in spinal cord arteries was also slower. It was especially noticeable when contrast material was added in the artery of Adamkiewicz. This occurred because spinal as well as cerebral dural malformations are found against a background of venous hypertension, which is considered the initial cause of dural malformations (Figs. 14 and 15).

If a dural AVM or AVF had antegrade epidural drainage, on MR images there were prominent venous pouches in the epidural space. These pouches had a round or ellipsoid form and stretched along the one to three vertebrae. The characteristic features of such lesions were insult-like ischemic changes of the spinal cord that appeared as an area of low MR signal intensity in the $T_1$-weighted and high intensity in the $T_2$-weighted sequences, and as an edema around the cord. Enlarged perimedullary vessels are not typical. Expanded radiculomeningeal arteriovenous malformations (AVMs) and arteriovenous fistulas (AVFs) were discovered on angiographic studies; these arteriovenous communications were shunted directly (in case of AVF) or through a vascular conglomerate into the expanded epidural veins, which drained through the veins passing through the intervertebral foramina.

**Epidural AVMs and AVFs.** These lesions were characterized by low-intensity-signal zones located epidurally on MR imaging, and had caused a compression of the dura mater. Selective spinal angiography obtained with contrast agents demonstrated the feeding vessels spreading out directly from the spinal branch or from the postcentral, prelaminal branches. The vascular conglomerate of the AVM or AVF was not large; it consisted of small vessels. In contrast to dural AVFs, these lesions were located, not in the region of the intervertebral foramen and dural sleeve of the nerve root, but in the epidural space. The lesions were drained rostrally or caudally through the epidural veins. Sometimes the drainage through the intervertebral veins at the level of the malformation passed to paravertebral veins (Fig. 16).
Intravertebral AVMs. According to the MR imaging data, intravertebral AVMs were discovered in the form of large vessels with intense blood flow, which were situated more often inside the vertebrae or with paravertebral spreading from these structures. Expanded epidural or paravertebral veins draining these AVMs were visible on MR images. Selective spinal angiography was used to identify AVM feeding vessels from the ventrolateral branches of segmental arteries (postcentral and prelaminar branches). The AVMs were drained through the epidural or the paravertebral veins into the ascending lumbar veins, the inferior vena cava, and the azygos and hemiazygos veins (Fig. 17).

Fig. 1. Sagittal (A) and axial frontal (B) MR images demonstrating an intramedullary diffuse AVM at the C-5 level. The open arrows designate the intramedullary AVM nidus.

Fig. 2. Vertebral angiograms demonstrating an intramedullary diffuse AVM at the C-5 level. A and B: Preoperative images, frontal and lateral projections. C and D: Angiograms obtained in frontal and lateral projections after the AVM resection. 1 = feeding vessels; 2 = the AVM nidus.

Fig. 3. Intraoperative photographs showing an intramedullary diffuse AVM at the C-5 level. A: A conglomeration of perimedullary draining veins is seen. B: The veins have been coagulated, and myelotomy has been performed in the root entry zone in front of the posterior roots. The roots have been moved aside dorsally with a rubber retractor. The vessels of the nidus within the spinal cord have been coagulated and cut off.
Combined Malformations. Combined lesions were situated in several adjacent anatomical structures. If combined glomus AVMs were located mainly intradurally, they received tributaries chiefly from the radiculomedullary arteries and had perimedullary venous drainage. In case of primary extradural localization, the combined glomera of the AVMs received tributaries from the spinal branches and were drained mainly by the epidural and paravertebral veins. However, these AVMs often had equally intradural and extradural locations (Figs. 18 and 19).

Surgical Interventions

Based on the MR imaging and angiography data we collected, the operative intervention was planned. The choice of surgical tactic depended on the localization, vascular structure, the ways of inflow and outflow, and hemodynamic peculiarities of the lesions. As a rule, in AVMs the feeding and draining vessels needed to be occluded and a resection of the malformation was necessary. In AVFs it was enough to occlude only the feeding vessels.

The indications for occluding only the feeding vessels were as follows: an intramedullary diffuse AVM (because the diffuse character of the AVM could lead to an extensive surgical injury to the spinal cord during an attempt to perform nidus resection); a perimedullary AVF; an intravertebral AVM; a combined AVM; or a dural AVF or AVM. In some cases of dural malformation it was optimal to occlude the draining vessels only.

The occlusion of the feeding and draining vessels and malformation resection were necessary in the following cases: intramedullary glomus AVM; perimedullary AVM; epidural AVM; and combined AVM.

The type of surgical intervention was determined in accordance with the chosen surgical tactics. Endovascular embolization was performed when there were indications...
for occlusion of only the feeding vessels. Embolization of the main vessels is possible only if they do not take part in the spinal cord blood supply, but provide blood to the vascular malformation only. In cases in which the main segmental vessel supplies the vascular malformation but takes part in supplying the spinal cord blood, only superselective obliteration of the malformation’s direct tributaries is possible.

Open microsurgery is necessary for resection of an AVM nidus. Open surgical interventions are advisable also when it is necessary to occlude only the feeding vessels, but one has failed to embolize them endovascularly, or if embolization of the main tributaries threatens to occlude the arteries feeding the spinal cord. Microsurgical interventions are also advisable if a vascular formation has the kind of structure in which there is a collateral blood flow and embolization of the main tributaries will not result in complete occlusion of the blood flow; or if selective spinal angiography does not identify all of the tributaries or there are suspiciously few of them, and their diameter is too small in comparison with the presumable one, according to the MR imaging data.

Combining surgical intervention with endovascular embolization that is performed before the microsurgical procedure is advisable if it is necessary to resect an AVM that has a high flow and numerous large feeding vessels running into it, or if after the endovascular embolization a mass effect due to AVM blood flow remains.

For endovascular embolization, two variations of the conventional technique were applied: 1) superselective obliteration of the feeding vessels; and 2) obliteration of the main tributary, usually the segmental artery. The first method was applied in cases in which either the main segmental vessel supplied blood simultaneously to the malformation and the radiculomedullary arteries or the malformation’s blood was supplied directly from the anterior or dorsal spinal arteries. If these arteries ended in the AVM or the AVF, it was possible to apply selective embolization. If these arteries extended branches to the AVM or the AVF and continued farther, feeding the spinal cord, we...
avoided embolization. The second method of embolization, a nonselective technique of obliteration of the segmental main tributary, was applied if this artery did not feed the spinal cord (Table 2).

If there were several tributaries, we performed an endovascular obliteration of all arteries during one operation. Later it became obvious that such a method led to neurological deterioration. That is why we now apply the method of staged obliteration of tributaries (no more than one per operation) with a few days between each stage.

Surgical approaches in cases of intramedullary AVM depended on the type of feeding vessels and the primary location of the nidus. In lesions with feeding vessels from the anterior spinal artery and the malformation’s location in the ventral regions of the spinal cord or its exophytic spreading, we use ventral approaches; a ventral paratrabecual cervical, transthoracic, or costotransversectomy approach in the thoracic spine. These approaches provide a direct route to the feeding vessels and the nidus, and they require no spinal cord traction.

In cases of tributaries from the dorsal spinal arteries when the AVM is situated in the dorsal regions of the spinal cord or when there is dorsal exophytic spreading, posterior approaches are chosen. In microsurgical resections of intramedullary glomus AVMs, two variants of nidus resection were applied: 1) first the tributaries and draining vessels near the nidus were isolated and coagulated, then the nidus was dissected from the spinal cord, and resection of the AVM was performed; or 2) the vessels of the AVM nidus were “untangled,” the feeding and the draining vessels were cut off in the nidus itself during its separation, and resection of the nidus was performed at the final stage. The last technique is more adequate if the AVM has feeding vessels from the anterior and posterior spinal arteries. First we cut off the tributaries from the posterior spinal arteries, and then we perform a myelotomy, identify, and cut off the individual vessels inside the nidus. Gradually, along the midline, we pass through the spinal cord ventrally, and in the ventral regions of the spinal cord we identify and cut off the feeding vessels from the anterior spinal artery. As a rule, these are expanded central branches of anterior spinal artery. In intramedullary diffuse AVMs, we cut off the feeding vessels near the nidus, then we performed a myelotomy, partially isolated the vessels in the nidus, coagulated and intersected them, but left them in situ. This provided the AVM with total occlusion, but taking into account diffuse penetration of the spinal cord by vessels and the presence of spinal tissue among the vessels, the method of leaving vessels in situ allowed us to reduce the spinal cord injury during operation. In cases of intramedullary glomus AVM of the conus medullaris, because of the possible pelvic disturbances, we only performed occlusion of AVM feeding vessels, leaving the malformation in situ (Figs. 1–6).

If perimedullary AVMs were present, the microsurgical technique included occlusion of the feeding vessels right at the nidus as the first step, then the draining perimedullary veins were cut off, and total resection of the AVM was performed. During this procedure, we tried to preserve the pial vascular plexus of the spinal cord. The perimedullary AVM in our studies had tributaries from the anterior and dorsal radiculomedullary arteries. We chose the dorsal approach because these arteries stretched along with the roots, which allowed us to isolate them and cut them off easily after facetectomy. In lesions with ventral drainage, ventral perimedullary draining veins were cut off via the dorsolateral approach (Figs. 7 and 8).

In cases of perimedullary AVF, the feeding vessels were cut off just before their inflow into the draining veins. When the blood flow was massive, the endovascular technique or combined interventions were used; the microsurgical technique was used when the blood flow was insignificant or moderate. When the fistula was fed from the posterior spinal arteries, the dorsolateral approach was chosen, and the feeding vessels were cut off at the point of their transition into the perimedullary veins, based on the angiograms. When the fistula was fed from the anterior spinal artery, the ventrolateral approach was chosen and, after the feeding vessels were cut off, a spine fusion was performed (Figs. 9–12).

With dural AVMs and AVFs, two variants of surgical technique were applied: 1) occluding the fistula or the malformation in the dural leaf of the spinal nerve root or cutting off the feeding vessels immediately outside the root; or 2) occluding the radicular vein, which provides
retrograde blood shunting from the AVF or the AVM into the perimedullary veins. The first method was more complicated; it required a facetectomy as well as wide isolation of the root and its dural leaf opening. This method sometimes resulted in irritation of the root and radicular pain postoperatively. In the second variant, the surgical technique was similar for dural AVM and AVF because we did not directly expose the fistula or the malformation. This variant technique essentially reduced the possibility of recurrences. With the use of the first method, the fistula or the malformation could recur due to the development of collateral vessels of the radicular arteries, which shunted into the radicular veins. Because of the lack of a shunting radicular vein, after application of the second method there was no possibility of recurrence. In all cases we used the dorsal approach. It is easier to cut off the dorsal radicular vein because it passes with the nerve roots toward the dorsal spine surface. The vein is coagulated and sectioned. The ventral radicular vein passes with the nerve roots toward the ventral spine surface. To uncover this vein, partial facetectomy and exposure of the ventral roots is necessary. We apply the first variant of surgical techniques only for dural AVMs and AVFs that have antegrade drainage into the epidural veins. In such cases, along with cutting off the tributaries, we coagulated the epidural veins into which the fistula drained (Figs. 14 and 15).

To occlude epidural AVMs and AVFs, only microsurgical techniques, dorsolateral approaches, and total or partial facetectomy were used. In the region of the intervertebral foramen, we coagulated and sectioned the direct tributaries: postcentral, prelaminar spinal branches. Sometimes we coagulated and sectioned the spinal branch of the segmental artery laterally at the point of its entry into the intervertebral foramen. We completely coagulated...
the intervertebral veins, and in addition we partially coagulated the epidural veins. Then we coagulated and sectioned the point of the fistula or removed the AVM. If the epidural draining veins compressed the dural sac, they were coagulated. When treating an epidural AVF feeding from the VA, we prefer the endovascular technique (Fig. 16).

For intravertebral malformations, we combine endovascular obliteration of feeding vessels and direct surgical intervention. To remove intravertebral vascular formations situated in the dorsal structures of the vertebrae, the standard dorsal approach was used. If the VB was affected but there was no body expansion and dura mater compression, for obliteration of vascular formations affecting the VBs, a percutaneous or intraoperative vertebroplasty was used. The VB was filled with bone cement in a volume up to 8 ml. For intervertebral malformations affecting only the VBs with their expansion, we performed ventral approaches with ventrolateral and spinal branches of the segmental arteries or the segmental branches themselves to occlude the vessels and resect the affected VB.

With combined AVMs, we applied either endovascular technology only, when there was primary extradural localization, or we used a combination of endovascular and microsurgical methods when there was mainly intradural location and spinal cord compression (Figs. 18 and 19, and Table 2).

In all 91 patients who underwent operation, total cutoff of the vascular malformation from the blood flow was a success. As mentioned earlier, in cases of AVF the intervention entailed cutoff of the fistula, in cases of AVM we performed total nidus resection, and in cases of diffuse intramedullary AVM (lesions of the spinal conus medullaris), only cutting them off from the blood flow was involved.

Actual results were estimated when the patients were discharged from the hospital. The duration of follow-up observations varied from 4 months to 8.2 years. As shown in Table 3, in 32 patients there was a considerable decrease or complete resolution of clinical symptoms immediately after the operation (Type I group), in 43 there was partial symptomatic relief (Type II group), in 10 the symptoms did not change essentially (Type III group), and in six the neurological symptoms were aggravated (Type IV group).

In the follow-up period, no aggravation of the neurological symptoms was noted. Among the six patients in the Type IV category, in two the neurological disorders reached the preoperative level, and in the other four, further slow resolution of symptoms was noted. Among the 10 patients in the Type III group, in the follow-up period relief of previous symptoms was noted in eight, and in two there was no change. Among the 43 patients in the Type II group, a further decrease of symptoms was noted in 27, and among the 32 patients in the Type I group, further relief of symptoms was noted in 19.

**Discussion**

Early classifications of spinal vascular processes included tumors and tumor-like diseases, such as hemangioblastomas and hemangiomas. In 1987, Rosenblum, et al., offered an up-to-date classification of spinal vascular malformations based on the data acquired during examination and treatment of 81 patients. These authors divided spinal AVMs into malformations and fistulas, and they...
distinguished intradural and dural vascular malformations. Intradural malformations were subdivided into intramedullary (glomus and juvenile AVMs) and extramedullary AVFs. These authors designated as dural AVFs the malformations that were situated in the dura mater and had retrograde drainage into the perimedullary veins.

In 1997 Bao and Ling distinguished intramedullary AVM, intradural AVF, dural AVF, paravertebral AVM, and Cobb syndrome (metameric localization of AVM) among the types of malformations. Intramedullary processes were subdivided into intramedullary and juvenile AVMs. Intradural AVFs were subdivided into Types I, II, and III, according to the intensity of the blood flow and the number of tributaries. These authors supposed that, for an adequate choice of treatment tactics with spinal AVMs, it was not enough to classify the lesions according to the previously established groups; it was necessary to take into account the data about the localization and structure of a specific malformation.

In 1995 Borden and colleagues subdivided spinal dural AVFs into three types by analogy with cranial dural AVFs: Type I, which had antegrade drainage into the epidural veins; Type II, which had both retrograde (into the perimedullary veins) and antegrade (into the epidural veins) drainage; and Type III, which had retrograde drainage into the perimedullary veins. In our observations, however, there were no malformations that had both antegrade and retrograde drainage.

Rodesch, et al., divided all spinal arteriovenous anomalies into AVMs or AVFs, with the latter classified as either micro- or macrofistulas. All of them corresponded to three categories, as follows: 1) genetic hereditary lesions (macrofistulas and hereditary hemorrhagic telangiectasia); 2) genetic nonhereditary lesions (all of which...
were multiple lesions with metameric or myelomeric associations); and 3) single lesions (which could represent incomplete presentations of one of the previous groups). According to these authors’ data, 81% were single lesions and 19% were multiple; among these, 59% were true intradural shunts with metameric features. Ten cases of Cobb syndrome, three of Klippel–Trenaunay syndrome, and two of Parkes–Weber syndrome, all with associated spinal cord lesions, were observed. Nineteen percent of all malformations were fistulas; 23% of those were macrofistulas, of which 83% were related to Rendu–Osler–Weber disease.17

In 2002, Spetzler, et al.,19 offered a new classification of spinal vascular processes. Among these entities the authors distinguished the following: spinal tumor vascular processes (hemangioblastomas, cavernous malformations), spinal aneurysms, AVFs, and AVMs. Among the AVFs they distinguished extradural and intradural lesions, with the following subclassifications: for ventral locations, A designated insignificant blood shunting, B was for moderate shunting, and C meant considerable shunting; for dorsal locations, A meant one tributary and B designated lesions with numerous tributaries. Among AVMs

Fig. 10. Intraoperative photographs. A: Perimedullary veins before occlusion of the fistula. B: Status during occlusion. Retrograde blood flow from the veins of the posterior fossa was blocked by clips.

Fig. 11. Selective spinal angiography. A–C: Angiograms demonstrating a perimedullary AVF with a blood supply from the anterior spinal artery at the L1–2 level. The anterior spinal artery receives its main inflow from the anterior radiculomedullary artery at the T-11 level (the artery of Adamkiewicz). D–F: Angiograms demonstrating that the fistula has been cut off at the L-1 level by clips placed at the site where the anterior spinal artery emptied into the perimedullary vein. (Clips are designated by the arrows.)
these authors distinguished extradural–intradural, intradural, and intramedullary lesions (compact, diffuse, or spinal cord conus). Intramedullary diffuse AVMs, which had been described earlier but were nowhere classified, as well as the first AVM distinguished in the spinal cord conus, were added to the classification.

From our point of view, the choice of surgical treatment depends on the localization, structure of the vascular network, and hemodynamic peculiarities of the AVM. Lengthwise (according to vertebral level) and axial (the anatomical relationship to the spinal cord) localization are the most important characteristics of a malformation. The types of feeding and draining vessels will depend on the axial and lengthwise localization because the blood supply to a malformation is always provided by the vessels passing nearby. The second most important characteristic is the structural type of malformation (AVF or AVM), taking into account compact or diffuse arrangement of the nidus vessels. The number of feeding vessels and the volume of blood flow depend on the dimensions of the AVM or AVF. We divided malformations into the following categories according to their axial localization: intramedullary, perimedullary, dural, epidural, intravertebral, or combined. In accordance with the hemodynamic data, three types of blood flow (A, B, or C) were distinguished in the malformations. To simplify the classification, we consider it necessary to indicate first the axial localization of the malformation and its structural peculiarities, and then the rest of its features (lengthwise localization, the type of inflow and outflow, and the intensity of blood flow) should be indicated as supplementary to the malformation’s characteristics.

We added the following new types of malformations to the proposed classification: the intramedullary diffuse AVM described earlier; the intradural perimedullary AVM described by Miyamoto, et al., and also found in our observations; and the dural AVM, epidural AVM and AVF, and an intervertebral AVM discovered in our clinical material. Epidural AVFs were described earlier by Asai, et al. We observed intramedullary AVMs of the conus medullaris, as did Spetzler, et al., but in our observations there were also perimedullary AVFs, which were situated in the conus region and received inflow from the arteries passing among the cauda equina. These cases are defined in our classification as correspondingly intramedullary or perimedullary AVM/AVFs with lengthwise localization in the conus medullaris. To classify perimedullary AVFs, we defined the following types: 1) so-called vertebral fistulas described earlier; 2) perimedullary AVFs localized in the thoracic spine; and 3) perimedullary AVFs occurring as lesions in the conus medullaris that were discovered in our clinical material, as perimedullary AVFs localized at 1) the cervical, 2) thoracic, and 3) conus medullaris regions, respectively. The fistulas that receive their blood supply from the VAs are also dissimilar; “low” and “high” AVFs are distinguished among them in the literature. The “high” vertebral fistulas receive inflow directly from the VAs, but they drain first into the veins accompanying the VA and then into the epidural veins. Therefore, they are mostly epidural AVFs, as opposed to “low” vertebral fistulas that drain through the radiculomedullary artery to perimedullary veins.

The division of spinal vascular malformations according to their localization and structural and hemodynamic principles favored the working out of a clear algorithm of treatment for these malformations.

For the intramedullary spinal AVM, endovascular as well as microsurgical interventions can be applied. However, endovascular treatment is not always safe, it is often impossible to cut off all the feeding vessels with this method, and the intramedullary nidus volume remains the same after treatment. Nevertheless, in 8% of observations Spetzler and coauthors failed to resect intramedullary AVMs completely. Therefore, in those authors’ opinion, when there is a considerable blood flow in an AVM, it is necessary to embolize it first and then resect it microsurgically. We used the same surgical principles: intramedullary AVMs with low and medium blood flow were treated microsurgically, whereas AVMs with a pronounced blood flow were embolized first and then resected microsurgically.
Perimedullary AVFs may be treated microsurgically or embolized, or a combination of the two methods may be used.\textsuperscript{5,7,10} When they are embolized, however, there is a great probability of recurrence, and the frequency of complications is high. On the other hand, if a microsurgical technique is used, the frequency of complications is lower and the efficacy of intervention is higher.\textsuperscript{5} At present, many authors prefer to treat perimedullary AVFs with low blood flow microsurgically, but to embolize AVFs with pronounced blood flow.\textsuperscript{7,10,14,16} Taking into account the fact that perimedullary AVMs consist of a conglomeration of vessels and often have several tributaries, only microsurgical interventions should be applied. During the operation we were trying to achieve radical removal of the AVM and to be sure to preserve the spinal arteries.

With dural AVMs and AVFs, microsurgical as well as endovascular intervention is possible.\textsuperscript{21} The relative simplicity of implementation and absence of complications...
FIG. 15. Intraoperative photographs. A: Arterialized perimedullary veins are depicted. The retrograde draining radicular vein is lifted slightly with a hook. B: After the coagulation and occlusion of the dorsal radicular vein (shown being lifted), arterIALIZATION OF THE PERIMEDULLARY VEINS HAS CEASED, AND THE VEINS HAVE TURNED BLUE AND HAVE BECOME FLAT.

FIG. 16. Angiograms revealing an AVF in the VA, which is mainly drained by the epidural veins. A and B: Direct projections. C and D: Lateral projections. E and F: Balloon occlusion of the VA supplying blood to the lesion. G and H: Angiograms demonstrating retrograde blood flow into the AVF from the opposite VA after occlusion of the VA supplying blood to the lesion and the fall of pressure in the AVF. 1 = VA; 2 = draining epidural veins; 3 = balloon used in occlusion; 4 = AVF with retrograde filling.
Spinal AVMs: new classification and surgical treatment

Fig. 17. Preoperative MR images demonstrating an intravertebral glomus AVM with extravertebral spreading.

Fig. 18. Preoperative MR image and angiograms demonstrating a combined, mainly intradural glomus AVM at the T9–11 level, with its blood supply arising mainly from the radiculomedullary arteries. A: Preoperative MR image acquired in a T₂-weighted sequence. B: Selective spinal angiography studies of the segmental artery, T-10 level, coursing from the left, early arterial phase. 1 = spinal branch, which passes into the posterior radiculomedullary artery; 2 = intramedullary/perimedullary AVM conglomerate; 3 = perimedullary draining veins; 4 = radiculomeningeal arteries; 5 = dural vascular conglomerate. C: Angiogram obtained in the final arterial phase. 1 = vessels of the intramedullary/perimedullary conglomerate; 2 = vessels of the dural conglomerate; 3 = perimedullary draining veins. D: Left injection of the T-9 segmental artery. The vascular conglomerate (2) is fed by the posterior radiculomedullary artery (1). E: Angiogram of the T-11 level of the segmental artery from the left. 1 = epidural conglomerate; 2 = draining epidural veins.
bral joint to be wide enough, which is often accompanied by postoperative radiculopathy. We prefer to cut off the radicular draining vein. This technique provides good clinical results and the recurrence of AVF/AVM has never been seen.

Different authors recommend endovascular as well as microsurgical treatment of epidural AVMs and AVFs in accordance with the volume of blood flow and the size of the feeding and draining vessels. With VA fistulas that have epidural drainage, endovascular occlusion is an optimum method.\textsuperscript{19} Epidural AVMs and AVFs in the thoracic and lumbar spines are better treated microsurgically, especially if they are situated in the artery of Adamkiewicz region.

With intravertebral AVMs, we combine embolization with a subsequent open operation. If necessary, intraoperative vertebroplasty is applied. Unless there is compression of nerve structures, we recommend applying endovascular embolization and/or transcutaneous vertebroplasty with bone cement. As a rule, combined AVMs require endovascular embolization followed by partial or complete resection of the malformation and decompression of the spinal cord.\textsuperscript{2,3,6,19,21}

Table 2: Methods of treatment used in 91 patients with AVMs

<table>
<thead>
<tr>
<th>Axial Localization</th>
<th>Structural Features of Lesion</th>
<th>Endovascular</th>
<th>Microsurgical</th>
<th>Combined</th>
<th>Total</th>
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<tbody>
<tr>
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<td>1</td>
<td>15</td>
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<tr>
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<td>1. glomus AVM</td>
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<td>2. AVF</td>
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<td>2</td>
</tr>
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<td></td>
<td>2. AVF</td>
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<td>0</td>
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<td>V. intravertebral</td>
<td>1. glomus AVM limited by vertebra</td>
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<td>0</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>2. glomus AVM w/ paravertebral spreading</td>
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<td>1</td>
</tr>
<tr>
<td>VI. combined</td>
<td>1. mainly intradural glomus AVM</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
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<td>2. mainly extradural glomus AVM</td>
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<tr>
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Table 3: Results of surgical treatment in 91 patients with AVMs

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<th>Axial Localization</th>
<th>Structural Features of Lesions</th>
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<th>II</th>
<th>III</th>
<th>IV</th>
<th>Total</th>
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<td>15</td>
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<td>2</td>
<td>3</td>
<td>28</td>
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<tr>
<td>IV. epidural</td>
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<td>2</td>
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<tr>
<td></td>
<td>2. AVF</td>
<td>0</td>
<td>1</td>
<td>0</td>
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<td>1</td>
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<tr>
<td>V. intravertebral</td>
<td>1. glomus AVM limited by vertebra</td>
<td>0</td>
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<td>1</td>
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<tr>
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<td>2. glomus AVM w/ paravertebral spreading</td>
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<td>1</td>
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<tr>
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<td>1. mainly intradural glomus AVM</td>
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<td>1</td>
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<td>0</td>
<td>3</td>
</tr>
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<td>2. mainly extradural glomus AVM</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
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<tr>
<td>total</td>
<td>32</td>
<td>43</td>
<td>10</td>
<td>6</td>
<td>91</td>
<td></td>
</tr>
</tbody>
</table>

* Roman numerals denote the following results: I, considerable or complete resolution of neurological symptoms; II, improvement of symptoms; III, no changes; IV, aggravation of symptoms.

Spinal AVMs: new classification and surgical treatment

Conclusions

For a successful surgical treatment of spinal AVMs, it is necessary to obtain as complete a set of data as possible about their localization, vascular structure, and hemodynamics. This information will promote the use of optimum surgical methods and the latest microsurgical and endovascular interventions, with treatments differentiated according to the type of malformation. One should try to apply the least invasive endovascular approach in these cases, where possible, to occlude the AVM or to reduce the intensity of blood flow by embolization. When performing an AVM resection or occlusion, one should use a direct approach to the malformation, blocking only the vessels supplying blood to the malformation and preserving the ones feeding the spinal cord. It is necessary to resect the AVM nidus sharply only along the border with the spinal cord. After the operation, it is always necessary to obtain an MR image and a selective spinal angiography study for follow-up evaluation. Only such a combination of neuroimaging methods can identify the remnants of a pathological vascular formation.2,3,19

References


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