A proposed classification for spinal and cranial dural arteriovenous fistulous malformations and implications for treatment

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A classification is proposed that unifies and organizes spinal and cranial dural arteriovenous fistulous malformations (AVFMs) into three types based upon their anatomical similarities. Type I dural AVFMs drain directly into dural venous sinuses or meningeal veins. Type II malformations drain into dural sinuses or meningeal veins but also have retrograde drainage into subarachnoid veins. Type III malformations drain into subarachnoid veins and do not have dural sinus or meningeal venous drainage. The arterial supply in each of these three types is derived from meningeal arteries.

The anatomical basis of the proposed classification is presented with several cases that illustrate the three types of dural AVFMs. A rationale for the treatment of spinal and cranial dural AVFMs according to their anatomical characteristics is discussed.

KEY WORDS • dural arteriovenous fistula • arteriovenous malformation • classification

There is debate regarding the correctness of the term “dural venous fistula” versus “dural arteriovenous malformation.” These malformations are comprised of one or more dural-based arteriovenous fistulas; use of the singular fistula to describe multiple fistulas therefore seems inappropriate. We prefer the term “malformation” because a single malformation can contain one or more fistulas. On the other hand, malformation suggests a developmental anomaly but dural fistulas are most commonly acquired. The term “dural arteriovenous fistulous malformation” (AVFM) would best describe all of these abnormalities whether they consist of one or more fistulas.

Dural AVFMs of the head and spine are uncommon, often appear to be acquired, and present later in life than arteriovenous malformations (AVMs) of the brain and spinal cord. It has been recognized that the venous drainage pattern determines the clinical presentation of cranial dural AVFMs. The most effective treatments of dural AVFMs of the head and spine are those directed primarily toward the venous side.

We are proposing a classification system that unifies spinal and cranial dural AVFMs and provides a rationale for modes of treatment. Our system is based upon the classification system for cranial dural AVFMs of Djindjian and Merland. It incorporates spinal dural AVFMs into that system and organizes their classification into three types that are relevant to treatment.

Spinal AVMs had been classified as intradural, extradural, or vertebral but were poorly understood until 1977, when Kendall and Logue pointed out that the majority of supposed intradural spinal AVMs were actually the venous drainage of dural AVFMs located on a nerve root sleeve. The striking, dilated venous drainage of some spinal AVMs was noted years ago by Hebold in 1885, Krause in 1911, and Elsberg in 1916, but the relationship between spinal extradural AVMs and spinal dural AVFMs that drained intrathecally has not been investigated. We suggest that they are related to one another in the same way that cranial dural AVFMs that drain into dural sinuses are related to those that drain into subarachnoid veins.
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Types of Dural AVFMs

Type I

Type I dural AVFMs are those that drain directly into the dural venous sinus or meningeal vein. These malformations consist of one or more fistulas that occur between a meningeal artery and a dural venous sinus or meningeal (dural) vein. The flow of the veins draining from the brain or spinal cord into the dural sinus or meningeal veins is in the normal direction (that is, anterograde). In the head, these malformations are often asymptomatic but may present with pulsatile tinnitus or cranial nerve deficits. They typically have a benign course. We consider carotid-cavernous fistulas (both posttraumatic and spontaneous) to be dural AVFMs.

We also classify as Type I dural AVFMs, malformations that involve Batson’s epidural venous plexus, which were previously considered extradural. This venous plexus, located between the spinal periosteum and dura, corresponds to the cranial dural venous sinuses and meningeal venous plexus that are located between the cranial periosteal and meningeal dural layers. In the spine, these malformations may present with myelopathy, radiculopathy, spinal bruit, or spontaneous epidural hematoma.

Type II

Type II dural AVFMs are those that drain into the venous sinus with retrograde drainage into subarachnoid veins. These malformations consist of one or more fistulas between meningeal arteries that drain directly into a dural sinus. However, the flow in subarachnoid veins is reversed: arterialized blood travels retrograde from the sinus into the subarachnoid veins. Type II dural AVFMs present with neurological deficits due to venous hypertension or hemorrhage. Spinal dural AVFMs that drain into both the epidural venous plexus and perimedullary veins are also classified as Type II dural AVFMs because the epidural plexus corresponds to an intradural sinus and the spinal perimedullary veins are subarachnoid.

Type III

Type III dural AVFMs are those that drain into the subarachnoid veins. These malformations consist of one or more dural AVFMs that drain into subarachnoid veins located at or on the wall of dural sinuses; these dural AVFMs drain retrograde through a vein that normally drains into the sinus. The sinus may be patent or occluded; however, when the sinus is patent, the arterialized vein and the sinus are not in communication at the point of the fistula. In some cases, the fistula is located directly between the meningeal artery and the draining subarachnoid vein. In other cases, the fistula is located between a meningeal artery and a segment of a dural venous sinus that is isolated, such as a dural venous sinus that is completely thrombosed on both sides of the arterialized segment. In still other cases, the fistula is located between a meningeal artery and a dural venous structure (that is, a vein or lake) which is similarly isolated from dural venous sinus drainage. In every case, the malformation drains through subarachnoid veins.

These AVFs present with hemorrhage or other sequelae of venous hypertension in superficial cortical veins, the deep venous system, and the spinal coronal or perimedullary venous plexus. Occasionally, infratentorial Type III malformations drain into spinal perimedullary veins or Type III spinal malformations drain intracranially. It is clear that the structures affected by venous hypertension and the location of the hemorrhage can be distant from the malformations.

The most common group of Type III dural AVFMs are spinal dural AVFMs that are supplied by meningeal branches of a radicular artery and that drain intrathecally through a subarachnoid vein into the coronal venous plexus. These malformations have previously been classified as angioma racemosum venosum or Di Chiro’s Type I intradural AVMs. However, it is clear that these malformations are not intradural and should not continue to be classified with intradural malformations.

Subtypes

Malformations can be subclassified as follows: those consisting of a simple fistula and those with multiple fistulas in a single region.

Subtype a: Simple Fistula. A simple fistula is a direct connection between a single meningeal artery and a draining vein or sinus. The fistula can drain directly into any of the following: a dural sinus or meningeal vein (Type Ia); a draining dural sinus or meningeal venous plexus with retrograde flow into subarachnoid veins (Type IIa); or only a subarachnoid vein (Type IIIa).

Subtype b: Multiple Fistulas. Other malformations have a more complex structure with multiple dural-based AVFMs that are fed by multiple arteries. Type Ib malformations drain into a single sinus or meningeal venous plexus. Type IIb malformations also drain secondarily into subarachnoid veins, and Type IIIb malformations drain solely into subarachnoid veins, usually into one but occasionally into several.

When multiple fistulas drain into a contiguous, arterialized dural venous structure, we consider them to be part of a single malformation, and when multiple fistulas drain into distinct subarachnoid veins, we consider them to be multiple distinct malformations. In certain conditions, such as when there is a global venous hypertension, the distinction between a single malformation that consists of multiple fistulas and multiple distinct malformations may become arbitrary.

Figures 1 to 7 demonstrate types of dural AVFMs of the superior petrosal sinus and spine. Table 1 summarizes each type of dural AVFM.

Illustrative Cases

Table 2 summarizes several cases that were recently treated. Cases that illustrate specific points are discussed below.

Case 5: Craniospinal Type II

This 64-year-old man presented with a subarachnoid hemorrhage, quadriaparesis, and respiratory arrest. Right vertebral angiography revealed an AVM of the extradural vertebral artery (transverse segment) draining into the epidural vertebral venous plexus at C-1 and intradurally into a subarachnoid vein, producing a varix anterior to the
medulla. Through a lateral suboccipital craniectomy and cervical laminectomy, the vertebral plexus was coagulated and stripped from the vertebral artery at C-1. The draining venous varix in the subarachnoid space was ligated.

**Comment.** This case demonstrates a dural AVFM with drainage into the vertebral venous plexus. Drainage was retrograde into subarachnoid veins along the brain stem. The transverse segment of the vertebral artery runs beneath the atlantooccipital membrane between the transverse foramen of C-1 and its dural entrance, and a portion of the vertebral venous plexus is confluent with the epidural venous plexus and marginal sinus of the foramen magnum. We classify this uncommon malformation as a Type II.

**Case 7: Superior Petrosal Type III**

This 57-year-old man presented with abrupt onset of obtundation and a dense spastic quadripareisis after a several-month history of personality change, memory loss, and gait deterioration. Computerized tomography scans of the head showed hydrocephalus and hemorrhage in the third and fourth ventricles. Magnetic resonance (MR) imaging showed edema (increased T₂-weighted signal) throughout the thalami, brain stem and cerebellum, and gadolinium enhancement in both thalami and the mesencephalon. A cerebral angiogram showed two dural AVMs, the first located at the tentorial incisura and drained through a dilated lateral pontomesencephalic vein into the basal vein of Rosenthal and the vein of Galen. The second malformation was located in the right anterior cranial fossa at the base of the superior sagittal sinus and drained through a cortical vein toward the superficial middle cerebral vein. The straight sinus was occluded; flow through the internal cerebral veins was retrograde (Fig. 8).

The petrosal fistula was approached via a subtemporal craniotomy. The meningeal supply from branches of the
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Intracavernous left internal carotid artery (ICA) was coagulated along the tentorium. After the tentorium was incised, the draining vein was identified on its undersurface along the lateral wall of the superior petrosal sinus. As the fistula was being excised, the superior petrosal sinus was found to be patent so it was packed with Surgicel. The draining vein was ligated between two clips.

Postoperatively, the patient has made a good recovery over several months. He has a mild residual left hemiparesis, but has become oriented with progressively improving memory deficits.

Comment. This case is important for several reasons. First, the patient had two Type III dural AVFMs. Second, it demonstrates that venous sinuses remote from a dural AVF can be thrombosed. In addition, it shows that intraventricular hemorrhage can occur distant from the site of the malformation. We hypothesize that the intraventricular hemorrhage developed from fragile subependymal veins that were placed under increased pressure. Although transverse sinus thrombosis has been related to the later demonstration of transverse sinus dural AVFMs, in this case it may be that the straight sinus either was previously thrombosed or became stenotic or thrombosed in reaction to chronic arterialization.

Case 11: Posterior Superior Sagittal Type III

This 61-year-old man had the onset of vertigo and episodic facial numbness 8 years prior to admission. An occipital bruit was found, and an angiogram showed dural AVFMs of the posterior superior sagittal sinus, which was supplied by the tentorial branches (Davidoff and Schecter) of both posterior cerebral arteries and the tentorial artery (Bernasconi and Cassonari) from the right ICA. Drainage was directed into a dilated, anastomotic occipital vein and ultimately into the vein of Galen (Fig. 9). Although the malformation was located on the superior sagittal sinus, it did not drain into it. Ligation of the arterialized draining vein and coagulation of the malformation on the wall of the sinus eliminated the malformation. Postoperative angiography showed stenosis but patency of the superior sagittal sinus.

Comment. This case demonstrates two points. First, Type III malformations may be associated with a patent underlying sinus; it has been previously stated that the
underlying sinus is always thrombosed. Second, this case demonstrates an occipital extracerebral venous anastomosis between the superior sagittal sinus (superficial system) and vein of Galen (deep system). Intracerebral venous anastomoses that drain into the subependymal collecting system frequently drain AVMs of the brain but have not been reported to drain dural AVFs.

Type I Dural Arteriovenous Fistulous Malformations

Many Type I malformations are asymptomatic but they can present with pulsatile tinnitus, cranial nerve deficit, or radiculopathies. Type I cranial malformations do not present with hemorrhage or venous infarcts but Type I spinal (epidural) malformations occasionally present with epidural hemorrhage. These lesions have a high rate of spontaneous thrombosis, and if they are asymptomatic, they do not need to be treated. However, when cavernous sinus malformations (that is, carotid-cavernous fistulas) present with progressive visual loss, they require urgent treatment.

Type I malformations may drain into either a major dural venous sinus or a minor dural venous channel (meningeal vein). When the malformation drains into a major venous sinus, it is important to preserve the venous drainage. For this reason, treatment is directed to the arterial side of the malformation.

In many cases, transarterial embolization with particles or glue is used to reduce blood flow through the malformation and can result in complete thrombosis of the malformation (approximately 60%). The best situation for transarterial embolization is a simple fistula (Type Ia), in which the blood supply is derived from a branch of the external carotid artery. When the supply is from arteries that supply important structures, embolization creates a risk of stroke. For embolization to be curative, glue should occlude all fistulas. When large dural collateral vessels exist between external and internal vessels, the injection force needed to perfuse a fistula can cause reflux across these collateral vessels into the brain. The malformations best treated by transarterial embolization consist of a simple fistula (Type Ia) supplied by a branch of the external carotid artery.
When a dural AVFM drains into a major venous sinus that cannot be safely occluded, it is often difficult to cure the malformation, especially if it has multiple fistulas. When the sinus must be preserved, the dural-based arterial supply can be significantly reduced by surgical isolation (skeletonization) of the sinus.\(^4\) Skeletonization is an option for treatment of multiple fistulas (Type Ib) when blood supply is from arteries that also supply important structures or when transarterial embolization fails. In most cases, the arterial supply cannot be completely eliminated unless the sinus wall is incised proximal and distal to the malformation. However, the low success rate of skeletonization combined with the benign natural history of Type I dural AVFMs makes this procedure an option to be considered only in rare circumstances.

When a dural AVFM drains into a meningeal vein or a sinus that can be occluded, it is best treated by occlusion or excision of the venous drainage at the site of the fistula(s). Spinal malformations that drain into epidural veins can be excised or coagulated. It may be difficult to identify the fistulas themselves but coagulating and/or excising all of the arterialized epidural veins is curative. Malformations that drain into the cavernous sinus can be treated by occluding the cavernous sinus from a direct surgical exposure\(^3,15,45,59,60,75\) or by using a percutaneous transvenous endovascular approach\(^18,38,39,56\).

**Type II Dural Arteriovenous Fistulous Malformations**

When the venous outflow of a sinus becomes restricted by stenosis or partial thrombosis within the sinus, the
pressure within the sinus becomes elevated. When the sinus is arterialized by a dural AVFM, the pressure can become markedly elevated and instead of venous blood flowing into a sinus from subarachnoid veins, arterialized blood flows out of the sinus through these veins. Once the subarachnoid veins become chronically arterialized, collateral venous drainage pathways form and the sinus can be safely thrombosed after the arterialized draining veins have been ligated.

We favor a combined surgical–endovascular approach for Type II malformations. When a percutaneous endovascular technique alone is used to treat these malformations, partial sinus thrombosis can redirect arterIALIZED blood into subarachnoid veins, thus risking a venous infarction. This risk is reduced by combining direct surgical exposure and ligation of the red arterIALIZED veins prior to endovascular sinus thrombosis. When the malformation drains into subarachnoid veins, it can also drain through intradiploic collateral vessels; as a result, profuse bleeding can occur during elevation of the bone flap. Preoperative transarterial embolization can be helpful to lessen this problem.

Type III Dural Arteriovenous Fistulous Malformations

Malformations that drain directly into subarachnoid veins cause venous hypertension, which produces neurological deficits by hemorrhage, brain edema, and/or ischemia. Neurological deficits can be severe and edema can be widespread. Many of these effects are reversible and the patients warrant treatment because they can recover from even profound neurological deficits.

Malformations that drain directly into subarachnoid veins (Type III) are not accessible via a transvenous approach through a sinus. Treatment of Type III malformations requires thrombosis or ligation of the draining vein as it exits the dura. Prior to ligation of the draining vein, the malformation can be coagulated, excised, or

FIG. 5. Illustration showing a Type III(b) dural arteriovenous fistulous malformation of tentorium cerebelli from the right subtemporal approach. The nidus on tentorial venous lake drains in retrograde fashion through inferior temporal lobe vein. The nidus is supplied by middle meningeal artery (MMA) and tentorial artery (taBC). The tentorial dural lake has aneurysmal dilatation and does not drain into superior petrosal sinus; the draining vein has multiple aneurysms. Number 3 = oculomotor nerve; number 4 = trochlear nerve; number 5 = trigeminal nerve; ITLV = inferior temporal lobe vein with multiple aneurysms; SPS = superior petrosal sinus; T = aneurysmal tentorial venous lake.
intraoperatively perfused with glue retrograde through the draining vein.\textsuperscript{5,40} The largest series of Type III dural AVFs are malformations of the spine, in which cases interruption of the draining vein alone is effective.\textsuperscript{50,64,82} Draining vein ligation is also effective in treating cranial malformations;\textsuperscript{37} however, when the fistulated dural segment is readily accessible, coagulation and excision of the dural base are frequently helpful to allow proper visualization of the dural junction of the draining vein prior to its ligation (Fig. 4). In contrast to AVMs of the brain, in which early ligation of a draining vein can produce swelling and hemorrhage, the draining vein of a Type III dural AVFM can be safely occluded with a clip at the dural junction. When properly clipped, the distal venous drainage gradually turns blue and loses its turgor. The vein is then ligated.

Type III malformations are usually fed by vessels that also supply important structures. For example, malformations at the anterior base of the superior sagittal sinus are fed by branches of the ophthalmic artery; malformations along the tentorium are fed by intracavernous branches of the ICA, and spinal malformations are fed by meningeal branches of the radicular arteries. When these arteries are embolized, there is a risk of stroke (approximately 10%).\textsuperscript{11,25,30,31,40,58,71} Because Type III malformations drain solely into subarachnoid veins and never into intradiploic veins, they do not carry the risk of operative bleeding seen in Type II malformations. The morbidity of ligating draining subarachnoid veins is very low.\textsuperscript{54,82}

\textbf{Fig. 6.} Illustration showing a spinal Type I(b) dural arteriovenous fistulous malformation. The nidus on nerve root sleeve is fed by meningeal branches of radicular artery. The dilated venous plexus causes nerve root and spinal cord compression; however, venous drainage of spinal cord is not affected. \textit{ara} = anterior branch of radicular artery; \textit{dsv} = dorsal spinal cord venous plexus draining into radicular vein; \textit{dvp} = dural venous plexus, arterialized and dilated; \textit{md} = meningeal (or true) dura; \textit{mra} = meningeal branch of radicular artery; \textit{ra} = radicular artery; \textit{rv} = radicular vein; \textit{sp} = spinal periosteum (or periosteal dura).

\textbf{Fig. 7.} Illustration showing a spinal Type III(a) dural arteriovenous fistulous malformation. The arteriovenous fistula on nerve root sleeve is fed by meningeal branch of radicular artery and drains into dorsal spinal venous plexus through subarachnoid anastomotic vein (arterialized). The dural venous plexus between spinal periosteum and meningeal dura does not drain the nidus. \textit{ara} = anterior branch of radicular artery; \textit{df} = dural fistula; \textit{dsv} = dorsal spinal cord venous plexus draining into radicular vein; \textit{dvp} = dural venous plexus; \textit{md} = meningeal (or true) dura; \textit{mra} = meningeal branch of radicular artery; \textit{ra} = radicular artery; \textit{rv} = radicular vein; \textit{pra} = posterior branch of radicular artery; \textit{sp} = spinal periosteum (or periosteal dura).

\textbf{Fig. 8.} Lateral arterial-phase angiogram demonstrating two dural arteriovenous malformations (arrows). The first is at the superior petrosal sinus supplied by the tentorial branches of the intracavernous carotid artery and draining through the petrosal vein into the deep venous system. The second nidus located at the base of the superior sagittal sinus is supplied primarily by ethmoidal perforators from the ophthalmic artery and drains into a cortical vein.
In our experience, dural malformations that present with hemorrhage have a high rate of early rebleeding. There is very little information in the literature on the rate of rebleeding of dural AVFs, but three of seven patients who presented with hemorrhage bled again within the first few days of hospitalization. Patients with dural AVFs who present either with hemorrhage or progressive neurological deficit should be treated expeditiously; this precludes the use of focused radiation.

**Discussion**

Yaşargil\(^8\) noted that Rizzoli, in 1881, was the first to describe an AVM that involved the dura mater and Sachs\(^7\) reported the first angiographic description in 1931. Subsequently, cranial dural AVFs have been most frequently described at the transverse sinus\(^27,52,81,83\) and cavernous sinus (being also known as carotid-cavernous fistulas\(^6,16,18,41,66,68,84\)), although they occur at every cranial dural sinus.\(^28,33,57\)

Krause\(^53\) first described angioma venosum racemosum of the dorsal thoracic spinal cord in 1910. Elsberg\(^32\), in 1916, was the first to describe a spinal epidural malformation (Type I) and was the first to successfully treat what we now understand to be a spinal dural AVM with intrathecal venous drainage (Type III) by ligation of its venous drainage. He performed a laminectomy on a patient who presented with a thoracic myelopathy and found a long, coiled vessel dorsal to the spinal cord. He ligated an apparent solitary thoracic arterial supply, and the patient made an excellent recovery. From the early descriptions of Krause and Elsberg, we can recognize these angioma racemosum as the subarachnoid venous drainage of a dural AVM. That these vessels were dilated veins was known to pathologists of the 1940s and 1950s\(^78,87\), but their pathophysiology was unclear and became more confused after their arteriovenous nature was demonstrated on spinal angiography.\(^20,21,23\) Di Chiuro classified these angioma racemosum as Type I intradural AVMs and subsequently these malfor-

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**TABLE 1**

**Summary of dural AVFM types**

<table>
<thead>
<tr>
<th>Category</th>
<th>Description</th>
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<tbody>
<tr>
<td>type I supply malformation</td>
<td>meningeal arteries</td>
</tr>
<tr>
<td>drainage</td>
<td>directly into dural venous sinus or meningeal vein</td>
</tr>
<tr>
<td>presentation</td>
<td>pulsatile tinnitus, cranial bruit, cranial nerve deficits, or radiculopathy</td>
</tr>
<tr>
<td>clinical course</td>
<td>often benign, high rate of spontaneous remission</td>
</tr>
<tr>
<td>type II supply malformation</td>
<td>simple AVFM, often high-flow, or complex/multiple fistulas</td>
</tr>
<tr>
<td>drainage</td>
<td>directly into dural venous sinus/meningeal vein with retrograde flow into subarachnoid veins</td>
</tr>
<tr>
<td>presentation</td>
<td>pulsatile tinnitus, cranial bruit, focal neurological deficit, seizure, hemorrhage, elevated ICP</td>
</tr>
<tr>
<td>clinical course</td>
<td>progressive neurological deficit or hemorrhage</td>
</tr>
<tr>
<td>type III supply malformation</td>
<td>meningeal arteries</td>
</tr>
<tr>
<td>drainage</td>
<td>subarachnoid veins, not through dural venous sinus</td>
</tr>
<tr>
<td>presentation</td>
<td>focal neurological deficit, seizure, hemorrhage, elevated ICP, myelopathy</td>
</tr>
<tr>
<td>clinical course</td>
<td>progressive neurological deficit or hemorrhage</td>
</tr>
</tbody>
</table>

\(^{AVFM} = \) arteriovenous fistulous malformation; ICP = intracranial pressure.
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mations have been considered together with other intradural AVMs as distinct from the extradural AVMs.\(^{89}\) Spinal dural AVFs have been reported to arise at all spinal levels from the foramen magnum up to and including the sacrum.\(^{22,24,58,69,87}\)

Houser, et al.,\(^{46}\) were the first to describe the importance of venous drainage in the clinical presentation of dural AVFs when they distinguished between drainage directly into the dural sinuses and the pial (subarachnoid) veins. Castaigne, et al.,\(^{12}\) described an additional type of malformation that drains into large venous lakes and presents with mass effect. Djindjian and Merland\(^{28}\) distinguished among drainage into venous sinus, sinus and veins, cortical veins, or dural lakes. Gobin and associates\(^{35}\) have recently extended this classification system to include intracranial dural AVFs that drain into spinal perimedullary veins.

We classify spinal dural AVFs with drainage into subarachnoid (intrathecal) veins as Type III dural AVFs because of their similarity to Djindjian and Merland’s\(^{28}\) Type III dural AVFs that have cortical venous drainage. We also classify as Type III malformations those dural AVFs that first drain into dural lakes and then into subarachnoid veins (Djindjian and Merland’s Type IV) because the treatment is the same as for other Type III malformations. In addition, those cranial dural AVFs that drain into spinal perimedullary veins (Gobin Type V\(^{35}\)) are classified as Type III because these veins are also subarachnoid and are treated in the same way. We classify all malformations that drain only through subarachnoid veins as Type III.

**Anatomical Basis of Classification**

The cranial dura consists of two layers: the outer or periosteal layer, and the inner or meningeal layer. At the foramen magnum, the periosteal dural layer forms the atlantooccipital membrane and continues caudally as the inner periosteum of the spine. The meningeal layer continues as the spinal dura. Thus the meningeal veins and sinuses located between the two fused layers of cranial dura correspond to the spinal epidural venous plexus, which is located in the space between the spinal periosteum and meningeal dura. The confluence between the cranial dural venous sinus system and the spinal epidural venous plexus was first described by Breschet, in 1824.\(^{7}\)

The intimate relationship between major cranial venous sinuses and the rich network of meningeal veins within dural structures such as the falx cerebri and tentorium cerebelli is well described by Browder and Kaplan.\(^{9}\) Djindjian\(^{23}\) and Gillilan\(^ {14}\) described the venous drainage of the spinal cord, including the intrinsic drainage and extrinsic coronal venous plexus, medullary anastomoses, and the epidural venous plexus.

Each artery that ultimately supplies the brain or spinal cord gives off a meningeal branch as it penetrates the dura. These arteries consistently penetrate the dura through a venous sinus or else are surrounded by a venous plexus. For example, the ICAs penetrate the dura through the cavernous sinuses, the vertebral arteries penetrate through the marginal sinus, and the radicular arteries are surrounded by the epidural venous plexus. The intimate relationship between the meningeal branches and the venous sinuses give ample anatomical opportunity for the development of fistulas. The meningeal arteries that tend to travel in vascular grooves between the skull and periosteal dural layer give off branches that supply the meningeal folds (that is, the tentorial and falcine arteries). Sinuses such as the superior sagittal sinus, the superior petrosal sinus, and the transverse sinus (which do not envelop major intracranial vessels) are located at these meningeal dural reflections (namely, the falx cerebri and tentorium cerebelli). The falcine and tentorial arterial branches pierce these sinuses, from their origin along the cranial vascular grooves to the meningeal dural folds. This again shows how easily fistulas could form.

Regardless of the type or location, dural AVFs occur at points at which meningeal arteries pierce the dura. At these points, meningeal arteries, which tend to run outside

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**Fig. 10.** Illustration showing the various types of dural arteriovenous malformations (AVMs) on left transverse sinus (ts) supplied by the middle meningeal artery (MMA). A: Type I(a) dural AVM with anterograde venous drainage. The fistula (f) is between a branch of MMA and the ts. B: Type I dural AVM. Thrombosis of sigmoid sinus causes reversal of flow within the ts. The ts drains normally across the torcular Herophili (tH). The vein of Labbé (VL) drains normally into the ts. C: Type II dural AVM. The proximal ts stenosis redirects outflow into VL which drains in retrograde direction. D: Type III dural AVM. Transverse sinus thrombosis with the patent channel draining f into the dilated, arterialized VL. E: Type III dural AVM. Transverse and sigmoid sinuses have partially recanalized. The f drains through the persistent channel into the VL. F: Type III dural AVM. After further recanalization of transverse and sigmoid sinuses, the sinus lumen is patent and venous blood flows anterograde from the tH to the jugular vein. The dural f is not in communication with lumen of sinus and persistently drains retrograde into the VL.
the dural layers, come much closer to meningeal veins, which tend to run between dural layers. The dura has a rich network of arterial anastomoses, which explains why malformations apparently supplied solely by external branches can quickly recruit blood supply from internal branches after embolization.

Etiology

Venous sinus thrombosis has been associated with cranial dural AVFMs, which explains why malformations apparently supplied solely by external branches can quickly recruit blood supply from internal branches after embolization.

We hypothesize that when thrombus forms on a sinus, the dural wall becomes inflamed and thin-walled vessels are formed in the process of neovascularization. During sinus recanalization, fistulas may form between abnormal thin-walled meningeal vessels and the venous lumen. Partial sinus thrombosis can redirect arterialized blood into subarachnoid veins and progressive thrombosis can form pathological fistulas as a result of sinus thrombosis. However, the exact mechanism for this formation has not as yet been described in detail.

TABLE 2

Summary of cases

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Type</th>
<th>Location</th>
<th>Presentation*</th>
<th>Arterial Supply</th>
<th>Venous Drainage</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>44</td>
<td>I</td>
<td>dorsal spinal (L-4)</td>
<td>progressive myelopathy</td>
<td>epidural defect on myelography, unknown branches of internal &amp; external carotid, multiple fistulas external carotid-cavernous fistula</td>
<td>epidural venous plexus</td>
<td>excision</td>
</tr>
<tr>
<td>2</td>
<td>56</td>
<td>I</td>
<td>transverse sinus</td>
<td>pulsatile tinnitus</td>
<td>Lt transverse sinus (stenotic)</td>
<td>none</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>54</td>
<td>II</td>
<td>cavernous sinus syndrome, meningeal inflammation, hemorrhage</td>
<td>cavernous sinus syndrome, meningeal inflammation, hemorrhage</td>
<td>thrombosed posterior cavernous sinus &amp; superficial middle cerebral vein, retrograde drainage into deep middle cerebral vein</td>
<td>none, patient died</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>58</td>
<td>II</td>
<td>convexity</td>
<td>hemorrhage</td>
<td>middle meningeal artery branches</td>
<td>drainage into meningeal vein draining into superior sagittal sinus</td>
<td>excision of dural ntidus</td>
</tr>
<tr>
<td>5</td>
<td>64</td>
<td>II</td>
<td>spinal (C-1)</td>
<td>hemorrhage, quadriparesis</td>
<td>meningeal branches of vertebral artery</td>
<td>vertebrobasilar plexus &amp; subarachnoid vein ventral to medulla</td>
<td>excision of venous plexus &amp; ligation of subarachnoid vein</td>
</tr>
<tr>
<td>6</td>
<td>64</td>
<td>III</td>
<td>frontal base of superior sagittal sinus</td>
<td>hemorrhage</td>
<td>bilateral ethmoidal branches of external carotid &amp; ophthalmic arteries</td>
<td>large venous aneurysms &amp; cortical vein draining into superficial middle cerebral vein</td>
<td>excision of dural ntidus &amp; ligation of draining vein</td>
</tr>
<tr>
<td>7</td>
<td>57</td>
<td>III</td>
<td>superior petrosal sinus</td>
<td>quadriplegia &amp; obtundation, SAH</td>
<td>bilateral ethmoidal branches of external carotid &amp; ophthalmic arteries</td>
<td>petrosal vein, lateral pontomesencephalic anastomotic vein &amp; vein of Galen. Straight sinus thrombosed. Retrograde flow through internal cerebral veins</td>
<td>excision of dural ntidus &amp; ligation of draining vein</td>
</tr>
<tr>
<td>8</td>
<td>39</td>
<td>III</td>
<td>superior petrosal sinus</td>
<td>facial pain</td>
<td>tentorial branches of internal &amp; external carotid arteries</td>
<td>petrosal vein, lateral pontomesencephalic anastomotic vein, &amp; vein of Galen</td>
<td>excision of dural ntidus &amp; ligation of draining vein</td>
</tr>
<tr>
<td>9</td>
<td>69</td>
<td>III</td>
<td>superior petrosal sinus</td>
<td>third ventricular hemorrhage</td>
<td>tentorial branches of internal &amp; external carotid arteries</td>
<td>venous varix ventral to midbrain ultimately into vein of Galen subarachnoid anastomotic vein to vein of Galen</td>
<td>coagulation of dural ntidus &amp; ligation of petrosal vein</td>
</tr>
<tr>
<td>10</td>
<td>61</td>
<td>III</td>
<td>posterior body of superior sagittal sinus</td>
<td>headaches, dizziness</td>
<td>bilateral middle &amp; posterior meningeal arteries</td>
<td>multiple subarachnoid veins from transverse sinuses</td>
<td>ligation of veins</td>
</tr>
<tr>
<td>11</td>
<td>66</td>
<td>III</td>
<td>torcular (bilateral transverse sinus)</td>
<td>infratentorial hemorrhage</td>
<td>posterior meningeal arteries</td>
<td>multiple subarachnoid veins from transverse sinuses</td>
<td>direct aneurysm puncture for Gisturco coil embolization</td>
</tr>
<tr>
<td>12</td>
<td>63</td>
<td>III</td>
<td>posterior inferior sagittal sinus at junction with straight sinus</td>
<td>mass effect, obtundation</td>
<td>anterior cerebral meningeal branches, tentorial branches of posterior cerebri</td>
<td>giant aneurysm of vein of Galen &amp; precentral cerebral vein, did not drain into inferior sagittal sinus or straight sinus</td>
<td>coagulation of ntidus &amp; ligation of draining vein</td>
</tr>
<tr>
<td>13</td>
<td>70</td>
<td>III</td>
<td>spinal (T-9)</td>
<td>myelopathy</td>
<td>T-9 radicular artery</td>
<td>coagulation of ntidus &amp; ligation of draining vein</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>71</td>
<td>III</td>
<td>spinal (L-3)</td>
<td>myelopathy</td>
<td>rt L-3 radicular artery</td>
<td>coagulation of ntidus &amp; ligation of draining vein</td>
<td></td>
</tr>
</tbody>
</table>

* SAH = subarachnoid hemorrhage.
transient Type I malformations into Type II malformations.\textsuperscript{54} We suggest that progressive sinus thrombosis can transform Type II malformations into Type III malformations. Partial recanalization of a thrombosed sinus may allow antegrade venous drainage through the sinus while a Type III dural AVFM on the sinus wall drains retrograde into a cortical vein, as noted, for example, in Case 11 (Figs. 9 and 10).

The relationship between meningeal venous thrombosis and the development of spinal dural AVFMs has not been established. Conditions associated with vascular fragility and the development of spinal dural AVFMs has not been established. Fibromuscular dysplasia\textsuperscript{4} and neurofibromatosis type 1\textsuperscript{17,44,76} (which is associated with abnormalities of the vertebral arteries) are associated with Type I dural AVFMs of the spine. Ehlers-Danlos syndrome has been associated with spontaneous carotid-cavernous fistulas.\textsuperscript{36,79} It is easy to see how trauma could lead to fistula formation between meningeal arteries and veins or dural sinuses but it is difficult to understand how complex malformations might develop by this mechanism unless sinus thrombosis and subsequent recanalization occur.

Summary

Spinal and cranial dural AVFs are classified into three types based on anatomical similarities. Type I dural AVFMs drain directly into dural venous sinuses or meningeal veins and can be treated by transarterial embolization or surgical skeletonization if venous drainage needs to be preserved. Both of these techniques are less than optimal, being associated with moderate risk, and should be used rarely, especially since Type I dural AVFMs usually have minor symptoms. If it is safe to sacrifice the venous drainage, the malformation can be treated by thrombosis of the sinus and coagulation or excision of the meningeal vein. Type II malformations drain into a dural sinus or meningeal vein and have retrograde drainage into subarachnoid veins. These malformations are treated by ligation of arterialized subarachnoid draining veins and thrombosis or excision of the sinus. Type III malformations drain directly into a subarachnoid vein without dural sinus or meningeal venous drainage. These malformations are treated by coagulation and/or excision and ligation of the draining subarachnoid vein. Unlike AVMs of the brain or spinal cord, the vein draining a dural AVFM may be safely ligated before the malformation is excised. In contrast to Type I dural AVFMs, which are usually benign, Types II and III dural AVFMs cause serious problems and need treatment. Our patients who have presented with hemorrhage from venous hypertension have had a high rate of early rebleeding.

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