Successful treatment of a group of spinal cord arteriovenous malformations by interruption of dural fistula

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As demonstrated by selective spinal cord arteriography, over 80% of spinal cord arteriovenous malformations (AVM's) occupy a predominantly extramedullary position. Current therapy frequently requires surgical stripping of the long dorsal intradural vessel(s) from the underlying spinal cord over many cord segments. The authors report six patients with a dural arteriovenous fistula fed by a cluster of abnormal epidural arteries. These vessels, which surrounded and were embedded into the dural covering of a thoracic nerve root, drained into a long sinuous intrathecal paramedullary vein(s). The angiographic and surgical appearance of the intradural component of these lesions was identical to that of lesions previously classified as Type I AVM's of the spinal cord. All patients had symptoms and signs of myelopathy. In five patients, surgery was limited to coagulation and excision of the extradural vessels and division of the intradural arterIALIZED vein. Progressive improvement began within days following surgery. No residual abnormality was demonstrated by postoperative selective spinal cord arteriography, which was performed in all five patients.

The findings support those of Kendall and Logue, that surgery restricted to elimination of the arteriovenous fistula at the intervertebral foramen is curative, and that more extensive surgery is unnecessary for this subgroup of AVM's of the spinal cord. These lesions comprise a sizable percent of all spinal AVM's. Resolution of myelopathy in these patients supports the hypothesis that venous hypertension causes chronic progressive myelopathy.

KEY WORDS • spinal cord • arteriovenous malformation • dural arteriovenous fistula • myelopathy

In 1914, Elsberg performed the first successful operation for a spinal cord arteriovenous malformation (AVM). The patient presented with severe paraparesis and sensory loss of the right side to the level of the T-9 dermatome. At surgery, a 2-cm segment of a large posterior vein traversing the dura adjacent to the T-8 nerve root was ligated at the dural opening and excised. The patient recovered complete neurological function within 3 months following the procedure. However, arteriography of the spinal cord was not to be available for another 50 years; thus, Elsberg was denied the assessment of the site of the arteriovenous communications(s) and of the afferent and efferent vascular channels of his patient's malformation.

The development of selective spinal arteriography in the early 1960's allowed precise assessment of the radiographic anatomy of these lesions, and their subsequent classification into juvenile, glomus, and a third and most common type. The lesions of the last type are usually entirely extramedullary and lie on the dorsal surface of the spinal cord, displaying slow flow and relatively low intravascular pressure. This type has been designated the "single coiled-vessel malformation," the "long dorsal arteriovenous malformation," "type I spinal arteriovenous malformation," and "angioma racemosum venosum" by various authors. In these malformations the area of transition from arterial to venous elements has not been clearly identified, although multiple communicating vessels between the dorsolateral arterial plexus of the spinal cord and the malformation have been considered as likely shunting sites. Surgical stripping of the "single coiled-vessel malformation" from the underlying pia and spinal cord, often requiring laminectomy and intra-arachnoid dissection over many cord segments, has been the surgical treatment most frequently employed. The reports of Kendall and...
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Fig. 1. Case 1. A: Myelographic serpentine filling defect from T-2 to T-8. An extradural mass at T5–6 indents the lateral margin of the dye column (arrow). B: Selective injection of the left fifth intercostal artery fills an extradural cluster of vessels within the T5–6 intervertebral foramen (arrows) and demonstrates early filling of the paramedullary coiled vessels from T-8 to T-2. C: Postoperative arteriogram fails to opacify the cluster of vessels within the intervertebral foramen (arrowheads) or the abnormal intradural vessels.

Logue and Merland, et al., suggest that most of these extramedullary “malformations” are arterialized extradural veins emerging from an extradural or dural arteriovenous (AV) fistula in the vicinity of a nerve root as it exits the dura.

In the past 23 months we have treated six patients, aged between 29 and 74 years, with a dural AV fistula fed by a cluster of abnormal extradural arteries which surrounded the dural covering of a thoracic nerve root and drained into a long sinuous intrathecal arterialized vein or veins. Five patients were treated surgically by coagulating and excising the cluster of abnormal epidural and dural vessels and dividing the intradural arterialized vein. All experienced progressive improvement of neurological function beginning within days after surgery. Our recent experience, and that of others suggests that the lesions previously considered to be extramedullary spinal AVM’s are caused by a dural AV fistula, and that the component previously designated as the AVM is actually the arterialized venous drainage of the AV fistula. To simplify the description of the following five cases and the ensuing discussion, we will use the term “paramedullary coiled vessel” to indicate the dilated serpentine vessel which lies on the surface of the spinal cord in these patients.

Case Reports

Case 1

This 56-year-old retired male restaurant owner developed impotence 7 years prior to admission. One year later he experienced urinary hesitancy, constipation, and burning dysesthesias of his right leg. The following year his left leg became weak and he began to limp. These symptoms gradually worsened and right-leg weakness and urinary dribbling began. On admission to the National Institutes of Health (NIH) Clinical Center, on December 29, 1980, the patient could walk only three paces.

Examination. Neurological examination showed that he had spastic paraparesis, hypalgesia below T-6, and hyperactive stretch reflexes in both legs. Bilateral plantar extensor responses were present, whereas abdominal and cremasteric reflexes were absent. Anal sphincter tone was normal. There was no spinal bruit or cutaneous hemangioma.

Myelography revealed a long coiled filling defect extending from T-2 to T-8. An epidural mass indented the lateral margin of the dye column at the level of the left T5–6 intervertebral foramen (Fig. 1A). Selective spinal cord arteriography showed a cluster of abnormal vessels in the left T5–6 intervertebral foramen (Fig. 1B) with contrast medium draining superiorly into a paramedullary coiled vessel lying ventral to the spinal cord. The dilated serpentine vessels extended from T-8 to T-1, where the contrast medium emptied into the dilated anterior median spinal vein, which could be followed superiorly as high as the foramen magnum.

Operation. Laminectomy of T4–6 and left T5–6 foraminotomy exposed the cluster of arteries surrounding the T-5 nerve root sleeve in the epidural space. These epidural arteries entered the dura adjacent to the T-5 nerve root, converged into a single vessel, and then traversed the subarachnoid space to drain into the paramedullary coiled vessel on the ventral surface of the spinal cord. The epidural cluster of vessels was coagulated and excised. The segment of the epidural
arterialized vein between the dural AV fistula and the ventrally located paramedullary coiled vessel was coagulated with bipolar forceps, and removed.

Histological examination of the excised vessels showed great variation in their wall thickness. Specific staining demonstrated no elastin in the walls of the intradural vessels, which were identified as arterialized veins.

Postoperative Course. The patient experienced rapid improvement of lower-extremity strength following surgery. Two weeks after the operation he walked 25 yards and climbed 10 steps before tiring. Postoperative spinal cord arteriography failed to opacify the cluster of vessels within the intervertebral foramen or the intradural vasculature (Fig. 1C). When seen 1 year later, the patient walked without assistance, but with a slight limp of the left leg. He had regained full bladder control but impotence persisted.

Case 2

This 29-year-old man complained of episodic right hip pain radiating into his thigh posteriorly. The first pain was noticed 2 years before consultation and became more severe in the few months prior to admission. He described it as an "electric shock" sensation that was elicited by prolonged sitting, rising from a sitting position, or walking long distances, and was relieved after several minutes by lying or standing. He occasionally experienced numbness and tingling in the same distribution. There were no symptoms of bladder or sexual dysfunction.

Examination. Neurological examination disclosed moderate spasticity at the knees, hyperactive stretch reflexes in the lower extremities, and an extensor plantar response on the left. Power, sensation, and gait were normal. There was no spinal bruit or cutaneous hemangioma.

A myelogram demonstrated tortuous abnormal vessels from the L4–5 interspace to the upper thoracic level. Dilution of the metrizamide at the upper level prevented assessment of the cervical area. Digital subtraction arteriography (DSA) with injection of contrast material into the aortic arch displayed prominent filling of enlarged sinuous spinal veins which drained both superiorly and inferiorly with centrifugal flow from a nidus at the T-6 level (Fig. 2A). These abnormal vessels continued to fill at the upper and lower limits of the image (C8–T10).

Selective spinal cord arteriography displayed feeding arteries from the right sixth and seventh intercostal arteries. Conglomerates of abnormal vessels located within the intervertebral foramina of T6–7 and T7–8 could be seen adjacent to the spinal canal (Fig. 2B and C). The drainage of the two clusters of vessels converged at T-6 to empty into the paramedullary coiled vessels, which covered both the ventral and the dorsal surfaces of the spinal cord and extended cranially and caudally the entire length of the visualized spinal cord (Fig. 2D).

Operation. Laminectomy of T5–8 and foraminotomy at T6–7 and T7–8 exposed the conglomerates of epidural vessels surrounding the dural covering of the T-6 and T-7 nerve roots (Fig. 3 left). Intradurally at the level of the pedicle of T-6, a single vessel, which ap-

FIG. 2. Case 2. A: Digital subtraction arteriogram demonstrates feeding arteries from the T-6 and T-7 intercostal arteries (arrows) filling the intradural coiled vessels (arrowheads). B and C: Selective injection of the right sixth (B) and seventh (C) intercostal arteries opacifies the clusters of abnormal extradural arteries within the intervertebral foramina of T6–7 and T7–8 (arrows) and fills the extensive complex of extramedullary vessels. D: Lateral view shows the paramedullary coiled vessels covering the ventral (small arrows) and dorsal surfaces of the spinal cord (large arrows) over an extensive length.
FIG. 3. Case 2. Operative photographs taken after removal of the laminae of T5–8 and dural incision. Left: The extradural conglomerates of abnormal arteries (arrows) adjacent to the spinal dura surround the nerve root sleeves of T-6 and T-7. The cloudy arachnoid covers the dilated serpentine extramedullary vessels. Right: View after removing the arachnoid. The arrow indicates the site of dural entry of the arterialized vein which fills the grossly distended coronal venous plexus of the spinal cord. The T-6 nerve root (arrowheads) penetrates the dura just cephalad to this site. Forceps grasp the dural margin.

The extradural conglomerates of abnormal arteries (arrows) adjacent to the spinal dura surround the nerve root sleeves of T-6 and T-7. The cloudy arachnoid covers the dilated serpentine extramedullary vessels. View after removing the arachnoid. The arrow indicates the site of dural entry of the arterialized vein which fills the grossly distended coronal venous plexus of the spinal cord. The T-6 nerve root (arrowheads) penetrates the dura just cephalad to this site. Forceps grasp the dural margin.

Postoperative Course. Following surgery, the pain in the right leg did not return. The patient had a paresthetic sensation in his left leg immediately postoperatively which has gradually resolved. Postoperative aortic DSA and selective spinal cord arteriography did not opacify the abnormal epidural or intradural vessels. He returned to his work as a drill press operator 3 months following the operation.

Case 3

This 56-year-old man developed urinary hesitancy and left leg weakness 5 months before admission. His condition remained stable for several months and he was able to walk limited distances but was no longer able to bowl or play tennis. His neurological deficit worsened 2 weeks preceding admission. He could then walk only a distance of several feet with the help of a cane. The perineum became numb and he developed urinary retention requiring placement of a bladder catheter. Myelography disclosed a long serpentine filling defect throughout the lumbar and the lower thoracic subarachnoid space.

Examination. Neurological examination revealed weakness of the left leg involving the proximal and distal muscle groups. Prominent spasticity to passive flexion was present in both knees and there was sustained clonus of the left ankle. Sensation to pinprick and cold was blunted below the L-4 dermatomal level. A left Babinski response was present. Muscle stretch reflexes were hyperactive at the knees and left ankle. The patient walked slowly with a wide-based and unsteady gait. There was no spinal bruit or cutaneous hemangioma.

Selective spinal arteriography displayed a cluster of abnormal vessels lying just lateral to the spinal canal in the T9-10 intervertebral foramen. These vessels were fed from the ninth right intercostal artery (Fig. 4).

Operation. Laminectomy of T-9 and T-10 and for-
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FIG. 4. Case 3. Selective injection of the right ninth intercostal artery fills a group of abnormal arteries in the intervertebral foramen (arrows) which drain into an elongated sinuous early-filling vessel in an intradural extramedullary location.

aminotomy at T9-10 exposed a cluster of epidural arteries surrounding the T-9 nerve root (Fig. 5 left). These vessels entered the dura where they converged to leave the inner surface of the dura as a single arterialized vein which drained the dural AV fistula into a paramedullary coiled vessel on the dorsal cord surface. The arterialized vein originated 89 to 2 mm superior to the dural entry of the sensory root of T-9 (Fig. 5 right). When this vein was cut between ligatures as it entered the subarachnoid space, the red color of the engorged vein changed to blue-gray, and the turgor of the vessel diminished but the vein did not collapse. The epidural vessels were then coagulated with bipolar forceps and transected. To ensure complete interruption of the arterial supply to the dural AV fistula, the nerve root was transected and several abnormal epidural arteries ventral to it were coagulated and divided. The dural fistula was coagulated after interruption of the entry and exit vessels.

Postoperative Course. Following surgery, the patient's neurological deficit improved daily. Within 2 weeks his strength returned to normal, the numbness of the left leg and perineum disappeared, he walked without difficulty, and urinary sphincter function returned. Postoperative DSA and selective spinal cord arteriography did not opacify the previously demonstrated abnormality. When seen 3 months postoperatively, he could climb five flights of stairs eight to 10 times per day and had normal sensory function, although paresthesias of the perineum persisted.

Case 4

This 74-year-old male lawyer suffered difficulty walking and numbness in the dorsum of his feet 3 years before admission. One year later he noted urinary dribbling, perineal numbness, and increasing leg weakness. His illness was attributed to diabetic neuropathy. He was admitted to the NIH Clinical Center 6 weeks after becoming wheelchair-bound.

Examination. Physical examination disclosed no cutaneous hemangioma or spinal bruit. Neurological abnormality was limited to lower extremity and sacral autonomic dysfunction. There was severe weakness affecting all muscle groups of the lower extremities. Sensation of pinprick and cold was absent below T-9; vibratory and proprioceptive sensation was absent in the feet. Muscle stretch reflexes were absent in the lower extremities and abdominal and lower extremity cutaneous reflexes were absent. He had no anal sphincter tone. The bladder contained 400 ml residual urine. To control overflow incontinence, he used a penile clamp.

Myelography showed a serpentine filling defect from T-10 to L-2. Selective spinal cord arteriography was performed with demonstration of a cluster of epidural vessels in the intervertebral foramen at L1-2. The draining channel was a large intradural early-filling vein lying on the posterior surface of the spinal cord from L-2 to T-10 (Fig. 6).

Operation. A T-10 to L-2 laminectomy and midline dural opening exposed a large thin-walled vessel which had the appearance of an arterialized vein and which pierced the dura of the root sleeve at L1-2. Foraminotomy at this level uncovered an extradural cluster of wormlike vessels surrounding the root sleeve. Both the extradural vessels and a segment of the paramedullary coiled vessel from L-1 to the upper level of T-10 were removed. Histological examination showed the intradural vessel to be consistent with an arterialized vein.

Postoperative Course. Repeat spinal cord arteriography, performed 6 months postoperatively, showed no residual filling of the abnormal extradural or intradural vessels. Over the first 6 months postoperatively, the strength of the patient's legs improved slightly, but he has remained severely paraparetic and incontinent of urine. There has been no further improvement over a 3-year postoperative period.

Case 5

This 61-year-old male dentist suffered the onset of paresthesias and weakness of his lower extremities in
1963. His weakness gradually progressed over the next 3 years to moderately severe paraparesis, requiring him to use crutches. A myelogram demonstrated an elongated serpentine filling defect extending from T-8 to L-3. He underwent a decompressive laminectomy, at which the diagnosis of spinal cord AVM was confirmed. However, his neurological condition continued to deteriorate, he developed bowel and bladder incontinence, and 7 months following the laminectomy he was admitted to the NIH Clinical Center (in 1966). Selective spinal cord arteriography demonstrated a large feeding vessel from the right eighth intercostal artery which filled what was then considered a Type I spinal cord AVM lying exclusively along the posterior cord surface (Fig. 7 left). At surgery, the feeding vessel was clamped intradurally. His neurological condition improved dramatically over the subsequent weeks and by 8 months after surgery, he was practicing dentistry, playing 18 holes of golf frequently, and running.

The patient’s condition remained stable until 1979, when he noted urinary frequency and occasional incontinence. In 1981, he began to drag his right foot and his gait became unsteady. These symptoms gradually progressed, and he was readmitted to the NIH Clinical Center in 1982.

**Examination.** At examination, there was no spinal bruit. The strength of his right lower extremity was mildly diminished. There was clasp-knife spasticity at the knees and unsustained clonus of the right ankle. Sensation of position, movement, and vibration was absent in the feet. The right plantar response was extensor. He walked with a wide-based unsteady gait, dragging the right foot. Epicritic sensation was normal.

Selective spinal cord arteriography opacified a minute cluster of vessels in the right T8–9 intervertebral foramen which drained into small paramedullary coiled vessels (Fig. 7 right). Reevaluation of his first spinal cord arteriogram (performed preoperatively in 1966) also revealed the epidural cluster of vessels (Fig. 7 left, arrows) emptying into a vein which coursed horizontally across the subdural space where it joined the long coiled vein on the dorsal surface of the spinal cord. These relationships had not been appreciated in 1966.

**Operation.** At surgery, the right T8–9 intervertebral foramen was explored after drilling away the bone.

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**Fig. 5.** Case 3. Operative photographs after removing the laminae of T-9 and T-10 and opening the dura. *Left:* Extra- and intradural abnormal vessels at the level of the T9–10 intervertebral foramen. Forceps grasp the dural edge that runs vertically the length of the photograph. Note the abnormal cluster of wormlike arteries lying extradurally (right) and the single dilated arterialized vein (arrow) draining intradurally. *Right:* The dura is retracted laterally revealing the relationship of the nerve root and the dural penetration of the arterialized vein which is typical of these lesions. Note the normal radicular vein on the nerve root.
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overlying the dorsal boundary of the foramen. The dural covering of the ganglion and the anterior and posterior roots as they joined the spinal dura were examined by microsurgical dissection. The spinal ramus of the intercostal artery entered the dura at the medial and ventral margin of the ganglion. It was somewhat larger than normal but its topography was normal. Multiple tortuous vermian vessels were found in the epidural areolar tissue surrounding the nerve root at its junction with the spinal dura which entered at this level. After opening the dura and dissecting the cloudy white arachnoid from the intradural vessels, a single vessel entering the dura 1 1/2 to 2 mm caudal to the level of exit of the T-8 nerve root fibers was exposed. The dorsal surface of the cord was covered by large collapsed white veins. The Olivecrona clip used to clamp the “feeding artery” 17 years previously was located 2 cm from the dural entry of the vessel draining the AV fistula. Between the dura and the clip, the vessel diverged into tortuous red-colored veins on the dorsal surface of the cord and intermingled with the larger collapsed white vessels. It appeared that the vessel drain-

Fig. 6. Case 4. Selective injection of the right first lumbar artery opacifies an extradural cluster of vessels within the L1-2 intervertebral foramen (arrows) which drains into a dilated tortuous intradural vessel.

Postoperative Course. The patient’s immediate postoperative neurological examination was unaltered from his preoperative status. However, his gait began improving and he stopped using a cane within 1 month after surgery. He returned to work 6 weeks following the operation. At that time, neurological examination showed improvement in gait and balance and increased power in the right lower extremity. There were no episodes of urinary incontinence postoperatively.

Case 6

This 60-year-old male dock worker developed weakness of his legs 1 year before admission. Five months later, he stopped working due to progressive weakness and frequent falls. Two months before admission to the NIH Clinical Center, he developed bowel and bladder incontinence and was admitted to a local hospital. Myelography demonstrated tortuous abnormal filling defects from the mid-thoracic to the mid-lumbar level. He was transferred to NIH after a diagnosis of a spinal cord AVM was made.

Examination. Except for his weight of 285 lb, the patient’s general physical examination was normal. There was no cutaneous hemangioma or spinal bruit. Neurological testing disclosed mild symmetrical weakness of his lower extremities with spasticity at the knees and clonus of the left ankle. Sensory examination revealed absent vibratory sensation in the legs and feet; pinprick and cold sensation were absent below the L-1 dermatome. He walked in a waddling manner with shuffling feet.

Digital subtraction arteriography was attempted, but the x-ray beam would not penetrate his massive torso. Efforts to demonstrate the AVM by selective spinal cord arteriography were unsuccessful on two occasions.

Operations. Because of the recent acceleration of the patient’s neurological deterioration, we performed surgery in an attempt to find and interrupt the arterial supply to the AVM. After a laminectomy of T9–12, the dura was opened exposing tortuous thin-walled red vessels covering both the ventral and dorsal surfaces of the spinal cord. The laminectomy was extended in stages to include T-5 to L-3. The area of dural penetration of each nerve root from T-5 to L-2 was examined by microsurgical dissection, but no vessel was found entering the paramedullary coiled vessels. No vessels of normal venous coloration were seen on the dorsal or
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Fig. 7. Case 5. Left: Selective injection of the right eighth intercostal artery performed in 1966 before surgical clamping of the intradural feeding vessel. The group of abnormal epidural arteries (arrows) was not recognized at that time. Right: Selective injection of the right eighth intercostal artery in 1983 shows filling of a small clump of epidural vessels (left arrows) which empty intradurally to opacify small paramedullary coiled vessels (right arrows).

lateral surfaces of the spinal cord over the 11 vertebral levels examined. A normal-caliber artery of Adamkiewicz was identified entering the dura with the left T-9 nerve root. A 3- to 4-mm thin-walled vessel carrying red blood was found ventral to the conus medullaris and followed caudally among the roots of the cauda equina to continue its path below the lowest level of exposure at L-3.

Three days postoperatively, the patient's legs became weaker and he developed urinary retention and bowel incontinence. At repeat selective spinal cord arteriography, contrast injection into the left L-3 artery revealed a small cluster of abnormal vessels in the L3-4 intervertebral foramen which emptied into a dilated abnormal intradural vessel. Flow of contrast material through this vessel eventually filled the dilated coiled paramedullary vessels extending from T-5 to L-5. Clearance of the contrast medium from the vascular lesion was quite slow.

At reoperation, a laminectomy of L-4 and foraminotomy of L3-4 exposed a cluster of abnormal vessels surrounding the L-3 nerve root. The spinal ramus of the intercostal artery, which was $1\frac{1}{2}$ to 2 mm in diameter, supplied the epidural vascular cluster and then entered the dura, covering the nerve root ventrally. After the dura was opened, a thin-walled vessel, 3 to 4 mm in diameter, carrying red blood was disclosed traversing the dura with the L-3 nerve root. It coursed upward in the same direction as the nerve root for 2 to 3 cm and then began its serpentine course among the roots of the cauda equina. This vessel was divided between ligatures at its dural entry, and the epidural vessels surrounding the L-3 nerve root were coagulated, dissected from the dura, and removed.

Postoperative Course. Within days postoperatively, the patient began regaining power in the lower extremities, and sensation to pinprick, cold, position, and vibratory testing improved. When evaluated 6 weeks postoperatively, he was walking with the help of a cane, and no longer suffered bladder or bowel incontinence.

Discussion

Following Elsberg's initial success, most early reports on spinal cord AVM's have concluded that the vascular constituents are predominantly venous. In his 1925 review of the literature, Sargent considered all but two of the 21 reported cases to be venous angiomas. That most spinal angiomas are venous was also concluded by Wyburn-Mason in his landmark monograph of 1943. Based upon a review of 80 cases from the literature and 30 of his own cases, he distinguished two main types of angiomas: an AV type and a purely venous type. The venous type, angioma racemosum venosum, he considered to be an abnormal mass of sinuous turgid blue pial veins on the posterior cord surface below the mid-thoracic region. This type made up 75 of the 110 cases in his report.

Selective spinal cord arteriography demonstrates that 80% of spinal AVM's are located on the dorsal cord
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surface with most occupying an entirely extramedullary position. The large majority occur in the middle and lower thoracic or the thoracolumbar spinal cord of adults. They extend an average of four to five cord segments, but may cover the entire length of the spinal cord and the cauda equina. In 1967, Di Chiro, et al., described the typical arteriographic appearance of spinal cord AVM's. This extramedullary “AVM” is generally composed of a single coiled vessel which is fed by one or two arteries that have an anatomically normal origin, course, and point of penetration of the vertebral canal. The arterial supply to the “malformation” is almost always distinct from the blood supply to the spinal cord. The circulation through the coiled vessel is slow, often requiring 16 to 20 seconds for clearance of the contrast medium. Until recently, the locus of the AV fistula has been somewhat enigmatic.

In 1977, Kendall and Logue reported their experience with a previously unrecognized group of extramedullary AVM's. They described nine patients with dural AVM's in which the efferent blood drained into the intradural spinal veins and elicited chronic myelopathy. Their patients were treated by excision of the epidural angioma and intradural interruption of the arterIALIZED vein connecting the angioma to the posterior venous plexus of the spinal cord. Seven of the eight patients improved. In 1980, Merland and co-workers reported an additional series of 13 patients with “intraspinal, extramedullary, radiculo-meningeal arteriovenous fistulae” draining into the intradural spinal veins. Eight of 10 patients treated by embolic interruption of the AV fistula improved.

Although in both reports the authors indicate that these lesions represent a large fraction of the retro-medullary AVM’s, many neurosurgeons, neurologists, and radiologists remain unaware of this subgroup of spinal AVM’s. In all six patients with extramedullary spinal AVM’s that we have treated since becoming aware of this type of spinal vascular abnormality, a dural AV fistula caused the engorged tortuous intradural vessels (the paramedullary coiled vessel).

In the arteriographic pattern characteristic of these lesions, the feeding vessel(s) disperses into a cluster of abnormal-appearing arteries adjacent to the spinal dura and within the intervertebral foramen. The transition from artery to vein (the AV fistula) can usually be recognized at the medial margin of this cluster of abnormal vessels. Classic intradural AVM’s, on the other hand, are always supplied by radicular or medullary arteries which typically take a hairpin turn after they enter the dura to proceed to their inosculation into the anterior or one of the posterolateral spinal arteries (Fig. 8). We have not seen this hairpin turn in an artery supplying the lesions we are reporting here. This is also consistent with an arterial supply of these AV fistulas by dural branches of the primary spinal ramus of the intercostal artery rather than radicular or medullary arteries.

Fig. 8. Selective spinal cord arteriogram demonstrating the hairpin turn (arrows) of the feeding vessel supplying a true intradural arteriovenous malformation, glomus type. We have not seen feeding vessels of this type in the patients comprising this report.

In two patients, aortic DSA demonstrated the approximate level of the AV fistula and early filling of the tortuous dilated spinal veins. This study permitted initiation of the selective spinal cord arteriogram in the general region of the fistula, and limited the longitudinal extent of the multiple selective catheterizations required. The epidural cluster of vessels in the interver-

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tebral foramen could not be clearly seen by DSA, however, and therefore this study alone does not allow one to confidently distinguish a spinal dural AV fistula draining intradurally from a true spinal cord AVM. 7

Currently, many surgeons treat extramedullary spinal “AVM’s” by intradural exposure and dissection of the entire length of the coiled extramedullary arterialized vein(s) from the underlying spinal cord by division of adherent pial and arachnoid fibers and interruption of tributary vessels emptying directly from the spinal cord. In many cases, this requires intra-arachnoid dissection over many cord segments, sometimes extending much of the length of the vertebral column. In patients with dural AV fistulas, the success of this approach depends upon including in the length of resected extramedullary vein the segment of arterialized vein that courses intradurally from the AV fistula and the venous drainage of the spinal cord. Moreover, this stripping of the extramedullary vein from the spinal cord also interrupts the drainage of intramedullary veins and coronal veins which carry the venous effluent from the spinal cord parenchyma. Considering that the radial veins have no anastomotic system within the cord tissue, 9 the patient treated by stripping the paramedullary coiled vessel from the spinal cord may be left with postoperative neurological deficits resulting from interruption of the normal venous drainage of the spinal cord.

An additional problem resulting from failure to recognize the dural AV fistula is that extramedullary “AVM’s” lying anterior to the spinal cord are often considered inoperable because they cannot adequately be exposed for surgical stripping. Recognition that the intradural extramedullary spinal venous abnormality is a secondary phenomenon and does not require resection allows therapy to be appropriately directed to the fistula alone in these patients.

The paramedullary coiled vessel is caused by drainage from the dural AV fistula flowing directly into the coronal venous plexus via a bridging vessel. Long-standing increased blood flow and higher intravascular pressure within the coronal plexus alter its configuration by causing elongation and dilatation of the component vessels (Fig. 9). Previous reports describing extraspinal AV fistulas that empty into the intrathecal venous system and produce arteriographic and myelographic appearances identical to those produced by Type I spinal cord AVM’s demonstrate that this acquired transfiguration of the coronal plexus can occur. 4,10,14

The bridging vessel which courses from the inner

![Diagram of normal and pathological conformation of coronal venous plexus](image)

**Fig. 9.** Schematic representation of normal vascular anatomy (left) and pathological conformation of coronal venous plexus resulting from a dural arteriovenous (AV) fistula (right) of the spinal cord, posterior view. Insets depict the normal course of the medullary vein draining the coronal plexus and a typical vessel draining the dural AV fistula intradurally into the coronal venous plexus. The dural branch of the radiculo-medullary-dural artery is not shown.
Dural fistulas in spinal AVM's

The epidural vessels often require transection of the epidural vessels, which in the cases reported here, it was found at surgery to be topographically identical with a medullary vein; the vessel always traversed the dura 1 1/2 to 3 mm rostral or caudal to the site that a nerve root pierced the dura, and its relationship to the nerve root was consistently like that of a medullary vein (it ran close to a nerve root and in the same general direction, but was always separate from the radicular vessel of the nerve root). The feeding artery of the dural AV fistula seems to arise from the dural branch of the radiculo-medullary-dural artery.

The myelopathy associated with spinal AVM's was attributed, by Aminoff, et al., to raised venous pressure; those authors contended that the flow from the AVM into the coronal venous plexus causes dilatation, congestion, and stagnation in the intramedullary veins. The subsequent recognition by Kendall and Logue, that dural AV fistulas cause neurological deterioration in these patients by arterIALIZATION of the vessels of the coronal plexus and venous hypertension led them to advocate removal of the AV fistula and interruption of the arterialized vein close to the AV fistula. Merland, et al., subsequently reported their detailed studies of the anatomy of these lesions at the intervertebral foramen by spinal angiotomography. They advocated embolizing the AV fistula with isobutyl cyanoacrylate (IBCA). Most of their patients (eight of nine cases) responded to embolic interruption of the fistula with improvement (occasionally dramatic) beginning within hours to days after treatment. However, one patient became paraplegic after embolization. They attributed this to a disturbance of medullary venous flow. Embolization was considered contraindicated in patients whose AV fistula was supplied by an artery which also contributed arterial supply to the spinal cord. Additional potential sources of complications exist with this therapy. If the IBCA does not polymerize until it flows beyond the AV fistula, it could occlude a segment of the arterialized coronal plexus, resulting in spinal cord dysfunction.

In the patients whom we have treated surgically, the epidural cluster(s) of wormlike arteries which surrounds the dura of the proximal spinal nerve and the intradural vein which links the dural AV fistula to the paramedullary coiled vessel have been quite easily exposed and identified. Interruption of the draining vein, coagulation and excision of the epidural conglomerate, and obliteration of the dural fistulous tract by bipolar coagulation is then a simple task. The complete obliteration of the epidural vessels often requires transection of the nerve root. In our patients, this was a thoracic nerve root and no symptoms or discernible sensory deficit could be attributed to this. Logue \( ^{10,11} \) has recommended excision of the dura covering the involved nerve root and removal of the AV fistula in continuity with a segment of the intradural vein issuing from the fistula. This often requires a dural graft. Either technique accomplishes the therapeutic goal, which is interruption of the arterial flow into the spinal veins and obliteration of the AV fistula. However, stripping the paramedullary coiled vessel from the spinal cord leaves the dural fistula intact and may permit recurrence of symptoms if the venous effluent again finds a path to drain intradurally, as occurred in our Case 5. These lesions require that the surgeon direct his attention and efforts to the epidural and dural components, in addition to the limited intradural dissection required.

Summary

Our recent experience with extramedullary spinal arteriovenous malformations demonstrates that many are actually dural AV fistulas which cause spinal cord dysfunction by draining intradurally and transmitting elevated venous pressure to the cord tissue. They are successfully treated by surgery limited to the area of the intervertebral foramen, and intradural dissection limited to the region of the AV fistula, and do not require stripping of the arterialized extramedullary vein from the spinal cord, which may be contraindicated.

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References


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