SPINAL EPIDURAL ARTERIOVENOUS MALFORMATIONS:
REPORT OF A CASE WITH DISCUSSION OF CLASSIFICATION AND TREATMENT
Caitlin M. Clark, B.A. and W. Craig Clark, M.D., Ph.D., FAANS, FACS, FICS

INTRODUCTION

True spinal epidural Arteriovenous Malformations (AVMs) are exceedingly rare abnormalities of the vasculature, with fewer than 100 cases reported in the literature. It should be noted that these are not to be confused with dural AV fistulas, and are not included in most common classification schemes. With both lesions, arteriovenous shunting occurs, and there is direct communication between the venous and arterial systems. AVMs are collections of dysplastic vessels supplied by feeding arteries and drained by venous channels, while AVFs consist of a feeder artery and draining vein without a nidus between them.[2]AVFs can be either intradural or extradural, while AVMs have nidi that are either intramedullary (glomus), or more extensive (juvenile or metameric).[5] Of all the different spinal vascular malformations, 15-20% are extradural, but most are cavernous hemangiomas and rarely are AVMs.[1]Patients often present with what is known as a syndrome of medullary venous hypertension[2], in which they experience distal to proximal paresthesias or dysesthesias, progressive weakness ranging from giveway to paraparesis, and sphincter dysfunction.[2]As a result of this lesion's rarity, treatment remains obscure, with presentation and treatment results only reported on a case-by-case basis with little ability to extrapolate or generalize the results. However, differentiating between an AVM and an AVF is extremely important, as management will change depending upon which type of lesion it is, regardless of further classification.With extradural AVMs or an AVF with intradural draining vein, one must obliterate the extradural component, as that is at least as important as simply disconnecting the intradural drainage alone. This added component makes these lesions very difficult to treat surgically or endovascularly.[8]The current case is described and used as a basis for review of the history of spinal AVMs, the most common classification schemes and suggested management strategies for each type of spinal vascular lesion.

CASE REPORT

Our patient is a 48 year old African-American male who presented to clinic with 4 months of back and right flank pain. Thoracic spine MRI showed an 8x8x15 mm epidural mass at T3-T4 that was hypointense, with no enhancement or dense calcification, as seen in Figure 1. A laminectomy was performed at T2-T4 with microsurgical resection of an epidural vascular mass, with no evidence of dural penetration. Upon resection, the large, dilated veins over the dorsum of the dura immediately normalized. However, the large veins in the lateral gutters remained dilated at closing. Postoperatively, the patient had no neurological deficits, and his wounds healed uneventfully. A postoperative MRI showed only postoperative changes, and no evidence of any remaining vascular abnormality. The pathology report showed an AVM, and spinal arteriography, including segmentals, was a normal study. About 5 weeks postoperatively, the patient began to develop a T10 sensory level. Repeat MRI showed a resolving small seroma, with no increased signal intensity within the cord, as seen in Figure 2. EMG/NCV performed at that time was a normal study. His symptoms continued to progress, and he developed allodynia in the soles of both feet. Treatment was initiated with gabapentin for his neuropathic-like symptoms. The symptoms remained sensory, with no motor involvement. After 6 months of treatment with gabapentin, he was able to wean off the drug with no sequelae.

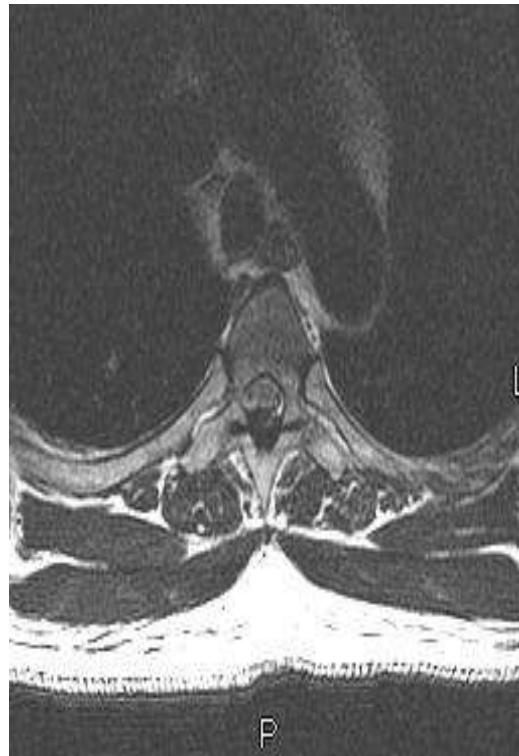Figure 1. Sagittal T2-weighted MRI showing flow void in thoracic region.

Figure 2. Axial T2-weighted MRI showing flow void in thoracic region.

DISCUSSION

Spinal AVMs were first described during an autopsy by Virchow nearly 150 years ago, and the first spinal vascular malformation of any kind was noted during an operation by Harvey Cushing in 1910. Stanley Cobb, his resident, was the first to discuss the association of the AVM with a nidus in the same somite.[5] Spinal epidural AVMs were first discussed in a 1914 case report by Dr. Charles Elsberg, who described a patient with severe spastic paraparesis with a T9 sensory level[4]. During the operation at T9, he noted an abnormal vessel penetrating the dura at T9, and, upon opening the dura, encountered abnormally large veins. After excision of a segment of the abnormal vessel at the dura, the patient recovered to normal over three months. This operation is significant because it was a successful treatment of a spinal epidural AVM, and foreshadowed modern operative treatment of these abnormalities.

Figure 3. Spinal Epidural AVM demonstrating ectatic vein from arterial flow. Reprinted by permission, Elsberg CA, 1910.

This case was followed by the next reported operation, performed by Spiller and Frazier in 1923, who were treating a 25 year old female with spastic paraparesis. They performed a laminectomy at T12 and L1, and noted a vascular lesion extending beyond the surgical exposure in either direction[12]. During the operation, they ligated a vessel thought to be pathological, and the patient was paraplegic postoperatively. This case is important because it and others form the basis for Frazier's recommendation of Laminectomy and Postoperative Irradiation without any attempt to deal with abnormal vessels when encountered.

Figure 4. Aneurysm of posterior spinal vessels. Reprinted with permission, Spiller and Frazier, 1923.

In 1944, Roger Wyburn-Mason identified 96 cases of spinal AVM in the literature at that time. He added 16 new cases to that list, and did not give any insights into treatment. This was the first critical mass of clinical material on spinal AVM, and was important because he distinguished between venous anomalies and arteriovenous malformations[13].Doppman and DiChiro, in 1965 and 1968 respectively, were among the first to report on selective spinal angiography, and the first to focus thinking on the nidus of abnormal vessels being the essential lesion in spinal vascular malformations[3]. They reported using 3mm stainless steel pellets for selective embolization of an arterial feeder to a spinal AVM. They were the first to conceptualize the nidus as the essential lesion of spinal AVM, and were also the first to perform endovascular selective embolization of a spinal AVM. In 1969, Krayenbuhl and Yasargil published their experience with the excision of spinal AVMs using the operating microscope and bipolar cautery[14]. This began the microsurgical era and allowed approaches to vascular lesions not only on the dorsal surface of the cord but also within the cord and on the ventral surface.
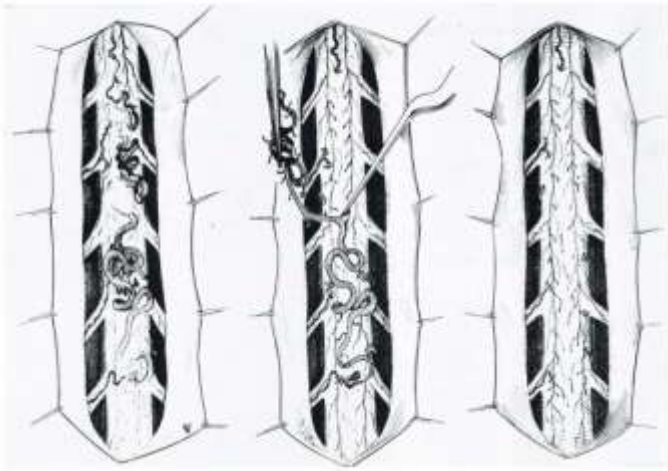
Figure 5. Diagram of the appearances and technique of removal of a dorsal arteriovenous malformation. Reprinted with permission. Yasargil MG, 1969.

Kendall and Logue reported ten cases of spinal AVM in 1977 where the nidus was located in the dura adjacent to a spinal root[7]. Logue excised the duralnidus, and left the abnormal vasculature on the surface of the cord along. Now, it is accepted that most spinal vascular malformations are dural A-V fistulae and are successfully treated by disconnection of the fistula and obliteration of the nidus. In 1983, Oldfield and Quindlen, who were working with Doppman and DiChiro at the NIH, published additional reports and duplicated the initial success[10]. As a result, advances in the treatment of this condition were clearly related to cooperative efforts of neurosurgeons and neuroradiologists. Detwiler and Spetzler developed first a classification scheme for intracranial AVMs, and then in 2002 developed a classification for spinal vascular lesions[11]. This was the first scheme to include and classify the perimedullary lesions like those described by Heros in 1986[6].

CLASSIFICATION OF SPINAL VASCULAR LESIONS

Spinal vascular lesions, as discussed above, were first classified in 2002 by Detwiler and Spetzler[11]. These lesions are divided into three large categories, and then divided from there. They are: spinal tumor vascular processes, spinal aneurysms, and intradural AVMs. Spinal tumor vascular processes include hemangioblastoma and cavernous malformations. Intradural AVMs are further subdivided into Type I-A, I-B, II, III, and IV. Type I are dorsal intradural AV fistulae, which are subdivided into I-A, with a single arterial feeder, and I-B, with multiple arterial feeders. In these Type I lesions, there is usually retrograde venous drainage occurring via the spinal medullary veins, which causes venous hypertension and engorged veins on the surface of the spinal cord, as seen in the figures below.[2]
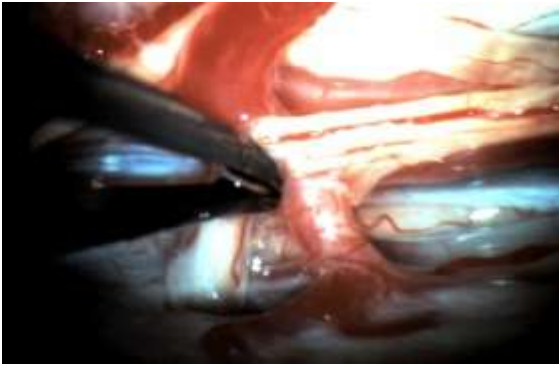
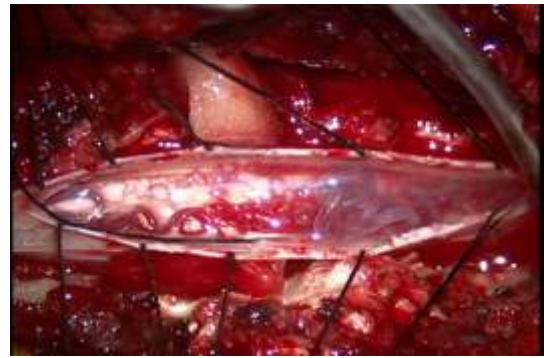Figure 6. AV fistula showing arterialization and nidus.

Figure 7. Ectatic veins showing arterialized flow.

Type II intradural AVMs, also called glomus, are true intramedullary AVMs, a distinct mass of dysmorphic arteries and veins without a capillary bed present.[2]When these lesions are completely resected, there is no hemorrhage; with an incomplete resection, there is no protection from hemorrhage. If an endovascular ablation technique is used, both complete and incomplete ablations resulted in no hemorrhage. There has been a critical mass of 196 cases of glomus extradural AVMs reviewed by Gross. Type III, also called juvenile, are intra or extramedullary, and are very rare congenital lesions. There have been 51 total cases reported of this type of lesion. It is often associated with aneurysms, and there is an association with this lesion between the presence of aneurysm and younger age with hemorrhage. These lesions can involve the spinal cord, as well as the vertebral and paraspinal tissues, giving them the classification of "metameric AVM."[2]Twenty-five of the fifty-one cases have been treated – the others were observed. Eight patients out of the twenty-five treated had an aneurysm. When the aneurysms were ablated, there was no hemorrhage. Type IV intradural AVMs, also called perimedullary AV fistulae, are ventral, intradural direct fistulas between the anterior spinal artery and a large vein. These Type IV lesions are further divided into A, which has insignificant shunting, B, which has moderate shunting, and C, which has significant shunting. There is no large reported experience with these lesions, and treatment usually involves a combined surgical and endovascular approach.Symptomatology with any AVM, including spinal epidural lesions, can be explained by one of three phenomena: the first is a "vascular steal" phenomenon, the second related to venous hypertension and relative ischemia, and the third is related to external compression and mass effect.[9]

CONCLUSION

We report an additional case of the very rare entity of spinal epidural arteriovenous malformation, and use this as an opportunity to review the classification and treatment of spinal vascular lesions. Most common classification schemes do not include true spinal epidural AVMs. Based upon our review of the limited case reports, we conclude that the preferred treatment would be either surgical or endovascular obliteration of the extradural component. It would appear that it is usually only necessary to interrupt any vascular connection to the intradural vasculature.

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