Endovascular management of spinal vascular malformations: history and literature review

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Spinal vascular malformations represent a complex group of heterogeneous pathological entities whose treatment paradigm continually evolves. Given the ever-increasing role of endovascular therapy, it is the goal of the authors to review the current literature regarding this therapeutic tool and to provide recommendations guiding management. A thorough literature search was conducted using Medline, with subsequent articles being identified through cross-referencing. The analysis revealed that, since its introduction in the 1960s, endovascular therapy has been used to manage the entire spectrum of spinal vascular malformations, during which period it has undergone considerable technological and technical evolution. As such, embolization has proved of growing therapeutic utility, largely resulting from the mounting evidence supporting its safety and efficacy, in addition to the inherent minimally invasive nature. This alternative to surgical intervention will be increasingly used as first-line therapy in spinal vascular malformations.


KEY WORDS • arteriovenous fistula • arteriovenous malformation • embolization • endovascular therapy • Onyx • spine

Spinal vascular malformations represent a rare and complex group of heterogeneous pathological entities whose optimal management remains in continuous evolution. Given the ever-increasing role of endovascular therapy, we endeavored to review the current literature regarding this therapeutic approach and to provide recommendations guiding management.

Endovascular Treatment of Spinal Vascular Malformations

Although variable in pathophysiology and clinical presentation, these lesions have been recognized for their considerable attendant morbidity since first described in the late 19th century by Hebold and Gaupp.4,6,7,19,22,34 Given the lesion’s natural history as an impetus for intervention, surgical correction has been attempted since the early 1900s; however, it was Dr. Charles Elsberg who provided the first report of a successful operation in 1914.2,19,34 A 23-year-old man was admitted to Mount Sinai Hospital with pain, weakness, and spasticity in the lower extremities severe enough as to prevent ambulation. At the time of surgery, T5–9 laminectomies were performed, and following durotomy, an enlarged left posterior spinal vein was revealed (Fig. 1). This vein was ligated at the dural opening and a 2-cm segment was excised (Fig. 2). The patient experienced an uneventful recovery and in 3 months was without neurological dysfunction.19

Following this initial success, continued therapeutic endeavors were undertaken, but it required the introduction and subsequent refinement of spinal arteriography by Djindjian,15 Doppman and DiChiro,16 and others2,18,34 to permit progression of knowledge concerning the pathophysiology and subsequent treatment of these lesions. In the presence of this improved comprehension of angioarchitecture, Doppman and colleagues17 generated successive advancements through the addition of percutaneous embolization to the therapeutic armamentarium, treating a 16-year-old boy by embolizing an arterial feeder with steel pellets. Subsequently, angiography, in addition to its position as the gold standard of diagnosis, has experienced a continued technical and technological evolution allowing for its increasing utility in the treatment of spinal vascular malformations, either alone or in combination with surgical intervention.

Nearly every facet of endovascular technology has undergone advancement over the past 40 years. Two principal components of therapeutic enhancement have been the advent of microcatheters and the continued in-
Introduction of novel substances for embolization. The newer microcatheters allow the neurointerventionalist to navigate small and tortuous vasculature with increased safety and to promote canalization of previously inaccessible vascular territories. Concerning the advancement in embolic agents, the use of liquid adhesives such as IBCA and, more recently, NBCA has resulted in substantial improvement in recanalization formerly demonstrated with particles such as organic fragments, silk, and PVA. Additional benefits include adjustable polymerization time and superior vessel permeation; however, despite these advantages, the liquid embolic agents are not without problem, including the risk of catheter adhesion, variability in physical behavior, and most significantly an increased risk of spinal cord infarction. Hopefully the recent application of Onyx, an innovative liquid embolic substance, will provide further improvements in this arena. With its properties, Onyx has additional advantages over NBCA as an embolic agent: it can be delivered slowly in a more controlled fashion over several minutes, carries a lower risk of premature venous occlusion or occlusion of a large intradural portion of the draining vein, and carries a low risk of a retained catheter. Although the experience in spinal vascular malformations has been limited, the results from intracranial application have proved promising.

Management by Classification

The literature regarding endovascular management of spinal vascular malformations is not extensive and is largely composed of case reports and small case series. The subject is further obscured because a plethora of classification systems evolved to categorize these lesions as more recent investigations permitted elucidation of their anatomical and pathophysiological basis. For the purposes of clarity, this review will largely adhere to the classification system proposed by Oldfield and colleagues, which divides these lesions based on their distinct biological characteristics.

Dural Arteriovenous Fistulas

Dural arteriovenous fistulas, also known as Type I lesions, are low-flow lesions composed of an aberrant connection between a radicular artery and medullary vein located in the dural sleeve of the nerve root. The resultant retrograde flow into the coronal venous plexus surrounding the spinal cord results in venous hypertension, chronic venous congestion, and subsequent cord ischemia. Physiological studies have demonstrated venous pressures averaging approximately 75% that of the systemic arterial circulation. These represent the most common form, accounting for 80–85% of all spinal vascular malformations. They typically occur in older adults (male predominance) and in most cases are thought to be acquired. Patients often present with vague complaints of back or radicular pain that frequently leads to significant diagnostic delays; however, this is followed by progressive lower-extremity paresis, sphincter dysfunction, and eventually paraplegia.

Early case series including that published by Morgan and Marsh suffered from the use of PVA particles for embolization. As such, despite safe and complete angiographic obliteration in 12 of 14 patients, over the follow-up course there were 13 recurrences. Subsequent reports after the introduction of liquid embolic materials were associated with improved success. Niimi et al. reported on their experience in treating 49 cases, 38 of which were treated utilizing microcatheter technology. In 33 (87%) of those 38 patients “adequate” angiographic results were obtained, as defined by penetration of the liquid embolic agent (both IBCA and NBCA) into the fistula or draining vein and the disappearance of the fistula or venous drainage by posttreatment angiography. In 3 of the 5 patients for whom this result was not feasible, a common pedicle existed from which the feeder vessel and spinal artery arose. All but one patient experienced symptomatic improvement following treatment, and there were no procedural complications in the group treated with microcatheters. Of the 35 patients in whom “adequate” embolization was achieved (including 2 treated with conventional catheters), 10 experienced symptomatic aggravation, 8 (23%) of whom required treatment for recurrent
DAVF. Two additional patients without evidence of recurrence underwent heparinization for suspected propagation of venous thrombosis and consequently worsening of spinal venous congestion. Niimi et al. concluded that because this procedure could be safely performed, embolization with liquid cyanoacrylate should be the first-line treatment provided that angiography does not demonstrate a common pedicle for both the feeder and spinal artery.

Given the demonstration of equivalent clinical outcomes following fistula obliteration regardless of methodology, other investigators have reached similar conclusions despite a substantial failure rate with embolization. In 1999, Ushikoshi and colleagues reported their experience in the treatment of 13 patients with DAVFs, 6 of whom underwent primary embolization with NBCA. In 4 of 6, complete obliteration of the fistulous connection was achieved, but in 2 this was not possible and subsequent surgical intervention was required. Two patients, 1 of whom had only partial embolization, developed transient worsening of their paraparesis. Symptomatic improvement (3 cases) or stabilization (3 cases) occurred in all patients at the 6-month follow-up. Song et al. retrospectively reviewed their institution’s data over an 8-year period and found that 27 patients were treated for a DAVF. Seventy-four percent (20 patients) had lesions amenable to NBCA-based endovascular intervention, and 90% (18 patients) had complete angiographic occlusion. Over an average follow-up period of 3.1 years (range 1 month–8.9 years) 15% of the patients suffered recurrence of the fistula. Evaluation with the Aminoff-Logue Disability Scale revealed improvement or stability of all patients with regards to gait and of 85% concerning micturition. Emphasizing the importance of cyanoacrylate penetration into the fistula and proximal vein, Guillemin et al. examined the predictive nature of NBCA location on CT scans following embolization. Twenty-six patients were included, in 19 (73%) of whom glue was observed within the dura. All of these patients experienced symptomatic improvement without angiographic evidence of recurrence during a mean follow-up period of 23.4 months. Of those without this CT finding, clinical deterioration in 5 of 7 required surgical intervention as a result of recanalization. Eskandar et al. reported their experiences over a 6-year period, during which time embolization served as first-line therapy for DAVFs. Twenty-six patients were included, of whom 23 (88%) were able to undergo liquid acrylic–based embolization as the primary treatment, with 3 patients undergoing surgery due to the presence of a common pedicle for both the feeding artery and spinal artery. Three complications occurred (acute myocardial infarction, toe ulcer due to distal extremity emboli, and an axillary hematoma), with all other patients experiencing symptomatic improvement or stabilization following the procedure. Nine of the patients (39%) eventually required surgical intervention due to recurrence.

Although the aforementioned results are encouraging, not all series have demonstrated equivalent success. Throughout a 15-year period, van Dijk and colleagues treated 49 patients in whom embolization was the initial therapy whenever feasible. Of the 44 patients who underwent endovascular therapy, only 11 (25%) had an angiographic cure; however, the authors neglected to expand upon the reasons for failure except in 2 cases in which arterial dissection necessitated cessation of treatment. Notably, penetration of NBCA into the proximal vein was required for “successful” intervention, and as such, no instances of recanalization occurred. Steinmetz et al. performed a meta-analysis of the published literature from 1966 to 2003 and, of the 10 studies included, definitive treatment was possible in only 46%. Notably, the use of NBCA was not an inclusion criteria, and the use of PVA and other embolic particles has been thoroughly demonstrated to result in recanalization.

In patients without a common origin for both the feeding artery and spinal artery, embolization with liquid acrylic agents represents a safe initial treatment option, providing durable cure in up to 70–85% of patients when the agent penetrates the proximal draining vein. Nevertheless, given surgery’s established efficacy and durability, it largely remains the gold standard for treatment. Perhaps with the use of Onyx, and with definitive long-term follow-up demonstrating the durability of liquid embolic agents, endovascular therapy may be adopted as first-line therapy. However, for individuals in whom embolization is not possible or when a fistula recurs, surgical intervention will continue to provide definitive management.

Glomus AVMs

Glomus malformations, also known as Type II lesions, are characterized by a compact nidus located completely or partially within the spinal cord. They are generally supplied by multiple feeding vessels from the ASA and posterior spinal artery, with high-flow and low-resistance drainage into medullary veins. In as many as 20–40% of lesions, an associated arterial or venous aneurysm is present. In contrast to DAVFs, they frequently occur in children or young adults without a significant sex predominance. Additionally, instead of progressive decline due to venous hypertension, they are more likely to hemorrhage with deterioration of spinal cord function. Ensuing rehemorrhage occurs at a rate of 10% within 30 days and 40% within the 1st year.

Therapeutic decision making requires consideration of the patient’s medical comorbidities, lesion location, and angioarchitecture, as both microsurgical excision and endovascular embolization carry significant risk of neurological injury. Many of the early series were performed during the period when PVA particles were used, thereby making the results noncomparable with those achieved with modern technology. However, the use of these particles may still have a role, especially when concerning lesions that are inoperable due to location (that is, ventral thoracolumbar lesions) or angioarchitecture, and/or when the nidus cannot be reached with current microcatheter technology. Particulate embolization, in addition to facilitating surgical intervention, reduces the risk of hemorrhage, provides symptomatic improvement, and can delay symptomatic progression. Biondi et al. evaluated the long-term outcomes in 35 patients with thoracic intramedullary AVMs, both glomus and juvenile type. During a follow-up period of up to 15 years, 158 proce-
dures were required to treat recanalization; however, 63% of the patients exhibited clinical improvement compared with baseline findings. Additionally, these procedures were performed with a low incidence of adverse effects for the 23 complications (15%), only 12 (8%) resulted in clinical worsening, all but 2 of which proved transient in nature. Given this success, albeit temporary, some authors have advocated the use of repeated embolization as opposed to surgical intervention for those lesions without characteristics favoring excision.

Corkill et al.13 published the first series to evaluate the efficacy of Onyx for embolization of intramedullary AVMs. The series included 17 patients, 11 of whom harbored glomus AVMs. Complete AVM obliteration was achieved in 5 patients (45%) and subtotal occlusion, defined as insignificant nidal remnant, in another 4 (36%); however, 3 patients required 2 separate treatments. Three complications occurred during the course of endovascular intervention without subsequent neurological sequelae, only 1 of which required cessation of the procedure. Six patients (55%) experienced acute neurological decline; however, all patients recovered to at least baseline status, with 9 (82%) experiencing functional improvement over the course of the follow-up period. Other authors have reported an incidence of permanent neurological deficits of up to 11%, the significance of which depends on whether treatment requires catheterization of the ASA. Nevertheless, these results still compare favorably with the natural history of these lesions.

Concerning the results of surgical intervention, Spetzler and colleagues31 reported a 68% rate of improvement in the treatment of 27 intramedullary AVMs. An additional 29% of the patients experienced symptom stabilization, with 8% suffering from further neurological deficit. Many of these were treated with preoperative embolization, and in 92% complete resection was accomplished. There does exist some ambiguity in the interpretation of these results, as the extent to which juvenile AVMs were included remains unclear.

Either therapeutic modality necessitates performance in centers well versed in its use; however, sufficient literature exists to justify an initial attempt at endovascular management, especially in the setting of deep lesions, those that are ventrally located, or those in patients who are poor candidates for surgery. Surgical intervention can be reserved for superficial lesions or patients in whom embolization fails.

Juvenile AVMs

Also referred to as Type III or metameric vascular malformations, juvenile AVMs are characterized by large, high-flow lesions with involvement of the paraspinal structures. Neurological deficits occur as a result of spinal cord compression, venous congestion, vascular steal phenomenon, and hemorrhage. The typical presentation is pain and progressive myelopathy in children or adolescents.

Of those series addressing sole endovascular management, many procedures were performed using PVA particles,8,55,56 but this potentially affords clinical stabilization or improvement as described for glomus malformations.8 Additionally, in the series described above by Corkill et al.13 6 patients with juvenile AVMs underwent embolization. Complete and subtotal obliteration was accomplished in 1 patient each, but the patient with initial complete occlusion experienced a recurrence and required further treatment. Fifty percent of the patients improved, 2 experienced symptomatic stabilization, and 1 had neurological worsening that persisted at the time of follow-up. No periprocedural complications occurred.

While the aforementioned discussion provides optimism regarding future therapeutic potential, the optimal management is certainly still best achieved through a multidisciplinary approach. Although reports of improvement and occasional case reports of cure do exist,22 treatment is largely considered palliative in nature.

Perimedullary AVFs

Perimedullary AVFs, or Type IV lesions involve an aberrant connection between the spinal arteries and veins without an intervening vascular network. Typically they are located near the conus medullaris in the ventral subarachnoid space and extend over multiple spinal levels. Given the significant flow present in these structures, aneurysms and ectatic venous malformation are frequent accompaniments.32,51,59 They manifest clinically most commonly in adults with either progressive decline secondary to venous hypertension or acute neurological deterioration as a result of aneurysm rupture.8,59 These lesions have been further subclassified by Merland and colleagues31 into Type I, II, and III lesions based on the size, length, and number of vessels feeding and draining the shunt. Type I perimedullary AVFs are small, low-flow lesions associated with only moderate venous hypertension, often located in the conus or terminal filum and deriving arterial supply from the ASA, whereas Type II perimedullary AVFs represent larger fistulous connections in the pia composed of several distinct shunts that are fed by an engorged ASA or posterior spinal artery. Finally, Type III perimedullary AVFs, the most common form, are giant lesions with multiple dilated feeding arteries and significantly dilated venous drainage from a single fistula.8,23,37,51 Although successful endovascular treatment of Type I malformations has been reported in several small series,11,37,45 these lesions are typically supplied by the ASA and are so small that they prohibit selective catheterization; thus, surgery has widely been accepted as the primary treatment. As such, further discussion will largely be limited to the role of embolization for Type II and III lesions.5,9,25,27,28,31

Halbach et al.25 treated 10 patients with Type II and III malformations (2 and 8 cases, respectively), 3 of whom received embolization alone and 7 of whom underwent combined therapy. After 16 embolizations and 8 operations, an angiographically confirmed cure was present in 7 patients (70%); of the remaining patients, 2 has some residual lesion (< 5% residual) and one refused follow-up. One patient experienced a procedure-related complication in the form of ASA rupture following deployment of a detachable balloon, but, despite transient worsening of the patient’s neurological status, they eventually returned to baseline. Only 1 patient experienced
symptomatic decline following treatment, with 4 others having some improvement and an additional 4 presenting with dramatic improvement. While this demonstrates the benefit of combined intervention, it provides little information regarding the utility of endovascular management as stand-alone therapy. In the same year, Mourier and associates reported their experience in 35 patients with Type I, II, and III lesions (in 4, 9, and 22 cases, respectively) treated both surgically and endovascularly. Five of the patients with Type II malformations underwent endovascularly, one with IBCA and 4 with particulate. In 2 patients an angiographic cure was obtained, with the remainder requiring surgical correction. In all patients with Type III lesions balloon occlusion was used as the primary therapeutic intervention, with complete obliteration being demonstrated in 15. In the other 6 patients only partial relief of the fistula was possible, and in 1 patient acute quadriplegia and subsequent death occurred. All patients in whom complete occlusion was possible exhibited improvement, but in those in whom only partial elimination was achievable, either symptomatic stigmatization (4 cases) or consequent worsening (2 cases) resulted. The difficulties of treating Type II abnormalities are a result of the multiplicity of feeding pedicles; therefore, realizing an endovascular cure remains improbable. In cases of Type III malformations, good clinical outcome was achieved in the majority (86%) of patients, thereby supporting the efficacy of this modality. While 1 death did occur, in addition to clinical deterioration in 2 others, the natural history provides significant impetus for intervention.

Further addressing this area, Ricolfi et al. reported on 12 patients in whom Type III lesions were managed with balloon occlusion or particle embolization. In 10 patients, complete occlusion was achieved, with the other 2 patients having either partial occlusion or no treatment due to difficulties with catheterization. Clinically, 6 (50%) had a good outcome following embolization; however, 2 patients had worsening symptoms, and one of them died within 2 days. Of the remaining 2 patients, 1 had a fair result and 1 had an initially favorable result, but only partial occlusion was accomplished and the patient subsequently experienced subarachnoid hemorrhage and paraplegia. Despite the associated morbidity, given the natural history and significant complications associated with attempted resection of these malformations, the authors continued to recommend endovascular therapy for initial management.

As with all spinal vascular malformations, the natural history provides significant impetus for intervention. Given their small size and sole supply through the ASA, Type I lesions continue to be best managed with surgical correction. Type II abnormalities possess multiple afferent pedicles and, as such, are difficult to cure using endovascular modalities; however, these remain as a therapeutic alternative for patients who are not surgical candidates, in addition to being an adjunct to operative therapy. Type III lesions generally benefit from initial embolization, with surgery reserved for patients in whom this fails.

Rodesch and colleagues published one of the largest clinical series on spinal vascular malformations treated during an 18-year period at the Bicêtre. One-hundred fifty-five patients were evaluated, of whom 69 (45%) underwent endovascular embolization, with exclusion criteria being unsuitable location and angioarchitecture for embolization (29%), as well as a lack of appropriate follow-up (26%). However, other than specifically excluding patients with DAVFs, the composition of their study population remains uncertain. While complete occlusion occurred in only 16%, 86% of the patients had >50% reduction in the size of their AVM, and clinical stabilization or improvement was noted in 83% of patients in a mean follow-up period of 5.6 years. Twenty-seven percent of patients developed neurological deficits postprocedure, 14% of which subsequently resolved with steroids or anticoagulation therapy. Notably, no recanalization was observed in patients treated with NBCA. Although comparison with other series by category of malformation is problematic, the lack of recanalization imparts considerable evidence supporting the efficacy and durability of cyanoacrylates.

Conclusions

Despite the considerable advancements made since their original description, spinal vascular malformations continue to pose significant therapeutic challenges. While not without controversy, a reasonable paradigm is to attempt embolization of any malformation with suitable angioarchitecture, knowing that at present resection remains a well-supported and durable treatment alternative. Additionally, one must recall that surgical and endovascular therapies have a complementary role in the management of complex lesions, and only continued clinical research will permit improved characterization of their appropriate capacities. Furthermore, continued advancement in catheter technology, as well as the introduction of novel embolic agents such as Onyx, will only serve to further expand the role of neurointerventional management for these demanding entities.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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