ORIGINAL ARTICLE

Spinal arteriovenous malformations: Is surgery indicated?

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ABSTRACT

Purpose: To identify clinico-radiological distinguishing features in various types of spinal arteriovenous malformations (AVM) with an aim to define the role of surgical intervention.

Materials and Methods: Hero's modified Di Chiro classification differentiated four types of spinal AVMs on digital subtraction angiogram (DSA) in 74 patients: I. Dural arteriovenous fistulae ($n = 35$, 47.3%); II. Glomus/intramedullary ($n = 13$, 17.6%); III. Juvenile/metameric ($n = 4$, 5.4%); and, IV. Ventral perimedullary fistula ($n = 21$, 28.4%). A patient with extradural AVM remained unclassified. Demographic profiles, DSA features and reason for surgical referral were recorded. Statistical comparison of discrete variables like gender, spinal cord level, presentation and outcome was made using Chi-square test; and, continuous variables like age, feeder number, duration of symptoms and number of staged embolizations by one way analysis of variance with Boneferoni *post hoc* comparison. Embolization alone ($n = 39$, 52.7%), surgery alone ($n = 16$, 21.6%), and combined approach ($n = 4$, 5.4%) were the treatments offered (15 were treated elsewhere).

Results: Type I-AVM occurred in significantly older population than other types ($P = 0.01$). Mean duration of symptoms was 13.18 ± 12.8 months. Thoracic cord involvement predominated in type-I and III AVMs ($P = 0.01$). Number of feeding arteries were 1 in 59.7%; 2 in 29.0%; and, multiple in 11.3% patients, respectively. Staged embolization procedures in type-III AVM were significant ($P < 0.01$). Surgical referral was required due to: Vessel tortuosity/insufficient parent vessel caliber ($n = 7$); residual AVM ($n = 4$); low flow AVM ($n = 3$); and, multiple feeders ($n = 2$). Check DSA ($n = 34$) revealed complete AVM obliteration in 26 and minor residual lesion in eight patients. Neurological status improved in 26 and stabilized in 25 patients.

Conclusions: Differentiating between Type I-IV AVMs has a significant bearing on their management. Surgical intervention should be considered as an important adjunct/alternative to therapeutic embolization.

Key words: Arteriovenous fistula, classification, embolization, radiology, spinal arteriovenous malformation, surgery

Introduction

Spinal arteriovenous malformations (AVMs) (constituting 3-4% of all spinal cord space occupying lesions),[1] if unrecognized and left untreated, may result in a major neurological disability within a short span of time after the onset of initial symptoms.

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A substantial improvement in the imaging technology and in super-selective catheterization has generated considerable success in the diagnosis and management of these rare lesions.[2] The existence of several classifications and therapeutic modalities (including surgery/endovascular therapy/combined therapy),[3-5] however, indicate that an unequivocal management strategy for addressing these lesions has not yet been crystalized. The surgical options include excision/coagulation of the nidus, disconnecting the draining vein, interruption of the intra- and extra-dural venous drainage or even stripping off the dilated and tortuous venous plexus along the cord.[1-5] Interruption of the feeding arteries alone may often prove ineffective due to subsequent recruitment of collateral flow.

In this study, in 74 patients with spinal AVM, the clinico-radiological features distinguishing different types of spinal AVMs were identified with an aim to better define the role of surgical intervention in the treatment of these lesions.

Materials and Methods

A review of 74 patients, between January 2001 and September 2012, who underwent a digital subtraction angiogram (DSA) to diagnose a spinal AVM, was undertaken. The clinical spectrum of the patients, mode of onset, extent and duration of clinical disability were noted. The type of AVM, the vertebral level involved and the number of feeding arteries and draining veins were studied from pre-operative DSA images. The therapeutic intervention performed (both embolism and/or surgery), and the post-operative outcome were recorded. A special emphasis, based upon the clinico-radiological correlation of the patients and the type of spinal AVM, flow status and the levels of spinal segments involved, was on ascertaining the particular reason for choosing one or more therapeutic modality for successfully dealing with these lesions.

The classification given by Di Chiro *et al.*[2] with modification from Heros *et al.*[6] was used to classify spinal AVMs into groups: Type I lesions are dural arteriovenous fistulas in which a dural branch from a radicular artery forms an abnormal communication with a dural vein, usually at the nerve root sleeve [Figures 1 and 2]; Type II lesions are glomus or intramedullary lesions [Figure 3]; Type III are juvenile or metameric AVMs associated with both intradural and extradural extension; and, Type IV lesions are ventral perimedullary fistulae supplied by the anterior spinal artery [Figures 4-7].

A statistical comparison was made between the types of spinal AVM using SPSS software (version 14). One-way analysis of variance test was applied with Bonferroni *post hoc* comparison for continuous variables like age, number of feeders, duration of symptoms and number of sittings of embolization. Association was tested for discrete variables like gender, region of spinal cord, type of presentation and clinical outcome using Chi-square test.

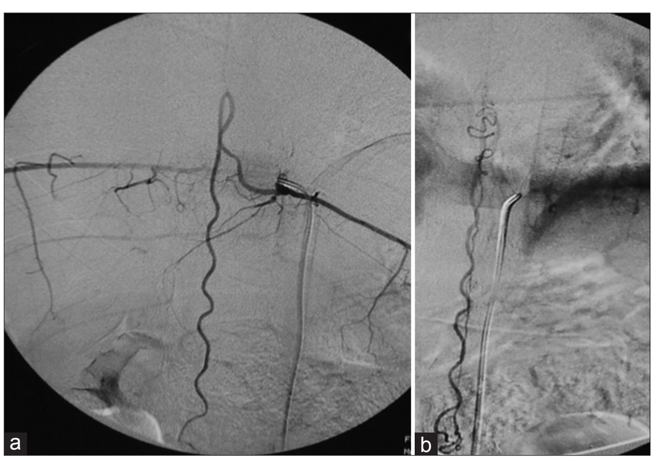

Figure 1: (Patient 1) Type I dural AV fistula (a) with the arteriovenous fistula situated in the D6 intervertebral foramen and a single large vein emerging from it running dorsally on the cord surface; and, (b) the dilated veins due to the venous congestion

Results

Clinical spectrum

There were 59 male and 15 female patients. Their age varied from 2 to 74 years with the mean age being 37.65 ± 18.7 years. The Type I spinal AVM was the most commonly encountered type (35 patients, 47.3%), followed by type IV (21 patients, 28.4%) and type II (13 patients, 17.6%). Type III was the uncommon type with inclusion of only 4 cases (5.4%). One of our patients had solely an epidural spinal AVM and was kept in the unclassified category since he could not be categorized into one of the known categories defined in the modified Di Chiro's classification.[2,7]

The age distribution was variable among the different types of spinal AVMs [Table 1]. Type I spinal AVM mainly

Table 1: Age of the patients having different types of spinal arteriovenous malformation

Age of the patient (in years)	Types of spinal AVM					Total
	Type I spinal AVM	Type II spinal AVM	Type III spinal AVM	Type IV spinal AVM	Unclassified	
0-9	0	1	0	5	0	6
10-19	0	1	2	4	0	7
20-29	1	8	2	4	0	15
30-39	3	2	0	3	0	8
40-49	7	0	0	3	1	11
50-59	16	1	0	2	0	19
60-69	6	0	0	0	0	6
70-79	2	0	0	0	0	2
Total	35	13	4	21	1	74

AVM – Arteriovenous malformation

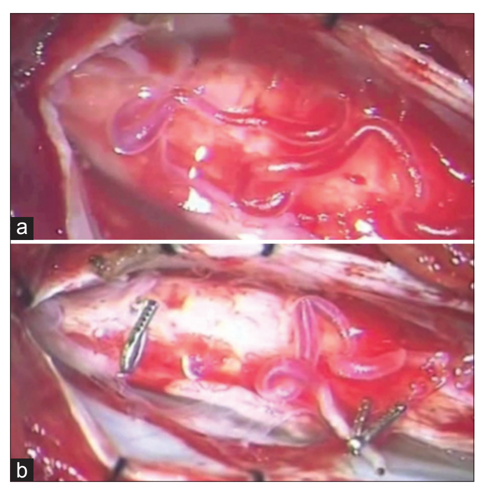

Figure 2: (Patient 1) Intraoperative photographs showing (a) the arteriovenous fistula with the artery directly draining into a single draining vein and leading to venous dilation at a lower level; and, (b) the interruption of the arteriovenous communication leads to lasting relief in the venous congestion. The ends of the divided arteriovenous fistula are coagulated and secured with Liga silver clips to prevent their recanalization

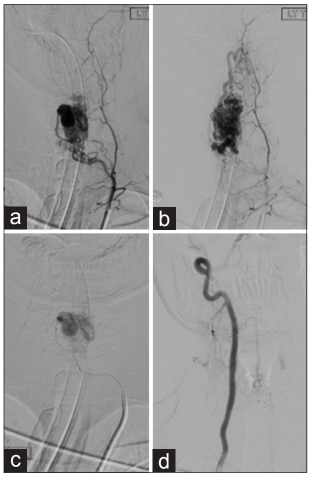

Figure 3: (Patient 2) Intramedullary arteriovenous malformation fed by three branches of the left thyrocervical artery. (a) One of the feeders reveals an aneurysm; (b) Early draining veins are also noted; (c) Superselective catheterisation of a feeding artery (with the aneurysm) at the time of glue embolisation; and, (d) Follow-up angiogram 9 months after glue embolisation reveals a small residual arteriovenous malformation nidus and early draining vein fed by the anterior spinal artery (arising from the right vertebral artery)

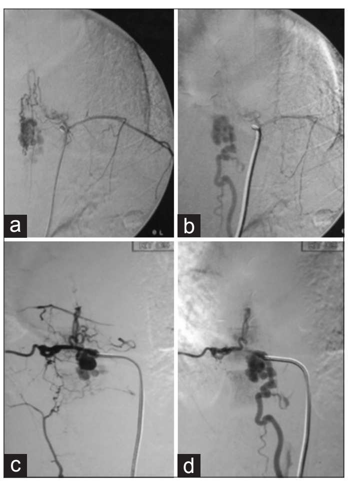

Figure 5: (Patient 3) Digital subtraction angiogram with selective catheterization of left D9 radiculo-medullary artery showing (a) the perimedullary arterial supply; and, (b) the tortuous perimedullary dilated veins; (c) selective catheterization of right D9 radiculo-medullary artery showing the venous aneurysm; and (d) the dilated veins draining into dorsally situated single large vein

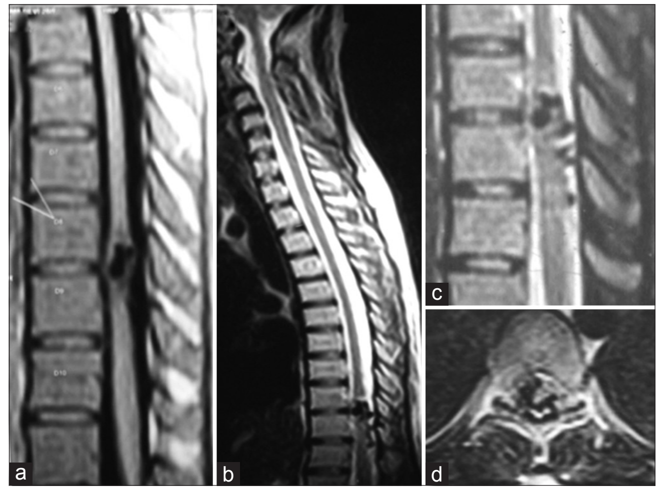

Figure 4: (Patient 3) (a) T1-weighted; (b) T2-weighted sagittal magnetic resonance (MR) images showing the flow voids of a perimedullary arteriovenous malformation; (c) Enlarged T2-sagittal; and (d) axial MR images showing the venous aneurysm situated on the right anterior aspect of the spinal cord with perimedullary flow voids

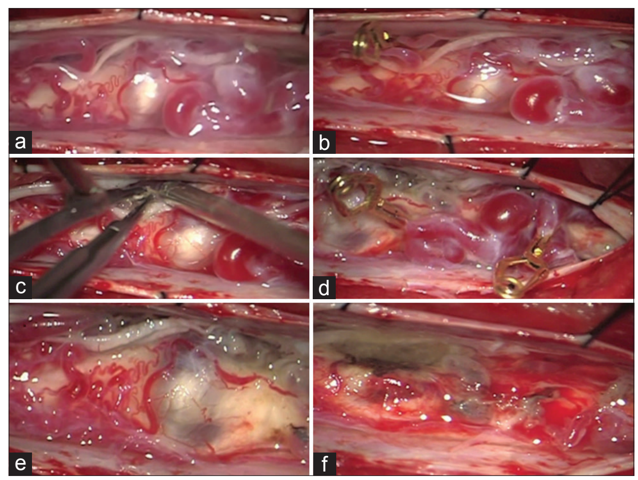

Figure 6: (Patient 3) Operative images showing (a) the dilated arteries and arterialized perimedullary veins along the spinal cord; (b) temporary clip applied to feeding artery to assess status of arteriovenous malformation; (c) coagulation of feeders to perimedullary arteriovenous malformation; (d) applying temporary clips to dilated perimedullary veins to assess for cord edema; (e) following excision of perimedullary sheath of vessels, bulge due to venous aneurysm becomes visible; and, (f) cord following coagulation of arterial supply, excision of dilated veins, and, excision of venous aneurysm

involved older patients with their age ranging from 28 to 74 years (mean age at presentation: 52.09 ± 10.7 years). The other three types of AVMs involved a relatively younger population (type II: Age range of 9-58 years with a mean age at presentation of 25.00 ± 11.5 years; type III: Age range of 13-24 years with a mean of 19.25 ± 4.6 years; and, type IV: Age range 2-54 years with a mean of 24.76 ± 17.1 years). Nine out of 21 patients (42.9%) with a type IV spinal AVM were in the pediatric age group (age $<$ 14 years). The patient having an epidural spinal AVM (who was kept under the unclassified category) was 41 years old. There was a significant difference ($P < 0.01$) in the age of onset of the clinical symptoms of type I spinal AVM when compared with other three types. There was no significant difference, however, in the age distribution in other three subtypes.

There was a significant difference in gender distribution between type I (34 male, 1 female patients) and type II (10 male, 3 female patients) ($P = 0.024$); and, between type I and type IV (11 male, 10 female patients) ($P < 0.01$) spinal AVMs. But the gender distribution was not significantly different between type I and type III (three male, one female

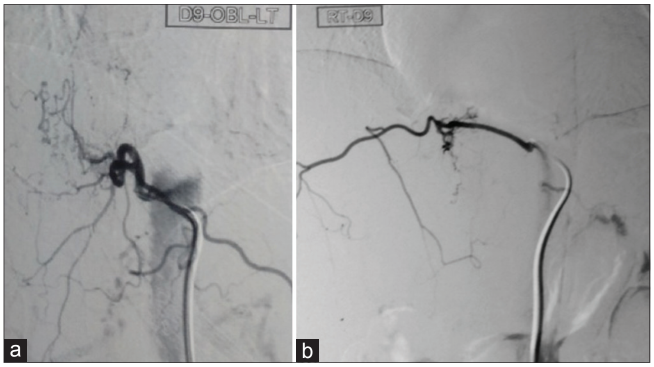

Figure 7: (Patient 3) Post-operative digital subtraction angiogram following selective catheterization of (a) left D9 and, (b) right D9 radiculo-medullary vessels showing complete excision of the arteriovenous malformation

patient). The patient with solely an epidural AVM was a male patient [Table 2].

Gradual progression of myelopathy occurred in 59 patients; in nine patients, however, there was a sudden onset of symptoms; and in six patients, the onset of symptoms could not be specified. Type IV spinal AVM had more instances of acute presentation (23.8%), as compared to the other types (type I: 2.9% and type II: 15.4%). One patient out of four of type III spinal AVM also had an increased incidence of sudden onset neurological disability. Our only case of epidural spinal AVM had a gradual onset of symptoms. The difference in the clinical presentation between types I and type IV spinal AVMs was significant ($P = 0.019$) but no significant difference in clinical presentation between other types of spinal AVMs was detected [Table 3].

The total duration of symptoms varied from 3 days to 5 years, with mean of 13.18 ± 12.8 months. There was no significant difference between the duration of presenting symptoms between the four types of spinal AVMs all of whom presented with long tract signs. There was no significant difference between presenting symptoms between different types of spinal AVMs [Table 4].

Radiology

In our patients, the thoracic spinal cord was most commonly involved (49 patients, 66.2%), followed by thoraco-lumbar (10 patients, 13.5%), lumbar (7 patients, 9.5%), and cervical (7 patients, 9.5%) spinal cord. Cervico-dorsal spinal cord affliction was present in 1 patient (1.4%). There was clear predilection for the thoracic level in type I (cervical 2.9%, thoracic 85.7%, thoraco-lumbar 5.7%, lumbar 5.7%), and type III (thoracic 100%) spinal AVMs. However, the thoracic level predominance was not so distinguishable in type II (cervical 23.1%, cervico-dorsal 7.7%, thoracic 38.5%, thoraco-lumar 23.1%, lumbar 7.7%), and type IV (cervical 9.5%, thoracic 47.6%, thoraco-lumbar 23.8%, lumbar 19.0%) spinal AVMs. The difference in the region of spinal cord involvement was significant between type I and type II ($P = 0.011$), and type I and type IV ($P = 0.025$) spinal AVMs. There was, however, no significant difference in the levels of involvement between other types of spinal AVMs [Table 5].

In 59.7% patients, there was single feeding artery, while in 29.0% patients, there were two feeding arteries. There were multiple feeding arteries in 11.3% patients. Most of the patients of type I spinal AVM had a single feeding artery (single feeder 73.3%, two feeders 23.3%, multiple feeders 3.3%), whereas nearly half of the patients of type II (single feeder 54.5%, two feeders 36.4%, multiple feeders 9.1%), and type IV (single feeder 52.9%, two feeders 29.4%, multiple feeders 17.7%)

Table 2: The male:female statistics in the series

Sex of the patients	Types of spinal AVM					Total
	Type I spinal AVM	Type II spinal AVM	Type III spinal AVM	Type IV spinal AVM	Unclassified	
Male	34	10	3	11	1	59
Female	1	3	1	10	0	15
Total	35	13	4	21	1	74

AVM – Arteriovenous malformation

Table 3: The onset of clinical presentation in patients with different types of spinal arteriovenous malformation

Clinical presentation	Types of spinal AVM					Total
	Type I spinal AVM	Type II spinal AVM	Type III spinal AVM	Type IV spinal AVM	Unclassified	
Sudden onset	1	2	1	5	0	9
Gradual onset	32	10	3	13	1	59
Not mentioned	2	1	0	3	0	6
Total	35	13	4	21	1	74

AVM – Arteriovenous malformation

Table 4: Duration of clinical presentation in the series

Duration of presenting symptoms (in months)	Types of spinal AVM					Total
	Type I	Type II	Type III	Type IV	Unclassified	
0-6	3	3	2	5	0	13
6-12	8	0	0	0	1	9
12-18	4	1	0	1	0	6
18-24	3	1	0	1	0	5
24-30	2	1	1	3	0	7
30-36	0	0	0	0	0	0
36-42	1	0	0	0	0	1
42-48	0	0	0	0	0	0
48-54	0	0	0	1	0	1
54-60	0	0	0	1	0	1
Total	21	6	3	12	1	43*

AVM – Arteriovenous malformation; *Patients for whom duration of symptoms was mentioned in records spinal AVM had two or more feeding arteries. All four patients of type III spinal AVM had multiple feeders. The difference in the number of feeders was significant between type III and other types of AVMs ($P < 0.01$) but there was no significant difference in other types of spinal AVMs.

Therapeutic intervention and post-intervention status

Embolization of the AVM was carried out in 39 (52.7%) patients; surgery in 16 (21.6%) patients; and, a combined approach, including both surgery and embolization, in 4 (5.4%) patients respectively. Fifteen (20.3%) patients, in whom a diagnostic DSA revealed a spinal AVM and further treatment in the form of surgery and/or embolization was advised did not report back for further treatment at our center. The therapeutic intervention in the different types of spinal AVMs were as follows: Type I: Embolization: 21 (60%), surgery: 9 (25.7%); combined modality 1 (2.9%); and, not reporting for further treatment after the diagnostic DSA: 4 (11.4%); Type II: Embolization: 5 (38%), surgery: 1 (7.7%); and, not reporting for further treatment after the diagnostic DSA: 7 (53.8%); Type III: Embolization: 1 (25%); combined modality 1 (25%); and, not reporting for further treatment after the diagnostic DSA: 2 (50%); and, Type IV: Embolization: 12 (57.1%), surgery: 5 (23.8%); combined modality 2 (9.5%); and, not reporting for further treatment after the diagnostic DSA: 2 (9.5%) [Table 6].

The reason for referral to surgery included failed embolization in 3 (15%) patients, residual AVM on check angiogram in 4 (20%) patients, difficulty in traversing the tortuous feeding arteries in 3 (15%) patients, an extremely low-flow arteriovenous fistula in 3 (15%) patients, close proximity of the fistula to the anterior spinal artery in 1 (5%) patient, multiple feeders in 2 (10%) patients and, embolization not being possible due to the close relationship of the fistula with the anterior spinal artery in 1 (5%) patient, respectively The reason for referral could not be ascertained in 5 (25%) patients [Table 7].

The operative procedures ($n = 16$), after the initial laminectomy, carried out in various types of spinal AVMs in the series included the following: (a) Type I: Disconnection/excision of fistula ($n = 4$, 40%), division/coagulation of feeding arteries ($n = 3$, 30%) and, application of clip to feeding vessel ($n = 3$, 30%); (b) Type II: Excision of nidus ($n = 1$, 100%); (c) Type III: Partial excision of nidus, followed by re-exploration and excision of the residual nidus in the same patient ($n = 1$, 100%); (d) Type IV spinal AVM: Disconnection/excision of fistula ($n = 4$, 57.14%), clipping of feeder ($n = 2$, 28.57%) and, partial excision but surgery abandoned due to profuse extradural bleeding ($n = 1$, 14.29%); and, (e) Extradural spinal AVM: Disconnection of fistula ($n = 1$, 100%).

One-time surgery or embolization was considered as one session of therapy. Performance of only a diagnostic DSA without further intervention was not considered as a therapeutic session. Most of the patients of type I (single session 90%, multiple session 10%) and type II (single session 83.3%, multiple session 16.7%) had a single therapeutic session. Most patients of type IV (single session 78.9%, multiple session 21.1%) and all patients of type III spinal AVM had multiple therapeutic sessions. The difference in the number of multiple sessions required was significant in type III compared with other types of spinal AVMs ($P < 0.01$).

Acrylic glue was the most commonly used embolic material. It was used in 35 patients, out of 42 patients in whom embolization was used as the sole treatment modality or as a part of combined therapy. Combined glue and coiling was used in two patients (one belonging to type I and the other to

Table 5: The level of spinal cord involvement

Region of spinal cord	Types of spinal AVM					Total
	Type I spinal AVM	Type II spinal AVM	Type III spinal AVM	Type IV spinal AVM	Unclassified	
Cervical	1	3	0	2	1	7
Thoracic	30	5	4	10	0	49
Lumbar	2	1	0	4	0	7
Thoraco-lumbar	2	3	0	5	0	10
Cervico-dorsal	0	1	0	0	0	1
Total	35	13	4	21	1	74

AVM – Arteriovenous malformation

Table 6: The type of therapeutic intervention performed

Intervention	Types of spinal AVM					Total
	Type I spinal AVM	Type II spinal AVM	Type III spinal AVM	Type IV spinal AVM	Unclassified	
Surgical	9	1	0	5	1	16
Endovascular	21	5	1	12	0	39
Combined	1	0	1	2	0	4
Only diagnostic DSA done, patient did not report for intervention	4	7	2	2	0	15
Total	35	13	4	21	1	74

DSA – Digital substration angiogram; AVM – Arteriovenous malformation

Table 7: Indications for surgery in the series

Reason	Number	Percent
Failed embolization	3	15
Residual AVM	4	20
Difficult cannulation of feeding artery	3	15
Low flow AVM	3	15
Multiple feeders	1	5
Embolization not possible due to proximity of AVM to anterior spinal artery	1	5
Not mentioned	5	25

AVM – Arteriovenous malformation

type IV spinal AVM categories. Combined glue and polyvinyl alcohol (PVA) particles were used in 1 patient of type II spinal AVM. In one patient of type I spinal AVM, combined coiling and PVA particles were used. PVA particles were the sole embolic material used for the embolization of type III spinal AVM; and, coiling, the only material used in 1 patient of type IV spinal AVM.

A check angiogram showed complete obliteration of the AVM in 26 patients, while in eight patients it showed a residual lesion. Records of check angiogram were not available in 25 patients.

There was a minimum of "Medical Research Council" one grade improvement in the neurological status of 26 patients at the time of discharge; while 24 patients remained in status quo and six deteriorated. The neurological status of three patients at discharge was not mentioned in the records [Table 8]. There was no significant difference in the results in different categories of therapeutic intervention performed [Table 9].

Discussion

Types of spinal AVMS and their treatment options

The type I spinal arteriovenous fistula was the most commonly encountered spinal AVM [Figures 1 and 2]. It was usually the easiest to treat among the various groups of spinal AVMs. It had a slow progressive development of myelopathy and also occurred in a significantly older group of patients than the other three types.[7-15] The gradual dilatation of the perimedullary veins, fed by a usually remote arteriovenous fistula, lead to venous congestion and edema. The fistula was often difficult to access due to its location at the nerve root sleeve in the intervertebral foramina. Interrupting the draining vein leading to the dilated perimedullary venous complex, however, often resulted in lasting neurological improvement. Usually the feeding artery and the draining vein in these patients was also single. This facilitated its easy control either by the endovascular or surgical technique. A 98% obliteration rate with less than 2% morbidity has been reported with this approach in these lesions.[16-18] In contrast, Type II [Figure 3] and III spinal AVMs had an intramedullary nidus that needed excision from within the spinal cord substance;[19] and, type IV lesions often had a component of vascular supply from the anterior spinal artery and multiple dilated peri-medullary vessels usually spanning multiple cord segments.[20] The direct involvement of the spinal cord in the type II, III and IV lesions clinically manifested as an earlier and more rapid onset of symptoms. These lesions usually had multiple feeding and draining vessels in contrast to the type I AVM. These factors considerably increased the degree of difficulty in their surgical excision. The Type IV perimedullary arteriovenous fistulae could be further subdivided into IVa: Simple, small fistula fed by minimally dilated, long thin anterior spinal or posterolateral artery with low flow velocity in the artery and vein and causing minimum mass effect. These were usually amenable to surgical excision or obliteration; IVb [Figures 4-7]: Medium sized fistula with one or two dilated feeding arteries, venous dilatation in the shunt region and dilated and tortuous draining veins. These were dealt with by embolization, surgery or combined therapy; and, IVc: Giant fistula with multiple feeding arteries of enlarged caliber and high flow associated with significant shunt volume and with multiple dilated veins with a winding course.[20-22] These usually required multiple stages of therapeutic embolization [Table 10].

Another interesting finding was the significant predilection of type I fistulae to involve the thoracic spinal cord when compared with the other subtypes. Jellema *et al.* has also reported a similar occurrence of dural AV fistulae in the thoracolumbar region with less than 6% occurring in the cervical or sacral regions.[9] Areas of thoracic spinal cord represent the watershed zone of vascular supply of the spinal cord where blood supply may often

Table 8: The clinical results (at discharge) in patients with different types of spinal arteriovenous malformation

Result	Type of spinal AVM					Total
	Type I spinal AVM	Type II spinal AVM	Type III spinal AVM	Type IV spinal AVM	Unclassified	
Improvement	15	0	1	9	1	26
Same	13	5	1	5	0	24
Deterioration	2	1	0	3	0	6
Not mentioned	1	0	0	2	0	3
Only diagnostic DSA done, patient did not report for intervention	4	7	2	2	0	15
Total	35	13	4	21	1	74

DSA – Digital substration angiogram; AVM: Arteriovenous malformation

Table 9: The clinical results (at discharge) in patients based on different types of therapeutic interventions undertaken

Result	Intervention				Total
	Surgical	Endovascular	Combined	Only diagnostic DSA done	
Improvement	10	13	3	0	26
Same	2	22	0	0	24
Deterioration	4	2	0	0	6
Not mentioned	0	2	1	0	3
Only diagnostic DSA done, patient did not report for intervention	0	0	0	15	15
Total	16	39	4	15	74

DSA – Digital substration angiogram

Table 10: Treatment options for various types of spinal arteriovenous malformations

Type of spinal AVM	Treatment
I	Surgery is preferred over embolization if introduction of microcatheter is difficult, Adamkiewicz's artery or anterior spinal artery originates from feeding radicular artery, if there are multiple small feeding arteries, if there is recurrence or if there is residual lesion remaining after embolization
II	Surgery±endovascular
III	Surgery+endovascular (palliative)
IV	Low flow-surgery
	High flow-endovascular±surgery

AVM – Arteriovenous malformation

be relatively deficient when compared to other cord segments. The thoracic spinal canal diameter is also the least capacious. In all likelihood, redistribution of vascular supply leading to an early cord ischemia or, venous congestion precipitating cord edema and thecal compression, manifested much earlier in the thoracic region. This probably led to the earlier and more frequent discovery of the underlying arteriovenous fistula/AVM in this region.

Radiological parameters

In our patients, the classical radiological hallmarks indicating the presence of a spinal AVM were the serpiginous flow voids in the spinal subarachnoid space and within the cord substance, their gadolinium enhancement, focal hyperintensity within the spinal cord that typically spanned several segments and intraparenchymal or subarachnoid hemorrhage.[23-26] However, the actual arteriovenous fistula/AVM detected on angiogram was often situated at a variable level when compared to the areas of flow voids detected on magnetic resonance (MR) imaging. There were often bilateral and multiple feeders associated with the spinal AVM. This phenomenon was particularly marked with the type III spinal AVM where all four of our patients had multiple feeders. It was usually impossible to detect the exact level of the AVM or its feeders solely on the initial MR imaging. Keeping this factor in mind and maintaining a high index of suspicion in getting an early DSA done improved the detectability rate of these relatively elusive lesions. The relatively long duration of illness in the present series points towards the delay in referral for treatment of these lesions due to their late detection.

Neurological deficits

Nine patients in the series had a sudden onset of symptoms; or, steps of rapid increase in neurological deterioration interspersed between progressive myelopathy. This may have been due to focal hemorrhage, rapid expansion of the AVM nidus, rapid redistribution of blood supply leading to development of ischemic areas, precipitating a "steal phenomena" or due to development of spontaneous thrombosis of vessels.[26] The potential causes of spinal cord hemorrhage in these lesions include ruptured arterial aneurysms or re-rupture of false arterial aneurysms, rupture of spinal cord subpial, subarachnoid veins and/or nidus or the fragile neovascularization and, the presence of hemorrhagic venous infarcts.[27] The preponderance of older patients in the series in this congenital anomaly, however, does point to the fact that the usual course is a slow progressive one as the AVM expands, parasitizes more arterial supply and causes venous dilatation, retrograde congestion and cord edema.

Therapeutic embolization

Successful endovascular treatment entails occlusion of the radiculo-medullary vein and/or the fistula itself in the type I arteriovenous fistulae; or, obliteration of the feeding vessels and/or filling up of the AVM itself with liquid embolic material in the other types of spinal AVMs.[5] As embolic materials, coils are not preferred, as they require rigid catheters that cannot be negotiated through tortuous vessels supplying the spinal AVM. They can only achieve a proximal embolization often leading to delayed collateralization of the nidus. This also has the disadvantage of not allowing future attempts to access the nidus for staged embolization. Particle embolization is performed in a stepwise fashion that has the advantage that the clinical effects of every stage of embolization are amenable to evaluation. However, there is a high rate of long-term recanalization. Its main role is, therefore, is in palliative treatment as it alters the natural history of the spinal AVM with a good clinical outcome.[28]

Liquid agents such as PVA have been used to permanently occlude the spinal AVM but occluding the primary area of the feeder without reaching the actual site of the fistula results in recanalization or formation of collateral vessels to the AVM. Liquid glue embolic agents such as isobutyl-2-cyanoacrylate or N-butyl cyanoacrylate facilitate distal vessel penetration with a more permanent AVM occlusion. However, these agents require micro-catheter access directly at the site of fistula. A rapid catheter withdrawal (within seconds) following injection of the embolic material is essential especially in slow flow AVMs since these agents tend to polymerize near the catheter tip. Ethylene vinyl alcohol (Onyx) has a relatively lower viscosity compared to other liquid embolic agents that helps to disperse the agents to parts of AVM/arteriovenous fistulae which the micro-catheter cannot access; and, the embolic material may be injected slowly (over a period of minutes) leading to its more accurate placement within the spinal AVM to secure a more complete embolization.[23] However, the remnant lesion may be difficult to visualize due to radio-opacity of tantalum mixed in onyx; onxy may be potentially toxic to neural and vascular tissue; and, a far greater reflux into feeding arteries (particularly the radiculo-medullary supply to the spinal cord) may occur during the execution of the endovascular maneuvers that facilitate deep penetration of onyx into the lesions.[29]

Indications for surgery

In patients with Type I spinal arteriovenous fistula, 25.7% underwent surgery and 2.9%, a combined modality of treatment. The ease of interruption of the dorsally placed type I arteriovenous fistula or of the dilated draining vein often makes surgery the "gold standard" of treatment.[18] The extreme tortuosity and sinuous course of the feeding arteries that made catheterization difficult; and an extremely low-flow fistula made surgery a favorable option in these patients. In type II AVMs, where 38% patients underwent surgery, the AVM manifested as an intramedullary mass lesion and surgical extirpation was the preferred option not only to alleviate the pressure effects on the cord but also to avoid compromising the important feeding vessels common to both the AVM and the cord substance.[19] Type III and type IV patients often had multiple feeders at variable levels and a diffuse nidus of vessels forming the AVM. Embolization often helped to occlude the primary vascular supply prior to the surgical maneuvers. The latter included additionally interrupting the dilated feeding arteries, excising or coagulating the nidus, stripping off the dilated perimedullary veins that were causing mass effect and venous engorgement or, obliterating the arterial/venous aneurysm.[21,22] In consonance with our findings, Zozulya *et al.* have defined the role of surgery in these lesions. They propose the general principle of occluding the feeding and draining vessels and resection of the malformation in AVMs; and, the occlusion of only the feeding vessels in arteriovenous fistulae.[5] Thus, occlusion of the feeding and draining vessels and excision of AVM was the preferred approach in most patients with type II intramedullary AVM with a focal nidus, epidural AVM and, an occasional type III AVM with intra- and extra-medullary component and type IVa or IVb perimedullary AVM (in the latter two groups usually after therapeutic embolization). Occlusion of only the feeding vessels was the preferred option in Type I dural arteriovenous fistula (interrupting of draining vein sufficed in most cases), type II intramedullary AVM with a diffuse nidus and, an occasional type IV perimedullary arteriovenous fistula or AVM and intramedullary diffuse AVM (where excision of the diffuse intramedullary nidus may lead to cord injury).[5] High flow type III and IVc lesions usually required solely therapeutic embolization of the feeding vessels with or without embolization of the nidus.

Limitations

The physicians evaluating results were involved in the primary treatment of the patients. The patients were not randomly assigned to either the surgical or embolization groups. Embolization was always the first option and the referral for surgery was made after the feasibility of therapeutic intervention had been carefully assessed. Greater recruitment of patients in all the groups would have led to a more meaningful statistical significance. The assessment of functional status of our patients was at discharge. There was a possibility of far greater improvement on a long-term basis. However, the emphasis of this study was on investigating the role of surgery in these lesions and not on assessing the clinical outcome of the patients on prolonged follow-up.

Conclusions

Differentiating between Type I-IV AVMs has a significant bearing on their management. Type I AVM occurs in a much older subset of patients often in a thoracic location when compared to patients in the other groups. Surgical intervention may be used to obliterate spinal AVMs/arteriovenous fistulae following the endovascular embolization of the feeding vessels; or when incomplete or failed embolization occurs especially when the vessels are tortuous and difficult to traverse or an extremely low flow AVM is present. It may also be a primary modality of treatment in the cases where a dorsally situated dilated vein leading from a remote arteriovenous fistula requires interruption; in situations where AVM behaves like an intramedullary mass lesion having a common blood supply with the spinal cord segment; and, to interrupt the arterial supply leading up to the AVM or to coagulate and strip off the dilated perimedullary veins and/or arterial/venous aneurysm causing mass effect or hemorrhage.

References

1. Bao YH, Ling F. Classification and therapeutic modalities of spinal vascular malformations in 80 patients. Neurosurgery 1997;40:75-81.
2. Rosenblum B, Oldfield EH, Doppman JL, Di Chiro G. Spinal arteriovenous malformations: A comparison of dural arteriovenous fistulas and intradural AVM's in 81 patients. J Neurosurg 1987;67:795-802.
3. Spetzler RF, Detwiler PW, Riina HA, Porter RW. Modified classification of spinal cord vascular lesions. J Neurosurg 2002;96:145-56.
4. Kim LJ, Spetzler RF. Classification and surgical management of spinal arteriovenous lesions: Arteriovenous fistulae and arteriovenous malformations. Neurosurgery 2006;59:S195-201.
5. Zozulya YP, Slin'ko EI, Al-Qashqish II. Spinal arteriovenous malformations: New classification and surgical treatment. Neurosurg Focus 2006;20:E7.
6. Heros RC, Debrun GM, Ojemann RG, Lasjaunias PL, Naessens PJ. Direct spinal arteriovenous fistula: A new type of spinal AVM. Case report. J Neurosurg 1986;64:134-9.
7. Westphal M, Koch C. Management of spinal dural arteriovenous fistulae using an interdisciplinary neuroradiological/neurosurgical approach: Experience with 47 cases. Neurosurgery 1999;45:451-7.
8. Huffmann BC, Gilsbach JM, Thron A. Spinal dural arteriovenous fistulas: A plea for neurosurgical treatment. Acta Neurochir (Wien) 1995;135:44-51.
9. Jellema K, Canta LR, Tijssen CC, van Rooij WJ, Koudstaal PJ, van Gijn J. Spinal dural arteriovenous fistulas: Clinical features in 80 patients. J Neurol Neurosurg Psychiatry 2003;74:1438-40.
10. Atkinson JL, Miller GM, Krauss WE, Marsh WR, Piepgras DG, Atkinson PP, *et al*. Clinical and radiographic features of dural arteriovenous fistula, a treatable cause of myelopathy. Mayo Clin Proc 2001;76:1120-30.
11. Tai PA, Tu YK, Liu HM. Surgical treatment of spinal arteriovenous malformations: Vascular anatomy and surgical outcome. J Formos Med Assoc 2001;100:389-96.
12. Narvid J, Hetts SW, Larsen D, Neuhaus J, Singh TP, McSwain H, *et al*.

Spinal dural arteriovenous fistulae: Clinical features and long-term results. Neurosurgery 2008;62:159-66.
13. Nagata S, Morioka T, Natori Y, Matsukado K, Sasaki T, Yamada T. Factors that affect the surgical outcomes of spinal dural arteriovenous fistulas. Surg Neurol 2006;65:563-8.
14. Ropper AE, Gross BA, Du R. Surgical treatment of Type I spinal dural arteriovenous fistulas. Neurosurg Focus 2012;32:E3.
15. Van Dijk JM, Ter Brugge KG, Willinsky RA, Farb RI, Wallace MC. Multidisciplinary management of spinal dural arteriovenous fistulas: Clinical presentation and long-term follow-up in 49 patients. Stroke 2002;33:1578-83.
16. Klopper HB, Surdell DL, Thorell WE. Type I spinal dural arteriovenous fistulas: Historical review and illustrative case. Neurosurg Focus 2009;26:E3.
17. Martin NA, Khanna RK, Batzdorf U. Posterolateral cervical or thoracic approach with spinal cord rotation for vascular malformations or tumors of the ventrolateral spinal cord. J Neurosurg 1995;83:254-61.
18. Steinmetz MP, Chow MM, Krishnaney AA, Andrews-Hinders D, Benzel EC, Masaryk TJ, *et al*. Outcome after the treatment of spinal dural arteriovenous fistulae: A contemporary single-institution series and meta-analysis. Neurosurgery 2004;55:77-87.
19. Velat GJ, Chang SW, Abla AA, Albuquerque FC, McDougall CG, Spetzler RF. Microsurgical management of glomus spinal arteriovenous malformations: Pial resection technique: Clinical article. J Neurosurg Spine 2012;16:523-31.
20. Sure U, Wakat JP, Gatscher S, Becker R, Bien S, Bertalanffy H. Spinal type IV arteriovenous malformations (perimedullary fistulas) in children. Childs Nerv Syst 2000;16:508-15.
21. Cho KT, Lee DY, Chung CK, Han MH, Kim HJ. Treatment of spinal cord perimedullary arteriovenous fistula: Embolization versus surgery. Neurosurgery 2005;56:232-41.
22. Hida K, Iwasaki Y, Goto K, Miyasaka K, Abe H. Results of the surgical treatment of perimedullary arteriovenous fistulas with special reference to embolization. J Neurosurg 1999;90:198-205.
23. Sivakumar W, Zada G, Yashar P, Giannotta SL, Teitelbaum G, Larsen DW. Endovascular management of spinal dural arteriovenous fistulas. A review. Neurosurg Focus 2009;26:E15.
24. Gilbertson JR, Miller GM, Goldman MS, Marsh WR. Spinal dural arteriovenous fistulas: MR and myelographic findings. AJNR Am J Neuroradiol 1995;16:2049-57.
25. Rodesch G, Hurth M, Alvarez H, Tadie M, Lasjaunias P. Spinal cord intradural arteriovenous fistulae: Anatomic, clinical, and therapeutic considerations in a series of 32 consecutive patients seen between 1981 and 2000 with emphasis on endovascular therapy. Neurosurgery 2005;57:973-83.
26. Krings T, Thron AK, Geibprasert S, Agid R, Hans FJ, Lasjaunias PL, *et al*. Endovascular management of spinal vascular malformations. Neurosurg Rev 2010;33:1-9.
27. Rodesch G, Hurth M, Alvarez H, David P, Tadie M, Lasjaunias P. Embolization of spinal cord arteriovenous shunts: Morphological and clinical follow-up and results – Review of 69 consecutive cases. Neurosurgery 2003;53:40-9.
28. Patsalides A, Knopman J, Santillan A, Tsiouris AJ, Riina H, Gobin YP. Endovascular treatment of spinal arteriovenous lesions: Beyond the dural fistula. AJNR Am J Neuroradiol 2011;32:798-808.
29. Niimi Y, Berenstein A, Setton A, Neophytides A. Embolization of spinal dural arteriovenous fistulae: Results and follow-up. Neurosurgery 1997;40:675-82.

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