ORIGINAL ARTICLES

Spinal Arterio- venous lesions: Variable Clinical and Angiographic Feature

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Abstract:

Background: Spinal Arterio-Venous Shunts are rare but treatable diseases. AVF has a direct shunt between artery and vein. AVM has a nidus between artery and vein. Differentiation is necessary because, the clinical presentation, angiographic architecture, and treatment options are different. Rationale: Presenting clinical features are nonspecific and vary in age and sex. MR images raise the suspicion of diagnosis. For proper understanding of disease and planning of treatment DSA is necessary. Objectives: To evaluate the nature of clinical feature, angiographic findings, and initial outcome after embolization in patients of spinal arterio-venous shunt. Materials & Methods: It was prospective study. Patients were referred for spinal DSA. Risk of complication was properly discussed with the patient and attendant and informed written consent was taken. Results: Among 9 patients, 5 were female and 4 were male. Male female ratio was 1: 1.25. Three patients were diagnosed as type I, three as type II, and three as type IV. All 3 patients (100%) of dural fistula were male, all 3 patients (100%) of pial fistula were female. Average age at presentation was 36 yrs. And mean age of dural AVF was 60.33 yrs, pial AVF was 18.33 yrs and spinal AVM was 29.33 yrs. Spinal DAVF occurred in patients ranging from 57 to 64 years of age, with an average age of 60.33 years. The average length of time between onset of symptoms and diagnosis was 16.44 months (ranging from 3 to 36 month). All 9 patients (100%) of patients had motor weakness, sensory disturbance was found in 66.66% (six of nine patients) and urinary disturbance was found in 77.77% (seven of nine patients). Progressive clinical course was followed in 100% of patients (all of nine patients). MRI findings revealed flow void in 77.77% of patients (seven of nine patients). Increased T2 signal in the spinal cord was present in 88.88% of patients (eight of nine patients). After DSA total 77.7% (seven of nine patients) feeder was located in low thoracic and lumbar region. Conclusion: Spinal AVM & AVF remain undiagnosed for a long period. They should be treated early for prevention of progressive morbidity and disability. MRI features of cord edema, contrast enhancement, and peri-medullary vessels may lead to the diagnosis of these vascular lesions.

Key words: Arteriovenous fistula, Arteriovenous malformation, Digital Subtraction Angiography

Abbreviation: AVF: Arteriovenous fistula, AVM: Arteriovenous malformation, DSA: Digital Subtraction Angiography

Introduction:

Spinal arterio-venous lesion represent different group of vascular anomalies. They are uncommon ¹. Based on hemodynamic criteria, spinal vascular lesion can be categorized into 2 distinct group: i) Spinal AVF (Direct shunt between the artery and vein) ii) Spinal AVM (Presence of nidus between the artery and vein)². They can be classified into 4 types: Type I: Dural arteriovenous fistulas (AVFs) Type II: Intramedullary glomus AVM

Type III: Juvenile or combined AVM

Type IV: Intradural perimedullary AVF

Spinal arteriovenous fistula is the most common among spinal vascular malformation ³. Approximately

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70% of spinal vascular lesions are dural AVF⁴. Spinal dural AVF consists of a lesion that makes a shunt which is located within the dura near the neural foramina and along the spinal canal⁵. It is an acquired lesion^{6,7}. It typically present after fourth or fifth decade and in more in male³. Symptoms are progressive in nature⁴. It is treatable and curable disease, so diagnosis should be early and in time⁸ to avoid morbidity⁹. If untreated, approximately 50% patients of dural AVF will be disabled⁴. Spinal AVM usually presents at younger age¹⁰. Perimedullary spinal cord AVF is not a common among spinal vascular malformation. It is mainly seen in the conus medullaris or cauda equina region ¹⁰.

Materials and Method:

In this institution, no department of Interventional Neurology exists. We, the interventionist, are working in different unit of Neurology Department. There are six units in this department. Patients are referred for angiogram or procedure from different units. We are working as operator. Angiogram have to be done in Paediatric Cardiac Cath Lab. Different unit asked for Spinal DSA to evaluate the patient whenever they need it. So, not every patient had MRI or MRI with contrast. From July 2012 to June 2015 was the study period. Total 12 patients were referred for Spinal DSA. Among 12 patients, angiogram of 3 patients was normal. We evaluated the findings of 9 patients.

Risk of complication of Spinal DSA was properly discussed with the patient and attendant and informed written consent was taken. Spinal DSA was done under local anesthesia through transfemoral route. Modified seldinger technique was used for sheath placement. Images were obtained at a rate of 2-4 f/sec and for 25-30 seconds. During DSA, segmental arteries were injected with iohexol (300 mg/ml) at 1 ml/sec. From the supreme intercostals artery to Lumbar 3 segmental artery were injected. In all cases, anterior spinal artery was identified. If no fistula was found, then additional injection was given to Carotid, Vertebral, Thyrocervical, Costocervical, Iliolumbar arteries

Results:

Table I & II showed total number of patients were 9(nine) and 5 were female and 4 were male. Male female ratio was 1: 1.25. Three patients were

sex, age and type of AVM									
patient	male	female	Ratio	Age average	Type I AVM	Type II AVM	Type IV AVM		
9	4	5	1:1.25	36 yrs	3	3	3		

Table-I

 Table-II

 sex. age and presentation of patients

Patient no	Sex	Age	Presentation
1	М	57	M, S, U
2	F	18	M, U
3	F	18	M, U
4	F	19	M, S
5	Μ	56	M, S, U
6	F	13	M, S, U
7	#	#	#
8	Μ	60	M, S, U
9	Μ	64	M, S, U
10	F	19	M, U
11	#	#	#
12	#	#	#

#: normal spinal angiography (not evaluated in study); M= Motor weakness; S= Sensory disturbances; U= urinary disturbances

diagnosed as type I, three as type II, and three as type IV. All 3 patients (100%) of dural fistula were male; all 3 patients (100%) of pial fistula were female. Average age at presentation was 36 yrs. And mean age of dural AVF was 60.33 yrs, pial AVF was 18.33 yrs and spinal AVM was 29.33 yrs. Spinal DAVF occurred in patients ranging from 57 to 64 years of age, with an average age of 60.33 years. The average length of time between onset of symptoms and diagnosis was 16.44 months (ranging from 3 to 36 month). All 9 patients (100%) of patients had motor weakness, sensory disturbance was found in 66.66% (six of nine patients) and urinary disturbance was found in 77.77% (seven of nine patients) (Fig.1). Progressive clinical course was followed in 100% of patients (all of nine patients). No patient had presented with an acute neurological deficit.

MRI findings revealed flow void in 77.77% of patients (seven of nine patients). Increased T2 signal in the

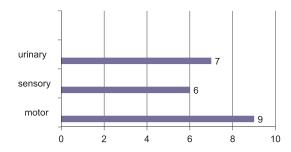


Fig.-1: distribution of patients on clinical presentation

spinal cord was present in 88.88% of patients (eight of nine patients). Hyperintense signal was homogenous and central in location that spared a thin rim of the cord peripherally. 5 patients were referred to us with MRI of Spine with gadolinium contrast. Among those 40% (two of five patients) had contrast enhancement.

After DSA, we found location of arterial feeder (Figure 2) in lumbar region was 44.4% (four of nine patients), in lower thoracic was 33.3% (three of nine patients) (Fig.2). One was located in cervical region and one was in mid thoracic region. Total 77.7% (seven of nine patients) feeder was located in low thoracic and lumbar region (Table III).

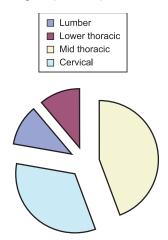


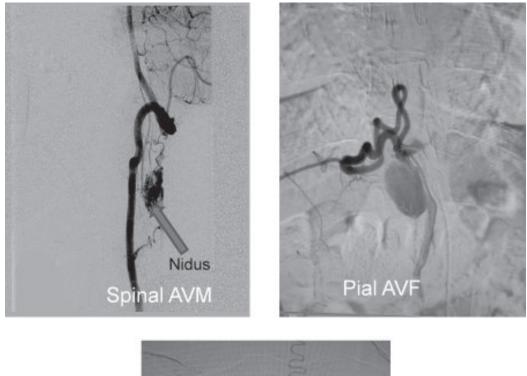
Fig.-2: distribution of respondents on location of arterial feeder in Spinal DSA

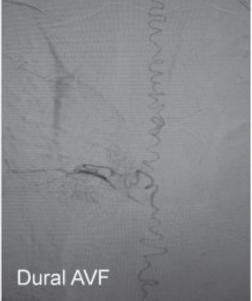
Angiographic findings of the patients									
Patient no	Arterial	Venous	Venous drainage	Angiographic					
	feeder	Aneurysm	Rostral/Caudal/Both	Diagnosis					
1	L1	0	В	Dural AVF					
2	L2	0	R	Pial AVF					
3	D11	1	R	Pial AVF					
4	D12	0	В	AVM (Intramedullary)					
5	C2	0	С	AVM (Intramedullary)					
6	D11	0	В	AVM (Intramedullary)					
7	#	#	#	#					
8	L2	0	В	DuralAVF					
9	D6	0	В	Dural AVF					
10	L1	1	В	Pial AVF					
11	#	#	#	#					
12	#	#	#	#					

 Table III

 Angiographic findings of the patients

C: Cervical; D: Dorsal; L: Lumbar. 0: Absent; 1: Present. B: Both; C: Caudal; R: Rostral. AVF: Arterio venous fistula; AVM: Arterio venous malformation





Discussion:

Spinal vascular lesions are rare disease and accounts approximately 2 to 4 % of spinal diseases¹¹. In our series, fistula was the most common spinal vascular malformation. 66.6% of patients had fistula and 33.3% had AVM. Dural fistula represented 68% of all spinal malformations in the study conducted by Gilbertson and his

colleagues⁶ and 63.33% (19 of 30 patients) in another study⁹. In our study, 33.3% patients had spinal AVM. In a review done by Patsalides A et.al.¹⁰ showed 20-30% had AVM in different studies. Mean age of dural AVF was 60.33 years in our study. Data from other studies showed mean age of 62 years (range from 38-87 years) in a study done by Guillevin R. and his colleagues¹¹. The late age presentation of dural fistula was also found in other series¹²⁻¹⁴. Dural AVF occurs most commonly in male¹⁵. In our study all patients were male. This male preponderance was also found in other series^{13,14}. In a review by Patsalides A et. al.¹⁰ showed that spinal AVM was typically seen in children and early adults. It was 29.33 years in our study. Corkill et. al.¹⁶ found female dominance, and Cullen et. al.¹⁷ and Rodesch et. al.¹⁹ found male dominance in case of spinal AVM. Casasco et. al¹⁸ reported two cases of female gender in Perimedullary AVF. Although Meng et. al.²⁰ and Cho et al.²¹ showed male preponderance. Mean age was 8.1 in those studies done by Meng et al. and 28 years in Cho et al. In our study all case of perimedullary AVF was female and mean age was 18.33 years. As dural AVF is rare and clinical features are non specific, diagnosis is delay²². In our study, mean period of diagnosis from time of onset was 16.44 months. This finding is comparable to other studies^{13,14}. It was 25 months in the study done by Guillevin R. and his colleagues among 26 patients¹¹. Song JK et al.⁸ found the mean time of 21 months (ranging from 3-60 months). In perimedullary AVF and intramedullary AVM duration between onset and diagnosis was 0.5 to 144 months (mean 16 months)¹⁰.

Clinical Feature:

The typical feature of fistula and spinal malformations had been described by many authors^{23,24}. In this study all patients had motor weakness, 66.6% had sensory disturbances, and 77.7% had urinary disturbances. Gemmete JJ. et al.⁴ found motor weakness in 87% and sensory symptoms in 75% of patients. These are also comparable to other studies (13,14,15). Bowen BC. et al.¹⁵ found 87.5% patients had motor weakness and 62.5% and 75% had sensory disturbances and urinary problem respectively. Song JK et al.⁸ found 75% had leg weakness, 70% had sensory disturbances as the presenting feature. But at the time of diagnosis almost all had the triad of weakness, sensory disturbance, and micturation problem. Most of the patient's symptoms were gradually progressive¹⁷. In our study, all patients had progressive course.

MRI finding:

Typical findings in MRI include T2 hyper intensity in central region, contrast enhancement within spinal cord, and vascular flow voids at the surface of the spinal cord⁵. Many authors described the abnormal T2 hyper intensity changes I the spinal cord in vascular lesion⁶. In our study 88.8% patients had T2 hyper intensity. In the study done by Gilbertson JR et al. T2 signal change was found in all patients and they concluded this as the most sensitive MR findings⁶. Prominent flow voids along the dorsal surface of the cord on T2 sequences is an important MRI finding⁶. In our study, this was present in 77.7% and this finding was comparable with other studies. Bowen BC. et al.¹⁵ found in 62.5% of patients. Song JK et al.⁸ found combination of perimedullary vessels and cord hyperintensity in 89% of patients. Gadolinium enhancement increases the sensitivity and specificity of MR finding ⁶. Gilbertson JR ⁶ found enhancement in 88% and Bowen BC et al.¹⁵ found in 45.45%. In our study five patients did MRI with gadolinium injection. Among them 40% (two of five) of patients had enhancement.

Angiogram:

MRI findings has minimal role for the localization and characterization of vascular malformation⁷. Combination of MRI and MRA (with contrast enhanced and 3D) has approximately 73% sensitivity to locate the fistula level ⁷. The sensitivity, specificity of MRI with MRA is 80-100% and approximately 80% respectively²⁴. Recently advancement in spinal MRA has been grown up with fast 3D contrast enhanced MRA with combination of a rapid bollus injection and good timing mechanism⁷. MRA help reduction of > 50% of radiation and use of contrast agent in DSA. 3D contrast enhanced MRA has the limitation of selecting the fistula level because of long acquisition time and low resolution. Fast (24 sec) contrast enhanced MRA can identify the level of fistula. But for detection of the level, repeated double/ triple MRA session is often required because of small field of view (FOV) Mull M. et al.⁹ found that MRA could identify the level of fistula in 14 out of 19 patients when compared to DSA. In AVM, MRA could identify 10 out of 11 feeding artery. Additional feeders in 5 patients were also missed by MRA⁷.CTA has also

role for detection of vascular diseases. In CTA, data acquisition must be done at the time when contrast agent fills the vessel to be imaged. Single detector CT has low speed: Multidetector has more speed and larger anatomical coverage with higher spatial resolution. MDCT can detect the feeding artery, fistula, and draining vein which correlate with conventional catheter angiography^{7.} Digital subtraction angiography (DSA) is the standard for spinal vascular lesion¹⁷. For classification and diagnosis of spinal vascular lesions, DSA is the definitive test. Both 3D contrast enhanced MRA and MD CTA are not suitable to differentiate the arterial feeder from draining vein in fistula. DSA has the role to distinguish them. Additionally to understand the character of the fistula, to identify any additional feeding artery and to determine whether the feeding artery and the anterior spinal artery arises from the same pedicle, DSA is the gold standard ⁷.

Conclusion:

Spinal AVM and AVF remain undiagnosed for a long period. They should be treated early for prevention of progressive morbidity and disability. MRI features of cord edema, contrast enhancement, and perimedullary vessels may lead to the diagnosis of these vascular lesion. DSA is the gold standard for characterization of the lesion and to determine the treatment modality of the vascular lesion.

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