Management of Vascular Myelopathy

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Abstract: Vascular myelopathies include several diagnoses that are often misdiagnosed or undertreated. Some represent neurologic emergencies, such as spinal cord infarction, and others can be disabling if they remain unrecognized, such as spinal dural arteriovenous fistulas. This article describes the clinical characteristics and current therapeutic strategies for the most common vascular myelopathies and emphasizes practical concepts for the clinician. Vascular myelopathies are infrequent, but their consequences to the patient's functional capacity can be devastating. Because of their relative rarity, these disorders are often initially misdiagnosed, and, in some cases, this delay in arriving at the correct diagnosis can prove very detrimental. Clinicians should be keenly aware of the clinical and radiologic features of the various vascular causes for acute or progressive myelopathy. Optimal management of patients with vascular myelopathies requires close collaboration with neuro-radiologists, neuro-interventionalists, and vascular neurosurgeons. Prognosis should be estimated with caution because functional outcomes over time may be better than initially expected. This article reviews the essential concepts of diagnosis and management of vascular diseases of the spinal cord.
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1. Introduction

Myelopathy describes pathologic conditions that cause spinal cord, meningeal or perimeningeal space damage or dysfunction. Traumatic injuries, vascular diseases, infections and inflammatory or autoimmune processes may affect the spinal cord (Hauser SL, 2005). There are many underlying causes for myelopathy. The most common causes are congenital stenosis and degenerative stenosis caused by spondylosis. Other causes are Ossification of the Posterior Longitudinal Ligament (OPLL), compression by the presence of a tumor, an epidural abscess or trauma. Myelopathy can also be caused by vascular injury to the cord by presence of vascular malformations or ischemic injury (Robertson CE et al., 2012).

Vascular myelopathies include several diagnoses that are often misdiagnosed or undertreated. Some represent neurologic emergencies, such as spinal cord infarction, and others can be disabling if they remain unrecognized, such as spinal dural arteriovenous fistulas. Spinal cord infarctions represent 5% of acute myelopathies. Aortic surgery is the most common cause of spinal cord infarction (Thron A et al., 2003). Spinal vascular malformations (SVM) are rare diseases that consist of true inborn cavernomas and arteriovenous malformations (AVM), and presumably acquired dural arteriovenous fistulae. Depending on the type of vascular malformation, initial symptoms may vary between acute intramedullary or subarachnoidal hemorrhages or subacute venous congestion leading to progressive myelopathy. The space-occupying nature of some of these lesions and a circulatory "Steal" phenomenon are additional possible pathomechanisms (Bostroem A et al., 2007).

When SVM is suspected, MRI should constitute the first diagnostic modality. It detect intramedullary pathologies such as intramedullary hemorrhages, cavernomas, edema or venous congestion, extramedullary intradural alterations such as dilated vessels or subarachnoidal hemorrhage. Magnetic Resonance Angiography (MRA) is a noninvasive technique of imaging based on MRI technology that creates images of blood vessels (Saraf-Lavi E et al., 2002). Selective spinal angiography is the next diagnostic step to define the type of vascular malformation and, thereby, to decide the appropriate therapy (Muralidharan R et al., 2011).

Treatment of vascular myelopathy shows a wide range of modalities and options. Patients with spinal cord cavernomas and perimedullary fistulae type I are surgical candidates (Saladino A et al., 2010). Dural arteriovenous fistulae can either be operated upon or can be treated by an endovascular approach. In spinal arteriovenous malformations, the endovascular

approach is the method of first choice (Gemmete JJ et al., 2013).

2. Materials and Methods

Patients

- Number of patients: Fifteen patients.
- Type of the study: Prospective and retrospective study.
- Inclusion criteria:

1. Age above 2 years old.
2. Motor power affection.
3. Upper motor signs.
4. Sphincteric disturbances.
5. Spontaneous non traumatic intramedullary cord hematoma.

- Exclusion criteria:

1. Age below 2 years old
2. Intact motor power.
3. Radiculopathy without myelopathy.
4. Significant compression on spinal cord due to prolapsed intervertebral discs, compression by the presence of a tumor, compression by an epidural abscess, trauma or compression caused by cervical kyphosis.

Methods

- Preoperative evaluation:
  - History taking:
    - Age and Sex.
    - Onset of complaint.
    - Previous misdiagnoses.
    - Course and progression of complaint.
  - Examination and assessment:
    - Neurological examination.
      - ✓ Motor power and Sensation.
      - ✓ Deep tendon reflexes and Muscle tone.
      - ✓ Sphincteric dysfunction.
  - Investigations:
    - Magnetic resonance imaging (MRI) with and without contrast.
    - Magnetic resonance angiography (MRA).
    - Computed tomographic angiography (CTA).
    - Selective spinal angiography.
  - Patient grading on admission:
    - Aminoff–Logue disability scale (ALDS) (table 1).

Table 1: Aminoff–Logue disability scale (ALDS).

Gait (0–5)		Micturition (0–3)	
G0	Normal.	M0	Normal.
G1	Leg weakness, abnormal gait or stance, but no restriction of activity.	M1	Hesitancy, urgency, frequency, altered sensation, but continent.
G2	Restricted activity but not requiring support.	M2	Occasional urinary incontinence or retention.
G3	Requiring one cane for walking.	M3	Total incontinence or persistent retention.
G4	Requiring two canes, or a walker.		
G5	Confined to wheelchair.		

- Procedure:

Surgical or endovascular management according to the cause.

- Evaluated parameters:
  - Pre and post-intervention motor power and sensory affection.
  - Pre and post- intervention ALDS.
- Follow up:
  - Evaluation of clinical outcome at the time of hospital discharge, then 3 months post procedure through Aminoff–Logue disability scale.
  - Selective spinal angiography after 6 months.

Statistical Analysis

Data was analyzed using SPSS (Statistical Package for Social Sciences) version15. Qualitative data was presented as number and percent. Comparison between groups was done by Chi-Square test. Quantitative data was tested for normality by Kolmogrov- Smirnov test. Normally distributed data was presented as mean ± SD. Non parametric data was presented as min – max and median. Mann-Whitney test was used for comparison between groups. Spearman's correlation coefficient was used to test correlation between variables. The ROC curve analysis has the ability to discriminate diseased cases from normal cases. $P < 0.05$ was considered to be statistically significant.

3. Results

The age of the patients of this study ranged from 8 to 63 years, with a mean of 39.27 years (± 18.43 SD). Eleven of the fifteen patients were males with a percentage of 73.3% and four were females with a percentage of 26.7% (Table 2) (Figure 1).

Table 2: Gender distribution of vascular myelopathy patients in the study.

Gender	No	%
Male	11	73.3
Female	4	26.7

Patients were sorted out into four groups according to definitive diagnosis: patients with spinal AVM, patients with dural AVF, patients with perimedullary AVF and patients with cavernous malformation (Figure 2).

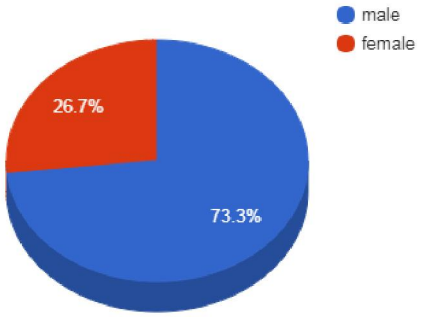

Figure 1. Gender distribution of vascular myelopathy patients in the study

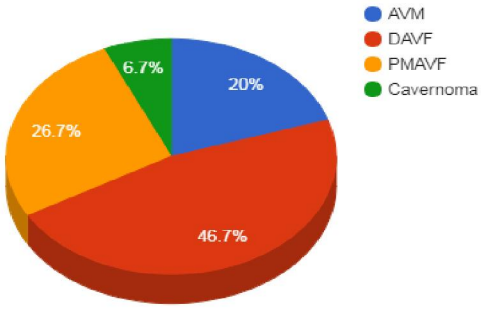

Figure 2. Distribution of various types of vascular malformations in the study.

Spinal AVMs group:

Three patients had spinal AVM with a percentage of 20% (Figure 2), their age ranged from 20 to 29 years, with a mean of 24.66 years (± 4.50 SD). Two patients were males (66.7%) and only one female (33.3%) (Table 3).

Table 3: Gender distribution of patients of spinal AVM group.

Gender	No	%
male	2	66.7%
female	1	33.3%

i. Clinical presentations:

All patients in the spinal AVM group were presented with acute onset of myelopathy. The most common initial presenting symptom was paraparesis (66.7%) followed by quadriparesis (33.3%). Two patients (66.7%) had a sensory disturbance during the course of their disease. Two patients experienced sphincteric dysfunction (66.7%) (Table 4).

Table 4: Distribution of clinical findings in patients of spinal AVM group.

Clinical presentations	No	%
Quadriparesis	1	33.3%
Paraparesis	2	66.7%
Hypoesthesia	2	66.7%
Sphincteric dysfunction	2	66.7%

Initial diagnosis and misdiagnosis:

The mean time from symptoms onset to diagnosis was 3 weeks (± 2 SD).

ii. Imaging findings:

All patients had an area of T2 high signal intensity in the spinal cord, suggesting venous congestion and cord edema. T2 perimedullary signal voids along the spinal cord were seen in two patients (66.7%). Hypertrophied spinal vessels on MRI were seen in two patients (66.7%). An intramedullary mass of turgid blood vessels on MRI was seen in all patients (Table 5). Spinal angiography was performed in all patients. The location of the AVM nidus occurred most frequently at the lower dorsal (D7–D12) level (66.7%), followed by the cervical region (33.3%). The feeders were mostly multiple (66.7%) (Table 6).

Table 5: Distribution of MRI findings in patients of spinal AVM group.

MRI findings	No	%
Intramedullary mass	3	100.0%
T2 perimedullary signal voids	3	100.0%
T2 hyperintensity	3	100.0%
hypertrophied vessels	2	66.7%

Table 6: Distribution of spinal angiography findings in patients of spinal AVM group.

Location	No	%	Feeders	No	%
Cervical	1	33.3%	single	1	33.3%
Dorsal	2	66.7%	multiple	2	66.7%

Treatment:

The preferred first-line treatment was endovascular embolization, all patients underwent endovascular treatment. Two patients achieved a complete elimination of their lesion (66.7%). One had a partially embolization and a remnant nidus (33.3%). The complete obliteration rate for endovascular therapy was 66.7%, and the success rate including partial embolization was 100%.

Outcome:

The mean pre-treatment ALDS was 5 (±1.73 SD). The mean post-treatment ALDS was 2 (± 1 SD). All patients had an improvement in their symptoms.

Dural AVFs group:

Seven patients had Dural AVF with percentage of 46.7 % (Figure 2), their age ranged from 25 to 63 years, with a mean of 51.42 years (±12.69 SD). Six patients were males (85.7%) and one female (14.3%) (Table 7).

Table 7: Gender distribution of patients of dural AVF group.

Gender	No	%
male	6	85.7 %
female	1	14.3 %

Clinical presentations:

The most common initial presenting symptom was paraparesis (42.9%); quadriparesis (28.6%) was seen associated with a fistula at the cervical level, followed by paraplegia (28.6%). Six patients (85.7%) had a sensory disturbance during the course of their disease. Sensory symptoms included hypoesthesia (57.1%) and sensory loss (28.6%). All patients experienced sphincter dysfunction in the form of urinary retention and incontinence (Table 8). The clinical course of SDAVF patients was also variable; chronic progression was the most common (71.4%), followed by acute onset (28.6%).

Table 8: Distribution of clinical findings in patients of dural AVF group.

Clinical findings	No	%
Acute onset	2	28.6%
Chronic	5	71.4%
Quadriparesis	2	28.6%
Paraparesis	3	42.9%
Paraplegia	2	28.6%
Hypoesthesia	4	57.1%
Sensory Loss	2	28.6%
Sphincteric dysfunction	7	100.0%

Initial diagnosis and misdiagnosis:

The mean time from symptom onset to diagnosis was 58.71 weeks (± 66.89 SD). Three patients (42.9%) were initially misdiagnosed.

Imaging findings:

All seven patients had an area of T2 high signal intensity in the spinal cord, suggesting venous congestion and cord edema (Table 9). The mean length of cord edema was 6.71 (±1.88 SD) vertebral body segments. A focal area of low T2 signal within the cord consistent with intramedullary hemorrhage was seen in one patient (14.3%). T2 perimedullary signal voids along the spinal cord, a characteristic finding of SDAVF, were seen in 6 of 7 patients (85.7%). Cord expansion on MRI was seen in 4 patients (57.1%). An intradural mass of turgid blood vessels on MRI was seen in one patient (14.3%).

Spinal angiography was performed in all patients (Table 10). The location of the fistula occurred most frequently at the lower dorsal (D7–D12) level (57.1%), followed by the cervical region (42.9%). The feeders were mostly single (71.4%).

Table 9: Distribution of MRI findings in patients of dural AVF group.

MRI findings	No	%
Intradural mass	1	14.3%
T2 perimedullary signal voids	6	85.7%
T2 hyperintensity	7	100.0%
Cord expansion	4	57.1%
Intramedullary hemorrhage	1	14.3%

Table 10: Distribution of spinal angiography findings in patients of dural AVF group.

Location	No	%	feeders	No	%
Cervical	3	42.9%	single	5	71.4%
Dorsal	4	57.1%	multiple	2	28.6%

Treatment:

The mean time from symptom onset to treatment was 60.85 weeks (±67.39 SD). Patients underwent either endovascular treatment or surgery (Table 11). The preferred first-line treatment was endovascular embolization (71.4%). All five patients who underwent endovascular therapy achieved a complete elimination of their fistula. Two patients received primary surgery (28.6%) due to technical difficulties for embolization. The two patients had a successful and complete ligation of their fistula. The complete obliteration rate for both endovascular therapy and surgical treatment was 100 %.

Table 11: Distribution of treatment modalities in patients of dural AVF group.

Intervention	No	%
Endovascular embolization	5	71.4%
Surgical obliteration	2	28.6%

The mean pre-treatment ALDS was 6.57 (±1.27 SD). The post-treatment ALDS was 3.28 (±2.81 SD). All patients had a lower ALDS after treatment. The majority of patients had an improvement in their symptoms. Two patients didn't show a significant improvement in their symptoms. Patients were classified into two groups according to post-treatment ALDS (Table 12).

We compared the differences in various factors between the two groups. A high Aminoff–Logue disability score before treatment, extensive cord edema on MRI and a long duration from symptoms

onset to treatment were identified as factors associated with a poor prognosis.

Table 12: Prognostic factors in the outcome of DAVF patients.

Prognostic factor	Good outcome group (post ALDS <6) n = 5	Poor outcome group (post ALDS score ≥ 6) n = 2	*P* value
Age, years, mean ± SD	47.4 ±13.03	61.5 ±2.12	0.159
Gender (M:F)	4:1	2:0	0.759
Pre ALDS, mean ± SD	6 ±1	8 ±0	0.008
Time to treatment, weeks, mean ± SD	26.6 ± 21.31	146.5 ± 70.00	0.012
Extent of cord edema, mean ± SD	5.8 ±1.92	7.5 ±0.70	0.010
Number of feeders (single: multiple)	3:2	1:0	0.065
Treatment modality (endo.; surgery)	3:2	2:0	0.180

Perimedullary AVFs group:

Four patients had Perimedullary AVF with a percentage of 26.7% (Figure 2), their age ranged from 8 to 50 years, with a mean of 26.25 years (±20.62 SD). Two patients were males and two were females (Table 13).

Table 13: Gender distribution of patients of perimedullary AVF group.

Gender	No	%
male	2	50 %
female	2	50%

Clinical presentations:

All patients in the perimedullary AVF group were presented with a chronic, progressive course of myelopathy. The most common initial presenting symptom was paraparesis (100%), three patients experienced sphincter dysfunction (75%) and one patient (25%) had a sensory disturbance during the course of the disease (Table 14).

Table 14: Distribution of clinical findings in patients of perimedullary AVF group.

clinical findings	No	%
Chronic	4	100.0%
Paraparesis	4	100.0%
Hypoesthesia	1	25.0%
Sphincteric dysfunction	3	75.0%

Initial diagnosis and misdiagnosis:

The mean time from symptom onset to diagnosis was 66 weeks (±36 SD). One patient (25%) was initially misdiagnosed.

Imaging findings:

Three patients underwent spinal MRI as the initial imaging study (75%). One patient, with a previous history of surgical excision of a cranial AVM and use of surgical clips that weren't MRI compatible, underwent CTA (25%) that revealed a vascular lesion in the dorsal spinal canal.

All three patients who underwent spinal MRI had an area of T2 high signal intensity in the spinal cord, suggesting venous congestion and cord edema (Table 15). Perimedullary T2 signal voids along the spinal cord were seen in all patients. Cord expansion on MRI was seen in one patients (33.3%). Hypertrophied spinal vessels on MRI were seen in two patients (66.7%). An intradural mass of turgid blood vessels on MRI was seen in two patient (66.7%). Spinal angiography was performed in all patients (Table 16). The location of the fistula occurred most frequently at the lower dorsal (D7–D12) level (75%), followed by the upper dorsal (D1–D6) level (25%). The feeders were mostly multiple (75%).

Table 15: Distribution of MRI findings in patients of perimedullary AVF group.

MRI findings	No	%
Intradural mass	2	66.7%
T2 perimedullary signal voids	3	100.0%
T2 hyperintensity	3	100.0%
Cord expansion	1	33.3%
hypertrophied vessels	2	66.7%

Table 16: Distribution of spinal angiography findings in patients of perimedullary AVF group.

Location	No	%	Feeders	No	%
Upper dorsal (D1-D6)	1	25.0%	single	1	25.0%
Lower dorsal (D7-D12)	3	75.0%	multiple	3	75.0%

Treatment:

The mean time from symptom onset to treatment was 57.33 weeks (±35.5 SD). Most patients (3/4) underwent either endovascular treatment or surgery. One patient refused any intervention after spinal angiography (Table 17). The preferred first-line

treatment was endovascular embolization. Two patients who underwent endovascular therapy achieved a complete elimination of their fistula. One patient received primary surgery due to technical difficulties for embolization. He had a successful and complete ligation of the fistula. The success rate for both endovascular therapy and surgical treatment was 100%.

Table.17: Distribution of treatment in patients of perimedullary AVF group.

Intervention	No	%
Endovascular embolization	2	50.0%
Surgical obliteration	1	25.0%
Patient refused intervention	1	25.0%

Outcome:

The mean pre-treatment ALDS was 5 (± SD 2). The mean post-treatment ALDS was 1.33 (±2.30 SD). All patients had an improvement in their symptoms.

Cavernous malformation:

One male patient, 50 years old, had Cavernous malformation with a percentage of 6.7 % (Figure 2). He was presented with progressive quadriparesis and urinary urgency for 5 months, Aminoff–Logue disability score was 2. MRI of the cervical spine revealed a well defined intramedullary lesion, associated cord expansion, high signal intensity on T1 and T2 suggestive of hemorrhage with a low signal intensity margin likely a hemosiderin ring. MRI findings were suggestive of intramedullary spinal cord cavernous malformation. Patient refused any further intervention and was discharged upon his request.

4. Discussion

Vascular myelopathy include several diagnoses that are often misdiagnosed or undertreated. Recent years have brought advances in diagnostic imaging and treatment alternatives as well as useful information regarding prognosis. Refinement in MRI technique now allows precise, noninvasive diagnoses of most vascular myelopathies and is crucial for the exclusion of differential diagnoses. Surgical and endovascular therapies are highly effective in treating spinal vascular malformations. Studies have shown that the prognosis of vascular myelopathy is more favorable than previously conceived and even patients with severe deficits can achieve meaningful recovery.

I. Spinal AVMs group:

Spinal cord AVMs account for 20 to 30% of all spinal arteriovenous malformation. 20% of the patients enrolled in this study were diagnosed with spinal AVM their age ranged from 20 to 29 years, with a mean of 24.66 years (± 4.50 SD). Our results are similar to the literature (Oldfield et al., 1988). In our study, there is a male predominance (66.7%) but in literature spinal AVMs occur in males and females at a nearly equal incidence (Rosenblum et al. 1997). This difference is mainly due to the small sample size in our study.

i. Clinical presentations:

Hemorrhage is seen in up to 50% of patients. About 25% patients present with motor and sensory symptoms. The risk of re-bleeding in patients presenting with hemorrhage is 10% at one month and 40% at one year. Non-hemorrhagic manifestations include back pain, radicular pain, motor/sensory deficits, sexual disturbance, sphincter disturbances and bruit. Five factors contribute to the spectrum of clinical manifestations in patients with spinal AVMs:

- Hemorrhage: Up to 50% of patients present with subarachnoid hemorrhage. These patients may present with moderate to severe backache and sudden neurologic deficit (van Beijnum et al. 2007).
- Venous hypertension: The arterialized veins have dysplastic walls and are not capable of handling high blood pressure. The resulting venous hypertension either leads to rupture of these vessels and hemorrhage or causes venous congestion and ischemia of the surrounding neural tissue (Germans et al. 2008).
- Venous thrombosis: Partial or complete thrombosis predisposes to ischemic damage of surrounding parenchyma and hemorrhage.
- Vascular steal: The phenomenon of vascular steal is seen in high-flow AVMs. The AVM vessels are dysplastic and do not respond to regulatory signals.
- Mechanical compression: The arterialized veins can cause compression of the surrounding parenchyma leading to progressive neurologic deficits.

In our study, all patients in the spinal AVM group were presented with acute onset of myelopathy. The most common initial presenting symptom was paraparesis (66.7%), hypoesthesia (66.7%) and sphincteric dysfunction (66.7%). One patient was presented with hemorrhage.

ii. Imaging findings:

MRI features suggestive of spinal AVMs include a conglomerate of dilated, peri- and intramedullary located vessels that are demonstrated on T2-weighted sequences as flow voids and on T1-weighted sequences, depending on their flow velocity and direction, as mixed hyper-hypointense tubular structures, a serpentine pattern of low signal on T1 and T2, scalloped appearance on T1 and a venous congestive edema may be present as an intramedullary hyperintensity on T2-weighted images with concomitant swelling of the cord (Thron et al. 2003). In our study, all patients had an area of T2 high signal intensity in the spinal cord. An intramedullary mass of turgid blood vessels on MRI was seen in all patients.

T2 perimedullary signal voids along the spinal cord were seen in 66.7% of patients. Hypertrophied spinal vessels on MRI were seen in 66.7% of patients (figure 3).

Spinal angiography is the gold standard for diagnosis and characterization of spinal AVMs. It also offers the opportunity to treat these lesions in the same sitting. These lesions are most often located in the cervicodorsal area, with multiple feeders. About 20-40% of spinal intramedullary AVMs are associated with aneurysms and their presence is associated with increased risk of bleeding (Caragine et al. 2002). In our study, the location of the AVM nidus occurred most frequently at the dorsal level 66.7%, followed by the cervical region 33.3%. The feeders were mostly multiple 66.7% (figure 4 A, B).

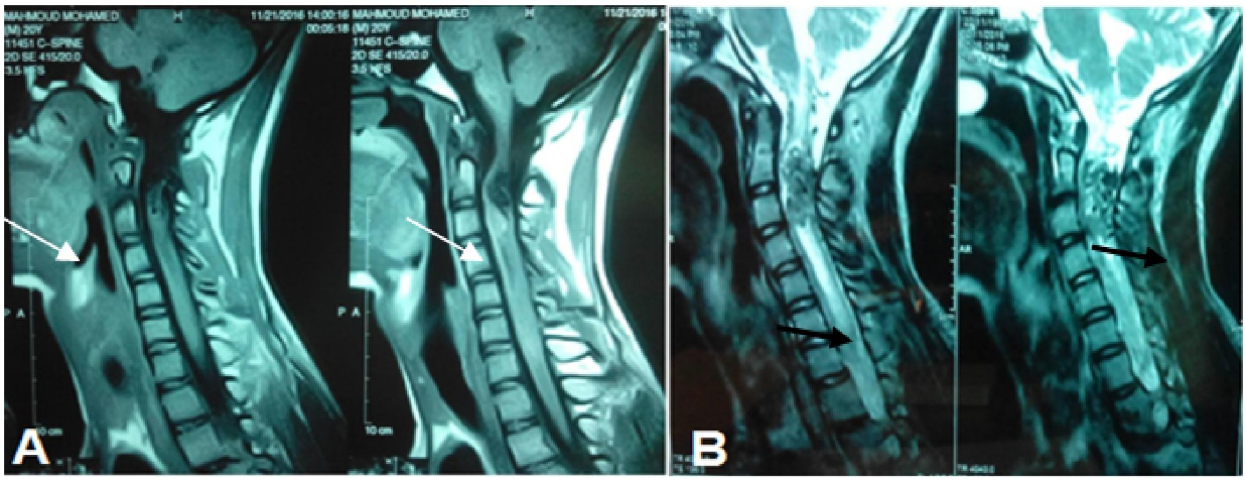

Figure.3 MRI cervical spine of case (1).
(A) T1 images show a lesion opposite C 2-3 (white arrow).
(B) T2 images show hyperintensity signal within the cord (black arrow) and internal flow voids.

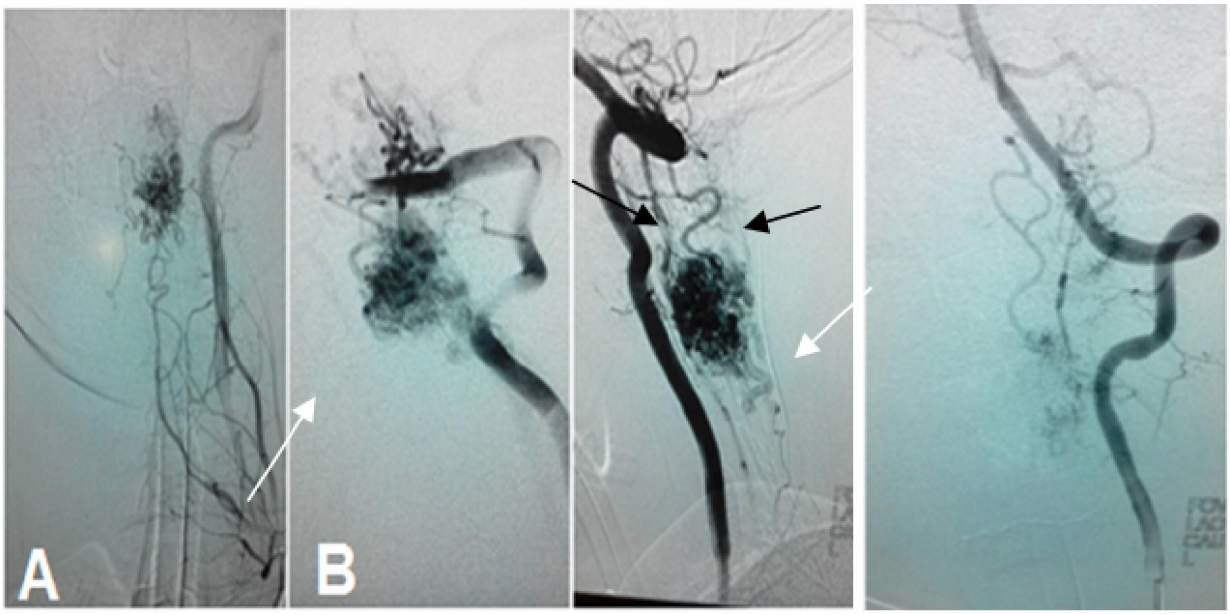

Figure.4 Pre-intervention DSA of case (1) shows: glomus intramedullary AVM (white arrow) with arterial feeder arising from thyrocervical trunk (A) and two arterial feeders (black arrows) arising from the left vertebral artery (B). (C) Post-embolization DSA shows: complete obliteration of the AVM

iii. Treatment:

First-line treatment is embolization because surgical treatment aiming to remove the nidus from the spinal cord carries a significant risk. In a surgical series of 20 intramedullary AVM cases, 20% patients suffered from worsening of postoperative neurological

symptoms (Bostroem et al. 2009). Although a few previous reports indicated that surgery is a feasible option in intramedullary AVM (Takai et al. 2015) surgery should only be indicated for symptomatic cases when embolization is too hazardous or if endovascular treatment results in an incomplete obliteration. In a recent report utilizing Onyx for 17 intramedullary AVM patients, complete obliteration was attained in 37.5% patients, whereas 82% patients experienced clinical improvements (Corkill et al. 2007). Radiosurgery is another treatment option for spinal AVMs. Hida et al. treated 10 patients with hypo fractionated linear accelerator stereotactic radiotherapy, all of whom presented with hemorrhage (Hida et al. 2003).

In our study, all patients underwent endovascular treatment. 66.7% of the patients achieved a complete elimination of their lesion (figure 4 C). 33.3% had partially embolization and a remnant nidus. There were no significant complications related to endovascular intervention. The complete obliteration rate for endovascular therapy was 66.7%, and the success rate including partial embolization was 100%. The mean post-treatment ALDS was 2 (± 1 SD). All patients had an improvement in their symptoms, even the patient with partial embolization.

II. Dural AVFs group:

SDAVFs represent 80% of spinal vascular malformations (Krings et al. 2010). 46.7% of the patients enrolled in this study were post-angiographically diagnosed with dural arteriovenous fistula. This difference is mainly due to the small number of cases in our study.

SDAVF usually affect males over 40 years. Our current data show age ranging from 25 to 63 years (mean age 51.42 ±12.69). These results are comparable to other published data (Donghai et al. 2013).

Our current data show that SDAVFs display a significant male predominance, similar to prior reports (Saladino et al. 2010, Wakao et al. 2012). The reason for this demographic feature is unclear, but differences in sex hormones might increase the likelihood of SDAVF in middle-aged men compared to women (Dorsett-Martin et al. 2007).

i. Clinical presentations:

The clinical manifestations of SDAVFs are progressive upper motor weakness of the lower limb in most patients, numbness, pain, sphincter disturbances and sexual dysfunction (Fugate et al. 2012). The underlying pathophysiological mechanisms result from shunting of high-pressure arterial blood into the spinal veins, leading to arterial steal, ischemia, venous congestion and progressive spinal cord necrosis. Acute neurological deterioration can occur due to hemorrhage from the fistula or thrombosis of the pathological veins, which is known as the "Foix-Alajouanine syndrome" (Song et al. 2010).

In our study the most common initial presenting symptom was paraparesis (42.9%); occasionally quadriparesis (28.6%) was seen associated with a fistula at the cervical level. The severity of motor weakness was variable, ranging from subtle weakness to paraplegia (28.6%). All patients experienced sphincter dysfunction in the form of urinary retention and incontinence. The clinical course varied in our patients from acute to a very chronic progression of myelopathy. Chronic progression was the most common (71.4%), followed by acute worsening (28.6%). Hemorrhagic complications from SDAVF are uncommon (Lucas et al. 2012). Only one patient was presented with intramedullary hemorrhage in our current study.

ii. Initial diagnosis and misdiagnosis:

SDAVF are associated with a long time to diagnosis compared to other myelopathies (Muralidharan et al. 2011). In our current study series, the longest diagnosis took 192 weeks. The mean time from onset to diagnosis was 58.71 weeks (± 66.89 SD), this similar to other published data (Jellema et al. 2006).

Delayed diagnosis and misdiagnosis of SDAVF is common. SDAVF can be commonly misdiagnosed as degenerative disc disease, myelitis, cord tumor, Guillain–Barré syndrome and other conditions (Donghai et al. 2013). In our study three patients were initially misdiagnosed. The difficulty in making an early diagnosis is largely explained by the rarity of the disease and the non-specific signs and symptoms, particularly in the initial phase.

iii. Imaging findings:

High signal intensity with a swollen spinal cord on the T2-weighted images is a common finding in SDAVF. In our patients the cord swelling was extensive, affecting more than three vertebral body levels. The sensitivity of T2 high signal intensity in the spinal cord in our study was 100% similar to prior published data (Luetmer et al. 2005). Perimedullary T2 signal void along the spinal cord is a characteristic finding of SDAVF. From our data, the sensitivity of a perimedullary flow void on a T2-weighted image was 85.7%. One study reported a sensitivity of 77% (Donghai et al. 2013), and another reported that 78 of 93 patients (83.9%) had vessel abnormalities on a reviewed MRI (Muralidharan et al. 2011). Another study reported a quite high sensitivity of 95% (Lee et al. 2016) (figure 5).

Spinal angiography is the diagnostic gold standard for SDAVF (figure 6 A, B). Like many other reports (Koch et al. 2003, Jellema et al. 2006) our current data also show that the dorsal level was the most frequently affected (57.1%). Most of our patients

have a single feeder (71.4%) this is comparable with other published data (Donghai et al. 2013). Spinal angiography is not only a diagnostic choice, but also an essential modality in the treatment strategy. Precise identification of the fistula level is important in both endovascular and surgical treatment.

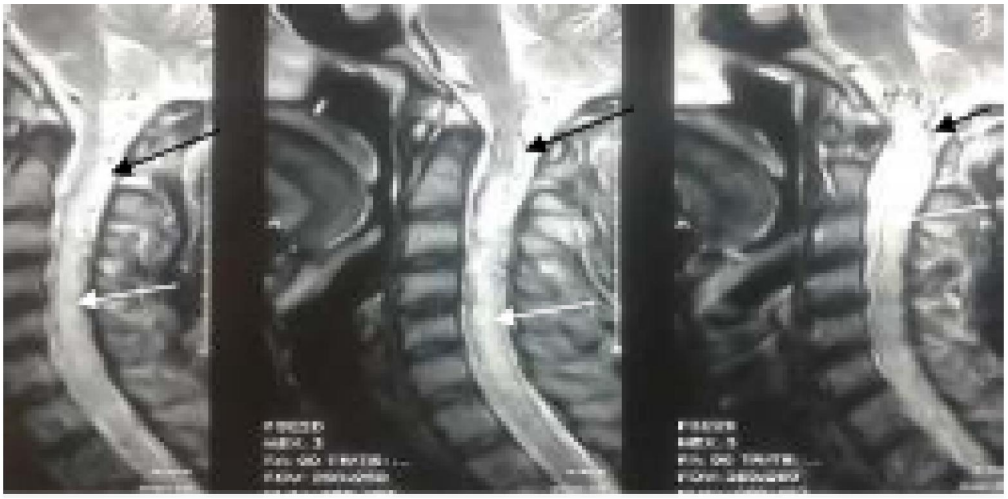

Figure.5 MRI cervical spine of case (10) T2 sagittal images show central cord hyperintensity (white arrow) and multiple signals void posterior to the cord (black arrow).

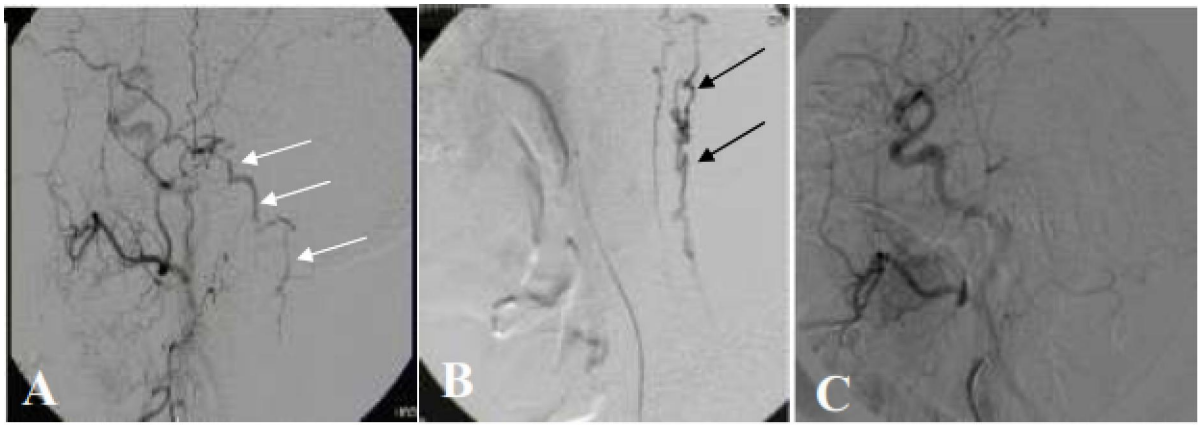

Figure.6 (A, B) Pre- intervention DSA of case (5) shows: a dural arteriovenous fistula (white arrows) arising from the left middle meningeal artery draining to spinal perimedullary veins (black arrows). (C) Post-embolization DSA shows: complete obliteration of the fistula.

iv. Treatment:

Both endovascular and surgical therapies are known to be effective in treatment of SDAVF. Controversies still exist regarding the best treatment modality for SDAVF; some authors favor surgery (Chibbaro et al. 2015, Nagata et al. 2006), whereas others recommend endovascular treatment (Medel et al. 2009, Andres et al. 2008).

Endovascular treatment is preferred, because it is safe and non-invasive. The technical difficulties of identifying a fistula, the possible risk of spinal cord infarction or unavailability of endovascular facilities are the main reasons for choosing surgical intervention (Gemmete et al. 2013).

In our study, the first-line treatment was endovascular embolization (5/7, 71.4%) (figure 6 C). All patients who underwent endovascular therapy achieved a complete elimination of their fistula. Two patients received primary surgery (2/5, 28.6%) due to technical difficulties for embolization.

Variable success rates have been reported for endovascular embolization (Patsalides et al. 2011). However, our study has shown a good success rate of embolization (100%) without significant complications. Our patients who received surgical treatment achieved a complete occlusion without any recurrence.

v. Outcome and prognostic factors:

Clinical outcome after intervention of a spinal DAVF is difficult to predict. It is difficult to assess the prognosis of every patient after intervention as regards to when and how much he will improve. Anatomical or angiographic cure of spinal fistula does not mean clinical cure. From the reported series in the literature, it seems that the most important factors affecting the outcome after successful treatment are the pre-treatment neurological condition, the time to intervention and the age of the patient (Cecchi et al. 2008), (Nagata et al. 2006).

In our series, there was a significant relationship (p=0.008) between the preoperative neurological condition and outcome. There was a significant relationship (p=0.010) between the extent of cord edema and the outcome. There was tendency to improve in younger age, but this was statistically insignificant (p=0.159). The same was found in the study of Schuss et al. 2015.

In our series, the shorter the time to intervention the better was the outcome (p=0.012). In some previous series, the duration of symptoms before intervention failed to show a significant correlation with postoperative outcome (Cecchi et al. 2008), (Nagata et al. 2006). In others, there was significant improvement with earlier intervention from the onset of symptoms (Inagawa et al. 2013), (Ofran et al. 2013).

Our data indicated that the most important factors in prognosis were severity of a preoperative deficit, the extent of cord edema, and the duration of symptoms. Our current results are largely consistent with the findings of previous studies (Muralidharan et al. 2011), (Iovtchev et al. 2015).

III. Perimedullary AVFs group:

PMAVF and accounts for 17-35% of spinal vascular malformations. They usually occur in young patients and mostly in second or third decade (Antonietti et al. 2010). 26.7 % of the patients enrolled in this study were post-angiographically diagnosed with perimedullary arteriovenous fistula. The age of presentation is very controversial in the literature. The mean age of presentation has been found to range from 19.5 to 45 years (Cho et al. 2005). Our current data showed age ranging from 8 to 50 years, with a mean of 26.25 years (±20.62 SD). In our study, Perimedullary AVFs were distributed equally between males and females, these results are comparable to other published data (Rodesch et al., 2005). However, in 2013, Gross et al. summarized 213 cases reported from 28 studies and found that patients PMAVFs were significantly older (mean age, 46.9 years) and primarily males (68% male) (Gross et al. 2013).

Although the etiology is not clearly determined in most instances, there are well-documented cases of both congenital and acquired PMAVFs (Meng et al. 2010). Most occurrences are presumed to be congenital lesions. PMAVFs may be part of a more complex vascular malformation syndrome in 10.5% (Meng et al. 2010), such as metameric Cobb syndrome and Hirschsprung's anomaly. Acquired fistulas have been described following excision of conus medullaris ependymoma and removal of teratoma of the cauda equine (Tender et al. 2005).

i. Clinical presentations:

All patients in the perimedullary AVF group were presented with a chronic, progressive course of myelopathy. In the series of Halbach et al. four of five patients suffered from acute onset symptoms (Halbach et al. 1993). Acute onset of symptoms seems to be more frequent in children (Meng et al. 2010). The two children in our current study were presented with chronic myelopathic symptoms. The initial symptoms may be leg weakness, sensory disturbance, back pain, or sphincter disorders (Wakao et al. 2012). In our current study, the most common initial presenting symptom was paraparesis (100%) followed by sphincter dysfunction (75%). The incidence of hemorrhage is about 10–40%. Rodesch et al. reported that children with PMAVFs had a greater tendency to bleed than adults did 70% vs. 45% (Rodesch et al. 2002). In our study, no patient had suffered any type of hemorrhage. The bleeding mechanism is unknown. Venous impairment probably occurs due to high flow and venous hypertension (Cho et al. 2005).

ii. Initial diagnosis and misdiagnosis:

Unless there are acute neurological findings or congenital anomalies indicative of intraspinal pathology, the diagnosing of PMAVFs will remain very difficult and will often be delayed by an average of 4.9 years (Scarff et al. 1979), and 9 years (Mourier et al. 1993). The mean time from symptom onset to diagnosis was 66 weeks in our current study and one patient was initially misdiagnosed.

iii. Imaging findings:

MRI is the preferred method for screening examinations on suspected patients. MRI shows perimedullary serpiginous structures of signal voids produced by rapid blood flow on spin-echo images. Hypertrophied spinal vessels on MRI were seen in 66.7% of the patients in PMAVFs group. T2 high signal intensity in the spinal cord was seen in all patients who underwent MRI (figure 7).

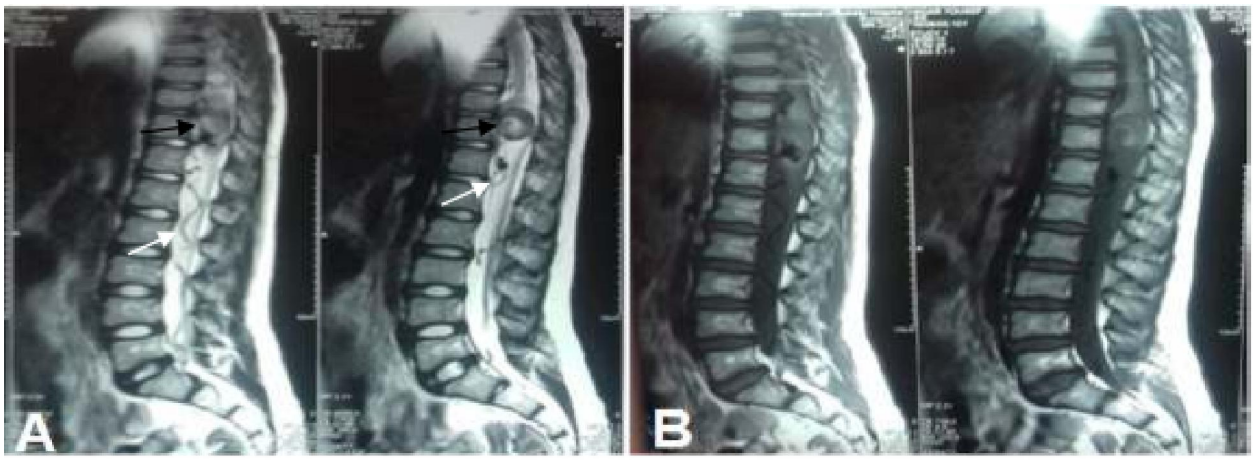

Figure.7 MRI dorsolumbar spine of case (11)
(A) T2 sagittal images show *a*n intradural extramedullary lesion (black arrow), central cord hyperintensity, and hypertrophied spinal vessels (white arrow).
(B) T1 sagittal images show hypointensity signal and cord expansion.

Multi-detector CT angiography (CTA) is a recent imaging technique that can provide high-resolution and high contrast images. This can locate the feeder vessels and the fistula, and greatly reduce the amount of time required for conventional angiography (Lai et al. 2006). CTA was very helpful in our current study; it was able to demonstrate a spinal vascular lesion in a patient who couldn't do MRI because of surgical clips that weren't MRI compatible (figure 8).

Spinal angiography remains the gold standard for characterizing the angioarchitecture of spinal vascular malformations. Only spinal angiography can accurately demonstrate a PMAVF by showing the high-flow shunt and the absence of interposed nidus between the arterial and venous sides of the vascular lesion. It is important to identify the exact number of arterial feeders. In our series, the majority of patients (75%) had multiple feeders, just as indicated by other reports (Meng et al. 2010, Rodesch et al. 2003). The fistula site may be identified by abrupt changes in the caliber of a blood vessel at the transition from feeder artery to draining vein (figure 9).

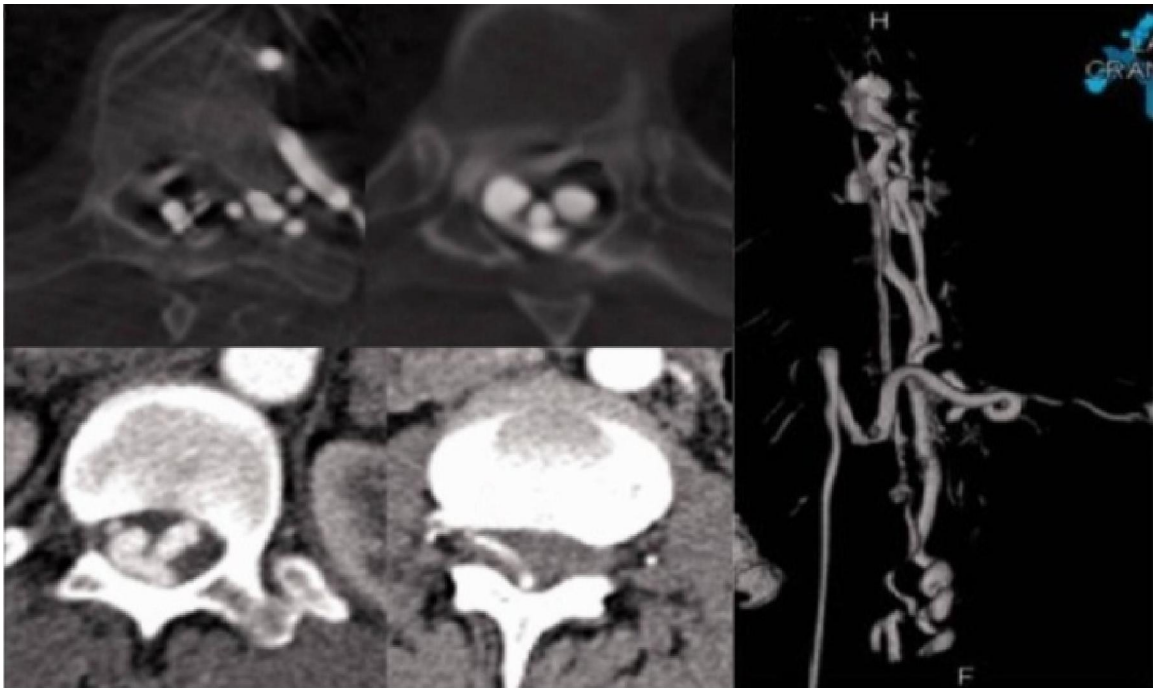

Figure.8 CTA dorsal spine of case (13) shows a vascular-enhancing lesion in the thoracic spinal canal

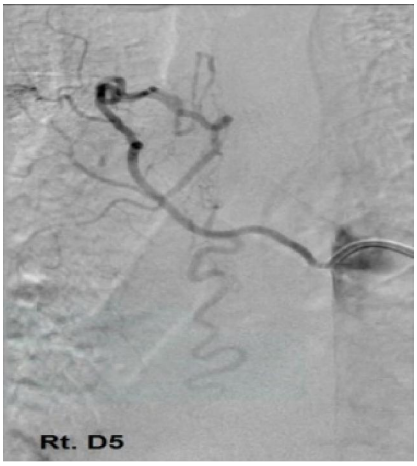

Figure.9: Pre-intervention DSA of case (14) shows: a perimedullary arteriovenous fistula arising from the Rt. D5.

These lesions can occur anywhere along the spinal axis but most commonly in the dorsolumbar region and, to a lesser extent, in the upper cervical region (Cho et al. 2005). In the present series, PMAVFs arise in the dorsal spine mainly at the lower dorsal (D7–12) level (75%), followed by upper dorsal (D1–6) level (25%).

iv. Treatment:

Management of PMAVFs is more demanding than SDAVFs. Surgery for a PMAVF is somewhat more challenging because of the anatomical location and involvement of the anterior or posterior spinal arteries in their formation (Oran et al. 2005). The aim of the treatment is to obliterate the fistula while preserving the normal arterial supply to the spinal cord. Closure of the proximal side of a feeder artery may not be enough to achieve complete cure of the AVF because many shunts are supplied by more than one feeder, as was shown in our series.

Recent advances in endovascular techniques and materials have allowed replacement of the complex and risky microsurgery of PMAVFs by endovascular embolization. Embolization presents the great advantages of continuous intraoperative angiographic monitoring and absence of direct maneuvers on the parenchyma of the spinal cord. Endovascular embolization is preferred for these lesions nowadays (Meng et al. 2010). However, difficulties may rise from the small size of the pedicles and from the possibility of selective occlusion of the AVF in relation to the functional spinal arteries (Inoue et al. 2006). When navigation through tortuous medullary arteries is not possible, transvenous techniques may allow access to the draining varix (Touho et al. 1995).

In our current study, two patients underwent endovascular therapy and achieved a complete elimination of their fistula. There were no significant complications related to endovascular intervention. The success rate for endovascular therapy was 100 % (figure 10).

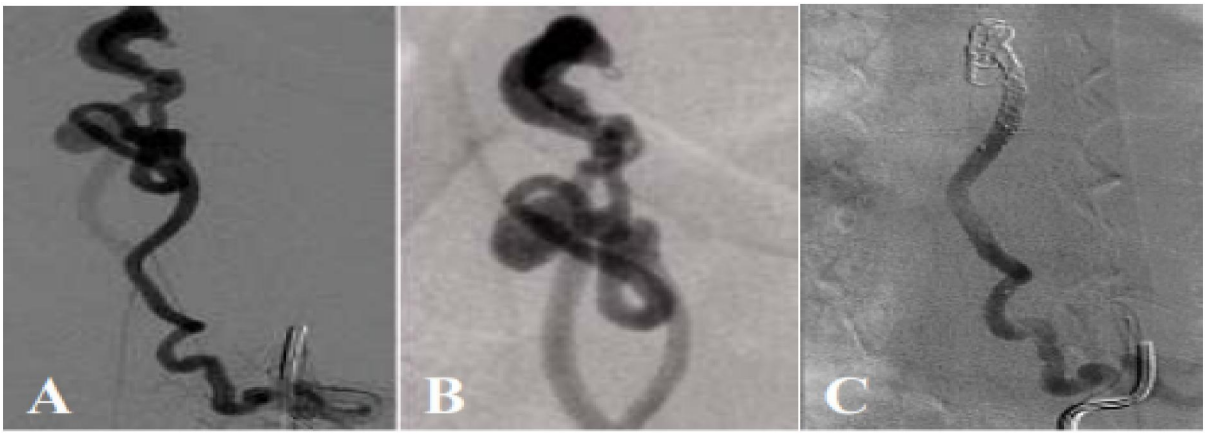

Figure.10 (A, B) Pre-intervention DSA of case (13) shows: a perimedullary arteriovenous fistula arising from the Lt. D9. (C) Post-embolization DSA shows: complete obliteration of the fistula.

With regard to surgery, the difficulties basically relate to the location of the AVF, when it lies against the anterior aspect of the spinal cord. A modified posterolateral approach for lesions involving the anterolateral pial surface of the spinal cord was proposed by Martin et al. (Martin et al. 1995). An anterior cervical approach involving corpectomy at two levels was successfully used by Hida et al. (Hida et al. 2002), while complete corpectomy by means of thoracotomy was advocated by Anderer et al. (Anderer et al. 2008) and lumbar corpectomy was used by Vitarbo et al. (Vitarbo et al. 2005) in one case. A sudden change in the caliber of the feeder artery going to the venous side demonstrates the fistula site.

In this current series, only one patient with dorsally located PMAVF was managed by surgery due

to technical difficulties for embolization. He had a complete ligation of the fistula. There were no significant complications related to surgery. The success rate for surgical treatment was 100 %.

v. Outcome:

Reports of the outcome of perimedullary fistulas are poor in the literature (Antonietti et al. 2010). The outcome of perimedullary fistulas versus dural fistulas is not clearly determined in the literature. However, we believe that perimedullary fistulas have better outcome than the dural type as shown in the results of this study, in PMAVFs patients The mean post-treatment ALDS was 1.33 ±2.30 while in DAVFs patients the mean post-treatment ALDS was 3.28 ±2.81, but the small number of cases has impacted the statistical power and further studies with larger series are therefore needed to support or deny this hypothesis.

IV. Cavernous malformation:

Cavernous malformations represent 5% to 10% of all spinal vascular abnormalities (Cosgrove et al., 1988). Cavernous malformations represent 6.7 % of vascular malformations in our study which is consistent with literature. According to the literature, mean patient age at time of presentation was 39.1 years (late 30s/early 40s) (Gross et al. 2010). The patient in our study was 42 years old which is consistent with literature.

Early reviews noted a preponderance of female patients (Zevgaridis et al. 1999). The literature then shifted toward a more balanced distribution, with no clear sex predilection (Gross et al. 2010). However, in more recent sizeable series, a slight predominance of male patients was found (Tong et al. 2012). There is a predilection of cavernous malformations for dorsal, followed by cervical, spinal levels (Gross et al. 2010). Our patient is a male and was presented with a cervical cavernous malformation.

The clinical course is mostly slowly progressive 55% followed by acute onset 45%, as reported by Gross et al. (Gross et al. 2010). The patient in our study was presented with progressive symptoms over 5 months. Progressive neurological decline is thought to result from enlargement of the lesion, which may be caused by micro-hemorrhages, intraluminal thrombosis, microcirculatory changes, or capillary proliferation. An acute pattern of clinical evolution is probably caused by frank lesion hemorrhage into the spinal cord parenchyma (Bian et al. 2009). The most common presenting symptoms in literature were motor weakness 60%, sensory affection 58%, pain 34%, and disturbance of bladder and/or bowel function 24% (Jetan et al. 2014). Our patient was presented with quadriparesis, paraesthesia and urinary urgency.

MRI is the most important diagnostic test for detection of cavernous angiomas. MRI typically demonstrates a well-delineated, low-signal intramedullary lesion with scattered heterogeneous areas of increased signal. The decreased signal is due to deposition of hemosiderin along the periphery of and within the malformation. On T2-weighted images, this mix of signal intensity produces the “target” configuration that is typical of cavernous malformations (Kivelev et al., 2010). Our patient showed the characteristic MRI findings of cavernous malformations (figure 11).

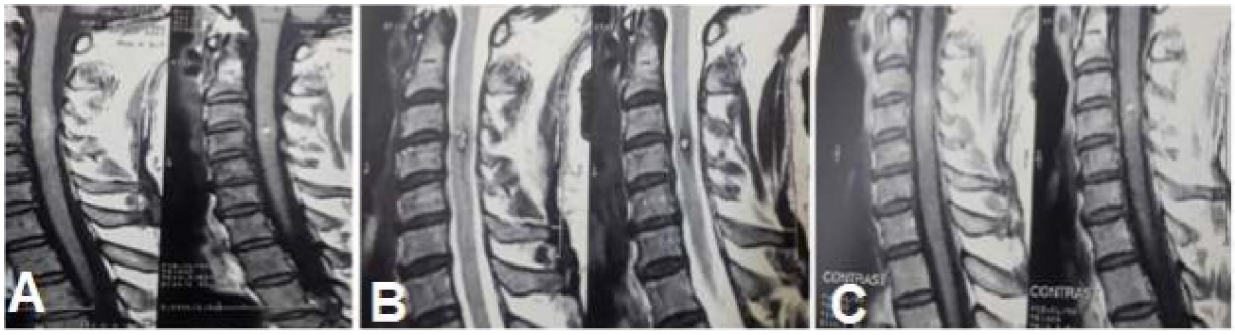

Figure.11 MRI cervical spine of case (15)

(A) T1 images show an intramedullary lesion opposite C 3-4, associated cord expansion and high signal intensity.

(B) T2 images show a lesion with high signal intensity surrounded with a low signal intensity margin.

(C) T1 with contrast images show contrast enhancing lesion.

It is generally accepted that patients with severe or progressive neurological deficits should be considered for surgical treatment. However, many patients with spinal cavernomas have lesions that produce less severe symptoms or are detected by MRI while asymptomatic. There is no compelling argument for treatment of asymptomatic cavernous angiomas of the spinal cord, and conservative management is an option for patients with less severe, nonprogressive manifestations (Moore et al. 2014). Our patient

refused any intervention and was discharged upon his request.

Techniques for removing cavernous angiomas are similar to those for excising benign intramedullary spinal cord tumors, but the margin of the cavernoma may be more adherent to the spinal cord. Because residual portions of cavernous angiomas, left in situ during surgery, tend to rehemorrhage and cause recurrent myelopathy, complete excision is mandatory (Steiger et al. 2010).

The outcome after surgery depends greatly on the patient's neurological function before surgery. The French Study Group of Spinal Cord Cavernomas reported the outcomes of 53 patients with spinal cord cavernomas, including 13 patients who did not undergo surgery. The average follow-up was 7.3 years. Of the 13 patients who did not undergo surgery, 4 experienced relapse of symptoms during the follow-up period (Labauge et al., 2008). In another study, none of the 10 patients with symptomatic cavernous angiomas of the spinal cord who were managed without surgery with a mean follow-up of 6.7 years had an acute intramedullary hemorrhage. In 9 of these 10 patients, neurological function at last evaluation was the same or better than their function at initial evaluation (DelCurling et al., 1991).

References

1. Anderer EG, Kang MM et al. (2008)."Successful management of an anterior Type IV spinal arteriovenous malformation with two associated aneurysms utilizing vertebrectomy." J Neurosurg Spine (9) 67–70.
2. Andres RH, Barth A et al. (2008)."Endovascular and surgical treatment of spinal dural arteriovenous fistulas." Neuroradiology (50) 869–76.
3. Antonietti L et al. (2010)."Long-term outcome in the repair of spinal cord perimedullary arteriovenous fistulas." AJNR Neuroradio (31) 1824-1830.
4. Bian LG et al. (2009). "Intramedullary cavernous malformations: clinical features and surgical technique via hemilaminectomy. Clin Neurol Neurosurg (111) 511–517.
5. Bostroem A et al. (2007). "Spinal vascular malformations-typical and atypical findings." 68(4):205-213.
6. Bostroem A et al. (2009). "Spinal glomus-type arteriovenous malformations: microsurgical treatment in 20 cases." J Neurosurg Spine 10: 423–429.
7. Caragine JLP et al. (2002)."Endovascular treatment of spinal cord vascular malformations."Semin Cerebrovasc Dis Stroke 2:236–244.
8. Cecchi et al. (2008)."Surgical treatment of spinal dural arterio-venous fistulae: Long-term results and analysis of prognostic factors." Acta Neurochir (Wien) 150:563-570.
9. Chibbaro S et al. (2015)."Surgical management of spinal dural arteriovenous fistulas." J Clin Neurosci;22:180–3.
10. Cho et al. (2005) Treatment of spinal cord perimedullary arteriovenous fistula: embolization versus surgery. Neurosurgery 56:232–241.
11. Corkill RA et al. (2007). "Embolization of spinal intramedullary arteriovenous malformations using the liquid embolic agent, Onyx: a single-center experience in a series of 17 patients." JNeurosurg Spine 7:478–85.
12. DelCurling OJ et al. (1991). "An analysis of the natural history of cavernous angiomas." J Neurosurg; 75:702-708.
13. Donghai et al. (2013). "The diagnosis of spinal dural arteriovenous fistulas." Spine;38:E546–53.
14. Dorsett-Martin et al. (2007)."Sex hormones and aortic wall remodeling in an arteriovenous fistula." Gend Med;4:157–69.
15. Fugate et al. (2012)."Clinical presentation and prognostic factors of spinal dural arteriovenous fistulas: An overview. Neurosurg Focus 32:E17.
16. Gemmete JJ, et al. (2013). "Spinal dural arteriovenous fistulas: clinical experience with endovascular treatment as a primary therapy at 2 academic referral centers." AJNR J Neuroradiology; 34(10): 1974Y1979.
17. Germans MR et al. (2008)."Spinal vascular malformations in non-perimesencephalic subarachnoid hemorrhage." J. Neurol; 255(12):1910–1915.
18. Gross BA et al. (2010)."Intramedullary spinal cord cavernous malformations. Neurosurg Focus 29(3):E14.
19. Gross BA et al. (2013). "Spinal pial (type IV) arteriovenous fistulae: A systematic pooled analysis of demographics, hemorrhage risk, and treatment results." Neurosurgery 73: 141-151.
20. Halbach VV et al. (1993)."Treatment of giant intradural (perimedullary) arteriovenous fistulas." Neurosurgery 33:972–979.
21. Hauser SL. (2005)."Diseases of the spinal cord." En: Harrison´s principles of internal medicine. 16th ed. New York; p. 2438-47.
22. Hida K et al. (2002)."Corpectomy: a direct approach to perimedullary arteriovenous fistulas of the anterior cervical spinal cord." J Neurosurg Spine 96:157–161.
23. Hida K et al. (2003)."Focal fractionated radiotherapy for intramedullary spinal arteriovenous malformations: 10-year experience." J Neurosurg 99: 34–38.

24. Inagawa et al. (2013)."Clinical results after the multidisciplinary treatment of spinal arteriovenous fistulas." Jpn J Radiol 31:455-464.
25. Inoue T et al. (2006)."Congestive myelopathy due to cervical perimedullary arteriovenous fistula evaluated by apparent diffusion coefficient values—case report." Neurol Med-Chir 46:559–562.
26. Iovtchev et al. (2015)."Late diagnosis of spinal dural arteriovenous fistulas resulting in severe lower-extremity weakness: a case series." Spine J;15:e39–44.
27. Jellema K et al. (2006). "Spinal dural arteriovenous fistulas: an underdiagnosed disease—a review of patients admitted to the spinal unit of a rehabilitation center." J Neurol; 253:159–62.
28. Jetan H et al. (2014)." Surgical outcomes and natural history of intramedullary spinal cord cavernous malformations: a single-center series and meta-analysis of individual patient data." J Neurosurg Spine 21:662–676.
29. Kivelev J et al. (2010)."Outcome after microsurgery in 14 patients with spinal cavernomas and review of the literature." J Neurosurg Spine; 13:524-534.
30. Krings et al. (2010)."Endovascular management of spinal vascular malformations." Neurosurg Rev 33(1):1-9.
31. Koch C et al. (2003)."Spinal dural arteriovenous fistula: clinical and radiological findings in 54 patients." Rofo Fortschr Geb Rontgenstr Neue; 175 (8):1071–1078.
32. Labauge P et al. (2008)." Outcome in 53 patients with spinal cord cavernomas." Surg Neurol.; 70(2):176-81.
33. Lai PH et al. (2006). "Multidetector CT angiography in diagnosis type I and type IVA spinal vascular malformations." AJNR 27:813–817.
34. Lucas et al. (2012). "Cervical spine dural arteriovenous fistula with coexisting spinal radiculopial artery aneurysm presenting as subarachnoid hemorrhage: case report." Neurosurgery;70:E259–63 [discussion E263].
35. Luetmer PH et al. (2005). "Pre-angiographic evaluation of spinal dural arteriovenous fistulas with elliptic centric contrast- enhanced MR angiography and effect on radiation dose and volume of iodinated contrast material." AJNR Am J Neuroradiol;26(4):711-8.
36. Martin NA et al. (1995)."Posterolateral cervical or thoracic approach with spinal cord rotation for vascular malformations or tumors of the ventrolateral apinal cord. JNeurosurg 83:254–261.
37. Medel R et al. (2009)."Endovascular management of spinal vascular malformations: history and literature review." Neurosurg Focus;26:E7.
38. Meng X et al. (2010)."Perimedullary arteriovenous fistulas in pediatric patients: clinical, angiographical, and therapeutic experiences in a series of 19 cases." Child Nerv Syst 26:889–896.
39. Moore SA et al. (2014)."Long-term natural history of incidentally discovered cavernous malformations in a single-center cohort." JNeurosurg;120:1188-1192.
40. Mourier KL et al. (1993)."Intradural perimedullary arteriovenous fistulae: results of surgical and endovascular treatment in a series of 35 cases." Neurosurgery 32:885–891.
41. Muralidharan R, et al. (2011). "The clinical and radiological presentation of spinal dural arteriovenous fistula." Spine; 36(25): E1641YE1647.
42. Nagata S et al. (2006)."Factors that affect the surgical outcomes of spinal dural arteriovenous fistulas." Surg Neurol;65:563–8 [discussion 568].
43. Oran et al. (2005). "Treatment of slow-flow (type I) perimedullary spinal arteriovenous fistulas with special reference to embolization." AJNR Am J Neuroradiol 26: 2582-2586.
44. Ofran et al. (2013). "Correlation between time to diagnosis and rehabilitation outcomes in patients with spinal dural arteriovenous fistula." J Spinal Cord Med 36:200-206.
45. Oldfield EH, Doppman JL. (1988)."Spinal arteriovenous malformations" Clin Neurosurg; 34:161-183.
46. Patsalides et al. (2011)."Endovascular management of spinal dural arteriovenous fistulas." J Neurointerv Surg;3:80–4.
47. Robertson CE et al. (2012)."Recovery after spinal cord infarcts: long-term outcome in 115 patients." Neurology; 78(2):114Y121.
48. Rodesch G et al. (2002)."Classification o spinal cord arteriovenous shunts: proposal for a reappraisal—the Bicetre experience with 155 consecutive patients treated between 1981 and 1999." Neurosurgery 51:374–379.
49. Rodesch G et al. (2003). "Spinal cord arteriovenous shunts: from imaging to management. " Euro J Radiol 46:221–232.
50. Rodesch G et al. (2005). "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 57: 973-983.

51. Rosenblum B et al. (1997)."Spinal arteriovenous malformations: a comparison of dural arteriovenous fistulas and intradural AVM's in 81 patients." JNeurosurg; 67:795-802.
52. Saladino A, et al. (2010). "Surgical treatment of spinal dural arteriovenous fistulae: a consecutive series of 154 patients." Neurosurgery; 67(5): 1350Y1357.
53. Saraf-Lavi E, et al. (2002). "Detection of spinal dural arteriovenous fistulae with MR imaging and contrast-enhanced MR angiography: sensitivity, specificity, and prediction of vertebral level." Am J Neuroradiology; 23:858–867.
54. Scarff et al. (1979)."Arteriovenous malformations of the spinal cord in children." Child Brain 5:341–353.
55. Schuss et al. (2015)."Surgical treatment of spinal dural arteriovenous fistula: Management and long-term outcome in a single-center series." World Neurosurg 83(6):1002-1005.
56. Song et al. (2010)."Spinal cord vascular malformations in children." Neurosurg Clin N Am 21(3):503-510.
57. Steiger et al. (2010). "Prognostic factors for the outcome of surgical and conservative treatment of symptomatic spinal cord cavernous malformations: a review of a series of 20 patients." Neuro Focus; 29:E13.
58. Takai K, et al. (2015)."Microsurgical resection of an intramedullary glomus arteriovenous malformation in the high cervical spinal cord: retrograde dissection techniques of the nidus located between spinal tracts." Acta Neurochir (Wien); 157:1659–1664.
59. Tender et al. (2005)."Spinal intradural arteriovenous fistulas acquired in late adulthood: absent spinal venous drainage in pathogenesis and pathophysiology. Report of two cases." J Neurosurg -Spine 3:488–494.
60. Thron A, et al. (2003). "Vascular malformations and interventional neuroradiology of the spinal cord." Neurological disorders. Course and treatment; pp 517–528.
61. Touho H et al. (1995)."Treatment of Type II perimedullary arteriovenous fistulas by intraoperative transvenous embolization: case report." Surg Neurol 43:491–495.
62. Tong X et al. (2012)."Clinical presentation and surgical outcome of intramedullary spinal cord cavernous malformations." J Neurosurg Spine 16:308–314.
63. Van Beijnum J et al. (2007)."Spinal arteriovenous shunts presenting as intracranial subarachnoid haemorrhage." J. Neurol.;254(8):1044–1051.
64. Vitarbo EA et al. (2005)."Split cord malformation with associated type IV spinal cord perimedullary arteriovenous fistula. Case report." J Neuro-Spine 3:400–404.
65. Wakao et al. (2012). "Clinical outcome of treatments for spinal dural arteriovenous fistulas: results of multivariate analysis and review of the literature." Spine:482–8.
66. Zevgaridis D et al. (1999)."Cavernous haemangiomas of the spinal cord. A review of 117 cases. Acta Neurochir (Wien) 141:237–245.

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