NEUROREHABILITATION AT PATIENTS WITH SPINAL DURAL ARTERIOVENOUS FISTULA VERSUS PATIENTS WITH SPINAL CORD ISCHEMIA

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Abstract

Background: Spinal dural arteriovenous fistulas (SDAVF) are the most common vascular malformation of the spinal cord, but still rare and underdiagnosed disease. The fistula causes a progressive loss of cord function: para- or tetraplegia, sensory loss, incontinence and pain. Because presenting the same clinical symptoms like SDAVF, spinal cord ischemia (SCI) is another entity which must be considered.

Patients and methods: Between August 2012- August 2013 we diagnosed 5 patients with SDAVF and 5 patients with SCI (only self- casualty). We collected information on history, time to diagnosis, neurologic examination, MR-angiography, MRI, surgical treatment and neurorehabilitation.

Results: In group A we have five patients with SDAVF. The median age was 50.6 years (range 28-71). The median time to diagnosis was 4 months (range 2 days to 12 months). The most frequent symptoms were paraplegia (100%) and micturition problems (100%). In our experience 80% of all SDAVF are located between T2-L5. We have one option in treatment of SDAVF: surgical occlusion of fistula.

In group B we have five patients with SCI. The median age was 40.8 years (range 19 to 60). The median time to diagnosis was only few hours (8 hours). The most frequent symptoms were paraparesis or plegia (100%), sensory loss (80%), back pain at onset (100%) and urinary problems (80%). The most common location to be affected is lumbosacral spine. The treatment of SCI was medical.

Conclusions: Patients with SDAVF develop a progressive myelopathy, which at the early stage of the disease often mimics a polyradiculopathy or anterior horn cell disorder. By the time, patients suffer from considerable neurological deficits. Patients with SCI suffer the same symptomatology; they are much younger that the patients with SDAVF and the median time to diagnosis was only few hours.

The diagnosis was confirmed by MR-angiography or MRI for both entities.

Prognosis: The effect of surgical treatment on activities of daily life was reported as better by 3 of 5 patients. At patients with SCI recovery is higher.

Key Words: Spinal dural arteriovenous fistula, spinal cord ischemia, neurorehabilitation.

Introduction

Spinal dural arteriovenous fistulas are the most common vascular malformation of the spinal cord but still rare and underdiagnosed disease.

A spinal dural arteriovenous fistula (SDAVF) is an abnormal shunt between a spinal radicular artery and corresponding radicular vein that drains the perimedullary venous system (Jellema K. at al, 2004).

The fistula causes congestion in medullary veins, which leads to decrease tissue perfusion with edema and progressive loss of cord function: para- or tetraplegia, sensory loss, incontinence and pain (Kendall. at al, 1977).

Because presenting the same clinical symptoms like SDAVF, spinal cord ischemia is another entity which must be considered. Weakness, flaccid paresis accompanied by diminished superficial and tendon reflexes below the level of the lesion, sensory loss, back pain and urinary complaints required catheterization were the most common symptoms of cord ischemia at the time of presentation. Maximum disability is observed within 12 hours of onset in a majority of patients (Bradley at al, 2008).

Surgical treatment aimed at closure the SDAVF. Embolization of SDAVF is not possible in our clinics.

The medical management of spinal cord ischemia is generally supportive and includes maintenance of adequate blood pressure, early bed rest, reversal causes such as hypovolemia and arrhythmias, antithrombotic therapy.

Physical and occupational therapy are useful in promoting functional recovery.

Significant subjective improvement was noted in walking and muscle power at patients with SDAVF. Muscle spasm and leg pain were reported; micturition and anal sphincter function tended to persist after surgical treatment.

Rates of recovery is high at patients with spinal cord ischemia; they have a favorable outcome defined as the ability to walk with none assistive device and no...
need urinary catheterization. Chronic pain tends to occur only at patients with spinothalamic sensory impairment.

**Patients and methods**

Between August 2012 - August 2013 we diagnosed 5 patients with SDAVF and 5 patients with spinal cord ischemia (only self-casuistry).

The patients with SDAVF were treated by surgery. The diagnosis of SDAVF was confirmed by MR-angiography which reveals “flow-void phenomena”, representing tortuous and dilated veins at the dorsal surface of the spinal cord; the central hyperintense lesions are difficult to interpret (Gogu et al, 2013).

The patients with spinal cord ischemia were treated with medical procedures. The diagnosis of spinal cord ischemia was confirmed by MRI. After spinal cord infarction, typical spin-echo magnetic resonance (MR) findings are cord enlargement and hyperintense signal on T2-weighted images initially (8 hours to several days), with or without gadolinium enhancement, followed by cord atrophy later (months). Abnormal signal and enhancement may demonstrate a double-dot (“owl’s eye”) pattern in the region of the anterior horns and H-shape pattern involving the central grey matter or a more diffuse pattern involving both grey and white matter (Bradley at al, 2008).

We collected information on history, time to diagnosis, neurologic examination, imaging, treatment and recovery. Patients were re-examined after three months, except one case who refused surgical treatment for SDAVF. Patients were asked to compare their current status with the worse before treatment; they graded changes in walking, muscle power, paresthesias, micturition problems, anal sphincter disturbances, pain, muscle spasm as worse, same or better.

**Results**

In group A we have 2 men and 3 women with SDAVF. The median age was 50,6 years (range 28 to 71). The median time to diagnosis was 4 months (range 2 days to 12 months). The most frequent symptoms were gait disturbances (paraplegia - all patients - 100%) and micturition problems (100%). Most fistulas are solitary lesions and are found in the thoracolumbar region. In our experience 80% of all SDAVF are located between T2- L5. Only one case has lesions at level T1-T2.

We describe three important cases with SDAVF:

**Case 1A**

A 57-year old female, M.B., with acute onset; the symptoms develop within minutes and mimic an anterior spinal artery syndrome: paraplegia, total sensory loss, urinary retention, bowel incontinence.

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![Fig.1A-a. MRA revealed: on T1 weighted sequences the cord edema is depicted as a centromedullary hyperintensity over T7-L1. At these level there is intraspinal haemorrhage. (a rare case report).](image-url)
Fig. 1A-b. T2 weighted MRA shows numerous “flow-voids” over the dorsal spinal cord, representing dilated perimedullary veins, between T10-L1 levels.

**Case 2A**- A 35-year old female, S.F., subacute onset in two weeks with gait difficulties, asymmetrical paraparesis and sensory symptoms; paresthesias in both feet, first with loss of pain and temperature sensation, gradually ascending to the T6 level; micturition disturbance

Fig. 2A-a. Hyperintensities on T2-weighted images represents perimedullary vessels which are dilated and coiled at T6-T11 levels.

Fig. 2A-b. The technique of first-pass gadolinium-enhanced MRA demonstrates the level of the shunt at T6.

**Case 3A**- A 71-year old male, H.I., with chronic onset in 3 months presented initial symptoms like difficulty in climbing stairs, gait disturbances, paresthesias and radicular pain which affect both lower limbs. These
neurologic symptoms are progressive with time and are ascending; later the patient was paraplegic with loss of all modalities of skin sensation below T2; urinary retention and constipation.

Fig. 3A-a. MRA revealed abnormal blood vessels on either the ventral and the dorsal side of the spinal cord below level T2.

Fig. 3A-b. The left foramen vascular routes – T2 have adherence from the subclavian artery and aortic cross, with left paravertebral intrathoracic extension and within left posterior - vertebral muscles.

Fig. 3A-c. MRA revealed extradural subacute hematoma T2 – T3.

Treatment modalities: the aim of treatment in SDAVF is to occlude the shunting zone (the most distal part of the artery together with the most proximal part of the draining vein) (Krings et al, 2009; Jellema et al, 2005; Van Dijk et al, 2002). There are two options in treatment of SDAVF: surgical occlusion of the intradural vein that received the blood from the
In group B, we have 3 men and 2 women with spinal cord ischemia. The median age was 40.8 years (range 19 to 60). The median time to diagnosis was only a few hours (under 8 hours). The most frequent symptoms were gait disturbances (paraparesis or paraplegia - 100%), sensory loss (80%), back pain at onset (100%) and urinary complaints requiring catheterization (80%). The most common location to be affected is lumbosacral spine (3 cases). Lower cervical lesions are less common (1 case) and we have one patient with midthoracic spinal ischemia. We describe the most important cases:

**Case 1B:** A 51-year old male, C.T., with acute onset: the symptoms develop within minutes with paraplegia, back pain and urinary retention. This patient has atrial mixoma and thromboembolism may cause spinal cord infarct.

![MRI showing hyperintense signal on T2-weighted images with gadolinium enhancement](image1.png)

**Fig. 1B-a.** MRI shows hyperintense signal on T2-weighted images with gadolinium enhancement at L1-L4 levels.

![MRI revealing a double-dot (“owl’s eyes”) pattern](image2.png)

**Fig. 1B-b.** MRI reveals a double-dot (“owl’s eyes”) pattern in the region of anterior horns.

**Case 2B:** A 32-year old female, C.M., with acute onset: asymmetrical paraparesis, sensory symptoms with loss of pain and temperature sensation first; proprioception changes accompany them; back pain and micturition disturbances. This patient has systemic lupus erytematosus with chronic treatment with Azathioprine.
Fig. 2B-a. MRI-findings are cord enlargement and hyperintense signal on T2-weighted images without gadolinium enhancement at T6-T12 levels.

Fig. 2B-b. MRI reveals an H-shape pattern involving the central grey matter and near T12 level we observe a more diffuse pattern involving both gray and white matter.

**Prognosis**
The changes in specific symptoms after treatment of spinal dural arteriovenous fistulas (SDAVF) or spinal cord ischemia (SCI) are summarized in the Table 1.
A few basic exercises that were used in this study:

1. The physical therapist initiates passive/passive-active movements in all diagrams, until the active movements is achieved.
2. Proprioceptive neuromuscular facilitation (PNF) – rhythmic stabilization (RS), contract-relax (CR), hold-relax (HR), repeated contractions.
3. Bridging – allows weight-bearing through the feet and is an important precursor to assuming the kneeling position and in developing sit-to-stand control. For this activity the patient is in a hooklying position (supine with hips and knees flexed and feet flat on the mat) and elevates the pelvis off the mat. This activity is particularly useful for facilitating pelvic motions and strengthening the low back and hip extensors in preparation for the stance phase of gait. In addition, bridging has several important functional implications, including bed mobility, pressure relief, movement from sit-to-stand, and stair climbing.
4. Quadruped posture – allows weight-bearing through the hips and is particularly useful for

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Table 1. The symptoms after treatment of SDAVF and SCI

<table>
<thead>
<tr>
<th>Rating</th>
<th>Walking disturbances</th>
<th>Muscle power</th>
<th>Paresthesias</th>
<th>Micturition problems</th>
<th>Anal sphincter problems</th>
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The effect of surgical treatment on activities of daily life was reported as better by 3 of 5 patients and as worse by 1 patient. Nearly one third of more severely affected patients required a wheelchair (2 cases). The most persistent symptoms were paresthesias, micturition problems, pain in legs and muscle spasm.

Neurorehabilitation

Recovery of independent locomotion (neurorehabilitation) is an important goal for many patients seeking physical therapy intervention. It is a functional skill that directly impacts performance of expected roles within the patient’s social, cultural, and physical environment. A general framework of preparatory exercises and locomotor training strategies has been presented that can be modified to meet the needs of an individual patient. Through a process of careful examination and communication with the patient, family, and/or care-givers, the appropriate preparatory activities and specific training strategies can be identified.

Enabling individuals to manage daily self-care is among the most important goals undertaken by the rehabilitation team. This is because such tasks relate directly to the business of living and their performance signifies a return to participation in the routines of daily life. Self-care tasks include dressing, eating, bathing, grooming, use of the toilet, and mobility within the home. These are basic tasks included within the general category of activities of daily living (ADL). Although able-bodied persons perform most self-care tasks routinely, such tasks can represent difficult challenges for persons with sensory, motor, and/or cognitive deficits. During the rehabilitation process, the family can have a considerable influence on functional outcome. A stable and supportive family unit can be of great assistance, whereas families that are functioning poorly can impede rehabilitation. In some cases, poor outcomes can be traced to a lack of family involvement in the rehabilitation process. In other cases, too much support can encourage dependency. This indicates that the family should be involved in all aspects of rehabilitation, including evaluation and the setting of rehabilitation goals and treatment strategies before and after discharge.

At patients with spinal cord ischemia, recovery is higher. Four patients had a favorable outcome; they walk with one assistive device or none and no need for urinary catheterization after subacute phase of disease. After three months all of them walk without assistive device. Chronic pain was a disabling consequence of spinal cord ischemia.
promoting control of the lower trunk and hips. For patients with spasticity, this posture can be used to provide inhibitory pressure to the quadriceps and long finger flexors (using an open hand position) to diminish tone.

5. Sitting – can be used effectively to promote static and dynamic postural control, reactive and anticipatory balance control, vertical midline orientation and postural alignment.

6. Sit-to-stand – movement transitions from sit-to-stand should emphasize symmetrical weight-bearing as well as coordination and timing of motor response. Initially, the patient shifts weight forward by using momentum and actively flexing the trunk forward (mass flexion pattern). The feet are placed back partially under the supporting surface to engage the dorsi-flexors in forward rotation.

7. Modified plantigrade - is an early weight-bearing posture that can be used in preparation for erect standing and walking. Modified plantigrade is an important precursor to walking inasmuch as it superimposes close to full weight-bearing on an advanced LE pattern. This pattern, required during gait, combines hip flexion with knee extension and ankle dorsiflexion.

Discussions

Patients, who are mostly middle-age, develop a progressive myelopathy which at the early stage of the disease often mimics a polyradiculopathy or anterior horn cell disorder. By the time involvement of upper motoneurons or sacral segments makes the diagnosis of SDAVF inescapable, patients suffer from considerable deficits (Jellema et al, 2006).

The fistula causes congestion in medullary veins decrease tissue perfusion with edema and progressive loss of cord function: paraplegia, sensory loss, disturbances of micturition and defecation, pain in legs and muscle spasms (Kendall at al, 1977).

Because presenting the same clinical symptoms like SDAVF, spinal cord ischemia (SCI) is another entities which must be considered; the most frequent symptoms were gait disturbances, back pain, sensory loss and urinary problems.

Recovery of motor control is an intrinsic person/nervous-system process.

This recovery is dependent on psychological, behaviourial, neurophysiological and tissue-related factors. Often many of these factors are interrelated.

The role of neuromuscular rehabilitation is to optimize the recovery of movement control, working with all these factors.

There are three main principles to consider in neuromuscular rehabilitation: functional movement, skill-and ability-level rehabilitation, and the code for neuromuscular adaptation (Lederman, 2010).

A functional approach promotes the use of what the patient already knows. Challenges to specific motor losses can be found within the person’s movement repertoire.

At skill-level rehabilitation the patient simply aims to do the movements they can’t do.

Cognition about injury and pain, persistent pain and fear of it, and behavioural factors can all be managed within skill-level rehabilitation.

Ability-level rehabilitation focuses on specific underlying motor losses that prevent the person from attaining their movement goals.
Conclusions

1. The diagnosis of SDAVF was confirmed by MR-angiography which reveals “flow-void phenomena”, representing tortuous and dilated vein at the dorsal surface of the spinal cord.
2. In our experience 80% of all SDAVF are located between T2-L5 levels.
3. Surgical treatment aimed at closure the fistula. Embolization of SDAVF is not possible in our clinics.
4. The patients with SCI are younger than the patients with SDAVF.
5. The median time to diagnosis was only few hours (under 8 hours) but at patients with SDAVF this time is longer (weeks or months).
6. The diagnosis of SCI was confirmed by MRI which reveals spinal cord enlargement and hyper intense signal on T2-weighted images (8 hours to several days). Gadolinium enhancement demonstrates a double-dot (owl’s eye) pattern in the region of the anterior horns.
7. The most common location to be affected by ischemia is lumbosacral spine (L2-S2). Historically, the literature has supported the notion of a spinal cord “watershed zone” of ischemic vulnerability centered at the mid-thoracic level (T4 to T6), describing the relative hypovascularity of this region (Duggal et al, 2002).
8. The medical treatment of spinal cord ischemia is generally supportive and includes maintenance of adequate blood pressure, early bed rest, reversal causes and antithrombotic therapy.
9. At patients with SCI, recovery is higher. They walk with one assistive device or non, and no need urinary catheterization but remains with chronic pain in legs. At patients with SDAVF a significant improvement was noted in walking and muscle power; muscle spasms and leg pain were reported but micturition and anal sphincter function tended to persist after surgical treatment.

References


