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Case Report

Diagnostic and Treatment of Spinal Dural Arterio-Venous Fistulae: Case Report and Literature Review

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Abstract

Dural arteriovenous (AV) fistula is a vascular malformation of the spinal cord that is usually found on the dural surface of the spinal cord and drain through the anterior or the posterior medullary vein. The incidence in the U.S population is estimated to be 5.9 women and 3.4 men per 100,000. Spinal cord edema may extend far away from the original site of the fistula. It is the most common spinal malformation, attributing to 80% of all spinal abnormalities. Spinal angiography is the diagnostic tool of choice to confirm the diagnosis of AV fistula. Patients usually present with upper motor neuron (UMN) and lower motor neuron (LMN) lesions [1]. The neurological symptoms progress gradually, including sensory and motor deficits as well as sphincter disturbances [2]. RASA 1 gene mutations is suggested to play an important role in the pathophysiology of the AV fistula [3]. Management includes surgical intervention or embolization. However, surgery is considered to be the best option, offering a 98% success rate [1].

Keywords: Spinal Dural Arterio-venous fistulae (SDAVFs); Upper Motor Neuron (UMN), Lower Motor Neuron (LMN)

Introduction

Vascular lesions are classified into AV fistulae and AV malformation [4]. Spinal intradural AV fistulae account 80% of all spinal abnormalities. Spinal dural AV fistula is usually located on the dorsal surface of the lower thoracic and lumbar spine [5]. They represent an abnormal connection between a spinal radicular artery and vein, near the exiting point of the nerve root [6].

Arterialization of the medullary vein leads to venous congestion in the coronal venous plexus of the spinal cord. Elevated venous pressure leads to hypoperfusion and vascular steal phenomena, causing ischemia [7]. Clinical presentation is usually non-specific and can be misdiagnosed with other spinal pathologies.

Hyper-intensity on multi-segmented T2-weighted images with associated subarachnoid flow voids is a diagnostic feature [1].

Case Description

A 74-year old right-handed male presented with gradual-onset progressive pain over 2 years, lower extremities weakness and numbness, sensory changes in the legs, imbalance, bladder and bowel incontinence. His past medical history was positive for hypertension, hyperlipidemia, diabetes mellitus and benign prostatic hyperplasia.

On examination the patient was neurologically intact except for diminished sensation in his lower extremities and bilateral hyperreflexia of both ankle and knee jerks. Electromyography (EMG) revealed L4-S1 radiculopathy for which he underwent physiotherapy and caudal epidural injection.

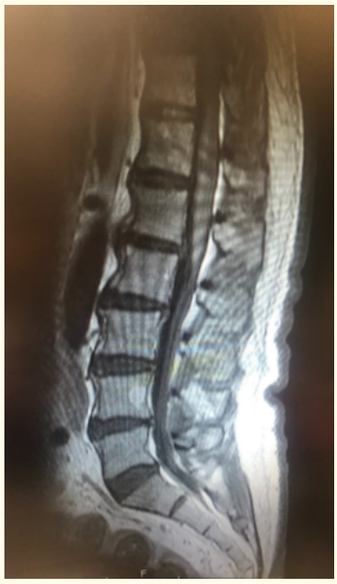
An initial contrast magnetic imaging (MRI) of Lumbar Spine demonstrates abnormal thoracic spinal cord expansion in the region of T12-S1, increased fluid signal within the cord of these levels causing local mass effect of the caudal equine nerve roots.

Non-contrast enhanced MRI of the lumbar spine was also performed and demonstrated multilevel broad based disc herniation's that extended bilaterally at the levels of T12-S1, causing stenosis.

A contrast-enhanced MRI of the thoracic and lumbar was performed which revealed edema within the spinal cord at T levels and an increased signal from the mid thoracic region to the conus medullaris.

The location of SDAF explained the spinal cord edema seen at T12-S1 levels, as well as the patient's sensation complaints and anal sphincter incontinence.

The gold standard to localize vascular malformation and confirming degrees of arterial drainage outflow is absolutely spinal angiography which demonstrated the dural AV fistula extended from T7 to the conus medullaris. It also showed patchy enhancement along the conus, as well as multilevel disc degeneration. Feeding arteries were located at a different level than the fistula point.



Figure

Discussions

Dural AV fistulas are arteriovenous shunts from a dural supply to a dural venous channel. This diagnostic entity represents a rare occurrence in pain management patients [4]. The etiology of these lesions is not fully understood, some are congenital and some are acquired whereas venous obstruction, aberrant angiogenesis and hypertension play a role as risk factors [8]. On the other hand, some cases of dural AVFs have no clear inciting event therefore, it is crucial to take a correct and thorough history [9]. Clinical presentation is non-specific, including a variety of symptoms ranging from mild to aggressive according to lesion location and pattern of venous drainage [5]. However, if the patient is not responding to physical therapy, as well as caudal epidural injections, it is important to refer out to the proper specialists, which in our case was a spinal surgeon. Successful treatment of dural AV fistula includes embolization of the artery. When endovascular embolization is technically difficult due to the difficult endovascular access route, or results noneffective then surgical treatment is required [10]. The rate of occlusion after initial treatment with endovascular embolization has not yet reached that of open microsurgery; moreover, the lasting effectiveness of open microsurgery is significantly superior to that of endovascular embolization because dural AVF recurrence has been more commonly reported after endovascular treatment. The success rate of permanent AVF occlusion was previously reported to be 98% for open surgery and 10% - 75% for endovascular embolization [11,12]. A high field magnet and contrast is usually required to properly diagnose rare conditions. Sometimes, such patients may present with comorbidities. SDAVFs can be easily misdiagnosed as radiculopathy, and ESI for relieving radicular pain can cause serious adverse effects such as paraplegia, clinicians should always keep in mind the possibility of SDAVFs and review the MRI before ESI, especially when the symptoms of myelopathy coexist [13]. Patients with SDAVF already have a high intradural and epidural pressure. Injection of even a small volume of drug in the epidural space could aggravate the neurological symptoms [14]. This is why it is imperative to think of more than one or two differentials to properly diagnose the patient. Undiagnosed SDAVFs can also present with paraplegia, urinary incontinence and gait problems following epidural and spinal anesthesia [15,16]. Expectant follow-up of these lesion should include serial magnetic resonance imaging for any evidence of changes in the Dural AVF anatomy. A radical cure of the dural AVFs is not needed unless symptoms are really debilitating or the DAVF is associated with leptomeningeal venous drainage [17].

Conclusion

A better understanding of these type of lesions has predisposed in early diagnosis and management of their clinical course. Therefore, considering other differential diagnoses in patients who aren't responding to traditional treatment is crucial. However, the clinical presentation apart from hemorrhage and progressive neurological deficits rarely orientates the physicians towards an aggressive behavior of the dural AV fistula.

The thorough diagnostic investigation and usage of MRA T2-weighted images are imperative to identify and confirm the disease. Treatment of dural AV fistula should be part of a multidisciplinary team with expertise and management of these lesions and variety of treatment options according to the characteristics of the

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