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Case Report

A case report of spinal dural arteriovenous fistula: origins, determinants, and consequences of abnormal vascular malformations

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ABSTRACT

A spinal dural arteriovenous fistula is an abnormally layered connection between radicular arteries and venous plexus of the spinal cord. This vascular condition is relatively rare with an incidence of 5–10 cases per million in the general population. Diagnosis of spinal dural arteriovenous fistula is differentiated by contrast-enhanced magnetic resonance angiography or structural magnetic resonance imaging, but a definitive diagnosis requires spinal angiography methods. Here, we report a case of a 67-year-old female with a spinal dural arteriovenous fistula, provide a pertinent clinical history to the case nosology, and discuss the biology of adhesive proteins, chemotactic molecules, and transcription factors that modify the behavior of the vasculature to possibly cause sensorimotor deficits.

Introduction

The spinal cord is composed of distinct cell types, including those cells that make up the anterior and posterior spinal arteries, spinal branches, and spinal veins. Vascular cells in the spinal cord depend on adhesive membrane proteins expressed, particularly, by endothelial cells or local extracellular chemotactic proteins for sculpting and maintaining a connective layered scaffold between the arterial and venous systems [1]. This requires that during central nervous system (CNS) development, differentiated cells become dispersed and positioned correctly, often over long distances, from sites in the early neural tube to final sites in the segmented spinal cord [2]. If this cell-to-cell guidance milestone fails to materialize, it is conceivable that a hugely disorganized and dilated vascular network generates upon full spinal cord maturation with serious pathologic implications (Fig. 1). For instance, during the formation of spinal dural arteriovenous fistula (SDAVF), an abnormally aligned connection between radicular arteries and venous plexus is established within ventral

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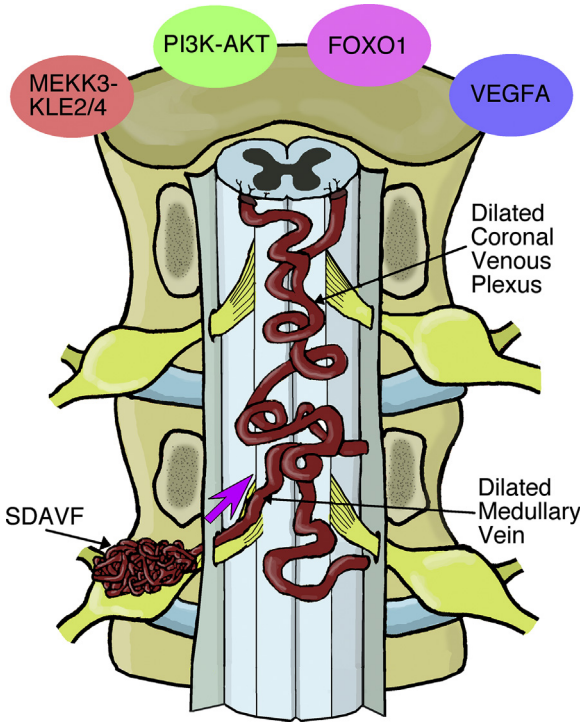

Fig. 1 – SDAVFs are localized to dural sleeves of nerve roots. This simplified schematic diagram shows that under pathologic conditions, blood is supplied to a SDAVF lesion by dural arteries (not shown) and then drained in a retrograde manner to normal venous flow through a medullary vein into the coronal venous plexus. Due to the aberrant arteriovenous architecture, the venous system (coronal venous plexus via the medullary vein) receives arterial blood that is "arterialized." This leads to venous congestion, which can put pressure on the spinal cord to produce clinical symptoms. Although direct evidence is missing, a modified vascular phenotype may be the result of changes in adhesive properties or the expression of specific differentiation-promoting (e.g., FOXO1) or differentiation-inhibiting factors that code for uncoordinated vascular hyperplasia (e.g., VEGFA; PI3K-AKT), vessel diameter malformation, and/or aberrant endothelial cell growth (e.g., MEKK3-KLF2/4). FOXO1, forkhead box O; VEGFA, vascular endothelial growth factor A.

(anterior; motor) and dorsal (posterior; sensory) aspects of nerve roots [3].

As its name implies, SDAVF is localized to the dural sleeve of a nerve root and the striking hyperplastic feature of the fistula leads blood from the arterial system to directly be shunted into the venous plexus with reduced vascular resistance and, importantly, without the inclusion of the capillary network to regulate unidirectional blood flow [4]. The end result of this abnormal blood flow is the "arterialization" of the venous plexus with subsequent impediment bouts of venous drainage that commonly lead to venous congestion, venous hypertension, intramedullary edema, and localized nerve cell hypoxia [3–6]. If this particular clinical phenotype goes unrecognized and left untreated, SDAVF can cause serious venous infarction, subarachnoid hemorrhage, and irreversible progressive ascending myelopathy [3,7]. Understanding the mechanisms of such variable expressivity will undoubtedly provide critical pathophysiological clues [8].

Rigorous attempts to diagnose SDAVF require using structural magnetic resonance imaging (MRI) approaches to reveal dilation of the perimedullary veins and discrete spinal cord enlargement gradients [9]. In addition, T2 signal abnormalities within the conus medullaris can also be used for precision diagnosis [3,5–7]. Although MRI is useful in identifying structural differences in spinal cord appearance among patients, spinal angiography is the gold standard in localizing vascular malformations and confirming degrees of arterial drainage outflow [3,6,7,10]. Although these imaging advances do not yet deliver a complete picture of the architecture of SDAVFs, there is sufficient information to draw some general conclusions.

SDAVFs are also known as type I spinal vascular malformations with a relatively low incidence of clinical presentations (5–10 individuals per million in the general population) [3,7]. Although small effect size common variants to larger effect size rare mutations are thought to provide casual anchors from which to understand SDAVFs, it is not known whether other, as-of-yet-unknown factors, may contribute to spinal cord vascular pathology with parent-of-origin effects. However, we cannot exclude the possibility of sporadic rather than familial estimates, contributing to disease onset and disease progression. Regardless of heritable estimates or rates of de novo point mutations, 80% of clinical cases are diagnosed in males, with 66% of these patients presenting with manifestations of vascular disease in their sixth or seventh decade [5,7]. Against this background, we describe here the clinical history of a female patient with SDAVF, highlight the challenges reflected in the heterogeneity of the syndromic diagnoses, and discuss treatment options and complications of disease pathology characterized by abnormally aligned endothelial blood vessels.

Case report

A 67-year-old white female presented to the emergency department with an acute onset of numbness and tingling in the hips and legs after going shopping. She then drove to work and was not able to exit her car upon arrival due to muscle weakness. This was accompanied by a short episode of 5/10 midsternal chest pain and anxiety. Two weeks prior, she had a similar episode of sensorimotor symptoms, which resolved spontaneously. The patient also reported a 1-month history of ON and OFF cold-like symptoms and oral/buccal cold sores. Past medical history included hyperlipidemia which was controlled with appropriate diet.

The patient was awake, alert, and oriented with signs of tachycardia with otherwise stable vital signs on physical examination. The neurologic examination in the emergency department was significant for decreased muscle strength (0/5) and decreased sensation (4/5) in the lower extremities, as well as decreased rectal tone. A lack of sensation in the lower extremities was found with the greatest degree of sensation