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CASE REPORT

Spontaneous Hemorrhage of Thoracic Cavernous Malformation Leading to Bilateral Lower Extremity Paralysis

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Abstract

Cavernous malformations (CMs) are abnormal clusters of thin-walled blood vessels located in the central nervous system. An 87-year-old male with a history of heart failure with reduced ejection fraction, coronary artery disease, and atrial fibrillation on rivaroxaban was admitted for acute onset, bilateral lower extremity weakness. He was found to have hemorrhagic transformation of a pre-existing spinal cord cavernous malformation located at the level of T9 of the thoracic column worsened by his use of anticoagulation. Surgical resection remains the only definitive management. Patients at high risk for surgical intervention are managed with supportive care and physical therapy.

Keywords: Cavernous malformation, CavMal, Hemorrhagic cavernous malformation, Paraplegia

1. Background

Cavernous malformations (CMs) are defined as abnormal clusters of thin-walled blood vessels located in the central nervous system[1] that are most prevalent in the cerebrum but on rare occasions can occur in the spinal cord. These may occur sporadically; however, most cases have an autosomal dominant inheritance.[2] CMs are thought to be benign slow-growing lesions but have the potential to acutely expand and become symptomatic. Diagnosis of spinal cord CMs requires a thorough history, physical exam, and imaging studies. Symptomatology of spinal cord CMs range from mild sensory deficits to paraplegia as a result of either compression or hemorrhage. Surgical removal remains the mainstay of therapy (see Fig. 1).

2. Objective

To describe an uncommon etiology of lower extremity paralysis requiring rapid evaluation and diagnosis to prevent long term complications.

3. Case report

An 87-year-old male with a history of heart failure with reduced ejection fraction, coronary artery disease, and atrial fibrillation on rivaroxaban was admitted for acute onset, bilateral lower extremity weakness. On the day of admission, he awoke and experienced a fall while attempting to get out of bed secondary to bilateral lower extremity weakness. He continued to experience progressively worsening weakness of bilateral lower extremities throughout the day prior to presenting to the ED. Neurological exam revealed 2/5 muscle strength in bilateral hips, knees, and ankles. Neurological exam also showed bilateral lower extremity hyperreflexia, increased muscle tone, positive Babinski, and decreased rectal tone. Fine touch and pain sensation was intact. The patient then underwent an MRI of the spine which demonstrated expansion of the mid and lower thoracic spinal cord with hyperintensity extending from T6-T8 levels and possibly T10-T11 level. After multidisciplinary discussion with neurology and radiology, the lesion was originally suspected to be indicative of transverse myelitis or other

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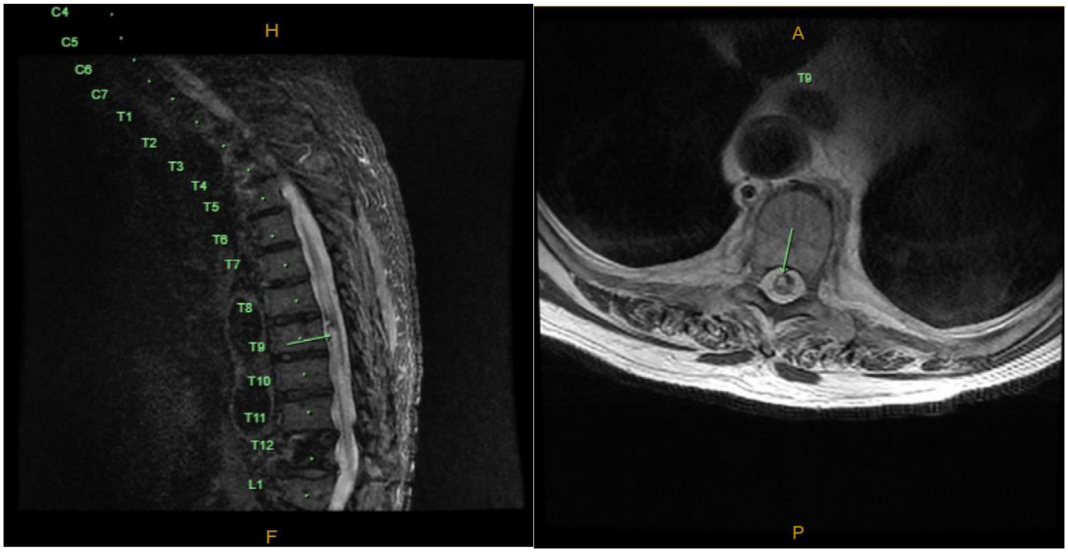

Fig. 1. MRI demonstrating cavernous malformation in Short Tau Inversion Recovery sagittal and T2 axial views.

inflammatory process with low suspicion for an active hemorrhage. He was started on a course of 1 g methylprednisolone daily and completed a 5-day course with minimal improvement in his symptoms. Due to the lack of improvement after a course of high-dose steroids, a lumbar puncture was performed to evaluate for infectious, rheumatologic, and paraneoplastic etiologies. Lumbar puncture revealed abundant red blood cells (17,211/mm3) which was initially attributed to a traumatic tap. The remainder of the cerebrospinal fluid analysis was unremarkable. Rivaroxaban was temporarily held following the suspected traumatic tap and was restarted the following morning per neurology recommendation. Extensive serum and cerebrospinal fluid studies showed no evidence of infectious, rheumatologic, or paraneoplastic processes. The patient continued to show minimal improvement despite aggressive physical therapy and having completed a course of high dose steroids. A repeat MRI of the thoracic spine was performed which showed a focus of low signal in the T9 spinal cord, increased signal inferior to this low signal lesion since previous MRI, and similar cord edema since previous MRI. These new findings were suggestive of a vascular lesion. The MRI was reviewed with both neurology and neurosurgery with the consensus that this lesion was most consistent with a cavernous malformation. The patient's clinical presentation was attributed to a hemorrhaging cavernous malformation with associated spinal cord edema. The patients anticoagulation was again discontinued to avoid exacerbation of the ongoing bleed. Neurosurgery was consulted although intervention was deferred given the patient's age and multiple severe comorbidities. He was managed with an aggressive physical therapy and rehabilitation program. He was discharged to an inpatient rehab facility for further physical and occupational therapy. Following extensive multidisciplinary discussion and patient involved decision making, anticoagulation was reinitiated prior to discharge given his elevated risk of embolic events. After 2 months from his initial presentation, the patient has regained some strength in his lower extremities; now 3/5 in bilateral hips, knees, and ankles. He remains dependent on a wheelchair for ambulation but has recently been starting to stand with assistance.

4. Discussion

Cavernous malformations (CMs) are defined as abnormal clusters of thin-walled blood vessels filled located in the central nervous system.[1] Most cases have an autosomal dominant inheritance secondary to loss of function of CCM genes which are essential for the stabilization of endothelial junctions.[2] Spinal cord CMs are thought to be rare with incidence rates ranging from 0.04 to 0.05 in general populations and comprising approximately 5–12% of all spinal cord vascular lesions.[3] Gross et al. found that 38% of spinal cord cavernous malformations were cervical, 57% were thoracic, 4% were lumbar and 1% were found in unspecified locations. The average age at presentation is 42.0 years and they are

believed to be equally common in both men and women.[4] CMs are thought to be benign slow-growing lesions but have the potential for acute expansion. It is thought that CM growth depends on the accumulation of small bleeding events creating cavitation around the vessels and resulting in hemosiderin deposition into the surrounding neurovascular tissue. Other described patterns of expansion include slow growth over time of the initial malformation and rapid growth secondary to acute massive hemorrhage with the possibility of mass-effect.[5] Typical clinical manifestations correspond to the size and location of the lesion. Symptoms can include peripheral motor deficits, sensory deficits and less commonly bowel or bladder dysfunction. Most cases are found to present in a chronic progressive setting with only about 30% presenting acutely.[4] Due to the absence of pathognomonic symptoms, the diagnosis is often overlooked or confused with other pathologies including demyelination, myelitis, and intramedullary tumors.[4] Diagnosis of spinal cavernous malformation requires a thorough history, neurological exam, and imaging studies. MRI remains the diagnostic test of choice demonstrating mixed-intensity lesions surrounded by a hypointense hemosiderin ring.[6] CMs have a singular location of symptoms and tend to be minimally responsive to steroids which helps distinguish them from multiple sclerosis and other demyelinating conditions. The major complication is hemorrhagic transformation of the cavernous malformation leading to worsening mass effect. The annual rates of hemorrhagic transformation of spinal cord cavernous malformations are estimated around 1.7–4.5%.[4] Our patient was found to have an acute hemorrhagic transformation of a likely preexisting spinal cord cavernous malformation located at the level of T9 of the thoracic column leading to severe lower extremity weakness. Of note, our patient was on chronic anticoagulation which may increase the risk of hemorrhagic transformation. Surgical resection remains the only definitive management but is considered high risk. If surgery is not feasible, patients are managed with supportive care and physical therapy. Unfortunately, our patient was not a candidate for surgical intervention given his age, multiple medical comorbidities, and the ventral location of the lesion.

Disclaimers

This article has not been submitted to other publications or presented at a conference or meeting.

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Statement of ethics

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

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Data availability

All data that support the findings of this study are included in this article. Further enquiries can be directed to the corresponding author.

Conflict of interest

The authors have no conflicts of interest to declare.

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