Spinal Arteriovenous Fistula, A Manifestation of Hereditary Hemorrhagic Telangiectasia: A Case Report

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Case Report

A 32-year-old woman with a history of HHT presented to the emergency department with acute partial paralysis of the right leg, urinary retention, and right-sided back and hip pain. Magnetic resonance imaging of the spine demonstrated multiple, dilated blood vessels along the cervical spine, diffuse AVMs in the lumbar and thoracic spine, and a new arteriovenous fistula at the twelfth thoracic (T12) vertebral level. Her symptoms improved after endovascular embolization of the fistula.

Conclusion:
Spinal AVMs are thought to be more prevalent in patients with HHT. Given the high morbidity of arteriovenous fistulas, early recognition and intervention are critical. [Clin Pract Cases Emerg Med. 2020;4(3):417–420.]

Keywords:
Hereditary hemorrhagic telangiectasia; arteriovenous fistula.
Clinical Practice and Cases in Emergency Medicine

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CPC-EM Capsule

What do we already know about this clinical entity?
Hereditary Hemorrhagic Telangiectasia (HHT) is an autosomal dominant disorder characterized by arteriovenous malformations (AVMs).

What makes this presentation of disease reportable?
A rare case of spinal arteriovenous malformations and fistula in a patient with HHT who presented with neurological symptoms.

What is the major learning point?
HHT can have neurological manifestations, and, although rare, spinal AVMs are thought to be more prevalent in these patients.

How might this improve emergency medicine practice?
Given the high morbidity of arteriovenous fistulas, early recognition and intervention are critical.

subsequent ED visits at other hospitals and was documented to have increasing weakness of hip flexion (4/5), knee extension (4/5), and ankle dorsiflexion (3/5). She eventually needed crutches for mobility. Across these three visits, the patient had negative radiographs of the femur, normal CT imaging of the abdomen and pelvis, and normal MRI of the pelvis.

Her pain and weakness progressed until she lost sensation in her right leg, was unable to bear weight secondary to weakness, and was subsequently bedridden. Additionally, she developed increased urinary frequency and difficulty fully evacuating her bladder. Upon her return to the ED, she was noted to have profound (0/5) weakness of right hip flexion, knee flexion, and ankle plantar- and dorsiflexion, as well as significant (2/5) weakness of right knee extension. She had no sensation to pinprick throughout her right thigh, leg, and foot, and had hyporeflexia throughout her right lower extremity. Her plantar reflex in both feet was normal, as were her mental status and cranial nerves. She had intact rectal tone and perineal sensation.

Based on her presentation, her providers were concerned for spinal cord compression, dural compression syndrome, and spinal cord ischemia. Neurosurgery was consulted, and MRI of her entire spine was performed. MRI of her cervical, thoracic, and lumbar spine demonstrated diffuse, dilated vessels extending from her brainstem to T12, consistent with her known dural AVF (Image 1). In addition to multiple arterial feeder vessels contributing to the AVF, she had two areas of AVM at T7 and T11-12, and an area of subacute/chronic hemorrhage at T11-12 (Image 2). There was also diffuse abnormality of the spinal cord signal extending from T6 to the conus medullaris that was concerning for multiple processes including edema and ischemia.

The patient underwent angiography and embolization of a feeder vessel to the dural AVF originating at T12 that connected to the AVM at T7, but this procedure incompletely treated the abnormal flow. After the procedure, she discussed further intervention with the neurosurgical service, and again declined surgical intervention. The patient spent four weeks in the hospital postoperatively and in rehabilitation, ultimately regaining normal bladder function and motor function in her right leg.

Two years after this visit, the patient developed left-sided radicular leg pain without loss of motor function. She continued to follow up with neurosurgery but postponed management of the residual AVM due to other complications of HHT, including anemia, recurrent epistaxis, and cardiomyopathy due to high-output heart failure. She developed progressive loss of function in the right leg, and eventually underwent successful embolization of the remaining fistula pouch nearly four years after the initial visit.

DISCUSSION

HHT is an autosomal dominant disorder characterized by AVMs in the gastrointestinal tract, central nervous system, and lungs, mucocutaneous telangiectasia, and recurrent epistaxis. Thought to be caused by changes in angiogenesis, HHT manifestations develop with increasing age and range from

**Image 1.** Sagittal magnetic resonance imaging T2-weighted image of the cervical and thoracic spine. Flow voids appear black against white cerebrospinal fluid, and indicate extensive dilated vessels suggestive of dural arteriovenous fistulas. Orange arrows identify vessels perpendicular to the plane of the image, blue arrow identifies vessel at an angle to the plane with relatively increased signal. The white arrows highlight areas of spinal cord being compressed or deformed out of plane by dilated vessels.
Conclusions: Although rare, vascular etiologies such as spinal arteriovenous fistula should be in the differential diagnosis for a patient with HHT presenting with neurological symptoms. Spinal AVF can cause significant morbidity, and given the effectiveness of embolization or surgical intervention, early recognition and treatment are critical.

Patient consent has been obtained and filed for the publication of this case report.

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