

Case Report

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Cauda Equina Syndrome due to Spinal Dural Arteriovenous Fistula

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Case presentation

A 80 year old man with a prior history of benign prostatic hyperplasia was referred to neuromuscular clinic for evaluation of 6 months of progressive sensory neuropathy. He developed numbness in his left leg which one week later progressed to involve his right leg and subsequently ascended over a month to the level of his groin. On further questioning, he was newly insensate of bladder and bowel movements requiring weekly suppositories for bowel movements. He had newly required self-catheterization for one month. He had absent sensation from his lower waist down as well as altered perineal sensation. On exam, the patient had moderate proximal and distal weakness in the bilateral legs and reduced or absent sensation in the distal legs to vibration, position, and monofilament. Pinprick was absent in the dorsal feet to the midfoot and posterior legs past the buttocks. Pin prick and vibration was absent below the level of T10 on his back. His gait was slow and ataxic requiring a cane.

Diagnosis and assessment

Nerve conduction study and electromyography showed absent bilateral sural sensory responses, normal peroneal and tibial motor responses, and mild acute on chronic neurogenic changes in the left gastrocnemius only; this was interpreted as a length-dependent sensory neuropathy and mild S1 radiculopathy with incomplete reinnervation. MRI of the thoracic and lumbar spine (Figure 1) showed confluent T2/STIR hyperintensity from the conus medullaris extending through the mid thoracic cord associated with patchy enhancement and serpiginous flow voids posterior to the spinal cord.



Figure 1: MRI (T2) of thoracic and lumbar spine on presentation, showing longitudinally extensive edema throughout the thoracic and lumbar spinal cord (solid arrow) and posterior serpiginous flow voids (dotted arrow).

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The imaging findings are pathognomonic for a dural arteriovenous fistula (dAVF) with associated myelopathy.

Management

Diagnostic angiogram showed a dAVF supplied by the right L1 segmental artery with a draining vein (Figure 2, left). The fistula was ligated following T12-L2 laminectomies. Follow up angiogram demonstrated obliteration of the spinal dural AV fistula without early venous drainage (Figure 2, right). Two-month post-operative imaging showed resolution of serpiginous flow voids, significant improvement in thoracic spinal cord edema (Figure 3), though still some patchy edema remained in the distal conus medullaris (Figure 4). At follow up, the patient had improved gait, strength, and sensation. He was able to walk but continued to have perineal numbness and constipation.

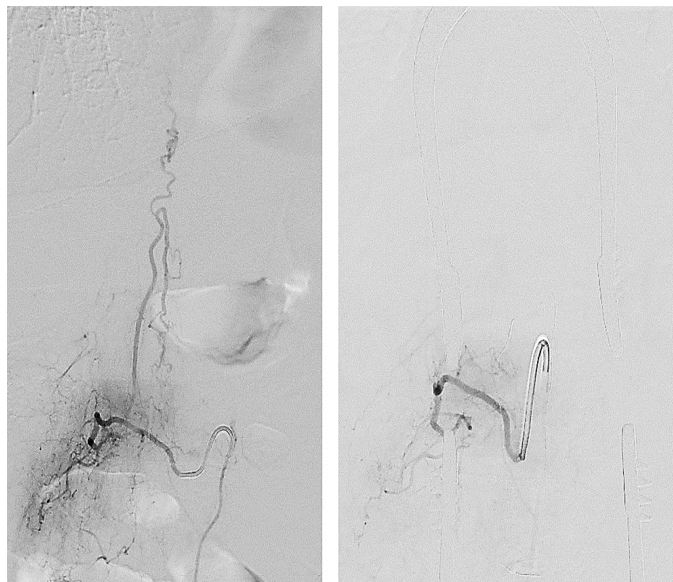


Figure 2: Initial angiogram of the right L1 segmental artery showing anomalous connection and abnormal draining vein (left), with resolution of the abnormal draining vein following surgical ligation (right).

Discussion

Spinal dural AV fistulas are the most common type of vascular malformation of the spinal cord. Overall, they are still a rare and reversible cause of progressive para- or tetraplegia. The annual incidence rate is estimated to be 5-10 cases per million [1]. The most common demographic affected is elderly men. Shunting from a radiculomeningeal artery into a radicular vein results in increased spinal venous pressure, congestion of normal spinal veins, and thus resultant spinal cord edema and dysfunction which are seen on MRI as a longitudinally extensive area of spinal cord edema, perimedullary flow voids and dilated vessels, and sometimes spinal cord enhancement. Treatment targets occlusion of the shunt, either by embolization or neurosurgical ligation. Following successful occlusion of the fistula, symptoms often improve. Presenting symptoms are often nonspecific, and diagnosis hinges on MRI imaging and diagnostic angiogram. Nevertheless, despite this being a treatable disease, the misdiagnosis rate is as high as 60% and delayed diagnosis is often associated with poor outcomes [2].



Figure 3: Comparison of pre (left) and post (right) surgical ligation MRI of thoracic spine. Axial sections are shown as insets at the levels indicated by localizer line.



Figure 4: Comparison of pre (left) and post (right) surgical ligation MRI of lumbar spine. Axial sections are shown as insets at the levels indicated by localizer line.

There are two important takeaways to highlight in our case. First, while cauda equina syndrome is often taught as an acute presentation and requires emergent evaluation, this presentation started with only sensory changes, and evolved over the next few weeks to months into a fulminant cauda equina syndrome picture. Second, initial presenting symptoms of spinal dural AVF are often nonspecific, and require a high degree of suspicion for accurate and timely diagnosis.

The patient's clinical syndrome of lower limb weakness, sensory changes, saddle anesthesia, and bowel and bladder incontinence are consistent with cauda equina syndrome. Sensory neuropathy is characterized by often symmetric loss of sensation to all modalities resulting in a sensory ataxia and loss of balance. While sensory neuropathy can be rapid and, with severe loss of proprioception, result in weakness due to inability to activate muscles, saddle anesthesia and bowel and bladder dysfunction are red flags for another process.

While most clinicians may be familiar that these red flags are concerning for cauda equina syndrome, the traditional teaching is that cauda equina presents acutely and requires emergent evaluation. However, an important feature to note in this case is that this patient's presentation started only with sensory changes and evolved over the next few weeks to months into a fulminant cauda equina syndrome picture. If clinicians are not aware that this syndrome can present on a more subacute time course, they may not pursue further investigation into an etiology. In this case, the patient's progressive sensory neuropathy was reflective of an insidious process within the spine, namely a spinal dural AV fistula, that could have been missed had the patient not been seen in follow-up and had imaging not been pursued.

Spinal dural AVFs have been described to initially mimic a peripheral nerve disorder [3]. Regardless of the level of the fistula, the caudal spinal cord is the first to be affected, resulting in initially nonspecific sensory and motor symptoms in the feet which might mimic a polyneuropathy or polyradiculopathy, as was seen in our case. By the time the classic hallmark of upper motor neuron findings with bowel and bladder dysfunction manifests which makes the diagnosis clear, patients often are already suffering from considerable deficits and delay in treatment can result in irreversible handicap.

This case demonstrates the importance of careful follow-up and attention to progression of symptoms in cases of sensory neuropathy. Also, while we frequently associate acute neurologic symptom onset with need for acute management, this is not always the case. Despite a subacute time course with progression of symptoms over 6 months, this patient had an underlying vascular etiology that required emergent management. The development of saddle anesthesia and bowel and bladder dysfunction should trigger the clinician to expeditiously evaluate the patient for cauda equina syndrome, irrespective of a more indolent time course. These patients should be immediately referred to neurosurgery for definitive management. Awareness of these principles demonstrated by this case can help avoid delays in diagnosis which translate to more negative neurologic outcomes.

Declarations

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Declaration of interest: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

1. Koch C. Spinal dural arteriovenous fistula. *Curr Opin Neurol.* 2006; 19: 69-75.
2. Ronald AA, Yao B, Winkelman RD, Piraino D, Masaryk TJ, et al. Spinal Dural Arteriovenous Fistula: Diagnosis, Outcomes, and Prognostic Factors. *World Neurosurg.* 2020; 144: e306-e315.
3. Jellema, K et al. "Spinal dural arteriovenous fistulas: a congestive myelopathy that initially mimics a peripheral nerve disorder." *Brain: A journal of neurology.* 2006; 129: 3150-3164.