Venous Congestive Myelopathy due to Chronic Inferior Vena Cava Thrombosis Treated with Endovascular Stenting: Case Report and Review of the Literature

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Abstract

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Objective—Impaired inferior vena cava (IVC) outflow can lead to collateralization of blood to the valve-less epidural venous plexus, causing epidural venous engorgement and venous congestion. Herein we describe a case of chronic IVC thrombosis presenting as venous congestive myelopathy treated with angioplasty and endovascular stenting. The pathophysiological mechanisms of cord injury are hypothesized, and IVC stenting application is evaluated.

Methods—Case report and review of the literature.

Results—IVC outflow obstruction has only rarely been associated with neurologic dysfunction, with reports of lumbosacral nerve root compression in the cases of IVC agenesis, compression, or occlusion. Although endovascular angioplasty with stenting is emerging as a leading treatment option for chronic IVC thrombosis, its use to treat neurologic complications is limited to one case report for intractable sciatica. Our case is the first description of IVC thrombosis presenting with venous congestive myelopathy, and treated successfully with IVC stenting.

Conclusion—Venous congestive myelopathy should be seen as a broader clinical condition, including not only typical dural arteriovenous fistulas, but also disorders of venous outflow. Therefore, identifying a rare, but potentially treatable, etiology is important to avoid permanent neurologic deficits. IVC stenting is proposed as a novel and effective treatment approach.

Keywords

Myelopathy; Inferior vena cava; Thrombosis; Endovascular; Stenting

Introduction

Venous hypertension from inferior vena cava (IVC) outflow obstruction causes increased blood flow into the collateral azygous–hemiazygous system and lumbar-lateral sacral venous plexus. This venous network communicates with the epidural venous plexus, providing optimal venous drainage of the spinal cord in physiological circumstances. However, as the epidural venous plexus is destitute of valves, outflow obstruction leads to epidural venous engorgement and venous congestion (Figure 1).

Rarely, abnormalities of venous return through the IVC have been associated with lumbosacral radiculopathy, presenting as sciatica [1,2], neurogenic claudication [3–5], and cauda equina syndrome [6–10] due to IVC agenesis [6,10], compression by pregnant uterus [1] or malignancy [1,3], as well as occlusion due to stenosis [1,2] or
thrombosis [1,4,7–9]. Signs of myelopathy in IVC disease have been described only once, in a female patient who presented with spastic monoplegia due to IVC compression from a pregnant uterus [11]. Neural foramen and spinal canal compression secondary to engorged epidural and paravertebral collaterals have been proposed as the major pathophysiological mechanism in radicular involvement [1,2,12,13].

Endovascular angioplasty and stenting is a treatment option in some cases of IVC thrombosis. However, to the best of our knowledge, there has been only one case where chronic IVC thrombosis with neurologic presentation (intractable sciatica) was treated successfully with endovascular stenting [2]. Herein we report the novel case of a patient with conus medullaris syndrome presenting as a complication of chronic IVC thrombosis, successfully managed with endovascular stenting. As opposed to external compression of nerve roots or spine by epidural venous plexus, venous congestive myelopathy (VCM) is hypothesized as the primary pathophysiological mechanism underlying this condition.

**Case report**

A 20-year-old male with no significant past medical history, apart from delayed umbilical cord healing, presented to our institution with urinary retention and bilateral lower extremity weakness (right worse than left). Four days prior to admission, while digging a hole for a pool, he developed sudden midline low back pain, which worsened with movement and straining. By the next day, he could barely walk. He noticed gradual ascending numbness and tingling that affected his whole body below the waist. He also reported constipation and numbness over the perineum when he wiped himself.

Neurologic examination was significant for mildly reduced strength in lower extremities. This was best appreciated when the patient attempted to walk on his

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**Figure 1. Spinal Venous Drainage.**

**A:** In the setting of normal venous drainage, blood from the spinal cord passes from the capillaries into the intrinsic venous system, the extrinsic (pial) veins, and then exits the dura via the valveless radicular veins. It subsequently joins with the extradural (vertebral) system and is carried away by efferent veins into the thoracic and lumbar intervertebral veins, finally reaching the azygous system and IVC, respectively (see inset). **B:** In the setting of IVC occlusion, venous drainage from the lower extremities, as well as the abdomen and pelvis, is redirected into the azygous system by way of paravertebral collaterals and the vertebral plexus. The vertebral plexus becomes engorged. Transmission of elevated venous pressure into the valveless radicular veins may result in congestive myelopathy due to slowed, static, or retrograde flow.
“tip-toes.” The right lower extremity was weaker than his left, and affected muscle groups included the iliopsoas, hamstrings, anterior tibialis, and gastrocnemius and soleus. Gait was wide-based. Reflexes were hyperactive in lower extremity with bilateral Babinski signs. Sensory was abnormal to pinprick in the perineal region only. Patient was unable to void and required urinary catheterization, draining initially 2.5 L. Clinically his exam was most consistent with conus medullaris syndrome.

MRI of the spine revealed epidural venous engorgement, T2 signal hyperintensity through the length of the thoracic spine, particularly inferiorly. B: Axial T2 weighted images through the lumbar spine demonstrate engorgement of the anterior internal venous plexus in the epidural space (white arrows) and basivertebral plexus (black arrow). C: Axial T2 weighted images show that the signal hyperintensity involves the central cord in the characteristic “snake eye” or “owl eye” appearance, representing injury of the gray matter. D: Postcontrast sagittal T1 images again demonstrate prominent epidural flow voids from the engorged spinal venous plexuses.

Over the next two days, the patient’s strength gradually improved. His lower back pain resolved, and weakness was undetectable. He reported that the lower extremity numbness also improved, roughly 80% normal at time of discharge, and he was able to void spontaneously. After 3 weeks, patient reported complete resolution of his neurologic symptoms.

Discussion

To the best of our knowledge, this is the first reported case of myelopathy due to IVC occlusion. The cause of this patient’s IVC occlusion is not entirely clear. The history of delayed healing of umbilical cord stump, fibrotic occlusion, and age of presentation, suggests a congenital thrombosis of the IVC. Most congenital IVC abnormalities remain asymptomatic due to development of collaterals, but collateral flow can become overwhelmed with aging, leading to venous insufficiency,
venous congestion, and epidural venous engorgement. As neuroimaging did not reveal compression of nerve roots or spinal cord, a mechanism of VCM can be hypothesized. This conclusion is supported with the patient’s clinical improvement following IVC recanalization and stenting.

Epidural venous congestion can reduce the arteriovenous pressure gradient within the cord, decreasing tissue perfusion and promoting chronic ischemia. This mechanism is similar to that of spinal epidural arteriovenous fistulas (AVF) [14,15]. In these cases, multiple small arterial vessels feed one or more epidural veins, leading to epidural venous plexus engorgement, which, after reaching a significant pressure gradient, can cause congestion or even reflux into the radiculomeningeal and then perimedullary veins. In our case, the mechanism of venous congestion is different from that of typical dural AVFs, so the classic imaging characteristics of VCM with serpentine enhancing veins on the cord surface (dilated perimedullary veins) were not present. Increased intrathoracic pressure, which occurs with exertion, such as digging holes, could acutely worsen venous return, and force pooling of blood into an already overwhelmed epidural plexus, increasing congestion or reflux to perimedullary veins with ensuing spinal cord ischemia. This is an accord with Kamerath and Morgan’s report of exercise-induced lower extremity numbness in a patient with IVC agenesis, wherein postexercise MRI showed a notable increase in the volume of the venous plexus [16].

In the 20 reported cases of patients with neurologic disorders in the setting of IVC involvement, one or more likely etiologies were found in 16 patients. These included IVC agenesis (2 cases) [6,10]; and IVC thrombosis attributed to oral contraceptive use (6 cases) [1,7,10], pregnancy (3 cases) [1,11], malignancy (2 cases) [1,3], Behcet’s disease (1 case) [1], Budd–Chiari syndrome (1 case) [2], and protein C deficiency (1 case) [1]. In cases of pregnancy, malignancy (1 out of 2), and protein C deficiency, treating the etiology with delivery, chemotherapy, and anticoagulation, respectively, was associated with good outcomes. In other cases of acute IVC thrombosis, other treatment approaches were performed with good outcomes reported, including venous thrombectomy (3 cases) [1,7,9]; intracatheter thrombolysis with balloon angioplasty (1 case) [7]; and systemic anticoagulation alone (10 cases) [1,8,10]. In the literature, there are only two reports of patients with chronic IVC occlusion presenting with neurologic symptoms. In one case of IVC agenesis, a patient presented acutely with cauda equina syndrome and underwent laminectomy, bilateral foraminotomies, and coagulation of epidural veins [6]. In another case, a patient with Budd–Chiari syndrome presented with intractable sciatica and underwent endovascular intervention with a 25-mm-wide 50-

Figure 3. Contrast-enhanced CT of the abdomen and pelvis demonstrates a: IVC narrowed and unopacified (black arrow) relative to the hepatic veins and B: large venous varices in the retroperitoneum (white arrows). C: Status post endovascular IVC angioplasty and stent placement, demonstrating preferential flow to the stented IVC.
mm-long self-expandable stent [2]. Both patients had complete resolution of symptoms. Angioplasty alone in chronic IVC thrombosis has been associated with venous recoil, low flow state, and rethrombosis. In patients with both long and short IVC occlusions, stenting helps maintain a large venous conduit, which promotes long-term patency [17,18]. Therefore, endovascular recanalization with stenting has emerged as the treatment of choice for chronic IVC thrombosis. Surgical procedures directed at decompression of the spine and resection of venous engorgement may not promote long-standing benefit, as venous hypertension will persist and affect the remaining collateral vessels. IVC occlusion needs to be considered in the diagnosis of patients with VCM, and these patients should be evaluated for potential etiologies of IVC occlusion, including structural abnormalities, portal hypertension, oral contraceptive use, thrombophilias, and malignancies.

**Conclusion**

Vascular myelopathy is a serious neurologic condition, and delay in treatment is associated with permanent neurologic deficits. It is important to identify potentially treatable etiologies, such as IVC thrombosis or other causes of venous outflow obstruction. Congestive myelopathy should be seen as a broader clinical entity, secondary to venous hypertension or reduction of the perfusion pressure gradient from both central and peripheral etiologies, not only due to previous well-described dural AVFs. In cases of chronic IVC obstruction presenting with neurologic symptoms, endovascular stenting appears to be a novel and effective approach. Evaluations for possible underlying etiologies of IVC occlusion should always be considered in order to prevent rethrombosis and recurrence of symptoms.

**References**


