Sacral preservation in cauda equina syndrome from inferior vena cava thrombosis

Case report

ALIK S. WIDGE, M.D., PH.D.,1 NESTOR D. TOMYCZ, M.D.,2 AND ADAM S. KANTER, M.D.2

1School of Medicine and 2Department of Neurosurgery, University of Pittsburgh, Pennsylvania

Acute cauda equina syndrome can occur due to a variety of causes. Inferior vena cava (IVC) thrombosis has been reported as the causal source of this phenomenon twice in the relevant literature, both cases of which presented in a form complete with a component of bowel and/or bladder dysfunction. The authors report an atypical case of cauda equina syndrome in a patient in a hypercoagulable state with an extensive IVC thrombosis, resulting in acute paraparesis in the absence of incontinence or perineal anesthesia. An increasing number of prophylactic and/or therapeutic IVC filters placed in the perioperative period should engender an increased clinical suspicion for IVC thrombosis in patients presenting with acute paraparesis. (DOI: 10.3171/2008.12.SPINE08389)

KEY WORDS • acute paraparesis • cauda equina syndrome • deep venous thrombosis • vena cava thrombosis

Cauda equina syndrome arises from an insult to the collection of lumbosacral nerve roots that arise from the caudal spinal cord. In its classic form, CES presents with symptoms of low-back and leg pain, lower-extremity weakness and sensory loss, perineal anesthesia, bladder/bowel incontinence, and erectile dysfunction.19 Bowel and bladder dysfunction are often considered essential to the diagnosis of CES,6,15,22,25 although many cases of CES with complete sparing of sacral dysfunction have been described.12 Patients with CES often exhibit subtle signs such as decreased perineal sensation and mild urinary retention, signs that may be concealed in the hospitalized patient.8 Thus, the manifestation of CES occurs across a broad spectrum, from dramatic complete forms with profound sacral dysfunction to more insidious partial forms that often contribute to a long delay before diagnosis.17

Lumbar disc herniations remain the primary pathology evoking CES, although hemorrhagic spinal tumors9,26 and arteriovenous malformations19 have also been described. Other rare causes include ischemia (such as aortic dissection21 and abdominal vascular surgery19), osteomyelitis/discitis impacting the epidural space,19 inflammatory polyneuropathy,13 vasculitis,11 and traumatic injury from retropulsed vertebral fragments8 or a penetrating foreign body.14 Acute CES presenting with severe lower-extremity sensorimotor and bowel/bladder dysfunction secondary to IVC thrombosis has also been reported. We report a unique case of acute CES caused by IVC thrombosis in a patient who presented with moderate lower-extremity motor and sensory disturbance, without saddle anesthesia or bowel/bladder incontinence.

Case Report

History and Presentation. This 55-year-old Caucasian male presented with a rapidly progressive paraparesis 16 days after admission to the hospital following an assault. His medical history included a warfarin-dependent factor V Leiden mutation, DVT, pulmonary embolus, IVC filter placement, and lumbar stenosis. The patient was neurologically intact upon admission. He subsequently developed a lower-extremity compartment syndrome necessitating multiple left lower-extremity fasciotomies. His anticoagulation therapy was switched from warfarin to low-dose subcutaneous heparin on hospital Day 10.

On the 16th hospital day, the patient awoke with right-leg weakness, which progressed to involve the left leg by early afternoon. On examination, he was insensitive in both lower extremities to light touch and pinprick stimulation. The lower extremities were cold to the touch, cyanotic, and edematous. A motor examination revealed hip flexor antigravity strength (3/5 on the Medical Research Council muscle grading scale) with total plegia of all muscle groups below the knee. Truncal and upper-extremity strength and sensation remained normal.

Operation and Postoperative Course. An emergency CT scan revealed severe thrombosis of the IVC with ex-
tension into both iliac veins. The patient was immediately taken to the operating room for a pharmacomechanical endovascular thrombectomy (Fig. 1). Postoperatively, he received a continuous femoral catheter tPA infusion and intravenous heparin. The patient’s neurological examination results improved significantly within 24 hours. Warfarin was restarted on postoperative Day 2. At that time, he experienced full return of lower-extremity sensation and improvement in his distal lower-extremity motor strength to 4/5. He was subsequently transferred on postoperative Day 8 to a rehabilitation service with full (5/5) lower-extremity strength and normal sensation.

Discussion

Inferior vena cava thrombosis is a rare cause of acute CES. The anatomical link between the IVC and the cauda equina is the venous connection, via the iliolumbar veins, between the IVC and the valveless Batson plexus surrounding the spinal cord and nerve roots. Branches of the Batson plexus run within the spinal epidural space and neural foramina along the nerve roots. In addition to the connection to the IVC, lumbar veins also ascend above L-2 and connect to the azygos venous system. Thus, thrombosis of the IVC necessitates that the lumbar plexus drain the lower extremities, leading to reversal of flow and venous dilation.\(^1\) This dilation can result in lumbosacral nerve root dysfunction via both ischemic and inflammatory mechanisms.

The cauda equina nerve roots have only a thin connective tissue sheath and limited vascularity, making them particularly sensitive to compressive forces.\(^{19}\) A vascular watershed zone exists within these roots; it has been shown that as little as 10 mm Hg of increased pressure can produce microischemic zones.\(^{15,19}\) This vulnerability is exacerbated by the venous stasis inherent in IVC thrombosis.\(^4\) In the patient presented here, preexisting lumbar stenosis may have additionally contributed to the CES by “precompressing” the cauda equina, thus reducing its functional reserve.

Local inflammation may also contribute to acute CES in the setting of IVC thrombosis. Thrombi provoke an intravascular inflammatory reaction with immune cell infiltration and resultant mass effect.\(^1\) Even in the absence of frank compression, venous inflammation and/or dilation may irritate cauda equina nerve roots akin to the neurovascular conflict observed in trigeminal neuralgia.\(^7,20\)

The 2 prior reports of acute CES from IVC thrombosis described a complete form of CES involving low-back pain, lower-extremity weakness and pain, perineal numbness, bowel/bladder dysfunction, and lower-extremity hyporeflexia.\(^3,16\) The symptomatology noted in our patient has only been previously described in more chronic presentations.\(^5,10,24\) The lack of sacral root symptoms in this case may be explained by the anatomical arrangement of the cauda equina nerve roots. The lumbar roots lie ventrally, particularly at higher lumbar levels, placing them closer to the anterior epidural and paravertebral venous plexus. Necropsy models using Young’s modulus have demonstrated that the linear strain on stretched nerve roots within the cauda equina may be least on the sacral roots, which may explain cases of CES such as this one in which sacral dysfunction was not evident upon examination.

The underlying cause of the IVC thrombosis leading to CES also remains inconsistent, but patients generally have a thrombotic predisposition. Reported causes include a factor V Leiden mutation,\(^5\) protein C deficiency,\(^20\) an antiplatelet antibody,\(^24\) mechanical compression of the IVC,\(^20\) and Behçet disease affecting the spinal vasculature.\(^20\) In addition to hereditary thrombophilia, the presence of an IVC filter may have been an independent risk factor for IVC thrombosis in this patient. In a retrospective review, Corriere et al.\(^3\) found that retrievable filters, especially those of a biconical design, increased the incidence of IVC thrombosis 200-fold. In the pediatric population, abnormalities of the vena cava, such as stenosis, must be considered as causative agents as well.\(^16,18,23\)

Symptoms and signs of DVT, including limb swelling,\(^3\) painful cyanosis (phlegmasia cerulea dolens),\(^3,4\) ab-

![Fig. 1. Computed tomography scan through the abdomen with intravenous contrast administration (left) shows a large thrombus occluding the IVC, with infiltrative changes in the vessel wall (arrowhead). After 2 consecutive attempts at thrombectomy with intervening tPA infusion, postoperative venography (right) demonstrated only a residual clot attached to the IVC filter (arrowhead).](image-url)
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dominal compartment syndrome, or simple paleness of a single extremity occur with most cases of IVC thrombosis. In one patient series, massive limb swelling was the most common sign, occurring in 7 of 10 patients. However, neurological signs of IVC thrombosis can develop over weeks or months, and may be unaccompanied by other signs or symptoms of acute thrombosis.

Cauda equina syndrome caused by IVC thrombosis may be diagnosed using MR imaging, although caution in using this modality is warranted because dilated epidural veins may be confused with a prolapsed disc or cystic tumor. An IVC thrombus can also be visualized on ultrasonography, but this method does not delineate the lumbar plexus. In clinically unstable or rapidly deteriorating patients, such as the one described in this report, a CT scan with intravenous contrast administration may be the most appropriate diagnostic tool. Both the dilated lumbar veins and the IVC may exhibit peripheral enhancement and resemble masses if there is sufficient local inflammation.

The definitive therapy for acute IVC thrombosis is thrombolysis. In cases of subtotal occlusion, intravenous heparin therapy with conversion to oral warfarin has been sufficient. Eight cases have been managed successfully with subcutaneous low-molecular-weight heparin alone. The most common treatment strategy is clot lysis with subcutaneous low-molecular-weight heparin prophylactically or following the diagnosis of DVT, may increase the risk of neurovascular compromise. Expedient thrombectomy with anticoagulation treatment has been shown to enable patients to achieve a full neurologic recovery.

Disclosure

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References


Address correspondence to: Adam S. Kanter, M.D., Department of Neurological Surgery, University of Pittsburgh, 200 Lothrop Street, Pittsburgh, Pennsylvania 15238. email: kanter@upmc.edu.