Cauda equina syndrome secondary to an absent inferior vena cava managed with surgical decompression

Case report

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The authors report on the case of a 24-year-old man who presented with back pain and radiculopathy due to epidural venous engorgement in the setting of a congenitally absent inferior vena cava. Despite initial improvement after steroid administration, the patient’s health ultimately declined over a period of weeks, and signs and symptoms of cauda equina syndrome manifested. Lumbar decompression was performed and involved coagulation and resection of the compressive epidural veins. No complications occurred, and the patient made a full neurological recovery. (DOI: 10.3171/2011.10.SPINE1121)

KEY WORDS • cauda equina syndrome • laminectomy • inferior vena cava • congenital abnormality

Cauda equina syndrome resulting from compression of the lumbosacral nerve roots exiting the caudal spinal cord classically presents with a constellation of acute low-back and leg pain, saddle anesthesia, lower-extremity weakness, and bowel or bladder dysfunction. There are numerous causes of CES including traumatic spinal injury, disc herniation, critical spinal stenosis, spinal cord neoplasms, inflammatory conditions, infectious conditions, iatrogenic causes, and vascular pathology.

There have been several reports of CES secondary to vascular pathology, and 2 reports of low-back pain and sciatica associated with an absent inferior vena cava with subsequent dilation of epidural vessels and compression of the thecal sac. There is, however, no previous description of CES due to an absent IVC.

Congenital absence of the IVC is uncommon but has been increasingly recognized with the development of cross-sectional abdominal imaging. The pathophysiology leading to this anomaly is not entirely clear. Theories range from true embryonic anomaly to sequelae of perinatal IVC thrombosis. The reported prevalence of congenital anomalies of the IVC ranges from 0.07% to 8.7%. In a small percentage of these individuals there will be a complete absence of the infrarenal or infrahepatic IVC.

Case Report

Initial Presentation. This 24-year-old man with a medical history of Type I diabetes and cigarette smoking presented to the emergency department with a 2-month history of low-back pain that started with an episode of heavy lifting. He also complained of bilateral lower-extremity pain and paresthesias. The patient described an additional history of slowly progressive weakness resulting in difficulty ambulating, rising from a seated position, and repeated falls that finally led to an inability to ambulate at presentation. At this time, he denied bowel or bladder dysfunction and exhibited none of the red-flag signs or symptoms of low-back pain. His examination at this time was remarkable only for Grade 3/5 strength throughout the bilateral lower extremities.
Due to the severity of his low-back pain combined with a radicular component, MR imaging was performed. The studies revealed flow voids in the spinal epidural space extending from T-12 through S-1 (Fig. 1). Given the patient’s severe low-back pain and its radicular component, we speculated that the symptoms were potentially the manifestation of nerve root irritation due to not only compression but also release of inflammatory cytokines in the perivascular space. Thus, the patient was started on a short tapering course of dexamethasone, with the understanding that dexamethasone could theoretically exacerbate the symptoms because it can lead to increased intravascular volume. Furthermore, the patient was admitted to the neurosurgical intensive care unit for close monitoring. A 28-vessel diagnostic spinal angiographic study revealed no evidence of a spinal AVM or fistula. Further review of the MR images suggested an IVC anomaly. This prompted us to order a CT scan of the abdomen and pelvis, which revealed an absent infrahepatic IVC (Fig. 2). It was concluded that the flow voids seen on MR imaging were consistent with dilated epidural veins resulting from congestion within the Batson venous plexus secondary to an absent IVC (Fig. 3). The vascular surgery service was consulted but no intervention was recommended. The patient was treated conservatively with a short dexamethasone taper, and he experienced symptomatic improvement. He was neurologically intact at the time of discharge. On hospital Day 6, the patient was discharged to home with instructions to taper his steroids and to undergo follow-up in the neurosurgical clinic.

Second Presentation. Three weeks after being discharged from the hospital, the patient returned to the emergency department, this time with a 4-day history of progressively worsening urinary retention and a single episode of fecal incontinence. He had worsening back pain and recurrence of his bilateral lower-extremity weakness. The neurological examination was notable for symmetric weakness in the lower extremities (Grade 3/5 in the iliopsoas, Grade 2/5 in the quadriceps, Grade 2/5 in the tibialis anterior, and Grade 2/5 in the gastrocnemius muscle). Decreased sensation in the lower extremities was also noted, both to light touch and pinprick, although a sensory level could not be reliably established. Perineal sensation was normal. Diminished bilateral patellar and ankle-jerk
Cauda equina syndrome due to an absent inferior vena cava

reflexes were found; the toes were down-going. Rectal tone was normal.

Operation. Given this fulminant presentation with urinary retention and weakness, surgical decompression was urgently performed. The patient was intubated, placed prone on a Jackson table, and fluoroscopic guidance was used to help make an incision that extended from L-4 to S-1. These levels corresponded to the levels of venous engorgement that caused significant stenosis, as observed on MR imaging. Conventional subperiosteal exposure was performed with electrocautery followed by laminectomy and bilateral foraminotomies at the aforementioned levels. After removal of the ligamenta flava, large, dilated epidural veins could be seen both lateral and anterior to the thecal sac (Fig. 4). Using the operative microscope, these enlarged veins were cauterized with bipolar coagulation and then resected with microscissors. We decided to coagulate the epidural veins intraoperatively based on the compressive nature of venous structures. First, we believed that a wide central posterior decompression might not alleviate the anterior and lateral compressive nature of the pathological entity. Second, we were concerned that the pathophysiology behind the patient’s clinical presentation might not be purely the result of compression alone but could potentially involve inflammatory processes related to the abnormal venous structures. At the conclusion of the procedure, the thecal sac and pertinent nerve roots were completely decompressed.

Postoperative Course. The patient’s pain and radiculopathy largely resolved following decompression. Furthermore, he experienced full recovery in motor function and sensation. By postoperative Day 2, he was voiding without difficulty and had had no further episodes or urinary or fecal incontinence. At a 6-week follow-up clinic visit, the patient was neurologically intact. Because the patient lived a significant distance from our institution, we conducted a 13-month follow-up via telephone. The patient stated that he continued to be neurologically intact and had had no further recurrence of symptoms.

Discussion

Neural compression due to dilated lumbar epidural veins is a rare but recognized phenomenon in the neurosurgical literature. Whether through a direct arteriovenous connection, as might occur with spinal AVF, or through globally impaired venous return (for example, in IVC thrombosis, Budd-Chiari syndrome, pregnancy, morbid obesity, or portal hypertension), venous hypertension precedes and promotes varix formation of the intervertebral and spinal epidural veins and results in compression of the nerve roots and thecal sac. Back pain and lumbar radiculopathies are the most common presenting symptoms, but more diffuse involvement may lead to epidural compression manifesting as neurogenic claudication or CES.9,18,21

As demonstrated in the present case, the congenital absence of the IVC may rarely be the cause of epidural venous congestion leading to symptoms of neural compression and should be considered in the differential diagnosis of lumbar radiculopathy or neurogenic claudication. The anatomical link is through the Batson venous plexus, which surrounds the spinal cord and drains via iliolumbar veins into the IVC. The normal embryological development of the IVC is a complex process that involves the formation, regression, and anastomosis of 3 longitudinal pairs of veins. Anomalies in its development may lead to variable presentations of abdominal and lower-extremity venous congestion.8,17,23,24

Presumably, in patients with congenital absence of the IVC, various collateral channels for venous return develop, initially preventing the venous engorgement that might lead to varix formation. Over time, and with the natural progression of venous insufficiency that accompanies increasing age, these collateral channels can become overwhelmed, leading to symptom onset. We considered the possibility of intradural venous congestion as a potential cause of neurological dysfunction in our patient. However, when neurological deterioration has been associated with intradural venous congestion, it has been due to spinal dural AVF and, less frequently, spinal intradural AVM; neither of these was present in our patient.14,22 In the present case, relatively diffuse, mostly anterolateral epidural varices and the progres-
In cases of congenital IVC absence, venous clots are common\(^1\) and should aggressively be sought, and if present, thrombolysis and anticoagulation should be considered in the treatment strategy. In the patient in question, no deep vein thrombosis or other thrombus was discovered. The appearance of extensive, abnormal flow voids on MR imaging in patients with associated neurological symptoms often raises the suspicion of a spinal AVM. In the present case, angiography demonstrated no fistulous connection. It is worth questioning the necessity of performing this invasive study in patients with dilated epidural veins if acute occlusion or the congenital absence of the IVC has also been recognized. Because the mechanism for AVF formation is not well understood and because it has been theorized that venous hypertension may be a factor,\(^1\) it is not inconceivable that these entities may coexist, although this has not been reported in the literature. Nevertheless, if symptoms progress and lumbar decompression is being considered, it is critically important to be certain about the etiology of venous engorgement because this will directly affect the surgical management. Dilated veins can and generally will be coagulated with impunity in patients with IVC occlusion whereas preservation of draining veins would be of paramount importance during open surgical disconnection of a spinal AVF. However, it should be kept in mind that the development of a large epidural venous plexus in our patient was most likely a response to an absent IVC, and coagulating these veins could potentially lead to other clinical manifestations, as seen in our patient. During the 13-month follow-up call when asked if he had experienced any new signs or symptoms since surgery, the patient did state that in the past several months he had developed “varicose veins” in his legs; he denied ulcerations related to these lesions. Varicose veins secondary to venous insufficiency in the setting of absent IVC has been described previously.\(^1,8\) Although these findings might represent the natural progression of the patient’s chronic venous insufficiency and that they were not present prior to surgery, one possible explanation is a compensatory mechanism in the setting of further diminished venous return after coagulation of the patient’s dilated epidural veins.

Lumbar decompression led to an immediate and durable resolution of symptoms in our patient. Preoperatively the epidural veins were found to be particularly engorged and were liberally coagulated to achieve decompression. In the case of acute IVC thrombosis causing epidural compression, endovascular thrombectomy is generally attempted initially and has been shown to lead to prompt symptom resolution.\(^8\) When the IVC is absent, the benefits of reconstructing the venous drainage via bypass must be weighed against the risks. The vascular surgery service was consulted in the case of our patient and did not believe that an endovascular or more extensive open surgical venous reconstruction was indicated.

**Conclusions**

Lumbar laminectomy is a simple and relatively low-risk intervention that should be considered as a first-line treatment option in patients with a congenitally absent IVC and severe symptoms of lumbar epidural compression. Steroids and mannitol, although unproven, may help decrease local edema and may be considered as temporizing measures until definitive surgical decompression is undertaken. In patients with frank CES, however, delay to treatment may result in permanent neurological deficits and poor outcomes.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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M. Ghiassi et al.
Cauda equina syndrome due to an absent inferior vena cava


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