

Bilateral lower-extremity numbness and ataxia in a young female runner

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Inferior vena cava (IVC) absence is thought to have either embryologic or developmental etiologies, depending on the degree of absence. Entire absence of the IVC is thought to be due to embryologic insult, whereas recent studies propose that infrarenal IVC absence is developmental, secondary to perinatal thrombosis. Here we report on an adolescent woman with infrarenal absence of IVC and common iliac veins. Clinically, she presented with bilateral lower-extremity numbness and ataxia following strenuous exercise (running > 1 mile). Symptoms resolved with 30 seconds of rest. Radiographically, MRI revealed extensive collateral vasculature that had developed within the paravertebral soft tissues and epidural space of the spinal canal; these collaterals coursed through the neural foramina and caused moderate stenosis at L4/5 and L5/S1.

Introduction

Absence of the infrarenal portion of the IVC is a very rare anomaly with very little reported literature; fewer than 20 cases have been proven by imaging (1). While the idea is controversial, it has generally been accepted in the literature that the most likely etiology for this anomaly is developmental secondary to thrombotic insult in the perinatal period, rather than that of congenital malformation.

Embryological development of the IVC proceeds separately as four segments: hepatic, suprarenal, renal, and infrarenal. Each of these segments develops respectively from embryologic structures during the first 6-8 weeks of embryonic life; the hepatic portion develops from the vitelline vein, the suprarenal from the right subcardinal vein, the renal from supracardinal and subcardinal anastomoses, and the infrarenal from the right supracardinal and postcardinal

veins. The postcardinal veins persist in the pelvis as the common iliac veins. Thus, absence of the infrarenal portion of the IVC has been thought to be due to abnormalities in the development of the supracardinal veins (2). It has been proposed that this anomaly is developmental in nature, in part because there have been no other associated congenital abnormalities reported with a missing infrarenal IVC (3). Additionally, it is difficult to pinpoint a single embryologic event that would lead to this anomaly. Because of these reasons, it has been hypothesized that a perinatal event is the most likely etiology (1, 4, 5). Additionally, a single case report in 2001 (3) actually showed evidence of perinatal deep vein thrombosis (DVT) of an infant who later (at the age of 12) presented with absence of infrarenal IVC.

IVC abnormalities leading to increased risk of DVTs have been reported in the literature as well (6-11); in one study, it was found that 5% of patients younger than 30 who presented with DVT had an absent IVC (12). However, these case reports have typically been associated with other IVC abnormalities, such as intrahepatic interruption of the IVC with azygous continuation, rather than infrarenal IVC absence. However, infrarenal absence has been associated with perinatal DVT in at least one case report (3) and suggested in another (13).

Here we report on a 14-year-old female athlete with absent infrarenal IVC and common iliac veins and neurologic symptoms, confirmed by CT and MRI. While other reports have commented primarily on the DVT risk of IVC

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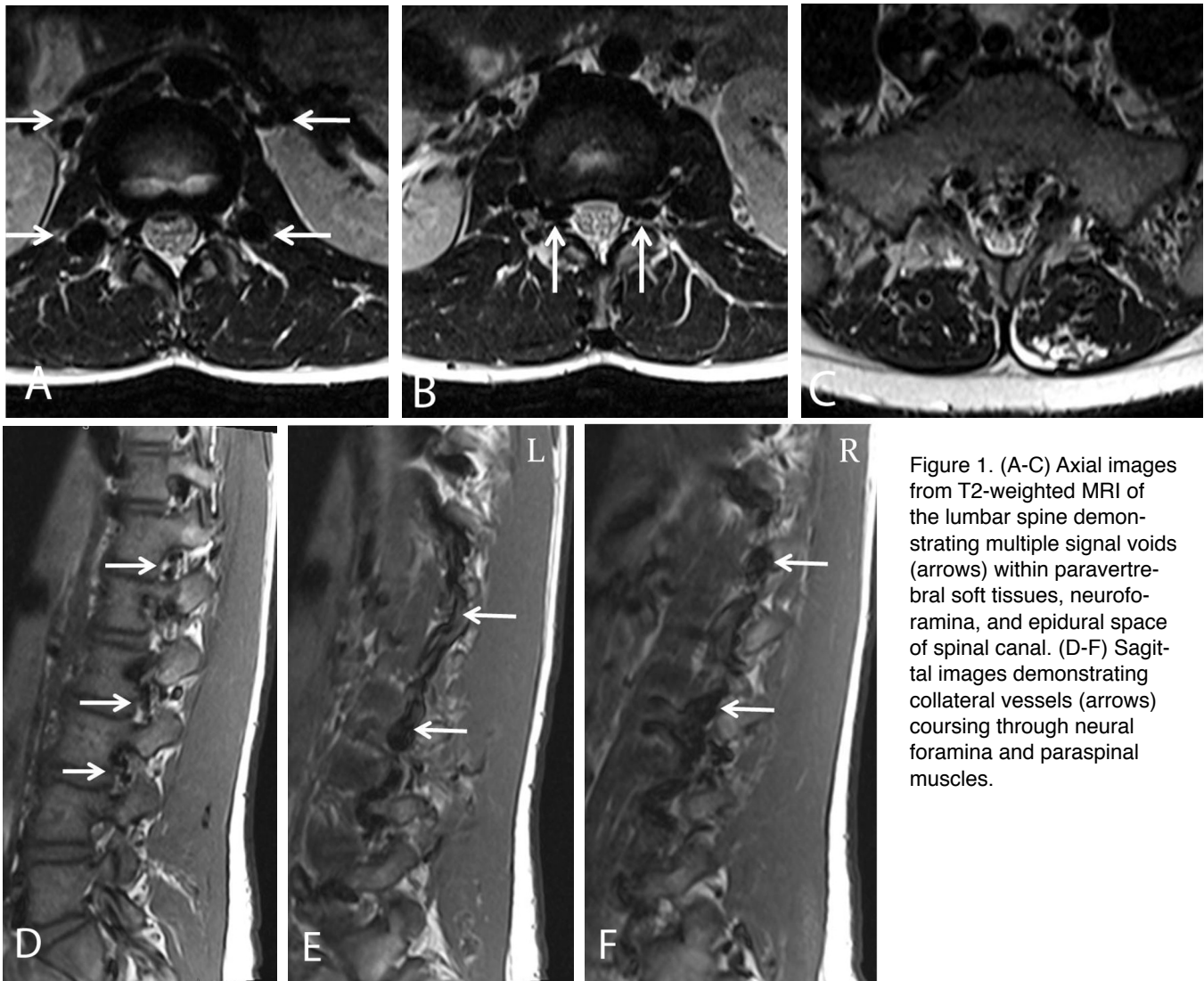


Figure 1. (A-C) Axial images from T2-weighted MRI of the lumbar spine demonstrating multiple signal voids (arrows) within paravertebral soft tissues, neuroforamina, and epidural space of spinal canal. (D-F) Sagittal images demonstrating collateral vessels (arrows) coursing through neural foramina and paraspinous muscles.

anomalies, we believe this is the first report that demonstrates neurologic symptoms secondary to an IVC anomaly and not specifically secondary to the effects of a DVT.

Case report

A 14-year-old female cross-country athlete presented to our institution's Center for Sports Medicine with complaints of an 8-week history of bilateral lower-extremity numbness and ataxic gait after running longer than one mile. These symptoms caused the patient to cease running, and the symptoms then resolved within 15-30 seconds. The patient denied any back pain, bowel/bladder dysfunction, or trauma. The patient's physical examination and laboratory values were normal. The patient endorsed no significant family or personal medical history. When seen at the sports medicine clinic, the patient ran on a treadmill until her symptoms and ataxia were reproduced and witnessed.

Radiographs of the lumbar spine revealed no abnormalities. A 2D echocardiogram was also normal. MRI of the lumbar spine showed numerous signal voids within the

paravertebral soft tissues and the epidural space of the spinal canal, suggestive of extensive collateral vasculature (Fig. 1). Collateral vessels coursed through neural foramina at multiple levels and resulted in moderate central canal stenosis at L4/5 and L5/S1. These findings raised concern for impaired venous return, potentially due to compression from a mass or congenital anomaly. Since the limited field for the lumbar MRI did not permit evaluation of the IVC or iliac veins, a contrast-enhanced CT of the abdomen and pelvis was performed. This revealed absence of the inferior vena cava and common iliac veins, beginning just inferior to the level of the renal veins. This anomaly caused engorgement of the internal/external iliac veins and forced drainage into collateral vessels, including the ascending lumbar veins and azygous/hemi-azygous systems. No mass lesion or adenopathy was identified (Figs. 2 and 3).

Based on the extensive collateral circulation that had developed, particularly in the spinal canal, it was thought that during exercise these venous systems would become enlarged, causing neural impingement and increased canal

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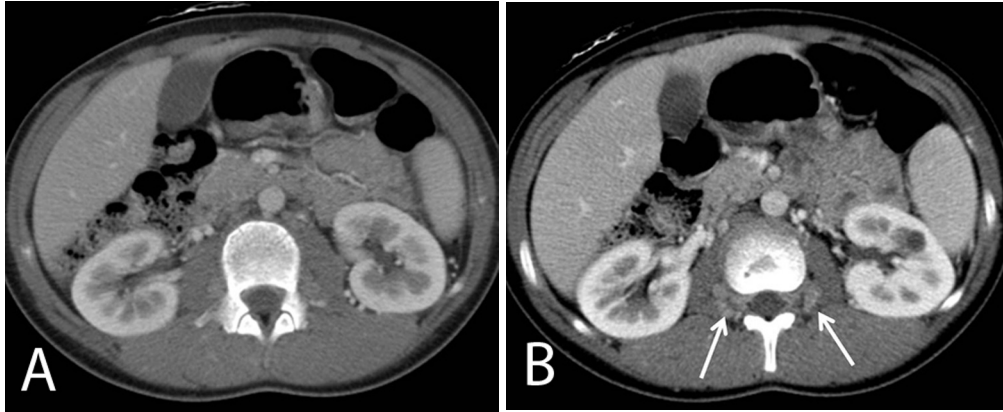


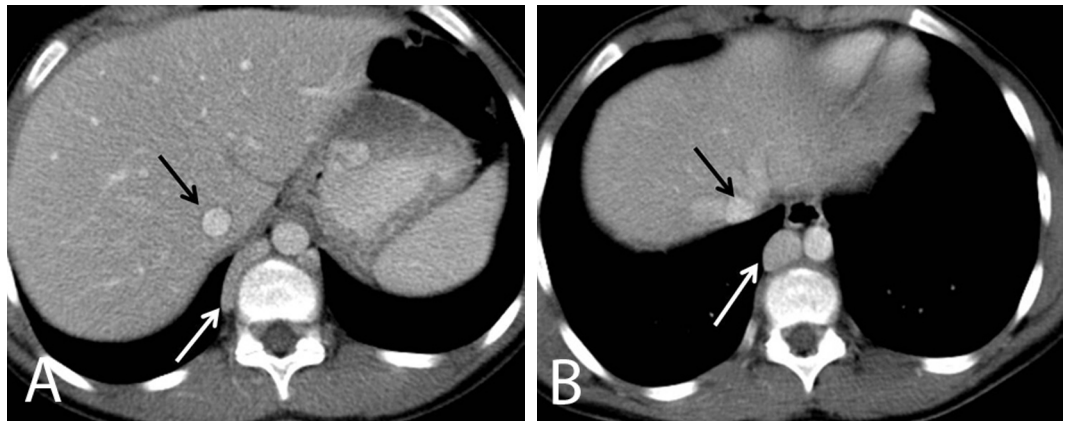
Figure 2. (A-B) Absence of infrarenal portion of IVC and collateral vessels (arrows) in the paraspinal tissues.

stenosis of the lumbosacral spine. This would be compatible with the reported bilateral leg numbness following exercise. Additionally, the poor venous return was thought to cause a transient decrease in cardiac output, producing ataxia. The patient was then advised to limit her participation in strenuous cardiovascular activities. She was also advised on her increased risk of DVT, and thus counseled to not smoke or take oral contraceptives; she was also advised on the risks of pregnancy. She was placed on prophylactic aspirin. Vascular surgery was consulted as well; they agreed with the treatment plan as laid out and advised delaying surgical intervention until the patient reached adulthood. At a 3-month followup, the patient had still not returned to exertional activity.

this has been reported in many case reports (many of which involve a missing intrahepatic IVC, however) and theoretically should apply to infrarenal anomalies as well. The collaterals that develop as a result of the absent IVC might be unable to fully accommodate the blood flow, especially during major exertion; this could in turn generate venous stasis, and possibly DVT (9).

Interestingly, the patient discussed in this case report did not have any imaging evidence of thrombosis, though nearly all of the case reports to date show patients being identified with IVC anomalies showing symptoms that were later explained to be secondary to DVT formation. Since the collateral vessels in this patient had developed in the epidural space of the spinal canal, and also at multiple lev-

Figure 3. A-B) Postcontrast CT shows engorgement of the azygous vein (white arrows) and a normal-appearing intrahepatic portion of the IVC (black arrows).



Discussion

It has generally been accepted that absence of the infrarenal portion of the IVC is due to developmental insult, most likely perinatal IVC thrombosis. This has been hypothesized and confirmed by at least two case reports (3, 13). We agree with this theory, as in our case, a single embryologic abnormality would not account for both a missing infrarenal IVC and common iliac veins; the origin for these vessels is separate (supracardinal and postcardinal, respectively). The primary clinically significant outcome of this anomaly has been thought to be increased risk of DVT;

els through the L4/5 and L5/S1 neural foramina, engorgement of these vessels was thought to have caused her neurologic symptoms. The ataxia she experienced was most likely due a decrease in cardiac output secondary to decreased preload from the poor collateral venous return. This finding is compatible with the fact that major exertional activities might not be able to be accommodated by the collateral development (9).

Treatment for this condition is typically limited to DVT prophylaxis (prolonged oral anticoagulation, elastic stockings) after evidence of DVT has been found on imaging; one study used prolonged Vitamin K antagonists as well (9). Regardless of DVT findings, however, patients likely do

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have an increased risk of DVT and should be counseled on the appropriate lifestyle measures—avoiding prolonged immobilization and observing oral contraceptive/pregnancy risks. In this patient in particular, since only exercise reproduced her symptoms, it was advised that she stop strenuous activities as well; development of DVT would likely be higher given the increased venous stasis secondary to decreased collateral accommodation that exercise would cause. Since she had not developed a DVT yet, we felt that aspirin prophylaxis was appropriate at this time. Given that the patient's symptoms were reproduced only by intense activity and not present at rest, surgical management was appropriately delayed at this time; a case report on the surgical reconstruction of an absent infrarenal IVC showed that it was efficacious for symptom relief, but that it should be performed only with a high level of symptom severity, and that oral anticoagulation would be the preferred treatment for the majority of cases (14).

Our case demonstrates that while the majority of IVC anomalies present clinically with symptoms related to DVT formation, this condition can actually have multiple effects depending on which structures the collaterals impact; thus, clinicians should keep IVC anomalies on the differential for young patients with unexplained lower-extremity neurologic symptoms. Additionally, we have shown the importance of CT and MR in the diagnosis of this condition, and primarily emphasize the importance of noting the signal voids seen on MR as evidence of collateral vessels. We also emphasize noting the path the collaterals take as seen on imaging, as they can significantly impact the clinical picture.

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