

Extensive Bilateral Lower Extremity Deep Vein Thrombosis in a Patient with Congenital Absence of the Inferior Vena Cava

Introduction

Variations and anomalies of the inferior vena cava (IVC), while rare, are well documented in literature. Occlusive bilateral lower extremity deep vein thrombosis is a serious medical condition which if not treated appropriately could lead to limb loss or severe complications. The combination of these findings can pose a challenging clinical situation.

In most reported cases of an absence or interruption of the IVC the finding is secondary to either other pathologies or during the process of attempting to identify pathologic states. There are cases of idiopathic and recurrent DVT along with venous ulcerations in young individuals that have been early indications of an abnormal venous outflow state.^(2,3) Due to the complexity of the embryologic development of the vena cava there are a large number of documented anomalies.⁽⁴⁾

Case Report

An otherwise healthy 23 year old white male was admitted with back pain progressing to 10/10 over seven days. Primary care physician was treating patient for sacroiliitis with prednisone; however, pain was worsening and an unusual venous pattern was evident.^(Fig. 1) The patient was sent for an MRI of the abdomen and pelvis. The results were concerning for “a compressed IVC by extensive presumed adeopathy” with “very prominent epidural venous engorgement throughout the visualized low thoracic and lumbar region with may serve as collateral drainage pattern”.

Physical exam was grossly negative other than fullness in the pelvis upon deep palpation and engorged circumflex and inferior epigastric veins^(Fig. 1) wrapping around to buttocks with tight thighs and supple calves. The patient and his family were informed of the need to be worked up for malignancy. A PET scan and scrotal ultrasound were ordered upon admission.



Fig. 1 Right pelvis and groin.

PET Findings

1. Mild to moderate FDG uptake associated with the lobulated left paraspinal soft tissue density masses at L3 and L4, which appear to extend into and widen the adjacent left neural exit foramina. While nonspecific, the findings are suspicious for an underlying neurogenic tumor, including schwannoma or paraganglioma. Low-grade lymphomatous involvement could have a similar appearance. A repeat MRI of the lumbar spine with contrast is recommended for further evaluation.

2. Expansile hyperdense material within the lower IVC, extending along the bilateral common iliac, internal iliac, external iliac, and visualized femoral veins, with adjacent fat stranding/edema, suspicious for acute expansile thrombus or thrombophlebitis. There appear to be extensive venous collaterals throughout the pelvis, retroperitoneum, and paraspinal regions. A dedicated contrast-enhanced abdominal/pelvic CT is recommended to further define the full extent of thrombus and evaluate for its potential etiology. A dedicated lower extremity venous Doppler examination could also be performed for further evaluation, as clinically warranted.

MRI Findings

Original abnormality on MRI does not represent adenopathy but represents thrombosed enlarged common iliac veins ^(Fig. 2) bilaterally as well as thrombosed and enlarged lumbar vein on the left. The degree of collateralization and the remodeling of bone suggest this is a long-standing process. Lumbar collaterals on the right are not thrombosed. Multiple nonthrombosed collaterals are noted in the retroperitoneum. CT will be helpful to further evaluate the IVC. A mass at the L3-L4 neural foramen is possible but this is thought most likely to represent an enlarged thrombosed venous branch. The possibility of a stenotic or atretic segment of IVC is raised with capacious common iliac veins and extensive collaterals. The acute symptomatology may be the result of thrombosis with propagation and hemostasis.

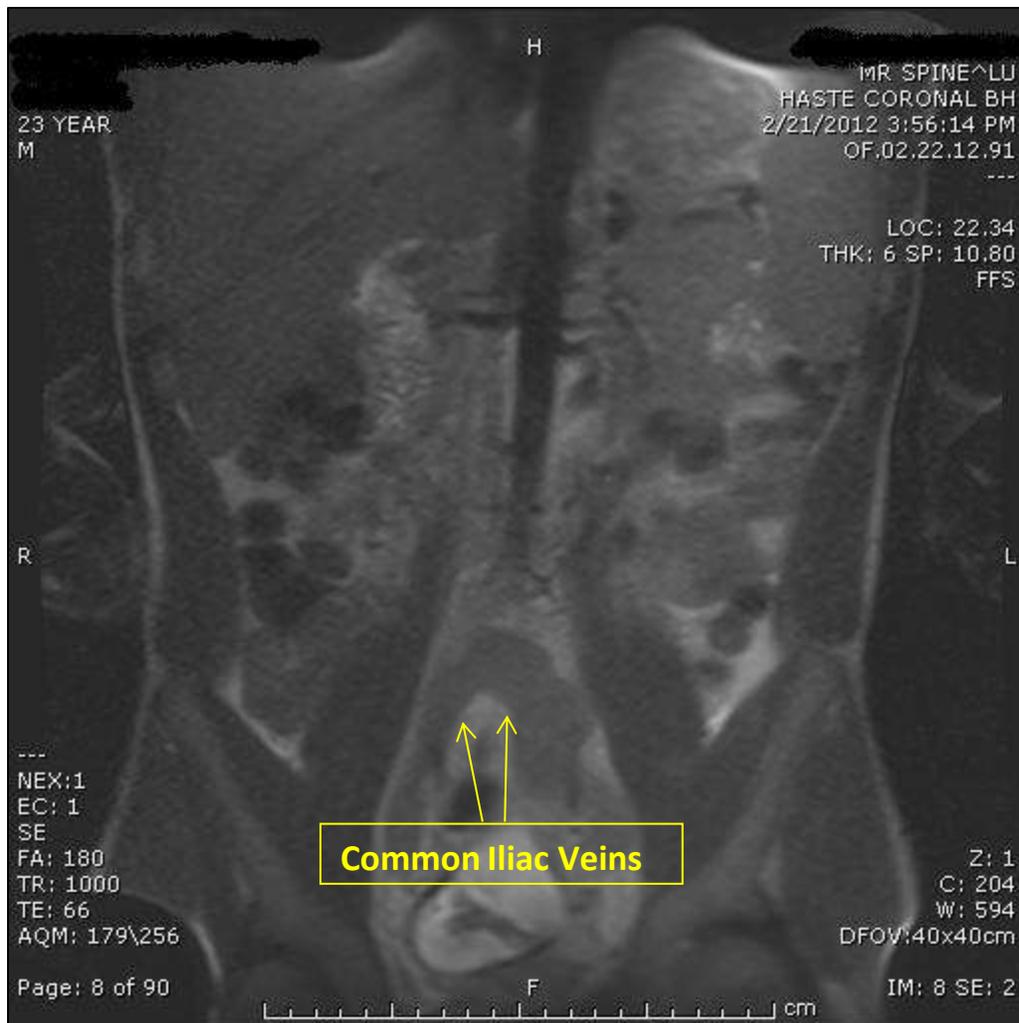


Fig. 2 MRI

Scrotal Ultrasound Findings

Negative for scrotal tumor.

CT Abdomen/Pelvis Findings

Extensive acute occlusive thrombus involving the infrarenal IVC, with expansile occlusive thrombus in the common, external and internal iliac veins and common femoral veins. Extensive venous collateralization via lumbar and renal veins with reconstitution of the intrahepatic IVC via collaterals. Negative for retroperitoneal mass.

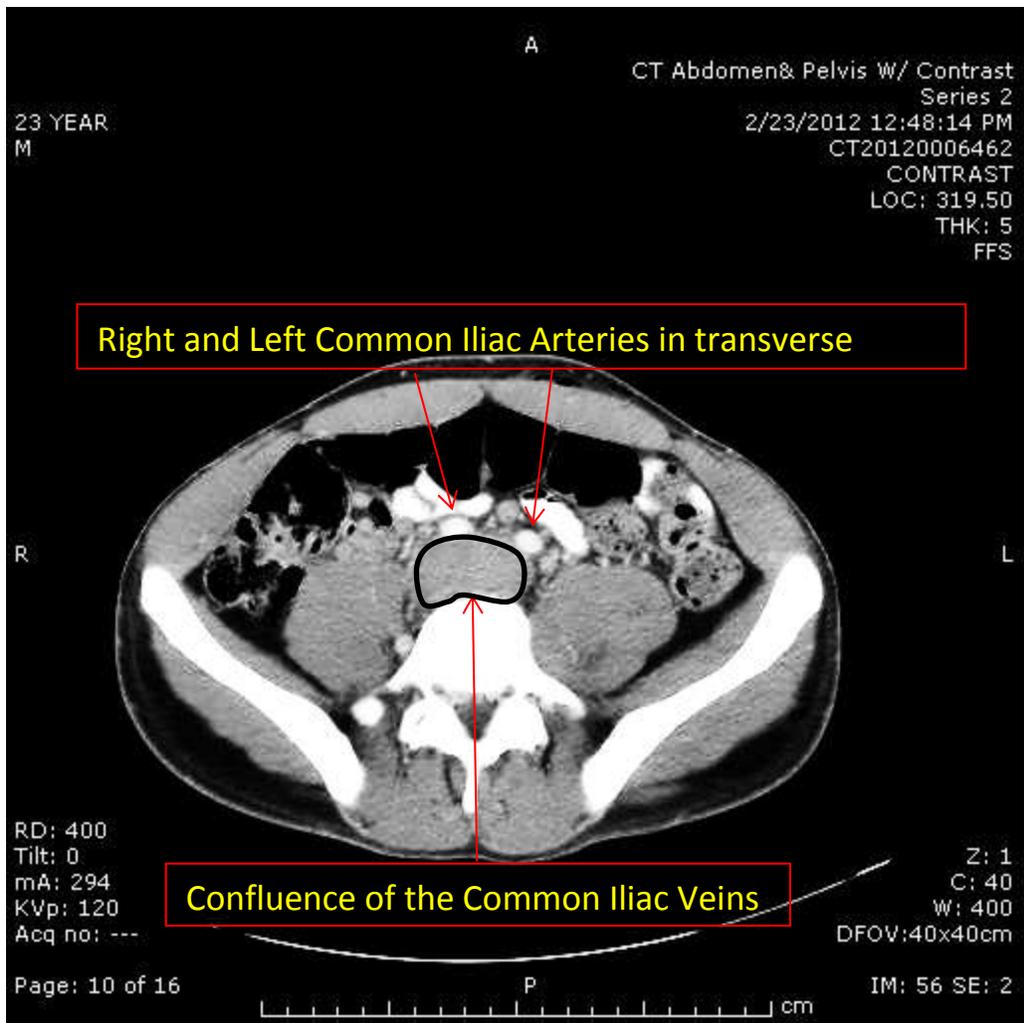


Fig. 3 CT

Lower Extremity Duplex Findings

Extensive, acute, occlusive DVT in the bilateral common and external iliac veins, common femoral and femoral veins. The distal and mid IVC also demonstrated DVT, being quite impressive in the distal IVC. The bilateral popliteal veins are currently patent. The posterior tibial and peroneal veins of each calf demonstrated response to augmentation; however, isolated calf vein thrombosis cannot be completely ruled out.

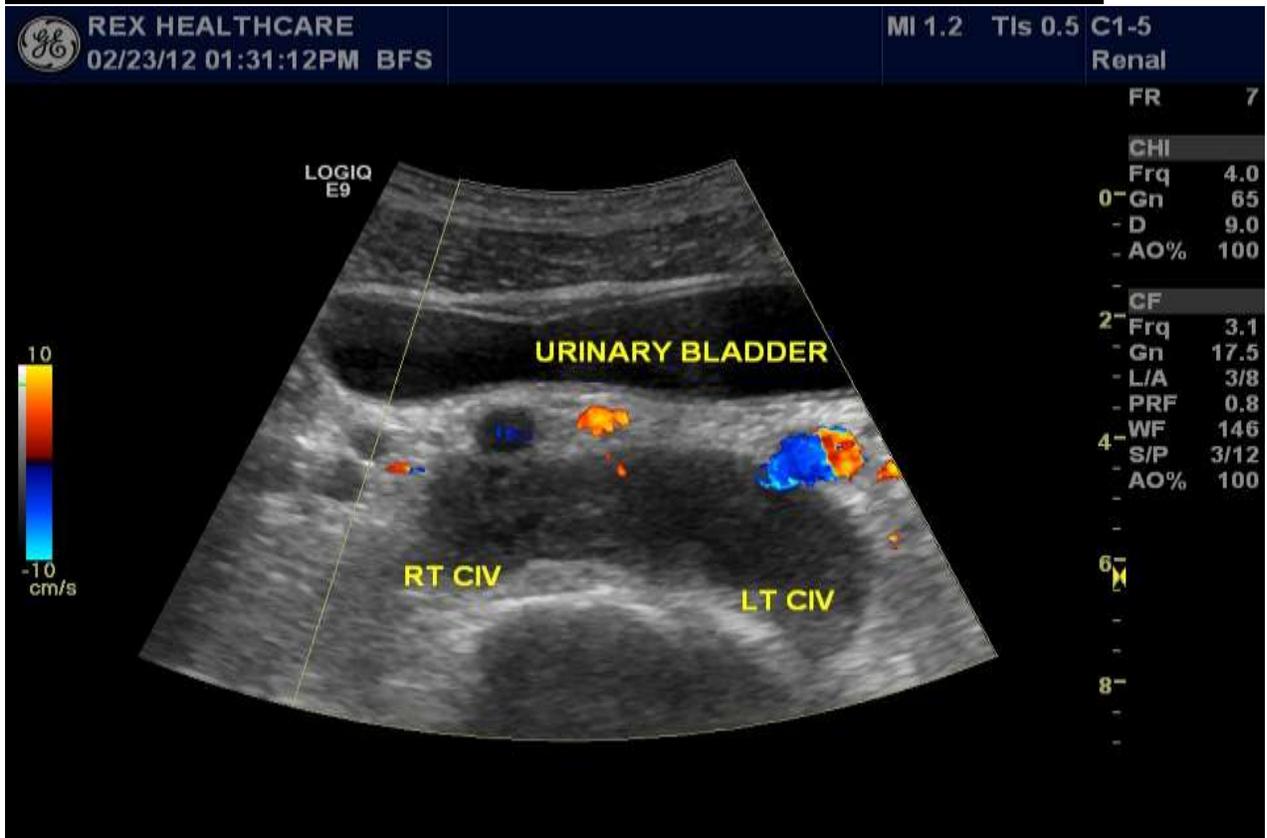
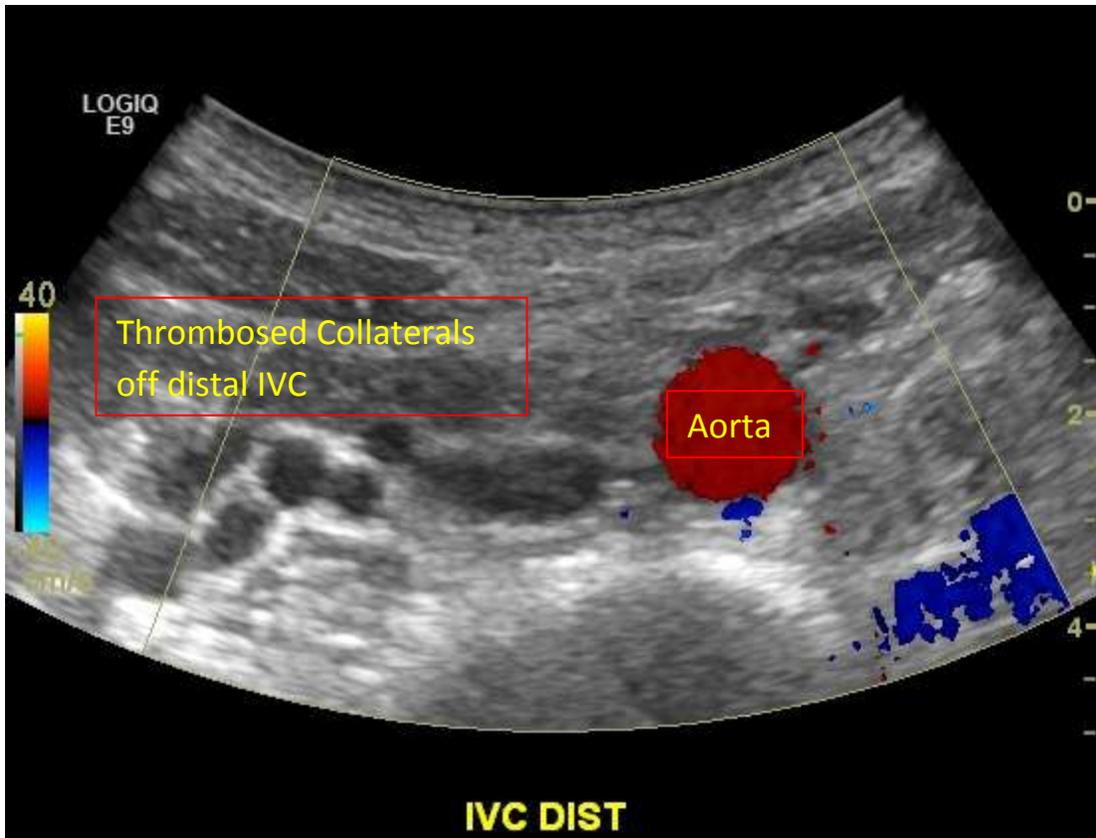


Fig. 4 Area of the distal IVC in transverse showing thrombosed collateral vessels
Confluence of common iliac veins with extensive thrombosis

Fig. 5

Medical and Surgical Management

Medical management of this particular patient's condition would not have been adequate alone. Traditional low molecular weight heparin and warfarin therapy would not have provided for clearance of the extensive nature of the thrombosis. Therefore, medical and surgical/mechanical treatment were applied. The patient underwent tissue plasminogen activator lysis of the bilateral lower extremities via access by the gastrocnemius veins through the popliteal veins and into the distal IVC. The lysis was administered for 48 hours followed by AngioJet mechanical thrombectomy due to the extensive residual thrombus burden in the bilateral lower extremities. After thrombectomy there was an approximate 90% reduction in thrombus load in both lower extremities. As well, angioplasty was performed on the left common femoral, femoral and iliac veins to improve residual stenosis.

Conclusion

Given the extensive nature of the thrombotic event of this patient and unclear cause, several modalities were used in tandem to correctly identify this rare occurrence of extensive occlusive bilateral lower extremity deep vein thrombosis in a patient with a congenitally interrupted inferior vena cava. In retrospect it was deemed most probable that this event was caused by an abrupt change in the patient's diet to include more green leafy vegetables and an increase in high intensity physical exercise. The increased vitamin K levels caused by diet in tandem with an increase outflow load from the patient's lower extremities created a situation in which the collateral pathways were not adequate and thrombosis resulted.

The patient returned for a follow up bilateral venous duplex ultrasound seven months after this event. Unfortunately there was still extensive chronic DVT throughout the patient's lower extremity deep venous systems. However, the patient reported that he is not suffering any ill effects with no noticeable lower

extremity swelling or edema and remains very physically active. He is on a life-long blood thinning regimen to avoid future total occlusive events.

References

1. Inferior Vena Caval Thrombosis; Luis G Fernandez, MD, KHS, FACS, FASAS, FCCP, FCCM, FICS; et. al. <http://emedicine.medscape.com/article/1933035-overview#showall> July 2011
2. Radiological evidence of anatomical variation of the inferior vena cava: Report of two cases; M.Artico, et. Al. 5 November 2003 Springer-Verlag
3. Congenital absence of inferior vena cava and thrombosis: a case report; Javaid Iqbal and Eswarappa Nagaraju; Journal of Medical Case Reports 12 February 2008
4. Minniti S, Visentini S, Procacci C. Congenital anomalies of the venae cavae: embryological origin, imaging features and report of three new variants. Eur Radiol. 2002 Aug;12(8):2040-55. Epub 2002 Mar 19. Review. PubMed PMID: 12136323.