

A Case of a Missing IVC: Always Absent or Adverse Event?

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Introduction

When taking care of the adolescent or young adult patient, it is important to consider both pathologies of childhood and adulthood. In this case, a patient was found to have significant thrombosis secondary to extensive venous collateralization. The formation of these collateral vessels may represent complications of central venous catheter (CVC) placement in childhood or a congenital anomaly of the inferior vena cava (IVC). Congenital anomalies of the IVC are presumably the result of abnormal development of the IVC. Potential embryologic events leading to this anomaly have been proposed, but no single embryologic event which fully explains this finding has been identified.¹ Thrombotic events in early life could also potentially cause regression of normal anatomy. In the neonatal population, 90% of deep vein thromboses (DVT) are suspected to be related to central venous catheter (CVC) use and one prospective cohort study of neonatal intensive care unit (NICU) patients concluded that the incidence of CVC-related thrombosis was 5.33 per 1000 of central line-days with DVTs identified in 22 of 264 cases.²

While congenitally absent structures are often expected to show signs or symptoms during childhood, an adult male patient has been reported in the literature to have an absent IVC that was not diagnosed until age 54 with his first DVT occurrence.³ Congenital absence of the IVC is rare, presenting in only 0.0005% to 1% of the general population, but is estimated to be present in about 5% of cases of young patients with DVT.⁴ The mechanism by which this association occurs is not completely understood but is presumably related to abnormal venous flow causing inadequate blood return via the collateral vessels and leading to chronic venous stasis in the lower extremities.³ Another incompletely understood element of congenitally absent IVCs is, as the case at hand will demonstrate, the etiology of the absent IVC—namely whether it is a consequence of an anomaly of fetal development or a complication of CVC use in infancy.

Case Presentation

A 16-year-old male was admitted with 1 month of worsening hip and left lower extremity pain with new erythema and edema of the left lower extremity (LLE) developing on the day of presentation. He had originally been seen by orthopedics and diagnosed with hip strain, but his symptoms failed to improve with conventional therapy. Past medical history revealed concern for a possible aortic arch abnormality identified on chest x-ray for which the patient was to follow up with cardiology as an outpatient. His mother also reported that while in the NICU the patient had a left femoral CVC which became “kinked.” Further investigation of this event revealed a CT scan from 2005 showing a calcified thrombus in the IVC at the site of the patient’s PICC line. He had no personal history of immobilization, surgery, DVT, or pulmonary embolism (PE). The patient’s exam was significant for nonpitting edema of the LLE to the mid-thigh as well as pain with dorsiflexion of the left foot. He also was found to have varicose veins on the left anterior thigh. Ultrasound revealed an extensive and occlusive DVT involving the left femoral, profunda, and left common femoral veins extending down through the left popliteal and posterior tibial veins. Subsequent CT venograms of the pelvis revealed no visible IVC with extensive venous collateralization. Hypercoagulability workup was significant for positive lupus anticoagulant.

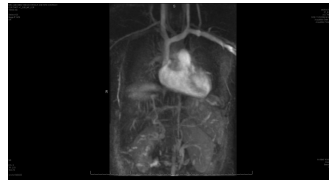


Figure 1. MRV showing severe atresia of the IVC

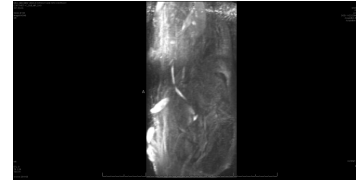


Figure 2. MRV demonstrating IVC remnant

After undergoing lytic catheterization and femoral venoplasty, he was discharged on therapeutic enoxaparin and aspirin (later transitioned to aspirin and rivaroxaban) with plans for lifelong anticoagulation. It remained unclear if his extensive collateralization was secondary to a congenitally absent IVC or complications of his prior central catheterization, though suspected to be related to neonatal CVC use given his neonatal imaging findings.

Conclusions

This is a case of a patient with a common presentation for DVT with an extremely rare finding of extensive venous abnormalities of unclear etiology. On one hand, an absent or interrupted IVC is often associated with other cardiac or visceral anomalies, which is suggestive of a possible congenital etiology. At the same time, a significant number of young patients with IVC thrombosis have had prior catheterization procedures, proposing the possibility of catheter-induced thrombosis as a cause of eventual agenesis of the IVC.⁴ A combined retrospective and prospective study of infants and children who experienced IVC thrombosis found that of 40 patients, 30 had had prior catheterization procedures performed and 17 had persistent iliac/caval venous obstruction during 18 year follow up.⁵ There are elements of this patient’s medical history to support either theory. While the presence of other possible cardiovascular anomalies in this patient adds support to the possibility that his absent IVC is congenital, the NICU history his family reported in conjunction with CT from the time showing a calcific thrombus of the IVC at the PICC line are more suggestive of the venous anomalies in this patient’s case being the result of a catheterization-related event. This case demonstrates the importance of considering congenital pathologies and birth history in the care of adolescent, young adult, and even older adult patients.

References

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