IVC atresia presenting as right-sided DVT

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Clinical Case

A 37-year-old female without pertinent medical history presented with four days of progressive, painful right lower extremity swelling that limited ambulation.

A right lower extremity ultrasound revealed extensive deep vein thrombosis (DVT) (Figure 1). Given her age and right-sided findings, there was high suspicion of an anatomical variant and a CT venogram was performed. The study confirmed the absence of an infrarenal IVC and showed extensive thrombi in the right external and internal iliacs, femoral and deep femoral veins.

Unsurprisingly, she had substantial collateral circulation with varicose veins, pelvic vessel congestion, and an enlarged azygous and lumbar venous system (Figure 2).

She underwent right iliofemoral DVT thrombectomy with restoration of vessel patency and persistent slow venous flow given her iliocaval atresia (Figure 3). She was started on enoxaparin but had re-thrombosis two days after her thrombectomy.

Approximately four months after her discharge from the initial hospitalization, the patient returned for elective reconstruction in which four stents were successfully placed (Figure 4). Her previously noted collaterals were not seen on venography post-reconstruction, supporting centralized venous flow.

Unfortunately, one month after post-reconstruction, she had asymptomatic thrombosis of the right iliac stent despite adherence to her outpatient enoxaparin regimen. She was given the option of watchful waiting or in-stent thrombectomy. She opted for thrombectomy at a later date and has yet to undergo the procedure. She continues to follow up with hematology for anticoagulation as an outpatient.

Unique Aspects and Recommendations

Our patient had a history of prematurity requiring a 6-month pediatric hospital stay with limited knowledge of the cause. We suspect that a perinatal/intrauterine injury may have contributed to her developing IVC atresia, but this information is limited.

Although asymptomatic for most of her life, her sedentary lifestyle and the addition of an oral contraceptive likely contributed to her initial presentation.

Anatomical variants should be considered with bilateral or right-sided DVTs in younger patients. This anomaly has a higher association with thrombophilia and a thorough hypercoagulable workup should be pursued. There is no consensus on therapy, but anticoagulation, commonly with enoxaparin, is the mainstay. Endovascular therapies, such as thrombolysis, thrombectomy, and stent placement, are preferable therapies based on patient characteristics.

Images

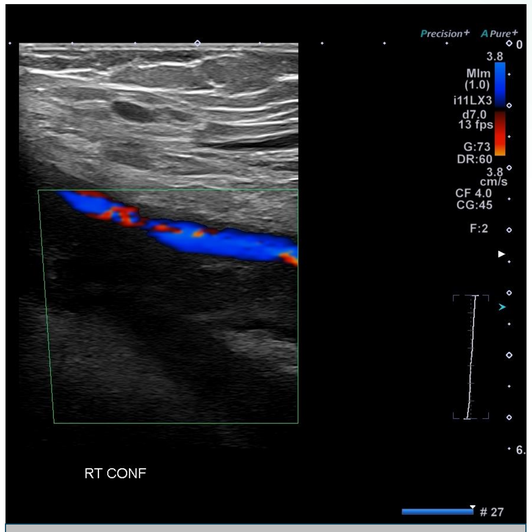

Figure 1. Doppler US demonstrating lack of flow in the femoral confluence.

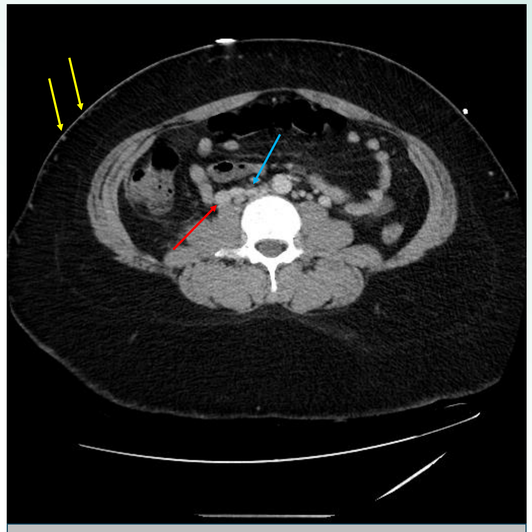

Figure 2. CT A/P axial view demonstrating atretic IVC (blue arrow), body wall collaterals (yellow arrows) and dilated gonadal vein (red arrow).

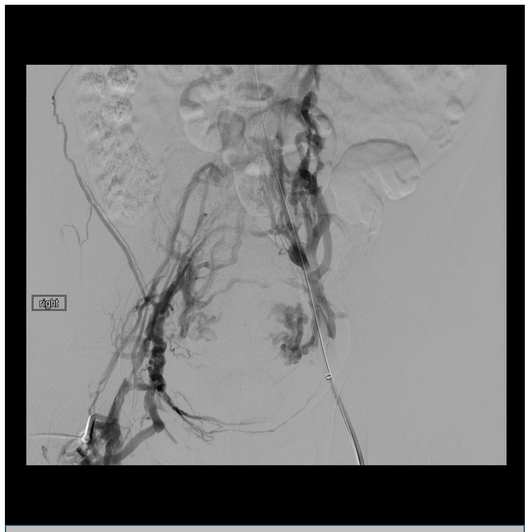

Figure 3. Subtraction venography demonstrating atretic IVC with paralumbar collateralization.

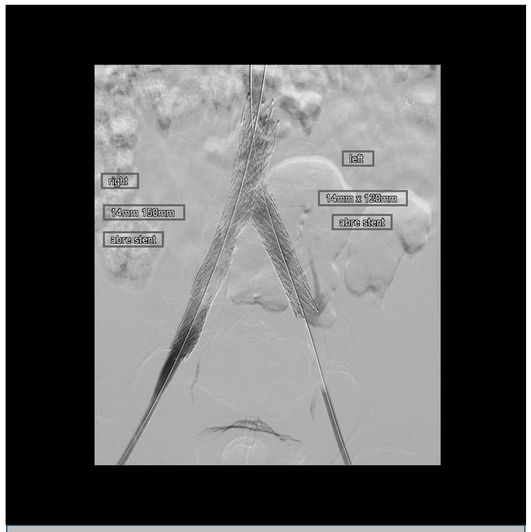

Figure 4. Post-reconstruction with balloon dilation and stent placement.

Literature Review

Congenital inferior vena cava atresia (IVCA) is caused by prenatal/intrauterine injury resulting in venous thrombosis or, more rarely, a disruption of embryogenesis. Patients compensate by developing a robust collateral system utilizing the azygous and hemiazygos circulation that can be seen on imaging. This altered anatomy predisposes patients to venous hypertension with findings of DVTs, venous ulcers, varicose veins, and pelvic congestion syndromes when the collateral circulation is unable to compensate adequately.

Our review of the literature revealed a number of case reports for patients who were ultimately found to have variations in their IVC anatomy resulting in atresia. All patients were treated with anticoagulation but not all patients underwent surgical. The most common presenting symptom or complaint was painful unilateral leg swelling.

References

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