Revascularization of acute inferior vena cava thrombosis and atresia in an adolescent

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ABSTRACT

Inferior vena cava (IVC) anomalies will remain silent until collateralized venous drainage has been lost. The initial signs can be subtle, including back pain, and are often missed initially until progressive changes toward motor weakness, phlegmasia cerulea dolens, and/or renal impairment have occurred. We have presented a case of acute occlusion of an atretic IVC and infrarenal collateral drainage in an adolescent patient, who had been treated with successful thrombolysis, thrombectomy, and endovascular revascularization for IVC stenting and reconstruction. (J Vasc Surg Cases Innov Tech 2022;8:331-4.)

Keywords: Deep venous thrombosis; Iliocaval stenting; Inferior vena cava atresia

CASE REPORT

A 16-year-old boy had presented to the emergency department complaining of 1 week of lower back pain and 2 days of lower extremity heaviness, swelling, and petechiae. He demonstrated short-distance claudication but was motor and sensory intact and had palpable femoral and pedal pulses. He had mild acute kidney injury, although the remainder of his laboratory test results were unremarkable, including a negative coronavirus disease 2019 test. Magnetic resonance imaging with time-of-flight angiography was obtained by the emergency department team, seeking to evaluate for any musculoskeletal etiology. The imaging study demonstrated complete interruption and thrombosis of an atretic infrarenal inferior vena cava (IVC) to the level of the right renal vein. The right renal vein and hepatic veins drained via a diminutive suprarenal cava, and the left renal vein drained directly via the paraspinous venous collateral vessels. Bilateral lower extremity venous drainage flowed exclusively into the collateral networks, many of which were also thrombosed, to reach the azygous vein and, ultimately, the superior vena cava. Computed tomography venography was considered but not performed secondary to the acute kidney injury.

The patient underwent heparinization and was admitted to the pediatric intensive care unit. His symptoms stabilized but did not improve during the initial 24 hours of admission using medical management with systemic heparinization and close observation. Therefore, we decided to proceed with bilateral lower extremity and IVC venography, pharmacomechanical thrombectomy, and evaluation for endovascular venous reconstruction. The bilateral saphenofemoral junctions were accessed under ultrasound guidance, and the initial venography confirmed substantial thrombus burden with no IVC filling and thrombosis of infrarenal collaterals (Fig 1). Additional supradiaphragmatic access was obtained via the right basilic vein, with venography confirming superior vena cava patency and a network of diminutive unusual IVC collateral vessels. Cannulation across the occluded atretic IVC was accomplished from the suprarenal cava to left femoral vein sheath, allowing for through-and-through access using a 7F trilobe EnSnare (Merit Medical, South Jordan, UT). A separate wire was placed up and over the iliac vein bifurcation. Over both wires, an 8F AngioJet (Boston Scientific, Marlborough, MA) was used with 30 mg of tissue-type plasminogen activator for power pulse with a 35-minute dwell time, followed by thrombectomy. Repeat venography demonstrated substantial clot disintegration and improved patency of both the atretic infrarenal IVC and the previously thrombosed infrarenal collateral vessels (Fig 2). Over the through-and-through caval wire, serial balloon dilation was completed to an 8-mm lumen, after which intravascular ultrasound was used to determine the stent diameter and length, using the perihepatic cava as a reference diameter. Three 18-mm × 10-cm Venovo venous stents (BD and Co, Franklin Lakes, NJ) were deployed from the suprarenal cava at the hepatic veins, overlapped through the left iliac vein, and ballooned to profile with a Kevlar balloon at nominal pressure. After left iliac and vena cava reconstruction, the outflow was significantly improved. However, the patient experienced substantial desaturation and a decrease in blood pressure that was concerning for pulmonary embolization. A catheter was advanced into the main pulmonary artery, and a pulmonary

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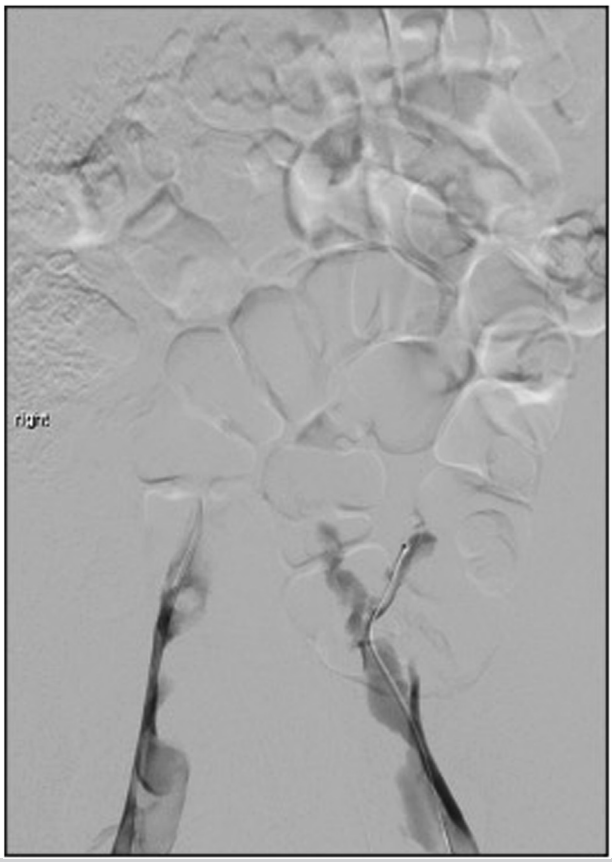

Fig 1. Initial venogram confirming iliocaval occlusion in the setting of inferior vena cava (IVC) atresia.

arteriogram was completed. Although no large pulmonary embolus was noted, there was concern for a smaller embolus, and an additional 10 mg of tissue-type plasminogen activator was administered directly into the pulmonary system. During the next several minutes, the patient's blood pressure normalized and his oxygen saturation returned to 100%. In retrospect, this was likely secondary to extreme washout from below the diaphragm. Residual thrombus in the right iliac vein was noted and removed using a Penumbra suction thrombectomy catheter (Penumbra Inc, Alameda, CA). To maximize right leg venous outflow, another 18-mm × 10-cm Venovo venous stent (BD and Co) was placed across the interstices of the stent at the peripheral IVC—left common iliac confluence and ballooned to profile (Fig 3). Completion venography and intravascular ultrasound demonstrated excellent stent expansion and in-line venous return, with attendant reductions in flow through the neighboring collaterals vessels.

Postoperatively, the patient's renal function recovered promptly, as did his index back pain and lower extremity edema. He was transitioned to apixaban and low-dose aspirin before discharge. His outpatient follow-up examinations have remained uneventful, and his recanalized IVC and bilateral iliac veins were patent 1 year postoperatively. The patient's guardian provided written informed consent for the report of the patient's case details and imaging studies.

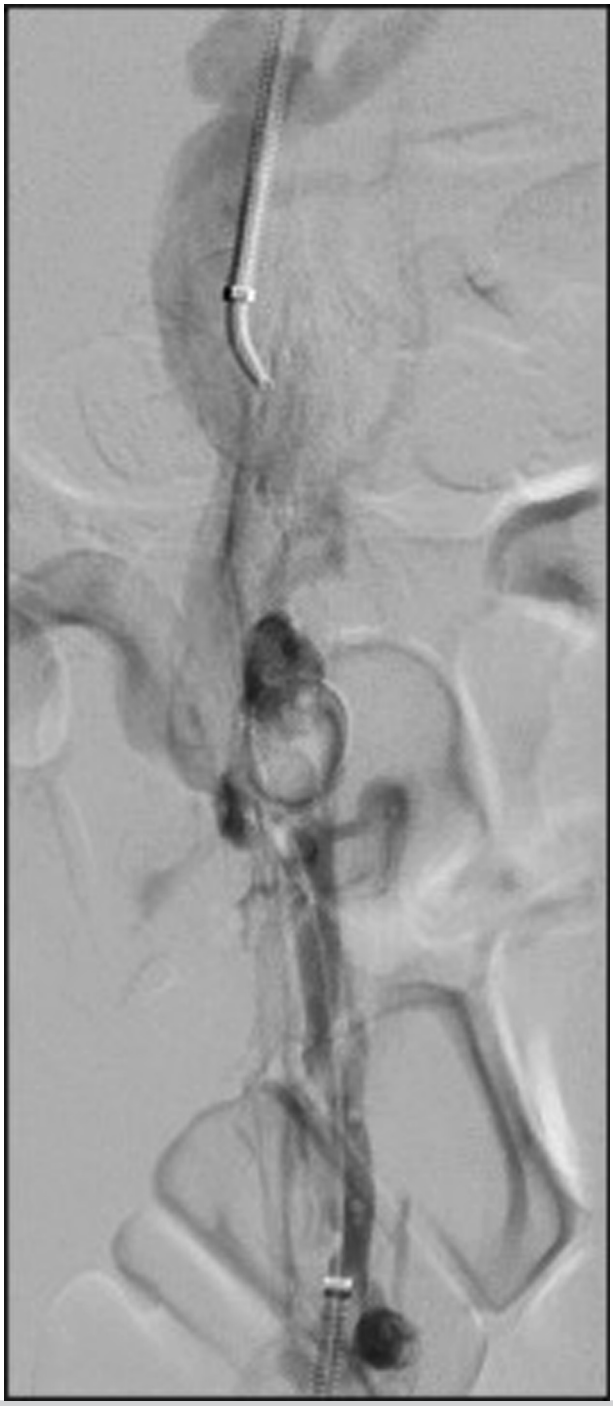

Fig 2. Venogram demonstrating extensive venous collateral vessels at the infrarenal and suprarenal inferior vena cava (IVC).

DISCUSSION

IVC anomalies occur with a prevalence of ~0.5% to 1.0% in the general population and ≤2% to 3% of those with congenital heart defects.[1,2] Congenital IVC anomalies can generally be classified into three anatomic categories[3,4]:

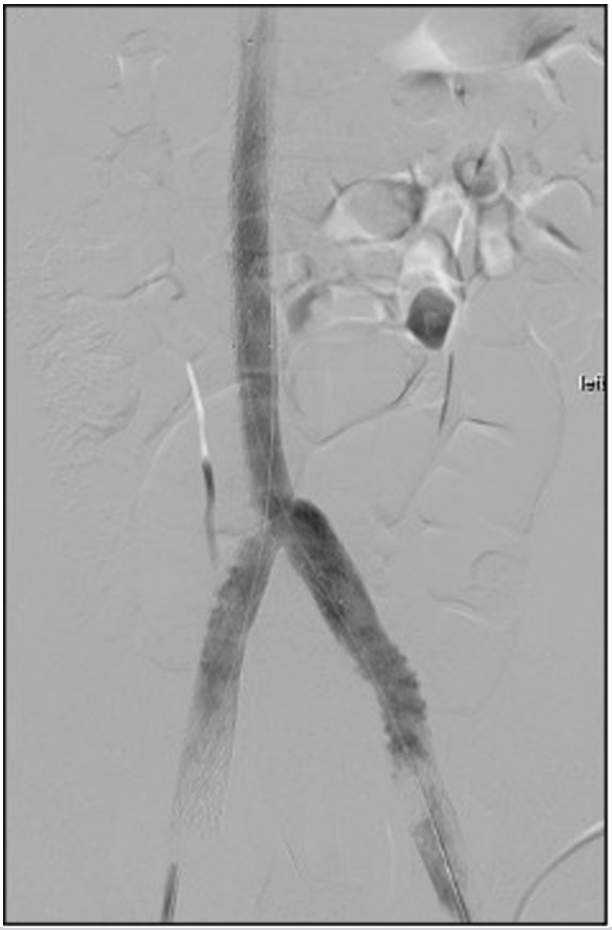

Fig 3. Completion venogram after inferior vena cava (IVC) recanalization, stenting, and bilateral iliac vein stenting with restoration of in-line venous outflow.

1. Infrarenal: duplicated IVC, preaortic IVC, left-sided IVC, or absence of the infrarenal IVC
2. Renal: retroaortic left renal vein, accessory left renal vein, or circumaortic left renal vein
3. Suprarenal: IVC membranes, congenital caval stenosis or atresia, or absence of the hepatic IVC with azygous continuation

Many IVC anomalies will remain clinically silent for years owing to well-developed collateral circulation or will be discovered incidentally on cross-sectional imaging studies. However, those with congenital IVC anomalies have a 60% to 80% lifetime risk of thrombotic events.[5,6] Thrombosis of the congenitally abnormal IVC, a prominent feeding vessel, or collateral channel can lead to acute or subacute proximal deep vein thrombosis, with associated acute and chronic sequelae, including phlegmasia cerulea dolens, pulmonary embolus, renal vein thrombosis, venous claudication, post-thrombotic syndrome, and ulceration.[7,8] The initial presentation will often be underwhelming with back pain and/or symptoms that mimic musculoskeletal complaints. It is our belief that the presentation will usually be preceded by a "second hit," such as a viral illness or dehydration, causing what were previously robust collateral vessels to thrombose. Motor dysfunction from venous hypertension is concerning, and higher venous pressures can lead to compartment syndrome.[9] Regardless of the etiology, the incidence of IVC atresia and adverse events has likely remained underestimated owing to the lack of standardized methods for detecting IVC occlusion.

Open treatment of proximal venous thrombosis is difficult. Esmarch thrombectomy, which works well in the extremities to remove venous clot, cannot be performed for proximal obstruction. Percutaneous thrombectomy offered the first paradigm shift in management, and cornerstones of treatment have subsequently focused on resuscitation, anticoagulation, thrombectomy, and/or thrombolysis.[10-15] Contemporary series of open and endovascular therapies have demonstrated comparable success, typically used in combination with iliocaval stenting.[11,16]

For future patients, it might be advisable to obtain a venous blood gas test of the venous blood below the occlusion and assess for potassium content and pH. For patients with a very low pH or high potassium levels, adjuncts might help prevent excessive washout. These adjuncts could include partial occlusion of venous return using a balloon vs division of venous blood through a continuous renal replacement therapy circuit before return. For cases with expected issues, the surgeon should consider this before full revascularization.

In the present patient, the motor symptoms and renal damage prompted aggressive attempts at revascularization to reestablish in-line flow and reopen renal drainage. Restoration of in-line venous return helped to further offload previous collateral venous hypertension. We previously reported the use of femoral–jugular continuous renal replacement therapy as a method of venovenous bypass for emergent offloading of elevated lower extremity venous pressure.[9]

The mid-term durability of IVC recanalization and stenting appears reasonable, with 4-year primary and secondary patency of 52% to 78% and >90%, respectively.[17,18] Recent venous stents seek to maximize the interstice size and flexibility, although previous stent designs still offer a wider variety, including sizes ≤24 mm. For the present case, the Venovo stent was selected, because it was available in our hospital in the sizes required.

CONCLUSIONS

We have reported successful venous thrombolysis, recanalization, and endovascular reconstruction of an atretic IVC with an unusual presentation of acute iliocaval occlusion.

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