From the Eastern Vascular Society

Extensive iliofemoral and femoropopliteal venous thrombosis in a young patient with iliocaval atresia

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ABSTRACT

Inferior vena cava (IVC) atresia is a rare congenital anomaly. Standardized treatment is not well defined due to its uncommon presentation, with this pathology associated with an increased risk of unprovoked lower extremity deep vein thrombosis (DVT). We present a case of a 32-year-old man who was admitted for bilateral lower extremity edema and pain and was found to have bilateral extensive iliofemoral and femoropopliteal DVT, absence of IVC filling, and extensive tortuous collateralization arising from the pelvic veins to the azygos vein. Bilateral mechanical thrombectomy and endovascular iliocaval reconstruction was performed. Three months later, the patient demonstrated widely patent iliocaval stents and the absence of DVT. Endovascular treatment of IVC atresia is feasible and optimizes the reduction of thrombus burden. (J Vasc Surg Cases Innov Tech 2024;10:101431.)

Keywords: Atresia; Congenital; Deep vein thrombosis; Iliocaval reconstruction; Inferior vena cava; Venous stenting

The occurrence of inferior vena cava (IVC) congenital anomalies is rare, ranging from 0.3% to 0.5% of cases in otherwise healthy young adults.[1] IVC atresia is an abnormality secondary to aberrant development of vein embryogenesis and thrombotic events in utero or early life, which is even more uncommon (<1% of the worldwide population).[2] This clinical condition is usually asymptomatic and diagnosed incidentally during workup imaging for other causes, with unprovoked lower extremity deep vein thrombosis (DVT) incidence rates of ≤5% for those aged <40 years, especially DVT due to venous stasis of the iliac vein and femoral veins.[3-5] The standardized treatment is not well defined due to its uncommon presentation. The present patient provided written informed consent for the report of his case details and imaging studies.

CASE REPORT

A 32-year-old man presented with a 5-day history of bilateral lower extremity (BLE) pain, numbness, and swelling. His medical history was significant for resolved COVID-19 (coronavirus disease 2019) infection 1 year before admission. The patient did not endorse limb weakness, trauma, strenuous exercise, back pain, DVT, or hypercoagulable disorders. Before admission at our institution, he was discharged from an outside hospital emergency department with presumed colitis. Antibiotic therapy with ciprofloxacin was prescribed and attributed as the cause of the BLE symptoms. Given the persistence of BLE edema and pain despite analgesic treatment, he was admitted to our institution. The pain was exacerbated with movement and weightbearing. Physical examination showed BLE pitting edema to the mid-thigh and pedal cyanosis, with preserved peripheral pulses and neurovascular function. The laboratory test results revealed a slight bilirubin elevation, an increased blood urea nitrogen/creatinine ratio, and elevated C-reactive protein. BLE duplex ultrasound (DUS) revealed the presence of noncompressible thrombus from the level of the common femoral veins to the tibial veins. Given concerns for iliofemoral DVT, emergent computed tomography venography (CTV) was performed, with findings of extensive bilateral femoropopliteal DVT, absence of IVC filling, and extensive tortuous collateralization arising from the pelvic veins to the azygos vein (Fig 1). Given the extent of thrombus burden and absence of IVC filling, bilateral 50-cm Cragg-McNamara iliac lysis catheters (Boston Scientific) were placed, and thrombolysis with infusion of heparin 500 IU every hour via each popliteal vein sheath and tissue plasminogen activator 0.5 mg every hour via each of his lysis catheters was performed. Despite 48 hours of thrombolysis, the findings from repeat CTV and his symptoms remained unchanged. Due to the persistence and extension of the thrombus burden and for prevention of long-term disability

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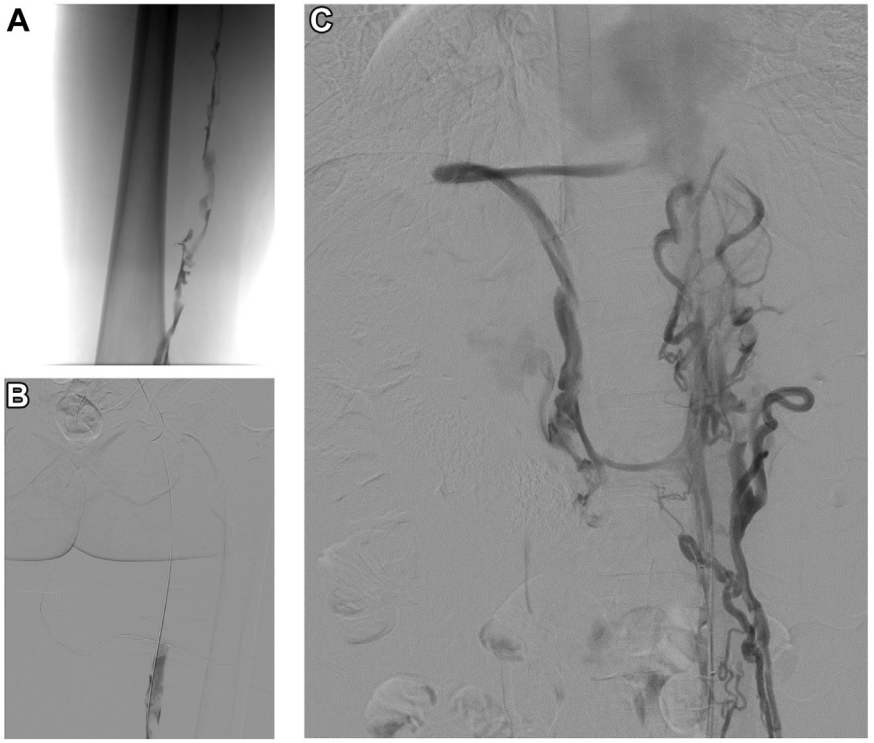

Fig 1. A-C, Computed tomography venograms demonstrating bilateral iliac vein thrombosis, absence of inferior vena cava (IVC) filling, and extensive tortuous collateral vessels with extension from the pelvic veins to the azygos vein.

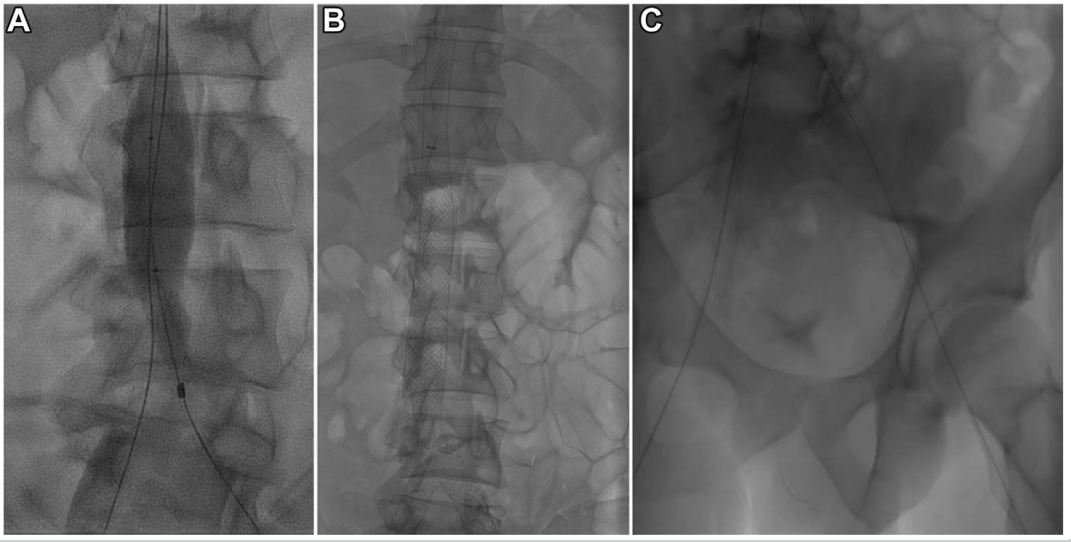

Fig 2. A-C, Operative venogram sequence showing angioplasty of the inferior vena cava (IVC) to the iliocaval confluence. Iliocaval reconstruction was performed with two wall stents, followed by deployment of bilateral common iliac vein stents in "top-hat" fashion and extension into the external iliac veins.

associated with iliofemoral DVT, we elected to proceed with mechanical thrombectomy of the BLE common iliac vein, external iliac veins, common femoral vein, femoral vein, and iliocaval reconstruction.

Under general anesthesia, the right internal jugular vein and bilateral previously accessed popliteal veins were used. From the right internal jugular vein access, a wire was advanced through the superior vena cava, and images revealed occlusion

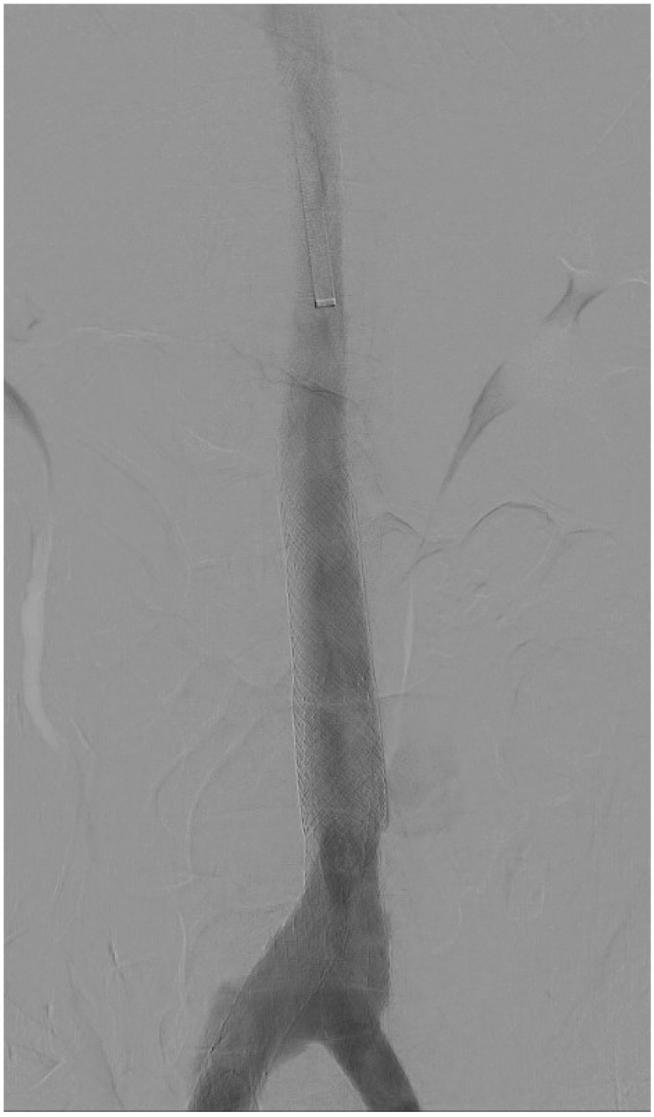

Fig 3. Completion venogram with presence of inferior vena cava (IVC) flow and adequate stent expansion.

of the suprahepatic, paravisceral, and infrarenal IVC, with extensive pelvic collateral circulation. Next, the wire was advanced to the iliocaval confluence, with subsequent insertion of a snare to catch the wires in the popliteal vein access and obtain right side body flossing. Next, the wires were snared from the left popliteal vein access, followed by BLE thrombectomy with an Inari ClotTriever Thrombectomy System (Inari Medical). After performing multiple passes, we interrogated the IVC and bilateral common femoral vein, femoral vein, external iliac vein, and common iliac vein with intravascular ultrasound (IVUS). After confirmation of removal of >90% of the clot, we proceeded with angioplasty of the IVC from the suprahepatic IVC to the iliocaval confluence with a 22-mm balloon, followed by BLE angioplasty of the common iliac vein, external iliac vein, and common femoral vein with 14-mm balloons. Next, we treated the IVC with two 24 × 70-mm wall stents (Boston Scientific) extended from 5 cm distal to the cavoatrial junction to the iliac vein bifurcation. Next, we deployed bilateral 14 × 140-mm Cook Zilver common iliac vein stents (Cook Medical) in a "top-hat" configuration with extension into the external iliac veins, with two additional 14 × 140-mm stents (Fig 2). With IVUS, we confirmed an adequate position of the IVC stents into the iliocaval confluence and visualization of the hepatic and renal IVC portions. A completion venogram was performed and demonstrated excellent stent expansion, patency, and IVC flow (Fig 3). The patient was transferred to the surgical intensive care unit in stable condition. He was discharged on postoperative day 2 with a prescription for lifelong apixaban and clopidogrel and a scheduled follow-up visit with DUS in 1 month. At the 1-month follow-up, the patient had relief of BLE pain and edema, with CTV demonstrating widely patent iliocaval stents (Fig 4). The 3-month venous DUS revealed patency of the IVC and bilateral common iliac vein and external iliac vein stents, with no evidence of DVT. The patient will continue to have 6-month DUS follow-up and 1-year monitoring with CTV.

DISCUSSION

Abernethy[6] described the first two cases of IVC anomalies in 1793, and Miller[7] described the first case of IVC atresia in 1925 after an autopsy of 2-month-old infant with underlying heart, gastrointestinal, and pancreatic duct malformations who died of bilateral bronchopneumonia. During weeks 6 to 8 of embryogenesis, three sets of paired veins are involved in the development of the IVC, corresponding to the postcardinal, subcardinal, and supracardinal veins. Anastomosis failure produces anomalies in the adult IVC via two mechanisms: (1) interruption resulting from agenesis or (2) a fusion defect between the suprarenal and hepatic segments of the IVC.[8] Other presumed etiologies for occlusion include intrauterine or perinatal thrombosis, coagulopathy, congestive heart failure, trauma, immobility, severe exertion, and iatrogenic causes (IVC filter implantation).[9,10] Clinical manifestations can range from an asymptomatic presentation due to the presence of well-developed collateral circulation since early gestation to the presence of abdominal collateral vessels, pelvic or lower extremity varicose veins, leg swelling, and pain with or without lower extremity DVT.[11] In the present patient, clinical suspicion for IVC atresia was higher given his unremarkable medical history for hypercoagulable disorders and a lack of a family history of thrombophilia, prolonged travel or immobility, strenuous exercise, or congenital malformations. The diagnosis was delayed due to workup for other conditions before his arrival to our institution and laboratory test results that were unrevealing for thrombophilia screening after hematology consultation. Extensive imaging workup allowed us to unveil significant iliofemoral thrombosis and pulmonary embolism and to consider de novo IVC atresia. To date, this is the third case of idiopathic BLE DVT with IVC atresia in a young adult treated with endovascular techniques.

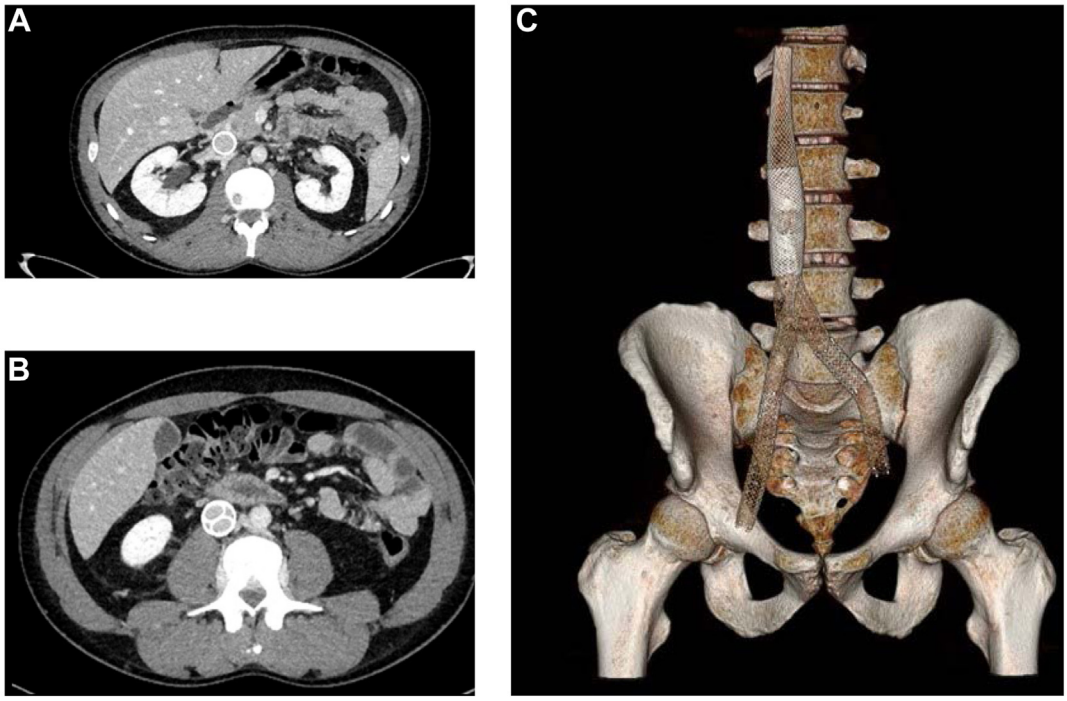

Fig 4. A,B Axial views of follow-up computed tomography venogram with wide patency of inferior vena cava and iliac vein stents. **C,** Three-dimensional postoperative reconstruction of iliocaval reconstruction.

The first step in the management of DVT secondary to IVC anomalies is anticoagulation. In the case of recurrent DVT, catheter-directed thrombolysis can be performed. If unsuccessful, iliocaval reconstruction is recommended. Wire- and catheter-based strategies, in conjunction with sharp recanalization techniques, are the main options for recanalization. Subsequent IVUS, balloon angioplasty, and stenting are mandatory.[12] In this case, we elected to proceed with endovascular treatment of the IVC atresia after failure of thrombolytic therapy and persistence of the extensive thrombus burden. Additionally, there was concern that the patient would develop post-thrombotic syndrome and the potential for DVT recurrence.

Multiple technical considerations are involved in performing iliocaval reconstruction. Blunt recanalization with a guidewire and catheter is the first step to cross through venous occlusions. Through-and-through access can be used subsequently, with adjunctive use of hydrophilic crossing catheters in cases of chronic obstructions. After confirming venous recanalization, sequential balloon angioplasty of the IVC, common iliac vein, and iliofemoral section is performed. Stenting of the caudal IVC with a buttressing stent, followed by iliocaval stenting in a "kissing" fashion, is preferred.[13] Stent options include nitinol self-expandable stents, which have been used to treat complete venous lesions with extension from the IVC to the external iliac vein. Bilateral extensions with nitinol stents are usually performed with parallel deployment in the vena cava stent with a 2-cm overlap. Alternatively, ballon-expandable stents can be deployed in the IVC stent to cover the common iliac veins and further extended in a lateral fashion with nitinol stents, which provide better bilateral symmetric inflow, stent integrity, and avoidance of extensive stent material in the iliocaval confluence.[14]

A series by Raju and Neglén[15] reported a technical success rate of 83% for iliocaval reconstruction for occluded iliac veins, with a 4-year primary and secondary patency rate of 35% and 72%, respectively. Various endovascular modalities have been used to treat the iliocaval confluence. One-stage double-barrel stenting is an excellent option for bilateral nonthrombotic disease, with less favorable outcomes in the setting of thrombotic disease. In scenarios with extensive disease involving the IVC (>5 cm) or the presence of fibrosis, the inverted Y technique with fenestration is an alternative.[16] Also, intraprocedural practice patterns can vary based on the etiology, provider, and preferred devices available for use.[17,18] As a result, technique selection should be determined according to the etiologic, clinical, and anatomic parameters to optimize the stent patency and reintervention rates.

Consensus for anticoagulation regimens remains to be determined given the uncommon nature of IVC atresia. Lifelong anticoagulation is considered on the basis of a caval anomaly as a nonmodifiable risk factor and the association of iliocaval thrombosis with acute and chronic

morbidity.[19] A systematic review of 1-year outcomes of deep venous stenting for iliofemoral venous outflow obstruction by Majeed et al[20] reported warfarin as the most common alternative, followed by direct oral anticoagulants and low-molecular-weight heparin in 33 of 50 studies. The treatment duration distribution was as follows: ≥3 months, 70%; 6 months, 48%; 12 months, 14%; and lifelong, 8%.[20] Attaran et al[21] described the patient outcomes with anticoagulant and antiplatelet therapy after iliocaval stenting. The overall stent restenosis or occlusion rate was 11%, with most occurring within the first 3 months. Patency was associated with larger nominal diameter stents ($P = .013$), regardless of the anticoagulation duration.[21] Thus, further studies are needed to determine the standardized anticoagulation therapy and duration to ensure optimal outcomes of adjunctive interventional approaches. Monitoring of iliocaval reconstruction for in-stent restenosis is performed at 6, 12, and 24 months after surgery with CTV.[12]

CONCLUSIONS

Iliocaval atresia is an uncommon anatomic variation of IVC congenital anomalies. The diagnosis can be challenging due to the asymptomatic presentation and absence of risk factors. Endovascular management, in conjunction with anticoagulation therapy, is feasible and optimizes reduction of the thrombus burden.

DISCLOSURES

None.

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