Congenital absence of the inferior vena cava with bilateral iliofemoral acute deep venous thrombosis

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Iliofemoral acute deep venous thrombosis (DVT) poses increased risk for post-thrombotic syndrome. Absent inferior vena cava (IVC) syndrome is a rare vascular anomaly that can be associated with idiopathic DVT in the young patient. It remains unclear whether endovenous thrombolytic intervention for DVT in patients with absent IVC can be successful, given the impaired venous outflow. This case report describes revascularization of bilateral iliofemoral and femoropopliteal DVT using endovascular pharmacomechanical thrombolysis and thrombectomy in a patient with underlying absent IVC syndrome to prevent post-thrombotic morbidity. (J Vasc Surg Cases 2016;2:193-6.)

Iliofemoral deep venous thrombosis (DVT) may be more prevalent than previously realized.[1] Acute iliofemoral DVT carries the highest risk of post-thrombotic morbidity. To prevent post-thrombotic syndrome, strategies for early removal of thrombi have been recommended and widely accepted regardless of the patient's age in the United States.[2,3] However, this treatment can be complicated by pre-existing venous disease, such as absent inferior vena cava (IVC) syndrome.

Congenital anomaly of the IVC and adjacent venous tributaries is an uncommon vascular malformation. This entity was first recognized by Abernethy, who described a congenital mesocaval shunt and azygos continuation of the IVC in a 10-month-old infant with dextrocardia in 1793.[4] The IVC and adjacent urogenital drainage system undergo complex embryogenesis between weeks 6 and 8 of embryonic life. Anastomosis and regression occur of three paired embryonic veins, including the posterior cardinal, subcardinal, and supracardinal veins. In a study of the development of the IVC, Huntington and McLure suggested 14 theoretical variations in its anatomy; 11 of the 14 variations have been observed in the domestic cat or in humans.[5] Whereas the duplicated IVC and retroaortic left renal vein has been widely recognized as a relatively common anomaly, reports of total absence of the IVC are scarce. Herein, we report a case of acute bilateral iliofemoral DVT in a young man with underlying absent IVC and subsequent endovenous treatment. Consent for this case report was obtained from the patient.

CASE REPORT

A 40-year-old man presented to the emergency department with acute-onset right lower extremity pain and swelling. The patient was a truck driver who recently drove a much longer route than usual, from Texas to Illinois, 10 days before presentation. During the trip, he experienced new-onset right lower extremity pain and swelling extending from the groin down to the posterior calf. He denied previous history of DVT or similar symptoms. He was an ex-smoker and denied illicit drug use or a family history of coagulopathy. Venous duplex ultrasound examination confirmed extensive acute DVT from the right common femoral vein to the peroneal and posterior tibial veins. He was admitted, and systemic administration of heparin was immediately started. Given the severity of his symptoms, he was referred for catheter-directed thrombolysis. An initial attempt at thrombolysis was made by another team on hospital day 2. Through right posterior tibial vein access, the wire and catheter were advanced to the right common iliac vein but could not cross the IVC or the venous collaterals. With use of an AngioJet Solent Omni catheter (Boston Scientific, Marlborough, Mass), 70 mg of tissue plasminogen activator (tPA) was power-pulse sprayed into the right iliofemoral and popliteal veins, followed by pharmacomechanical thrombectomy after 60 minutes of dwell time. Although the iliofemoral and popliteal vein DVT had improved, no central venous outflow was visualized. A 30-cm infusion catheter was placed across the iliofemoral segment, and tPA infusion was started at 0.5 mg/h. The next day, venography demonstrated recurrent and increased clot burden within the right common and external iliac veins with persistent lack of central venous outflow. Catheters were removed, and a second opinion was requested. A computed tomography (CT) scan of the abdomen and pelvis was then obtained, which revealed evidence of persistent bilateral iliofemoral DVT in addition to absence of the IVC (Fig 1).

He underwent two pharmacomechanical thrombolysis and thrombectomy treatments with vascular surgery. We hypothesized that lack of treatment focus on the outflow venous collaterals caused failure of the first intervention. The first treatment on hospital day 5 addressed the more severe right iliofemoral

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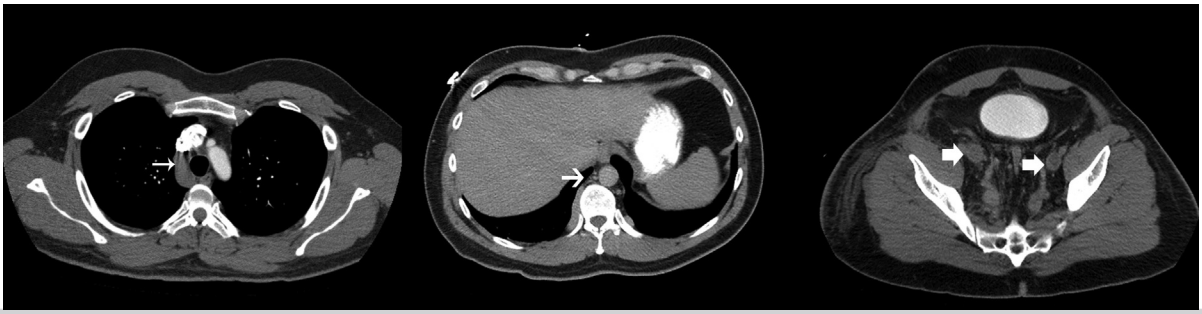

Fig 1. Left, Right lower extremity preintervention venogram. **Middle,** Right lower extremity postintervention. **Right,** The *arrows* indicate right common iliac vein with azygos darinage of venous flow.

and femoropopliteal acute DVT. After the right popliteal vein was accessed from a posterior approach with the patient in the prone position, stiff Glide wire and angled Glide catheter (Terumo Interventional Systems, Somerset, NJ) were advanced to the L4 level. The AngioJet Solent Omni thrombectomy catheter was introduced, and pharmacomechanical thrombolysis using power-pulse spray with 20 mg of tPA was performed. In particular, the majority of the tPA volume was administered in the IVC collaterals, including the azygos vein. After 30 minutes of dwell time, pharmacomechanical thrombectomy was performed using the same device for 2 minutes. Completion venography demonstrated successful revascularization of the entire femoropopliteal and iliofemoral vein segments with <10% residual stenosis (Fig 2). This also visualized an excellent network of retroperitoneal collaterals providing outflow to compensate for the absent IVC.

The second treatment was performed 3 days later to treat the left iliofemoral and femoropopliteal DVT. The patient complained of left leg swelling and pain as well. We used the same pharmacomechanical thrombolysis and thrombectomy technique. Completion venography demonstrated a near-occlusive chronic DVT in the left common femoral vein. Therefore, venoplasty was performed using a 10-mm × 4-cm and 12-mm × 4-cm balloon, each inflated to 14 atm for 3 minutes. Completion venography demonstrated <30% residual stenosis, not flow limiting, of the left common femoral vein (Fig 3). Plain old balloon angioplasty is the authors' primary modality for treatment of residual chronic DVT, with stents reserved for flow-limiting residual stenosis in suitable anatomic locations. This location at the common femoral vein would not be suitable for stenting and posed high risk for fracture.[2]

The patient was transitioned to warfarin with planned lifelong therapeutic anticoagulation. He was discharged home on hospital day 11 with 30 to 40 mm Hg knee-high compression stocking therapy. He returned to truck driving 2 months postoperatively, restricted to local driving only and the requirement of a break every 2 or 3 hours. Serial follow-up at 21 months revealed no significant lower extremity pain, edema, or stasis dermatitis. Surveillance venous duplex ultrasound did not demonstrate normal respirophasic flow as expected in this patient with absent IVC but did confirm long-term patency without restenosis of all lower extremity veins.

DISCUSSION

Absent IVC is an infrequent venous malformation often managed without surgical intervention. Prevalence of the disease is estimated to be between 0.3% and 0.5%.[6,7] Two theories explain this condition: failure of development of a connection between the right subcardinal vein and hepatic sinusoids; and IVC thrombosis in an early phase of embryogenesis with subsequent collateral formation. As a result, venous return from the lower body is rerouted through the azygos vein and retroperitoneal venules that join the superior vena cava in the right paratracheal space. In contrast, hepatic segmental venous flow directly drains into the right atrium.

The availability of CT, magnetic resonance imaging, and venography facilitates the diagnosis of IVC anomalies. CT venography and magnetic resonance venography are considered imaging modalities of choice; venography should be reserved for confirmation of the diagnosis and concomitant treatment using mechanical thrombectomy, thrombolysis, and angioplasty.

Absent IVC syndrome is recognized as a risk factor for lower extremity DVT. Absent IVC accounts for about 5% of idiopathic DVT in young, healthy patients without associated risk factors, such as thrombophilia, cancer, or use of oral contraceptive pills.[8,9] Few hypotheses regarding the pathophysiologic process of the formation of the DVT in this setting have been discussed in the literature; these include chronic venous hypertension and stasis and impaired venous return. This condition in several patients in previous case reports was complicated by thromboembolism[10-12] or concomitant cardiac or urinary malformation.[6,13]

Some reports suggest that indolent DVTs in the setting of IVC malformation can be successfully managed conservatively using systemic anticoagulation. Few reports exist documenting the efficacy of surgical or endovascular intervention for treatment of DVT in this

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