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Case Report

A case of inferior vena cava atresia complicated by bilateral deep vein thrombosis ☆,☆☆,★,★★

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ABSTRACT

In this report, we describe a case of deep vein thrombosis with suspected congenital inferior vena cava atresia treated with thrombolysis, angioplasty, and bilateral “kissing” iliac stent placement. An 18-year-old male presented with left common iliac vein thrombus and suspected congenital inferior vena cava atresia. He was treated over 4 days and discharged on anticoagulation which was continued long-term. These treatments were shown to be clinically successful in treating and preventing re-thrombosis in the context of inferior vena cava atresia initially presenting with symptomatic bilateral lower extremity deep vein thrombosis.

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Introduction

Deep vein thrombosis (DVT) is relatively uncommon and thought to occur in 0.1%–0.2% of the general U.S. popula-

tion [1]. Inferior vena cava (IVC) atresia is associated with increased occurrence of DVT [2]. Few trials have evaluated the treatment of extensive DVT associated with IVC atresia. We present a case of successful DVT thrombolysis and IVC recanalization requiring bilateral iliac vein stenting and long-term anti-coagulation in an 18-year-old male with IVC atre-

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sia complicated by extensive bilateral lower extremity DVT. Consent to publish this case report was obtained from the patient.

Case report

An 18-year-old male presented with a one-week history of bilateral leg cramping and lower abdominal pain. He had no significant past medical history and family history was negative for DVT or coagulopathies. Venous duplex ultrasound found thrombus in the left common iliac vein extending to the IVC bifurcation and suspected congenital IVC atresia with dilated retroperitoneal collateral veins. After a low molecular weight heparin (LMWH) bridge to warfarin, the patient was discharged home and achieved a therapeutic INR 3 days later.

The patient returned 2 weeks after discharge with acute worsening of his left leg pain. His INR was within therapeutic range. Repeated duplex ultrasound examination revealed extension of the thrombus which prompted consultation of Interventional Radiology for further evaluation and management.

Procedure details

Via the popliteal vein, venography was performed, which showed extensive thrombus and irregularity of the walls of the popliteal, femoral, and common femoral veins with markedly enlarged left pelvic and paralumbar collaterals (Fig. 1). Additionally, thrombus filling the left common iliac vein and involving the right external and common iliac veins without contrast was seen extending into a native IVC.

Mechanical thrombectomy was performed with the AngioJet Solent Omni catheter (Boston Scientific, Marlborough, MA) from the level of the left popliteal vein to the right common iliac vein and/or external iliac vein junction. Subsequent venography revealed improved flow with some deep increase in distal clot burden. Catheter-directed thrombolysis (CDT) was performed using two multi-sidehole infusion catheters with tissue Plasminogen Activator (tPA) at a rate of 0.5 mg/mL for a total dose of 1 mg per hour overnight.

The following day, repeat venogram showed improved flow in the popliteal, superficial femoral, and common femoral veins, although residual thrombus was noted. A trace amount of contrast was seen extending into a short caudal segment of the atretic native IVC. Additional mechanical thrombolysis, pharmacomechanical thrombolysis (PMT), and balloon angioplasty was performed. After which, flow of contrast improved through a majority of the affected vessels (Fig. 2). However, the infrarenal IVC was unable to be traversed by multiple guidewires. Digital subtraction angiography showed persistent irregular narrowing of the left and right common iliac veins, with collateral flow through pelvic and paralumbar lumbar collateral vessels. This constellation of findings suggested a more chronic component to the thrombosis remaining in the bilateral iliac veins, which would not respond to further tPA administration. In order to maintain antegrade flow in

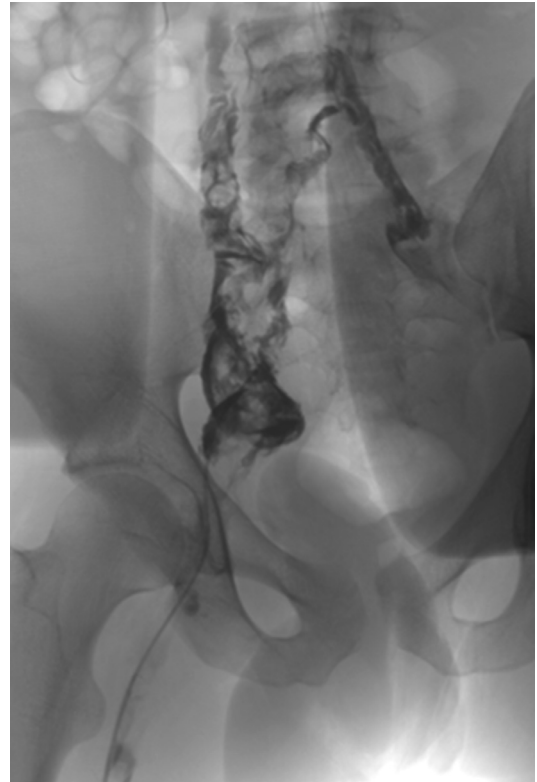


Fig. 1 – (Day 1) Initial venogram via left popliteal access showing flow into extensive collateral vessels that contain significant thrombus.

the iliac veins and decrease risk of recurrent DVT, stents would need to be placed. The length and expected diameter of the iliac veins and IVC were determined from the venogram images and appropriate sized wall-stents were delivered the next day.

On day 3, the atretic IVC was traversed with a microguidewire and serial balloon angioplasty dilation of the IVC was performed up to 18 mm (Fig. 3). Ultimately, “kissing” 14 mm self-expanding Wallstents (Boston Scientific, Marlborough, MA) were placed in the common iliac veins. A 12 mm Wallstent was also placed in the left external iliac vein. Due to the left femoral and popliteal vein re-thrombosis that occurred overnight, despite the patient receiving therapeutic anticoagulation, a multi-sidehole infusion catheter was again placed for overnight tPA infusion.

On day 4, the thrombus was nearly resolved with adequate antegrade flow through both iliac veins and the native IVC (Fig. 4A and B). The patient’s pain completely resolved, swelling improved, and he was discharged home the following day. Anticoagulation with LMWH and warfarin was again initiated.

The patient was continued on long-term anticoagulation. He has followed-up clinically for the past 3 years and remains asymptomatic with no symptoms of post-thrombotic syndrome (PTS). CT of the abdomen and pelvis, performed 5 and 9 months after the procedure, and ultrasound imaging, performed 11 months after the procedure, showed



Fig. 2 – (Day 2) Venogram showing flow into significantly atretic IVC.

no recurrent thrombosis with patent IVC and patent iliac stents.

Discussion

Anticoagulant therapy alone is the treatment given to most patients with uncomplicated acute DVT. However, extensive iliofemoral DVT warrants aggressive management with catheter directed thrombolytic therapy and/or mechanical thrombectomy as anticoagulant therapy alone rarely results in venous recanalization [3]. The residual thrombosis and stenosis in the iliac veins and IVC after CDT and PMT warranted multiple rounds of balloon angioplasty and stenting of the iliac veins to re-establish antegrade flow, decrease stasis, and prevent recurrent thrombosis [4]. Studies demonstrate the safety and efficacy of balloon angioplasty and

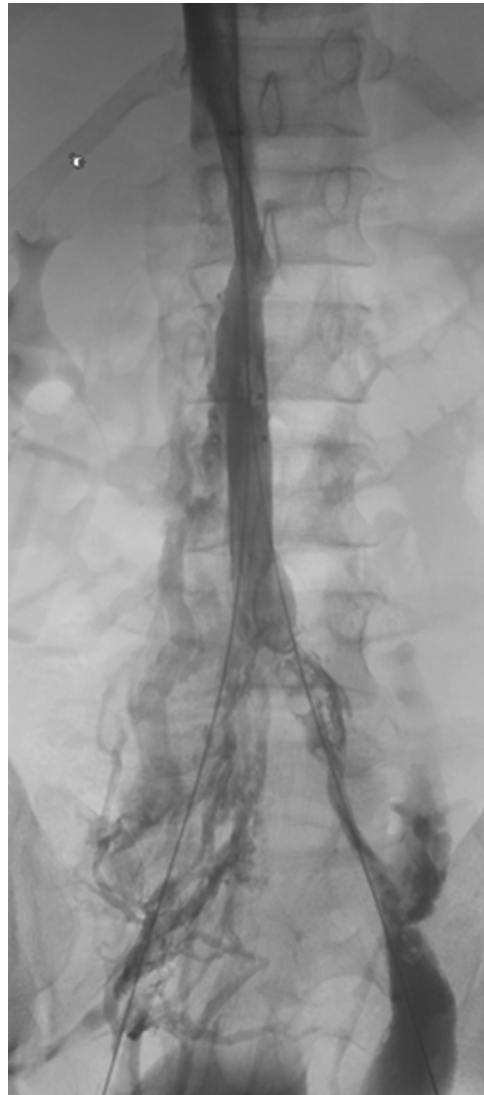


Fig. 3 – (Day 3) After successful subsequent IVC canalization and angioplasty, there is significant improvement in IVC caliber and flow, but iliac veins remain stenotic with significant collateral flow.

stenting of nonmalignant IVC and associated iliofemoral obstruction [5,6]. One systematic review showed no clear evidence for improved clinical outcomes for post stent anticoagulation after reviewing mostly small prospective and retrospective studies [7]. Other studies show that appropriate anticoagulation serves to reduce the risk of PTS [8]. The Cardiovascular and Interventional Radiological Society of Europe guidelines recommend post-procedural anticoagulation for ilio caval stenting [9]. A recent survey showed that experts prefer to use long-term anticoagulation after iliac vein stenting for post thrombotic iliac vein occlusion [10]. This case report suggests benefit from PMT with balloon angioplasty, “kissing” stent placement, and anticoagulation in an 18-year-old male with IVC atresia and extensive bilateral lower extremity DVT.

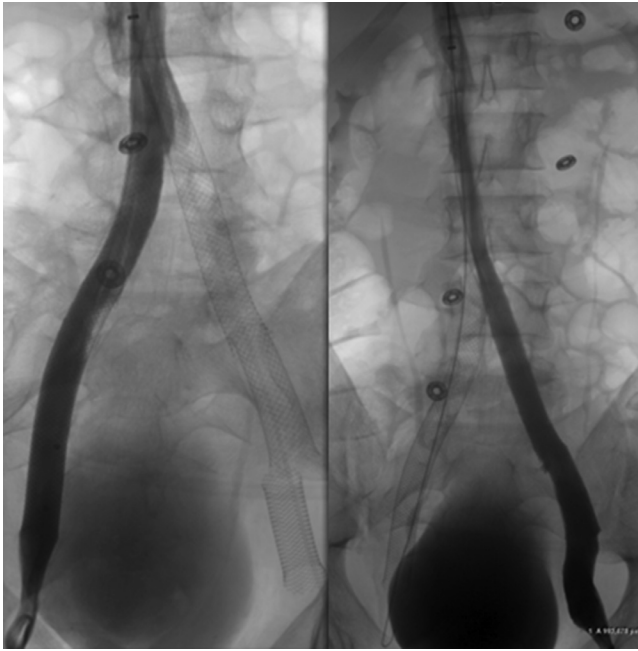


Fig. 4 – A, Figure 4B: (Day 3) Venography following stent deployment shows iliac vein and IVC patency and improved antegrade flow with marked decrease in collateral flow.

Conclusion

DVT is associated with IVC anomalies and evaluation for IVC anomalies should be explored in younger patients who experience DVT when they are without risk factors for DVT. Anticoagulation therapy alone may not be sufficient to prevent progression of thrombus in severe DVT. While CDT and PMT can be additional effective means for treatment of acute thrombus, angioplasty and stenting may be required when residual thrombus and stenosis persists. Post stent anticoagulation

can be used to prevent recurrent thrombosis, but additional studies are needed to provide evidence for best practice.

Patient Consent

Consent to publish this case report was obtained from the patient.

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