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

Alexander P. Nissen, MD, Ama J. Winland, MD, David W. Schechtman, MD, Joseph M. White, MD, Marlin W. Causey, MD, and Brandon W. Propper, MD

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 paraspinal 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.

From the Department of Surgerya and Division of Vascular Surgery,b San Antonio Military Medical Center, Fort Sam Houston; the Department of Surgery, Uniformed Services University of the Health Sciencesc; and the Department of Surgeryd and Division of Vascular Surgery,e Walter Reed National Military Medical Center, Bethesda.

Author conflict of interest: none.

Correspondence: Brandon W. Propper, MD, Division of Vascular Surgery, Walter Reed National Military Medical Center, Uniformed Services University of the Health Sciences, 4494 Palmer Rd, Bethesda, MD 20814 (e-mail: Brandon.w.propper.miil@mail.mil).

The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

© 2022 Published by Elsevier Inc. on behalf of Society for Vascular Surgery. This is an open access article under the CC BY-NC-ND license [http://creativecommons.org/licenses/by-nc-nd/4.0/].

https://doi.org/10.1016/j.jvscit.2022.04.006
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 x 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 balloon 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.

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 categories3–6:

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

Fig 2. Venogram demonstrating extensive venous collateral vessels at the infrarenal and suprarenal inferior vena cava (IVC).
1. Infra-renal: duplicated IVC, preaortic IVC, left-sided IVC, or absence of the infra-renal IVC
2. Renal: retro-aortic 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. 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. 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. 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. Contemporary series of open and endovascular therapies have demonstrated comparable success, typically used in combination with iliacal stenting. 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. 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. 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 iliacal occlusion.
REFERENCES

1. Chee YL, Culligan DJ, Watson HG. Inferior vena cava malformation as a risk factor for deep venous thrombosis in the young. Br J Haematol 2001;114:878-80.

2. Chuang VP, Mena CE, Hoskins PA. Congenital anomalies of the inferior vena cava: review of embryogenesis and presentation of a simplified classification. Br J Radiol 1974;47:206-13.

3. Spentzouris G, Zandian A, Cesnebasi A, Kinsella CR, Muhlenan M, Mirzayan N, et al. The clinical anatomy of the inferior vena cava: a review of common congenital anomalies and considerations for clinicians. Clin Anat 2014;27:1234-43.

4. Truty MJ, Bower TC. Congenital anomalies of the inferior vena cava and left renal vein: implications during open abdominal aortic aneurysm reconstruction. Ann Vasc Surg 2007;21:86-97.

5. Alkhouli M, Morad M, Narins CR, Raza F, Bashir R. Inferior vena cava thrombosis. JACC Cardiovasc Interv 2016;9:629-43.

6. Sitwala PS, Ladia VM, Brahmbhatt PB, Jain V, Bajaj K. Inferior vena cava anomaly: a risk for deep vein thrombosis. N Am J Med Sci 2014;6:601-3.

7. Kahn SR. The post-thrombotic syndrome: the forgotten morbidity of deep venous thrombosis. J Thromb Thrombolysis 2006;21:41-8.

8. Alkhouli M, Morad M, Narins CR, Raza F, Bashir R. Inferior vena cava thrombosis. JACC Cardiovasc Interv 2016;9:629-43.

9. Kahn SR. The post-thrombotic syndrome: the forgotten morbidity of deep venous thrombosis. J Thromb Thrombolysis 2006;21:41-8.

10. Elliot MS, Immelman EJ, Jeffery P, Benatar SR, Funston MR, Smith JA, et al. The role of thrombolytic therapy in the management of phlegmasia caerulea dolens. Br J Surg 1979;66:422-4.

11. Koopmann MC, McLafferty RB. Advances in operative thrombectomy for lower extremity venous thrombosis. Surg Clin North Am 2018;98:267-77.

12. Markel A, Manzo RA, Strandness DE Jr. The potential role of thrombolytic therapy in venous thrombosis. Arch Intern Med 1992;152:1265-7.

13. Nagarsheth KH, Sticco C, Aparajita R, Schor J, Singh K, Zia S, et al. Catheter-directed therapy is safe and effective for the management of acute inferior vena cava thrombosis. Ann Vasc Surg 2015;29:1375-9.

14. Patel NH, Plorde JJ, Meissner M. Catheter-directed thrombolysis in the treatment of phlegmasia caerulea dolens. Ann Vasc Surg 1998;12:471-5.

15. Tung CS, Soliman PT, Wallace MJ, Wolf JK, Bodurka DC. Successful catheter-directed venous thrombolysis in phlegmasia caerulea dolens. Gynecol Oncol 2007;107:140-2.

16. Kolbel T, Lindh M, Holst J, Uher P, Eriksson KF, Sonesson B, et al. Extensive acute deep vein thrombosis of the iliocaval segment: midterm results of thrombolysis and stent placement. J Vasc Interv Radiol 2007;18:243-50.

17. Erben Y, Bjamason H, Oladottir GL, McBane RD, Gloviczki P, Endovascular recanalization for nonmalignant obstruction of the inferior vena cava. J Vasc Surg Venous Lymphat Disord 2018;6:173-82.

18. Murphy EH, Johns B, Varney E, Raju S. Endovascular management of chronic total occlusions of the inferior vena cava and iliac veins. J Vasc Surg Venous Lymphat Disord 2017;5:47-59.

Submitted Dec 21, 2021; accepted Apr 4, 2022.