

Management of phlegmasia cerulea dolens caused by a giant leiomyoma

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ABSTRACT

In the present report, we describe the case of a young woman with a large uterine leiomyoma causing phlegmasia cerulea dolens with thrombosis of the left common and left external iliac veins. She underwent tissue plasminogen activator catheter thrombolysis and mechanical thrombectomy to temporize the condition until she could be evaluated by a gynecologic oncologist to remove the cause of the venous obstruction. Before the hysterectomy, a supracaval inferior vena cava filter was placed. However, <12 hours after the hysterectomy, she developed recurrent thrombosis involving the left common and external iliac veins. She underwent repeat mechanical thrombectomy with wall stent placement in the left common iliac vein with resolution of her symptoms. (*J Vasc Surg Cases Innov Tech* 2022;8:240-3.)

Keywords: Phlegmasia cerulea dolens; Thrombectomy; Uterine leiomyoma

Uterine leiomyomas are the most common pelvic tumor found in premenopausal women.¹ In the present report, we describe the case of a young woman who presented with phlegmasia cerulea dolens secondary to compression of the left common iliac vein (CIV) by a large uterine leiomyoma. She underwent mechanical thrombectomy to temporize her condition before total abdominal hysterectomy. The patient provided written informed consent for all procedures and the report of her case details and imaging studies.

CASE REPORT

A 46-year-old woman presented to the emergency department with a complaint of left lower extremity pain and swelling of 2 days' duration. Her medical history was significant for oral contraceptive use and a known 10-cm uterine leiomyoma. The physical examination revealed left lower extremity swelling extending from the foot to the mid-thigh, with erythematous discoloration of the skin (Fig 1). Biphasic Doppler signals in left posterior tibial and dorsalis pedis arteries were present, with triphasic signals on the right side. Motor and sensory function was intact in both lower extremities.

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Duplex venous ultrasound imaging showed extensive acute deep vein thrombosis (DVT) of the left common femoral, femoral, popliteal, gastrocnemius, posterior tibial, peroneal, and great saphenous veins. The left external iliac vein (EIV) close to the junction with the common femoral vein showed evidence of thrombosis. However, the left CIV could not be evaluated using duplex venous ultrasound imaging. Our computed tomography (CT) pulmonary embolism protocol showed right lower lobe segmental and subsegmental emboli. CT venography of the abdomen and pelvis showed a large heterogeneous mass measuring 18.6 × 15.5 × 9.9 cm (Fig 2). A mass effect was noted on the inferior vena cava (IVC) and left CIV, with evidence of thrombus in the left EIV, internal iliac vein, and CIV. Intravenous heparin was started.

A left lower extremity venogram with access through the left popliteal vein demonstrated thrombus within the left femoral and common femoral veins and chronic near complete occlusion of the left EIV and the entire portion of the left CIV. A 30-cm infusion length UniFuse catheter (AngioDynamics, Inc, Queensbury, NY) was placed in the treatment area from the proximal IVC to the distal femoral vein. Overnight, thrombolysis was initiated with tissue plasminogen activator. The infusion catheter was removed after 12 hours. A left lower extremity venogram demonstrated persistent thrombus within the left femoral vein, with persistent lack of flow within the distal left EIV and left CIV.

Angioplasty using a 4 × 40-mm Armada balloon (Abbott Cardiovascular, Abbott Park, IL) was performed within the IVC, left CIV, and EIV for predilatation before mechanical thrombectomy using a 105 cm × 16 mm ClotTriever (Inari Medical, Irvine, CA) to achieve access into the lumen. Mechanical thrombectomy of the proximal IVC, left CIV, EIV, common femoral vein, and femoral vein, with retrieval of a large amount of acute-, subacute-, and chronic-appearing clot (Fig 3). A completion venogram demonstrated complete removal of the thrombus within the left femoral vein, common femoral vein, and the visualized portions of the proximal EIV. Flow was present within the left



Fig 1. Photograph showing left lower extremity swelling and skin color changes significant for phlegmasia cerulea dolens.

CIV, with contrast seen entering the IVC. Chronic stenosis was present in the left CIV and EIV secondary to the mass effect from the uterus.

Three days after initial mechanical thrombectomy, a suprarenal Option ELITE IVC filter (Argon Medical Devices, Frisco, TX) was placed before surgery for excision of the pelvic mass. Systemic heparin was withheld 6 hours before surgery. She underwent total abdominal hysterectomy with bilateral salpingectomy. The pathologic examination revealed a benign leiomyoma measuring 18 × 18 cm. The patient was started with 5000 U of heparin every 8 hours immediately after surgery, with a therapeutic heparin infusion given at 24 hours after surgery.

At ~8 hours postoperatively, the patient started complaining of recurrent left calf tightness and swelling. Increased pitting edema was present with palpable pulses, the compartments were soft, and she had no pain with passive range of motion. Repeat duplex ultrasound showed acute DVT of the left common femoral, femoral, posterior tibial, and peroneal veins.

Venography showed diffuse thrombosis of the left femoral, common femoral, external, and CIVs, with high-grade stenosis involving the CIV. Mechanical thrombectomy was repeated using the ClotTriever (Inari Medical). The venogram did not show residual thrombus. Intravascular ultrasound demonstrated an 80% stenosis involving the mid to distal left CIV that had likely been missed on prior imaging owing to overlying thrombus. Balloon angioplasty with a 14-mm Atlas balloon (Bard Medical, New Providence, NJ) was performed within the CIV, followed by placement of a 16 × 120-mm Vici stent (Boston Scientific, Marlborough, MA). Completion venogram

demonstrated adequate inflow and outflow through the left CIV stent with no significant residual thrombus or stenosis (Fig 4).

Her postoperative course was complicated by adynamic ileus that resolved with conservative management. She required a blood transfusion owing to a steady decrease in her hematocrit, likely secondary to delayed bleeding from the hysterectomy that was further exacerbated by administration of the high-intensity heparin drip after mechanical thrombectomy and stent placement. CT arteriogram of the abdomen and pelvis showed a large pelvic hematoma with extravasation. However, angiography failed to show any active bleeding. Anticoagulation therapy was withheld until her hemoglobin had stabilized. She was discharged on hospital day 15 with oral apixaban 5 mg twice daily. Her left lower extremity edema had significantly improved (Fig 5). She returned 2 months later to undergo IVC filter removal. A follow-up office visit 3 months later showed a patent left CIV and EIV stent.

DISCUSSION

Phlegmasia cerulea dolens is a precursor of venous gangrene. It is a limb- and life-threatening condition with a mortality of 20% to 40%, with 30% of those deaths caused by pulmonary embolism.² DVT will be diagnosed in ~2 million Americans annually. However, phlegmasia cerulea dolens is rare and more prevalent in men, with a male/female ratio of 1.5:1.³ Patients will present with pain followed by significant swelling and cyanosis secondary to collateral vein involvement, causing a massive fluid shift into the interstitium, obstruction of the arterial flow, and, ultimately, capillary involvement, leading to venous gangrene.⁴

May-Thurner syndrome constitutes compression of the proximal segment of the left CIV by the right common iliac artery. The most common variant, found in ~20% of the population, is compression of the left iliac vein by the right iliac artery against the lumbar spine, causing collagen deposition in the vein, known as venous spurs.⁵ The consequential venous stasis increases the risk of DVT.

The risk factors for May-Thurner syndrome overlap with the risk factors for DVT. These include female gender, especially with oral contraceptive use and during the postpartum period, scoliosis, and thrombophilias. The present case did not involve this anatomic defect of compression of the iliac vein by the iliac artery. Instead, a large fibroid had caused extrinsic compression of the IVC extending into the left CIV. The resultant fibrotic changes in the vein ultimately led to this acute presentation of thrombosis superimposed on chronic stenosis. The external iliac pathology was likely due to chronic thrombosis and, therefore, was also stented.

CONCLUSIONS

Patients with DVT will generally be prescribed systemic anticoagulation therapy to prevent further thrombosis. In the present case, catheter-directed thrombolysis and

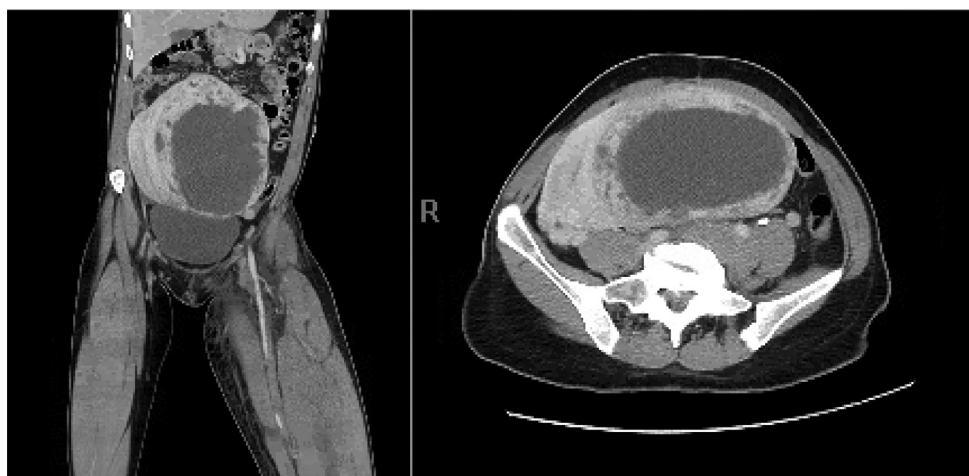


Fig 2. Coronal (**Left**) and axial (**Right**) computed tomography (CT) views of an 18.6 × 15.5 × 9.9-cm pelvic mass causing a mass effect on the pelvic structures.

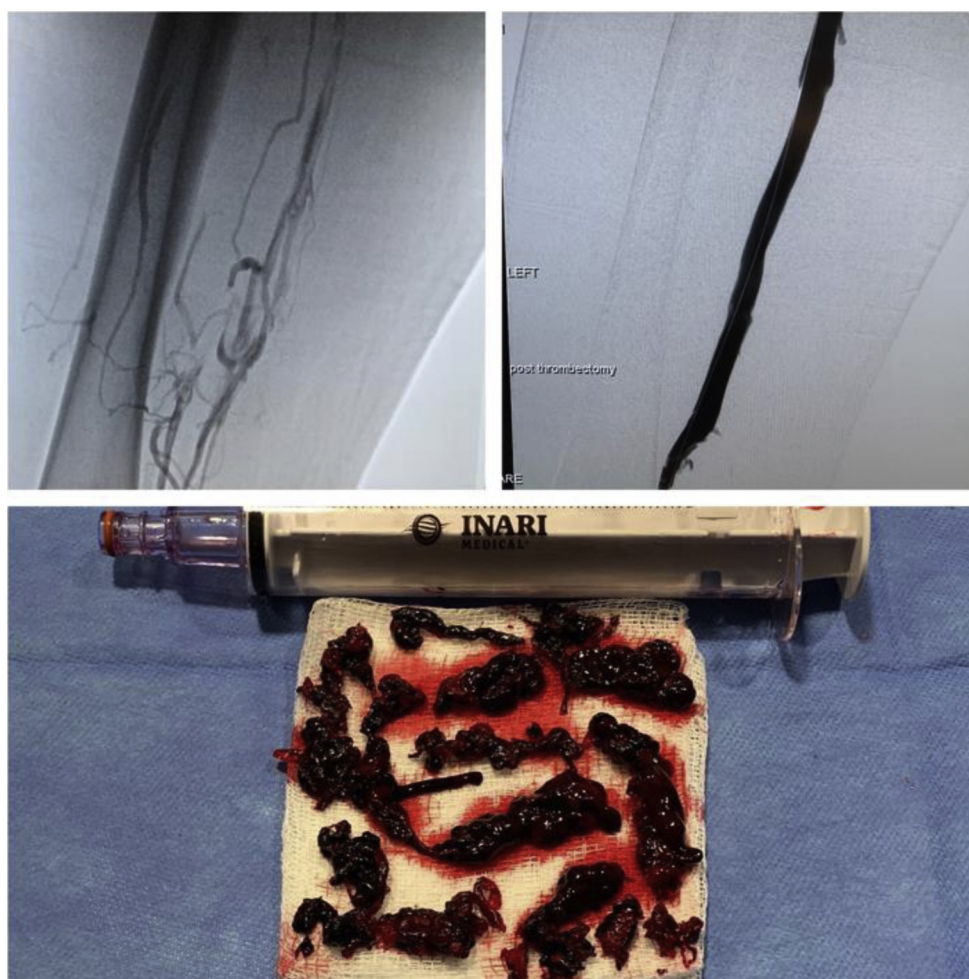


Fig 3. Top Left, Imaging study after tissue plasminogen activator administration and before mechanical thrombectomy. **Top Right**, Imaging study after mechanical thrombectomy. **Bottom**, thrombus extracted using the ClotTriever device (Inari Medical, Irvine, CA).



Fig 4. View of the left common iliac vein (CIV) stent.



Fig 5. Photograph showing the left lower extremity at hospital discharge.

mechanical thrombectomy were used until hysterectomy with bilateral salpingectomy had been performed, addressing the etiology of her extensive DVT. Several cases have been reported of a uterine mass causing occlusion to venous flow. However, it is difficult to effectively treat the phlegmasia and venous thrombosis without treating the underlying cause, such as a pelvic mass.

REFERENCES

1. Stewart EA, Laughlin-Tommaso SK. Uterine leiomyomas (fibroids): epidemiology, clinical features, diagnosis, and natural history. UpToDate 2021. Available at: www.uptodate.com/contents/uterine-leiomyomas-fibroids-epidemiology-clinical-features-diagnosis-and-natural-history?search=uterinefibroid&topicRef=5478&source=related_link#H4. Accessed November 19, 2021.

2. Chaar CIO, Satoskar S, Dardik A. Phlegmasia alba and cerulea dolens: background, pathophysiology, epidemiology. Medscape 2020. Available at: <https://emedicine.medscape.com/article/461809-overview#showall>. Accessed November 19, 2021.
3. Weaver FA, Meacham PW, Adkins RB, Dean RH. Phlegmasia cerulea dolens: therapeutic considerations. South Med J 1988;81:306-12.
4. Hari Krishna Reddy M, Sangeetha B, Aruna M, Sarat Chandra V, Venkata Kumar AC, Ram R, et al. Phlegmasia cerulea dolens: complication of femoral vein catheterization. CEN Case Rep 2016;5:184-7.
5. May R, Thurner J. The cause of the predominantly sinistral occurrence of thrombosis of the pelvic veins. Angiology 1957;8:419-27.

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