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Case Reports

Surgical Intervention for Phlegmasia Cerulea Dolens in a 61-Year-Old Cancer Patient

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Phlegmasia cerulea dolens, a rare and potentially fatal complication of acute deep vein thrombosis, is characterized by substantial edema, intense pain, and cyanosis. Phlegmasia cerulea dolens may compromise limb perfusion and lead to acute ischemia, gangrene, amputation, and death. We present the case of a 61-year-old woman with a history of breast cancer who had signs and symptoms of phlegmasia cerulea dolens in her left leg. She was treated promptly with open surgical thrombectomy and sequential distal compression with use of an Esmarch bandage to ensure complete thrombus extraction. These techniques restored venous flow and saved her leg. Open surgical thrombectomy should be considered in the presence of limb-threatening acute deep vein thrombosis presenting as phlegmasia cerulea dolens. ***(Tex Heart Inst J 2022;49(1):e207400)***

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Venous thromboembolism can cause death or substantial morbidity, and associated healthcare costs are high.[1] When diagnosed accurately and promptly, deep vein thrombosis (DVT) is a preventable cause of in-hospital death. A rare complication of DVT is phlegmasia cerulea dolens (PCD), which presents with limb swelling accompanied by acute ischemic pain and discoloration. Untreated, it can lead to arterial ischemia, gangrene, amputation, and death.[2] Treatment options range from percutaneous endovascular interventions to open surgical thrombectomy.[3] Although endovascular therapies are being used more frequently as first-line treatment for lower extremity DVT, their results in the presence of PCD are unclear,[4-7] and there is no consensus on whether endovascular or surgical treatment is more appropriate. The primary objective is to save limbs and lives, so new evidence supporting treatment results is needed.

We present the case of a cancer patient who presented at our institution with PCD, whom we treated surgically because of urgency and a lack of endovascular equipment at our institution.

Case Report

A 61-year-old woman who had undergone chemotherapy and radiation therapy for breast cancer presented at our emergency department with acute swelling of her left lower leg. Violaceous discoloration of the limb was caused by cyanosis. Physical examination revealed substantial cyanotic edema that extended from her foot to her left inguinal area (Fig. 1). She reported a current 6-hour history of intense, pulsating, oppressive left gluteal pain that radiated to the ipsilateral popliteal fossa, accompanied by left foot paresthesia. Her blood oxygen saturation was 94% on room air; blood pressure, 100/70 mmHg; and pulse rate, 70 beats/min. Her left leg was noticeably cooler to the touch than her right leg. Vascular examination revealed palpable pulses and audible Doppler signals in the femoral and popliteal arteries; the dorsalis pedis and posterior tibial pulses were diminished with normal Doppler signals. Sensory and motor functions were preserved in the left leg. Laboratory results included the following: hemoglobin, 14.5 g/dL; leukocytes, 16.9 × 10^3/μL; platelets, 271 × 10^3/μL;

Fig. 1 *Photograph at presentation shows severe edema and cyanotic color changes in the patient's left lower leg.*

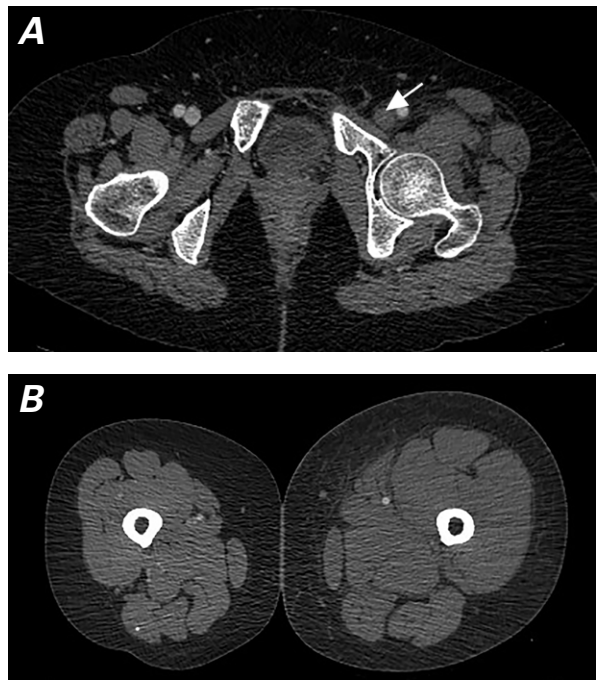

Fig. 2 *Computed tomographic angiograms show* ***A****) left femoral vein thrombosis (arrow) and* ***B****) substantial edema with inflammatory changes in soft tissues of the left lower leg.*

C-reactive protein, 11.78 mg/dL; and creatinine, 1.79 mg/dL. Our presumptive diagnosis was PCD. Computed tomographic (CT) angiograms showing a left femoroiliocaval thrombosis confirmed the diagnosis (Figs. 2 and 3). A continuous heparin drip was immediately started, and the patient was transferred for surgery.

Surgical Technique

The patient was placed under general anesthesia and in supine decubitus position. A 5-cm longitudinal incision was made in the left inguinal area. After careful dissection, the common femoral vein was identified. Vascular control was gained with use of silastic vessel loops on the common femoral vein, femoral vein, and deep femoral vein. A longitudinal venotomy was made with use of a #11 scalpel blade and Potts scissors. Proximally, fresh thrombus was removed with the sequential use of 3F, 4F, and 5F Fogarty arterial embolectomy catheters (Edwards Lifesciences Corporation). Distally, fresh thrombus was removed by compression; an Esmarch latex bandage was placed on the patient's lower leg, which was elevated at a 20°-to-30° angle. This maneuver was performed 3 times until venous backflow was observed. Figure 4 shows the fresh proximal and distal thrombus removed from the lower leg. Femoral vein distention resulting from the DVT enabled the primary closure of the venotomy without compromising the lumen, so after adequate femoral inflow and outflow were verified, a continuous primary venorraphy was performed with use of a double-armed 5-0 polypropylene suture. Hemostasis was achieved, and the skin incision was sutured. Femoral flow was verified by using continuous-wave Doppler ultrasound. The total operative time was 90 minutes.

Immediately after the operation, the temperature of the patient's leg was adequate, and the color had returned to normal (Fig. 5); the dorsalis pedis and posterior tibial pulses were also normal. At her one-month follow-up examination, her foot paresthesia, pain, and edema had resolved completely, and her muscular strength and extremity movement were normal. At 3 months, she remained asymptomatic.

Discussion

Untreated venous thromboembolism can lead to death or serious morbidity. In two-thirds of symptomatic patients, DVT will develop.[7] The condition is associated with several risk factors, including a history of venous thrombosis, older age, prolonged immobilization, cancer, surgery, trauma, pregnancy, the use of oral contraceptives or hormonal therapy, iliac vein compression, and venous insufficiency. Specifically, cancer has been associated with the development of acute massive venous

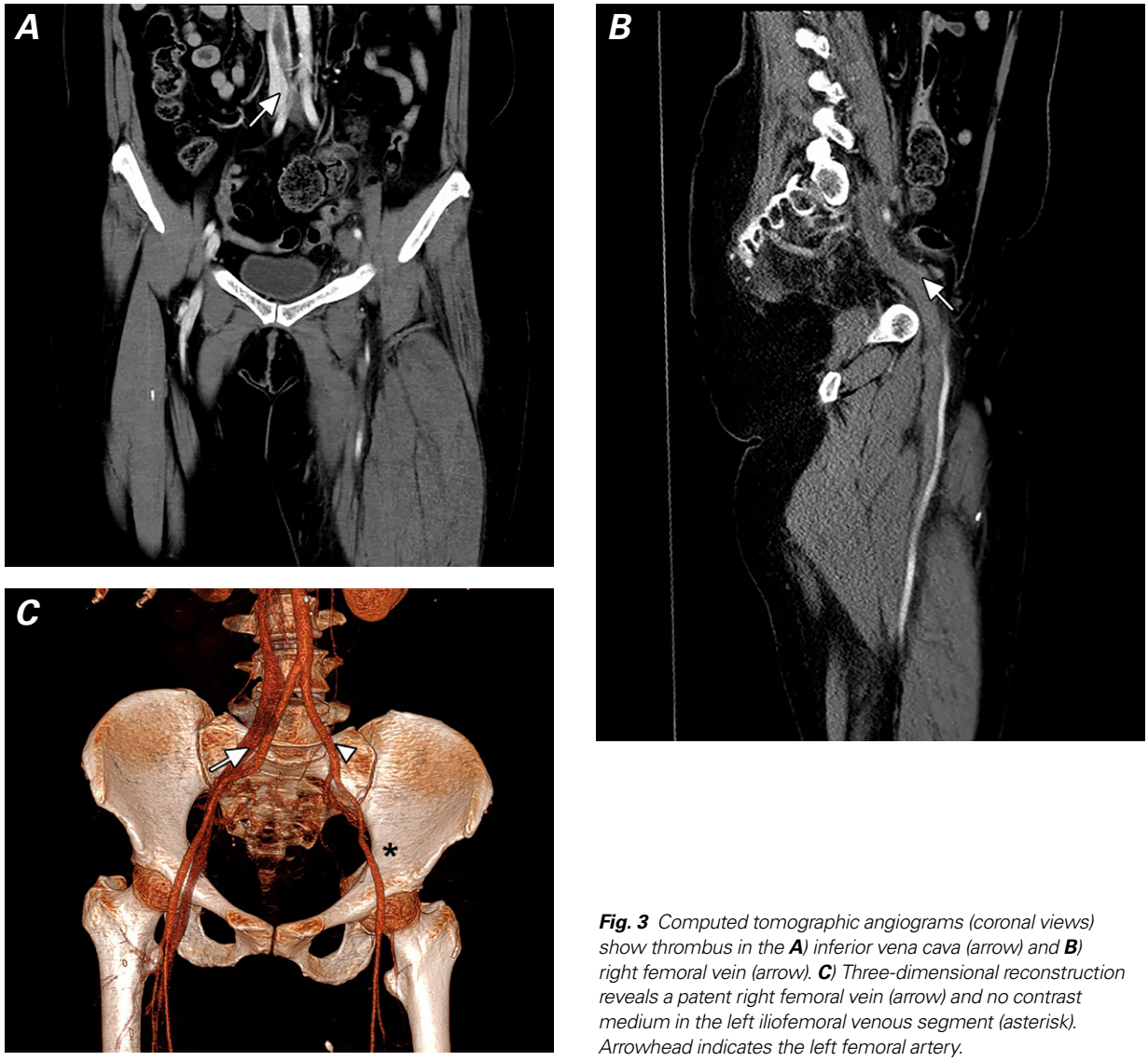

Fig. 3 *Computed tomographic angiograms (coronal views) show thrombus in the* ***A****) inferior vena cava (arrow) and* ***B****) right femoral vein (arrow).* ***C****) Three-dimensional reconstruction reveals a patent right femoral vein (arrow) and no contrast medium in the left iliofemoral venous segment (asterisk). Arrowhead indicates the left femoral artery.*

thrombosis.[8] Our 61-year-old patient's history of breast cancer put her at high risk for massive DVT.

Phlegmasia cerulea dolens, a potentially devastating complication of complex DVT, is characterized by extensive thrombosis of the major axial deep veins of the lower extremity.[3] Typical signs and symptoms are severe pain, edema, decreased extremity temperature, and cyanosis.[9] Our patient had all of these. Prompt diagnosis is crucial, because PCD can lead to impaired arterial circulation with consequent tissue ischemia, limb gangrene, or both. Amputation rates of 20% to 50% and mortality rates of 25% to 40% have been reported.[8] Although the diagnosis should be clinically integrated, it can be confirmed by using Doppler ultrasound or CT angiography. Imaging findings include noncompressibility or extensive thrombus in the deep venous system.

Therapy for PCD should be aggressive and focus on removing or reducing the thrombus load, improving venous patency, and limiting the progression of thrombosis in order to reduce venous hypertension.[8] Treatment options include systemic or local thrombolysis, pharmacomechanical thrombectomy (PMT), and surgical thrombectomy,[5,8] but no standard or verified therapeutic algorithm has been developed.

Vedantham and colleagues[4] revealed that the clot-removal rate with PMT was 82%, compared with 26% for surgical thrombectomy alone. Ockert and associates[10] reported a primary technical success rate of 100% with surgical thrombectomy and concluded that cardiovascular surgeons should include it in their repertoire. Pharmacomechanical thrombectomy is most beneficial in young, functionally active patients who have extensive and proximal thrombus and who present with acute DVT (<14 d) or with PCD.[11] Some disadvantages of PMT are major bleeding, microemboli, bleeding at intervention sites, and device cost.[5]

Fig. 4 *Photograph shows fresh thrombus surgically extracted from the patient's left lower leg.*

Fig. 5 *Photograph shows the immediate postoperative resolution of edema and discoloration.*

Several endovascular PMT devices are available in our country. These include the Indigo CAT mechanical thrombectomy catheter (Penumbra, Inc.), the AngioJet Peripheral Thrombectomy System (Boston Scientific Corporation), and the EKOS EkoSonic Endovascular System (Boston Scientific). Penumbra's Indigo Aspiration system involves constant vacuum aspiration with separator-assisted mechanical extraction of thrombus and débris. The AngioJet uses high-velocity water jets to dissolve thrombus in accordance with the Bernoulli principle. The EKOS system accelerates thrombus resolution in minimally invasive fashion by combining lytic therapy diffusion with a localized acoustic field. The AngioJet and EKOS can be used concomitantly with thrombolytic therapy.

Unfortunately, our institution does not stock these endovascular devices. Procuring them entails contacting distributors and arranging shipment, as well as collecting full device-related costs from the patient. Our patient had rapidly progressing DVT, so we decided on surgical thrombectomy as the fastest way to restore venous flow.[12] In a different clinical situation, however, endovascular therapy would have been a viable option.

Venous thrombectomy cannot clear distal thrombus, which is a major drawback to surgical treatment. We therefore complemented the patient's treatment by using lower extremity compression with an Esmarch bandage, which enabled us to retrieve much additional fresh thrombus. We performed a longitudinal venotomy because the femoral vein was distended and large, and this did not affect the venous lumen as a transverse venotomy would have. Fasciotomies to treat compartment syndrome secondary to PCD are rarely reported.[12] Our patient presented at the emergency department early in her disease course; in addition, her pedal and posterior pulses were diminished but palpable, and the absence of calf pain prompted us to avoid prophylactic fasciotomy during surgery. The patient's edema resolved almost immediately after thrombectomy, making fasciotomy unnecessary.

Constructing a small arteriovenous fistula (AVF) may assist in maintaining patency by increasing the flow velocity through a thrombogenic iliofemoral venous segment and promoting collateral development; however, we have found no solid evidence in the literature to support creating an AVF during open surgical thrombectomy. Doing so can increase the risk of venous hypertension and postthrombotic syndrome. In addition, creating an AVF may heighten surgical risk because of longer operative times and the need for reintervention. Finally, most of our patients are lost to follow-up, which precludes AVF ligation after 2 to 3 months. In a controlled hospital network with strict patient follow-up, AVF creation can be useful in individual patients.

Because our patient had a history of cancer and no previous venous insufficiency, we did not expect to find an underlying causal lesion; an intraoperative ultrasonogram showed none in the iliofemoral segment. A venogram, obtained later in the catheterization laboratory, showed no residual lesions.

Conclusion

In PCD, rapid diagnosis and treatment are crucial for restoring venous circulation, preserving limb function and integrity, and perhaps saving patients' lives. Open venous thrombectomy should be considered a viable treatment option for PCD.

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