A Case of Severe DVT with Phlegmasia Cerulea Dolens and Maythurner Syndrome treated with Endovascular Intervention

Sajjad Haider MD, Sameer Saleem MD, Aniruddha Singh MD, Muhammad S Akbar MD, Avnish Tripathi MD
Western Kentucky Heart and Lung Foundation, The Medical Center of Bowling Green

Objective

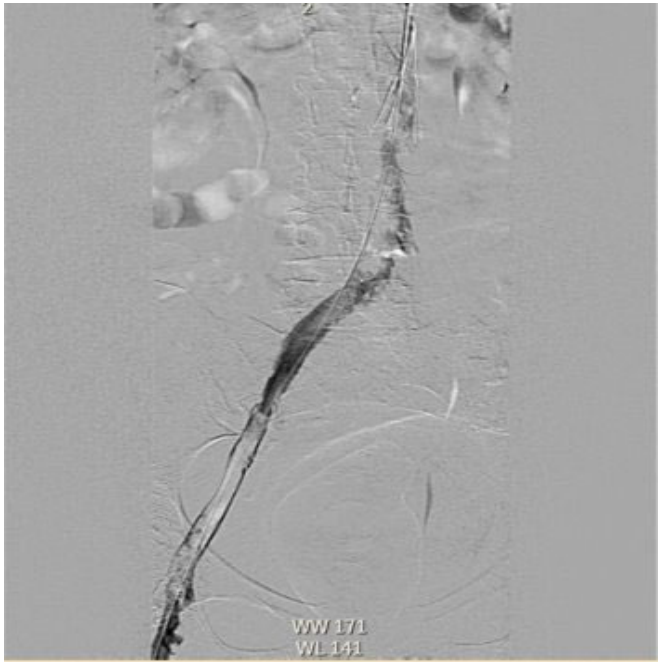
To understand limitations of traditional treatment (anticoagulation) in cases of severe DVT. To underscore importance of prompt recognition of limb threatening conditions including phlegmasia cerulea dolens (PCD) in such cases and swift treatment with endovascular interventions.

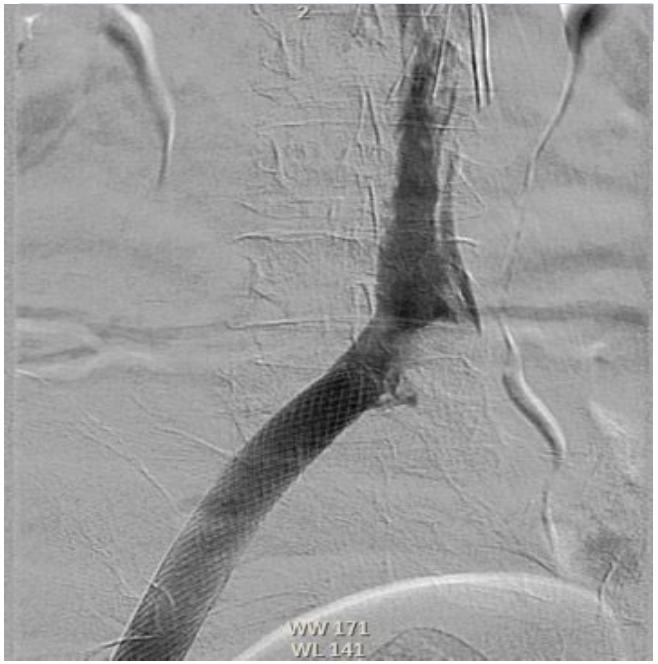
Introduction

PCD is an uncommon presentation of severe DVT which can cause irreversible ischemia and gangrenous limb necrosis. While many cases of DVT are adequately managed with anticoagulation alone, Phlegmasia cerulea dolens is a medical emergency requiring escalation of therapy beyond simple anticoagulation to prevent sequelae such as post-thrombotic limb. May-Thurner syndrome can present as a spectrum ranging from asymptomatic stenosis to severe DVT. May-Thurner syndrome should be considered as etiology in cases of phlegmasia cerula dolens. Timely identification of PCD is pertinent to initiate systemic anticoagulation & is an indication of thrombolysis and/or thrombectomy. We present a unique case of May-Thurner syndrome causing severe DVT and phlegmasia cerulea dolens following cardiac catheterization.

Case Presentation

A 70-year-old female with past medical history of CAD, remote DVT, colon cancer s/p hemicolectomy, hypertension and hypercholesterolemia presented with chest pain. She was diagnosed with NSTEMI and underwent cardiac catheterization through right femoral approach with DES placement to mid LAD. POD 1, she developed pain in bilateral lower extremities. CT angiogram of abdominal aorta with run off showed patent visceral and bilateral lower extremity arteries. Venous doppler of thelower extremities showed extensive DVT of right common femoral vein and saphenous vein with some flow seen on color doppler. Occlusive DVT in the right proximal, mid, and distal femoral vein and popliteal vein. Right deep femoral and posterior tibial vein showed normal flow. On the left, occlusive DVT in the common femoral, proximal greater saphenous vein, throughout the femoral vein and popliteal vein. She was initiated on heparin, but patient continued to complain of worsening bilateral extremity pain. Bluish discoloration was noted in her left lower extremity suspicious for phlegmasia cerulea dolens. CT venography was unavailable at our facility. Patient thus underwent venogram of infra-renal, left iliofemoral and left femoral-popliteal system which showed extensive acute thrombosis of left lower extremity of involving the left common iliac vein, left common femoral vein, left superficial femoral vein and left popliteal vein with. Left common iliac vein had stenosis from May-thurner syndrome caused by compression from right common iliac artery. Mechanical thrombectomy was performed from infra-renal IVC to the left popliteal vein using anigojet catheter followed by balloon angioplasty of the venous system from left common iliac vein to the left popliteal vein and deployment of wall stent endoprosthesis. Her left lower extremity pain and phlegmasia cerulea dolens resolved post procedure. Patient was continued on oral anticoagulation with Apixaban at the time of discharge.





Discussion

Phlegmasia cerulea dolens is an advance complication of severe DVT and can cause frank venous gangrene. Typical presentation of PCD include Extreme pain, severe swelling, cyanosis, and discoloration of the involved extremity due to near total venous occlusion. It is a life-threatening condition with amputation rates of 12 to 50% and mortality rates of 20 to 40%, thus it is crucial to recognize this condition promptly and accurately as treatment is time sensitive. Many risk factors including hypercoagulable syndrome, trauma, heart failure, inflammatory bowel disease, malignancy, and May–Thurner syndrome predispose to PCD. Our patient had two risk factors including May-Thurner syndrome and history of colon cancer. May-Thurner syndrome is stenosis of iliac vein caused by compression from iliac artery. Contrast venography demonstrating a flattening or narrowing of the iliac vein at the pelvic brim is the gold standard for May–Thurner syndrome diagnosis. It is rarely considered in the differential diagnosis of DVT, particularly in patients with other risk factors. Systemic anticoagulation alone is insufficient treatment, and a more aggressive approach is necessary to treat severe DVT and prevent recurrent DVT. In patients with phlegmasia cerulea dolens, May-thurner syndrome should always be considered as a differential. Aggressive treatment including catheter-directed thrombolysis, mechanical thrombectomy, angioplasty and stent placement, fasciotomy, and placement of an inferior vena cava filter should be considered in patients with PCD caused by May-Thurner syndrome.

Conclusions

Although rare, occlusive thrombosis of the lower extremity deep veins associated with May-Thurner syndrome can lead to significant morbidity, including arterial compromise, gangrene, and, ultimately, amputation. In patients with extensive lower extremity thrombosis as in the present case, prompt diagnosis and aggressive endovascular treatment can result in optimal outcomes.

Contact

Sajjad Haider MD
U of Ky COM,
Bowling Green.
Email:sajjadhaider@uky.edu
Phone:270-745-1000

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