CLINICAL VIGNETTE

May-Thurner Syndrome: A Pelvic Anatomic Variant Predisposing Individuals to Venous Thromboembolism

Christie Masters, M.D., MBA, MHA

Introduction

An elderly male presented with a second occurrence of venous thromboembolism (VTE) associated with immobility and inactivity. Hypercoagulable workup was negative, however, imaging revealed May-Thurner syndrome (MTS), an anatomical variant resulting in segmental narrowing of the left common iliac vein with predisposition to venous thrombosis. Clinicians should include MTS in the differential of seemingly unprovoked left lower extremity deep vein thrombosis in young or otherwise healthy individuals.[1] This case presentation reviews MTS and the approach to management.

Case Report

A 73-year-old male presented to the Emergency Department (ED) with twelve hours of left lower extremity pain. His past medical history is significant for left lower extremity deep vein thrombosis (DVT) in 2011 treated with six months of warfarin. His previous DVT was attributed to inactivity. The patient retired from computer programing in 2000. Since retiring, he spends most of his time lying in bed working on his computer. He first noticed recurrent swelling in his left lower extremity the day before this Emergency Department (ED) presentation. His left leg also became painful with ambulation. The persistent pain and swelling caused him to come to the ED. On review of systems, he reported increasing shortness of breath without associated chest pain for the past few weeks when climbing stairs or walking short distances. Review of systems was negative for fevers, chills, night sweats, and unintentional weight loss.

His family medical history was negative for VTE and hypercoagulability. He had no recent travel, trauma, or surgery. He does not smoke. D-dimer was elevated at 7910 ng/mL. Duplex ultrasound showed acute DVT of the left lower extremity involving the common femoral, proximal profunda femoris, superficial femoral, popliteal, posterior tibial, peroneal, and gastrocnemius veins. Arterial ultrasound of the left lower extremity was unremarkable. Computed tomography angiography of the chest revealed acute pulmonary embolism (PE) with normal transthoracic echocardiogram (TTE) and negative troponins. Antithrombin III factor V Leiden, protein C and S, cardiolipin, factor VIII homocystine, prothrombin, and prostate-specific antigen returned within normal limits. He was started on heparin infusion in the ED.

Pulmonary and interventional radiology were consulted for consideration of thrombolysis of his PE. Thrombolytic therapy was not recommended for the PE given the patient's clinical stability and benign TTE. Instead, inferior vena cava (IVC) filter and thrombolysis of his left lower extremity were recommended due to the extent of clot burden and risk of post-thrombotic syndrome (PTS). Venogram of his left lower extremity reports acute thrombus throughout the popiteal veins through the common femoral vein, as well as chronic thrombus within the common and external iliac veins, compatible in appearance with May-Thurner Syndrome. Venogram also reveals acute on chronic thrombus in his left lower calf.

The patient underwent catheter directed thrombolysis of left lower extremity via the common femoral vein and popliteal vein approach using the EKOS EndoWave Infusion Catheter System with tissue plasminogen activator (tPA) running at 0.25 mg/h per catheter for a total dose of tPA 0.5 mg/h. He additionally continued on low-dose heparin and was transferred to the intensive care unit for close observation while receiving tPA.

Post-intervention left lower extremity venogram two days later revealed restoration of venous outflow within the left lower extremity and widely patent femoral and popliteal veins. However, residual segmental narrowing in the left common iliac vein remained, secondary to May-Thurner syndrome. Stenting and balloon venoplasty of the left common iliac vein were performed with post-stent venogram showing normal caliber of the stented proximal left common iliac vein and resolution of the segmental narrowing with excellent antegrade flow. The patient was started on rivaroxaban 15mg orally twice a day for three weeks followed by rivaroxaban 20mg orally daily for lifelong therapy due to recurrent DVT. He was discharged home without complications and returned four weeks later for removal of IVC filter.

Discussion

Venous compression syndromes occur when adjacent structures extrinsically compress a vein in tight anatomical spaces.[2] Clinicians can suspect extrinsic compression syndrome in patients who present with presumably unprovoked venous thrombosis who are otherwise healthy or young.[1-3] Examples of extrinsic venous compression syndromes include Paget-Schroetter Syndrome associated with compression of the subclavian vein, Nutcracker Syndrome affecting the left renal vein, Popliteal Venous Compression, and May-Thurner Syndrome (MTS), which is also known as iliac vein compression syndrome, Cockett's syndrome, and iliocaval compression syndrome.[2,4] May-Thurner syndrome occurred in 2-5% of patients undergoing evaluation of DVT or venous disorder.[4]

The case above describes MTS, which is a venous compression syndrome causing iliofemoral deep venous thrombosis due to an anatomic variant in which the right common iliac artery compresses the left common iliac vein against the lumbar spine. In 1851, Virchow first reported a greater incidence of left-side DVT than right.[1,3] In 1957, May and Thurner further defined the anatomic variant causing Virchow's finding.[2,4] Virchow's Triad included stasis of blood flow, endothelial injury, and hypercoagulability. The anatomical variant associated with MTS results in repeated endothelial trauma to the vein by chronic pulsations from the overlying artery.[3] Venous intimal hyperplasia develops from this chronic trauma and predisposes the affected individual to venous stasis and thrombosis, as well as chronic venous insufficiency, varicosities, or formation of venous collaterals.[1-3]

MTS is more prevalent than once thought. According to Peters et al,[1] many left lower extremity DVTs are not correctly attributed to MTS if a patient has an identifiable cause for DVT, and US confirms the presence of thrombosis. Failure to diagnose MTS can lead to recurrent DVT or PE, both of which occurred in the patient described. Other complications that can result from undiagnosed or inadequately treated MTS include chronic venous stasis, non-healing ulcers, severe venous claudication, or iliac vein rupture.[1,4] Venogram, magnetic resonance imaging, or intravascular ultrasound are required to diagnose MTS due to the location of the variant high in the pelvis.[1,4]

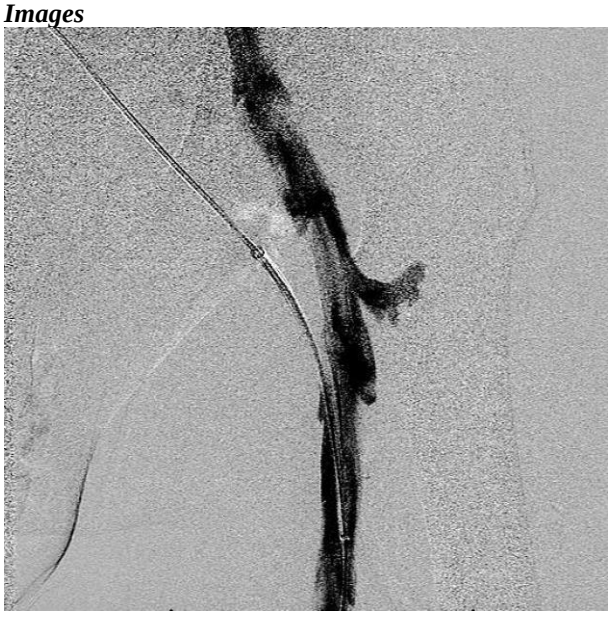
Management of MTS involves treating the thrombus and preventing recurrence. According to Hsu et al,[4] the principle treatment is to relieve the vein narrowing caused by the extrinsic compression. Surgical interventions to correct or bypass the anatomic variant were once recommended but have shown variable long term results.[4] Anticoagulation helps to prevent recurrent thrombosis and thrombus extension, it but does not dissolve acute thromboembolism or correct the underlying anatomic variant causing the problem in MTS.[5,6] Standard of care in the treatment of MTS is evolving to include catheter-directed thrombolysis to quickly dissolve the DVT and decrease the risk of post-thrombotic syndrome, as well as intravenous stenting to repair the venous narrowing.[1,5,7] The use of IVC placement and lifelong anticoagulation are debatable. The patient in this case underwent both catheter-directed thrombolysis and intravenous stent placement, as well as IVC filter placement with subsequent removal four weeks later given the extent of this acute and chronic thromboses.

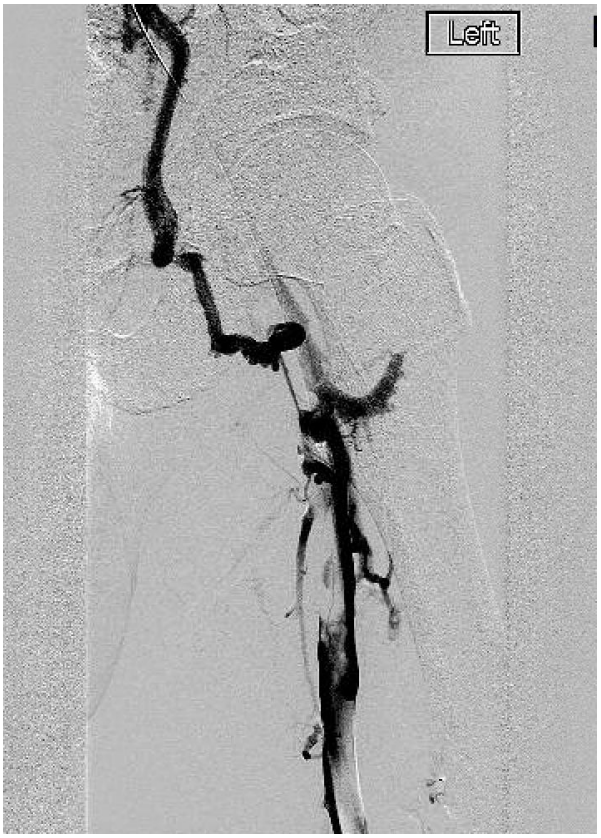
Post-intervention imaging 48-72 hours after thrombolysis is required to assess any remaining clot-burden and the degree of remaining venous narrowing after dissolution of the thrombus.[1,6] Post-intervention left lower extremity venogram for the patient in this case revealed residual segmental narrowing of his left common iliac vein that required an intravascular stent to prevent recurrent thromboembolism due to the presence of MTS.[1] Lifelong oral anticoagulation is recommended for this patient given his sedentary lifestyle, extent of clot burden, recurrent VTE, and PE. Short-term anticoagulation following thrombolysis and repair of the anatomic variant has been recommended in other cases of MTS or venous compressions syndromes.

Conclusion

May-Thurner syndrome is a cause of iliofemoral deep vein thrombosis due to an anatomic variant most commonly causing compression of the left common iliac vein between the overlying right iliac artery and lumbar spine. MTS should be suspected in young or otherwise healthy patients who appear to have an unprovoked left lower extremity DVT. Treatments include anticoagulation to prevent recurrent thromboembolism and extension of the clot, catheter-directed thrombolysis to decrease the risk of post-thrombotic syndrome, and stent placement to further correct the anatomic variant causing the thromboembolic disease. Benefits of IVC filter placement and lifelong anticoagulation are debatable. The choice of treatment for an individual patient depends on the risks and benefits associated with each case presentation.

Images





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