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CLINICAL VIGNETTE

May Thurner Syndrome

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A 19-year-old female presented with chief complaint of left leg swelling for one day. She denied fevers, cough, dyspnea, chest pain, recent trauma, or immobility.

She had no significant past medical or surgical history, no allergies, and no significant family medical history. Of note, she had started an estrogen-progesterone containing oral contraceptive 3 weeks prior to the visit but was not taking any additional prescribed or over the counter products. She denied any alcohol, tobacco, or recreational drug use.

Her physical exam showed a well-developed female in no apparent distress with normal vitals except for a resting tachycardia of 115.

Her HEENT, oropharyngeal, lung, and abdominal exams were unremarkable. Her cardiac exam showed a tachycardia without murmurs, gallops, rubs, or extra heart sounds. Inspection of her left lower extremity showed left thigh edema. Her left thigh measured one inch larger in circumference compared to her right thigh. Skin overlying her left thigh was tender to touch, but there was no overlying erythema or warmth. No palpable cords or varicosities were identified. She did not have calf or ankle edema. Neurologically, she was intact, and her dorsalis pedis and posterior tibial pulses were normal.

Given clinical suspicion for left lower extremity deep venous thrombosis, she was sent to the emergency department where she underwent a left lower extremity Doppler. This imaging did not show any sonographic evidence of acute deep vein thrombosis though slow flow was noted in the high left common femoral and popliteal vein. A CT angiogram of her chest ruled out pulmonary embolism. Lastly, she had an abdominal CT with IV contrast that showed the May Thurner anatomic variant and nonenhancing left common iliac, external iliac, internal iliac, and left common femoral veins.

Review

May Thurner syndrome or “iliac vein compression syndrome” is a rarely diagnosed condition in which patients develop iliofemoral deep venous thrombosis (DVT) due to an anatomical variant in which the right common iliac artery overlies and compresses the left common iliac vein against the 5th lumbar vertebrae.^{1,2} Virchow in 1851 observed that iliofemoral deep venous thrombosis (DVT) was five times more likely to occur in the left leg as compared to the right.² Years later in 1957, MTS was first described by May and

Thurner who noted that 22% of 430 cadavers possessed this iliac vein anatomical variant.³ They described the development of “venous spur” in the left common iliac vein caused by chronic pulsation of the overlying iliac artery leading to venous stasis, intimal fibrosis, and ultimately venous thrombosis.³

Patients with May Thurner syndrome typically present with left lower extremity swelling and pain. The clinical prevalence of MTS-related DVT is low, reportedly occurring in only 2% to 3% of all lower extremity DVT's.¹ This may be an underestimation as other risk factors for DVT are more easily recognized, including pregnancy, use of combination contraceptives, immobilization, and prolonged travel.¹ In addition, traditional imaging studies for DVT evaluation like the lower extremity doppler will miss the underlying anatomic variant.

If MTS is suspected, contrast venography, magnetic resonance imaging, or intravascular ultrasound should be performed as the anatomic defect occurs in the pelvis and may be missed with the use of a lower extremity Doppler.⁴ While systemic anticoagulation is commonly used for lower extremity DVT's, as monotherapy is not sufficient to prevent recurrence of thrombosis caused by MTS. Treatment is focused on clearing the thrombus present to prevent post-thrombotic syndrome and to correct the underlying compression of the left iliac vein.² The standard of care involves the combination of thrombolytics and endovascular intervention.² Stent placement has proven highly successful in MTS with 1-2 year iliac vein patency rates reported between 95-100%.^{1,4} Following stent placement, systemic long-term anticoagulation is recommended for at least 6 months.⁴

My patient underwent percutaneous pharmacomechanical thrombectomy of the left common iliac vein, external iliac vein, and common femoral vein and thrombolysis. A stent was placed in the left common iliac vein, and she was anticoagulated with warfarin for 6 months. She is currently on Plavix and recently had a 12-month follow-up showing stent patency.

In conclusion, May Thurner syndrome is an important cause to consider in the differential of DVT risk factors. This is true in particular when presented with a case of unilateral DVT especially in the younger age population (age group of 20-40 years). If the anatomic variant is missed, the recurrence of thrombosis, pulmonary embolism, and post thrombotic syndrome will lead to significant morbidity and mortality. The

key to successful treatment in MTS related DVT is to fix the anatomical lesion through endovascular stenting along with removal of the clot and use of anticoagulation.

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