When You Cannot Go With the Flow: A Case Report of May-Thurner Syndrome

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ABSTRACT

Introduction: May-Thurner syndrome (MTS) is caused by extrinsic compression of the left iliac venous system, most commonly between an overlying right iliac artery and fifth lumbar vertebra, and is seen mainly in women between 20 and 50 years of age. This compression may be asymptomatic but can lead to the formation of venous thrombi causing left lower extremity pain and swelling.

Case Presentation: We report a case of MTS in a patient who initially presented with back and left lower extremity pain and swelling whose initial duplex venous ultrasound was negative for deep venous thrombus. Subsequent computed tomography (CT) revealed a venous thrombus of the left common, internal, and external iliac veins.

Discussion: In the case of unilateral lower extremity swelling with a negative initial duplex venous ultrasound, consider further investigation with CT venography to evaluate for possible MTS.

INTRODUCTION

May-Thurner syndrome (MTS) is a vascular phenomenon that occurs secondary to compression of the left iliac vein by an overlying right iliac artery and usually presents with signs and symptoms of deep venous thrombosis. Although first described by Rudolph Virchow in 1851, MTS was coined by May and Thurner in 1957 after they found that up to 32% of cadavers they studied had their right iliac artery compressing the left iliac vein.1 Interestingly, MTS has been shown to be the primary cause of deep venous thrombosis (DVT) only 2%-5% of the time.2 We present a case of a 71-year-old woman presenting for evaluation of lower back and left leg pain with swelling. The patient’s initial duplex venous ultrasound was negative for venous thrombosis, but she was subsequently diagnosed with MTS via computed tomographic venography.

CASE PRESENTATION

A 71-year-old woman with a history of osteopenia, cataracts, and a 25 pack-year smoking history presented to the outpatient clinic with a 1-day history of back and left leg pain. The patient initially woke up with lower back pain and then developed left leg pain and subjective erythema. Other than her sex and smoking history, she had no risk factors for thromboembolic disease. Her review of systems was negative other than the presenting symptoms. On physical examination, the entire left leg was mildly swollen and erythematous compared with the right (Figures 1 and 2), but there was no tenderness to palpation, no weakness, a negative straight leg raise test, and palpable pulses. Given the high clinical suspicion for DVT, she was sent for a duplex venous ultrasound. High-resolution images of the common femoral through popliteal veins of the left lower extremities were obtained in conjunction with doppler duplex sonography, augmentation, and compression with additional images to identify the peroneal, posterior tibial, and anterior tibial veins. No left deep venous thrombosis was identified, and the contralateral right common femoral vein was found to be patent with symmetrical phasic flow.

The following day, the patient presented to the emergency department (ED) for left-sided back pain radiating to her left anterior thigh, which worsened on ambulation. Physical examination at this time found the left leg to be 3 cm larger in circumference compared with the right leg. Her laboratory findings were consistent with an elevated D-dimer (> 4.0 μg/mL). Given the physical examination and pertinent laboratory findings, the original lower extremity doppler ultrasound was thought to be a false negative. The patient was therefore given a single dose of enoxaparin and instructed to get a second ultrasound the following morning. Repeat duplex venous ultrasound revealed fully compressible deep veins with slow flow and loss of phasicity in the left common femoral waveform. These findings raised concern for a more proximal compressive lesion, so the patient underwent computed tomography (CT) venography of the abdomen and pelvis. This study revealed the presence of expansile intraluminal material in the left common, internal, and external iliac veins, consistent with venous thrombus and the diagnosis of MTS.

After discussion with Interventional Radiology, the patient was started on rivaroxaban and subsequently underwent left iliac vein thrombolysis followed by Vici venous...
self-expanding stent placement. She was started on a 1-month course of enoxaparin with a transition to rivaroxaban alongside a daily baby aspirin. She was educated on the importance of smoking cessation and adequate cancer screening, which ultimately came back negative. Repeat CT venography 8 months post-thrombolysis showed no evidence of DVT in the left common, internal, and external iliac veins, so the patient was taken off direct oral anti-coagulation and maintained on lifelong aspirin therapy with no further recurrence of symptoms to date. A timeline of the case appears in Table 1.

DISCUSSION

Acute unilateral lower extremity swelling is a concerning physical finding that requires prompt evaluation. The differential diagnosis for acute unilateral leg swelling includes, but is not limited to, DVT, muscle strain or tear, cellulitis, venous insufficiency, and MTS. Once the diagnosis of DVT is ruled out, strong clinical suspicion is warranted to evaluate for underlying MTS. MTS is caused by extrinsic compression of the left iliac venous system, most commonly by an overlying right iliac artery (Figure 3). Such compression may be asymptomatic, but chronic and complete compression can cause endothelial injury within the vessel walls, leading to intimal hyperplasia and the formation of venous spurs and thrombi. The clinical stages of iliac vein compression are further detailed in the article by Kim et al, and the stages of MTS formation are described by Ibrahim et al.

Because most individuals with MTS are asymptomatic, the exact incidence of the syndrome is unknown. A study done by Kibbe et al retrospectively evaluated 50 helical abdominal CT scans of patients presenting to the ED for abdominal pain and found an overlying compression of the left iliac venous system in 22%-24% of the patient population. Despite this prevalence, MTS is thought to be the cause of DVTs at a rate of only 2%-5%. In patients with strictly left lower extremity DVTs, the incidence has been shown to be as high as 76%.
A 71-y-old woman with a history of osteopenia, cataracts, and a 25 pack-y smoking history presented to the outpatient clinic with a 1-d history of back and left leg pain.

<table>
<thead>
<tr>
<th>Date</th>
<th>Summaries from initial and follow-up visits</th>
<th>Diagnostic testing</th>
<th>Interventions</th>
</tr>
</thead>
<tbody>
<tr>
<td>September 26, 2019</td>
<td>Presented to clinic with left leg and lower back pain and swelling</td>
<td>Duplex venous ultrasound negative for a deep vein thrombus</td>
<td>Instructed to return to clinic or present to the emergency department (ED) if symptoms worsen; provided smoking cessation counseling</td>
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<tr>
<td>September 27, 2019</td>
<td>Presented to ED with worsening leg pain and swelling</td>
<td>D-dimer &gt; 4000; repeat duplex venous ultrasound reveals slow flow in the left-sided deep veins</td>
<td>Given single dose of enoxaparin and scheduled for outpatient computed tomographic (CT) venography of the abdomen and pelvis</td>
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<tr>
<td>October 1, 2019</td>
<td>CT venography revealed venous thrombus in left common iliac, internal, and external iliac veins consistent with May-Thurner syndrome</td>
<td>No additional tests given</td>
<td>Started on rivaroxaban and provided smoking cessation counseling; consult placed to Interventional Radiology for lysis procedure</td>
</tr>
<tr>
<td>October 3, 2019</td>
<td>Undergoes interventional radiology-guided left iliac vein thrombolysis and Vici venous stent placement.</td>
<td>No additional tests given</td>
<td>Started on 1-mo of enoxaparin and aspirin with plan for transition to ivaroxaban.</td>
</tr>
<tr>
<td>October 14, 2019</td>
<td>Presented for post-thrombolysis follow-up with primary care provider with complete resolution of symptoms</td>
<td>No additional tests given</td>
<td>Instructed to finish 1-mo course of enoxaparin before transitioning to rivaroxaban and to continue daily aspirin.</td>
</tr>
<tr>
<td>June 8, 2020</td>
<td>Presented for interventional radiology follow-up after taking rivaroxaban for over 6 mo</td>
<td>Repeat CT venography to assess patency of iliac veins and stent revealed no deep vein thrombus and patent stent</td>
<td>Instructed to stop rivaroxaban and continue aspirin for life with no further follow-up warranted</td>
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</table>

Although the most common symptom of MTS is left lower extremity swelling, patients may present with venous claudication, skin discoloration, or ulceration. As in our case, claudication of the patient’s left thigh led to her presenting to the ED despite having a normal initial duplex ultrasound, ultimately requiring further workup with a repeat ultrasound. Although these clinical features may hint at the diagnosis of MTS, definitive diagnosis requires radiological evidence of stenosis in the appropriate anatomical location. As in our case, determining the clinical probability of DVT is essential using D-dimer and Wells criteria. The elevation in our patient’s D-dimer on presentation to the ED raised the index of suspicion for the presence of a thrombus requiring further imaging.

It is important to note that even though doppler ultrasound is the most widely used technique to diagnose DVT, with a sensitivity and specificity greater than 90%, it is unable to detect proximal iliac vein spurs or thrombi due to their anatomical location. As such, other diagnostic modalities should be considered, such as computed tomography and magnetic resonance venography, both of which have a sensitivity and specificity greater than 95%. In addition to identifying proximal iliac vein compression, CT venography can help rule out other causes of left lower extremity swelling apart from MTS, including cellulitis, hemATOMA, and venous obstruction from cancer-related metastatic lymphadenopathy. An important limitation to consider is that CT venography does not account for the volume status of the patient, thereby raising the risk of false negatives due to the increased degree of iliac vein compression in hypovolemic patients. We report an interesting case of MTS where initial diagnostic imaging results were incongruent with the clinical presentation. In such instances, we propose that elevated clinical suspicion and probability for a DVT warrant further imaging of more proximal vasculature using CT or magnetic resonance venography to evaluate for MTS.

The management of MTS aims to dissolve the clot burden and relieve the iliac venous compression. Catheter-directed thrombolysis is the first-line therapy for removal of the thrombus. In patients with mild symptoms or contraindications to lytic therapy, lifelong anticoagulation has been a common treatment modality. This form of therapy, however, has been shown to be less effective in the long run of decreasing the clot burden compared with catheter-directed thrombolysis. In patients with no such contraindication, thrombolysis with urokinase or tissue plasminogen activator have proven efficacious in clot dissolution. Following thrombolysis, several studies have demonstrated the importance of stent placement as a promising option over angioplasty alone for post-thrombotic iliac vein obstruction. Without stenting after thrombolysis, recurrent rates have been shown to be higher than 70%. In our case, we decided to proceed with an iliac vein thrombolysis followed by venous stent placement with excellent clinical outcome.
CONCLUSION

We conclude that MTS should be considered in the differential diagnosis of patients presenting with unilateral lower extremity swelling, particularly of the left lower extremity. CT venography of the abdomen and pelvis may prove useful in patients with signs and symptoms concerning for a deep venous thrombus but with an initial negative venous ultrasound.

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Author Contributions
Adnan Khan, MD, researched the topic and wrote the manuscript. Jeffrey Wang, MD, aided with researching the topic and editing the final manuscript. Leslea Brickner, MD, Nirmala Ramalingam, MPP, and Nicole Tran, MD, PhD, provided critical feedback and contributed to the final editing of the manuscript.

References