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Case of Deep Venous Thrombosis Secondary to May-Thurner Syndrome

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Abstract May-Thurner syndrome (MTS) remains an underdiagnosed cause of venous thromboembolism (VTE) because most individuals with MTS anatomy are asymptomatic. The most common variant of MTS is due to compression of the left iliac vein between the overlying right common iliac artery and the fifth lumbar vertebrae. Clinical features include chronic venous hypertension that usually presents as lower extremity pain and swelling. Acute VTE almost exclusively affects the left lower extremity. In cases of acute VTE of an unknown etiology, MTS should be considered when other more common pathologies have been ruled out. A 65-year-old African American gentleman with past medical history of hypertension and coronary artery disease presented with left lower extremity pain and swelling of the leg for five days. He had no history of leg trauma, recent surgery, bed rest, travel, malignancy, previous clotting episodes or family history of hypercoagulable disorders. Patient regularly ambulates. He is a lifetime non-smoker and does not take any medication. His left lower extremity was swollen from the calf down to the ankle and foot, tense, erythematous and tender to palpation. Dorsalis pedis and posterior tibial pulses were weakly palpable. Homan's sign was appreciated while the rest of the physical exam was unremarkable. Duplex ultrasound of the left lower extremity showed thrombi in the left popliteal, posterior tibial and peroneal veins. CT abdomen and pelvis with IV contrast demonstrated significant compression of the left common iliac vein as it crosses posterior to the left internal iliac artery, consistent with MTS. Spiral chest CT was significant for subsegmental emboli in the bilateral lobe pulmonary arteries. Patient was started on anticoagulation, then he was referred to an advanced vascular center to consider the need for angioplasty and stenting and for possible thrombolysis. MTS was first described in 1908 by Virchow, who observed that iliofemoral vein thrombosis was five times more likely to occur in the left leg than in the right leg. May and Thurner discovered an anatomical variant where the right iliac artery compressed the left iliac vein against the fifth lumbar vertebra. Clinicians should have a high index of suspicion for MTS in the presence of unprovoked DVT in the left lower extremity, and/or signs of chronic venous hypertension. Angioplasty and stenting of the affected lesion is the definitive treatment for MTS, while anticoagulation management is similar to patients with provoked VTE. Therefore, it can be argued that in patients with an unexplained cause of VTE, investigation for MTS if clinically suspected can impact management decisions.

Keywords: *May-Thurner syndrome, Venous Thromboembolism, Anticoagulation, Deep Vein Thrombosis, Pulmonary Embolism*

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1. Introduction

A 65-year-old male with no prior history of venous thromboembolism (VTE) who presents with 5 days of left lower extremity pain and swelling. A deep vein thrombosis (DVT) was then confirmed with duplex ultrasonography. He was started on therapeutic enoxaparin. As he had no historical provoking factors for VTE he was further investigated with a CT abdomen with IV contrast. This revealed the cause of the DVT - significant compression of the left common iliac vein as it crosses posterior to the left internal iliac artery, consistent with May-Thurner syndrome (MTS). He was subsequently discharged with apixaban with referral to a vascular center for angioplasty and stenting.

MTS continues to be a cause of VTE that is underdiagnosed. Investigating this provoking phenomenon in patients with unexplained VTE should be considered as definitive treatment entails angioplasty with stenting and short-term anticoagulation. Most MTS cases report compression of the left common iliac vein between the right common iliac artery and the fifth lumbar vertebra, however less common anatomical variants have been reported, such as our case, where the left common iliac vein is compressed between the left internal iliac artery and the first sacral vertebra.

2. Background

The most common variant of MTS is due to compression of the left iliac vein between the overlying right common iliac artery and the fifth lumbar vertebra. The exact incidence and prevalence of MTS are unknown but are likely to be underestimated given that most individuals with MTS anatomy do not have symptoms and require no treatment [1]. Risk factors include female gender, scoliosis, dehydration and hypercoagulable disorders. Clinical features include acute lower extremity pain and swelling, with the swelling usually involving the entire limb. Claudicating pain can also be present. Diagnosis of MTS may be suspected based upon clinical features and initial diagnostic testing with venous duplex ultrasound, CT or MRI venography, however more invasive studies including catheter-based venography and intravascular ultrasound (IVUS) might be done in the event of diagnostic uncertainty. Treatment of MTS depends on whether a deep venous thrombosis (DVT) is present. In the absence of DVT, treatment is conservative with compression stockings. For advanced nonthrombotic MTS with symptoms/signs of advanced chronic venous insufficiency, treatment is targeted toward reducing the severity of the stenotic venous lesion using angioplasty and stenting of the affected segment. Here, we present a patient who was found to have what was initially suspected to be an unprovoked DVT, only to confirm on further imaging the presence of MTS upon evaluation for predisposing conditions.

3. Case Presentation

A 65-year-old African American male with a past medical history of hypertension and coronary artery disease status post stent in 2010 presents with left lower extremity pain and swelling for the past 5 days. Pain began in the popliteal region. He has no history of any leg trauma, recent surgery, bed rest, travel, malignancy, previous clotting episodes or family history of hypercoagulable disorders. He denies any dyspnea or chest pain. Patient is regularly ambulatory. He is a lifetime non-smoker, occasional alcohol use, occasional marijuana and cocaine use. He does not take any home medications. Lung and cardiac exam unremarkable. His left lower extremity was tense, erythematous, tender to palpation and swollen from the calf down to the ankle and foot. Dorsalis pedis and posterior tibial pulses were weakly palpable. Homan's sign was present.

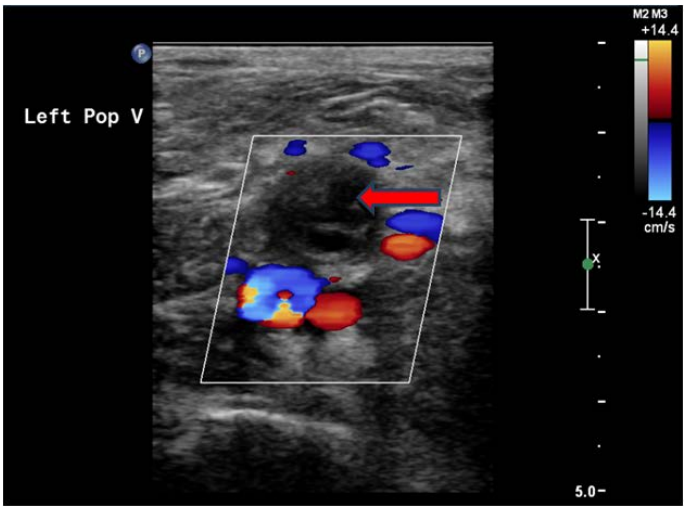

Figure 1. Color doppler ultrasonography image of patient's left popliteal vein. Left popliteal vein is not compressible due to occlusive thrombus (arrow).

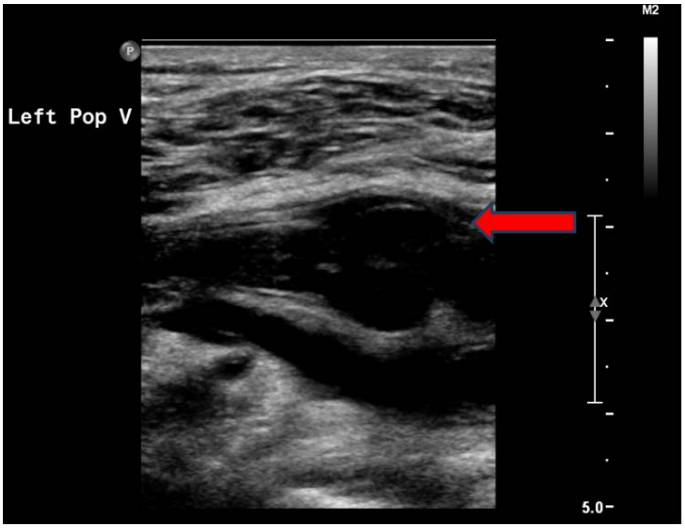

Figure 2. Color doppler ultrasonography image of patient's left popliteal vein. Left popliteal vein is not compressible due to occlusive thrombus (arrow)

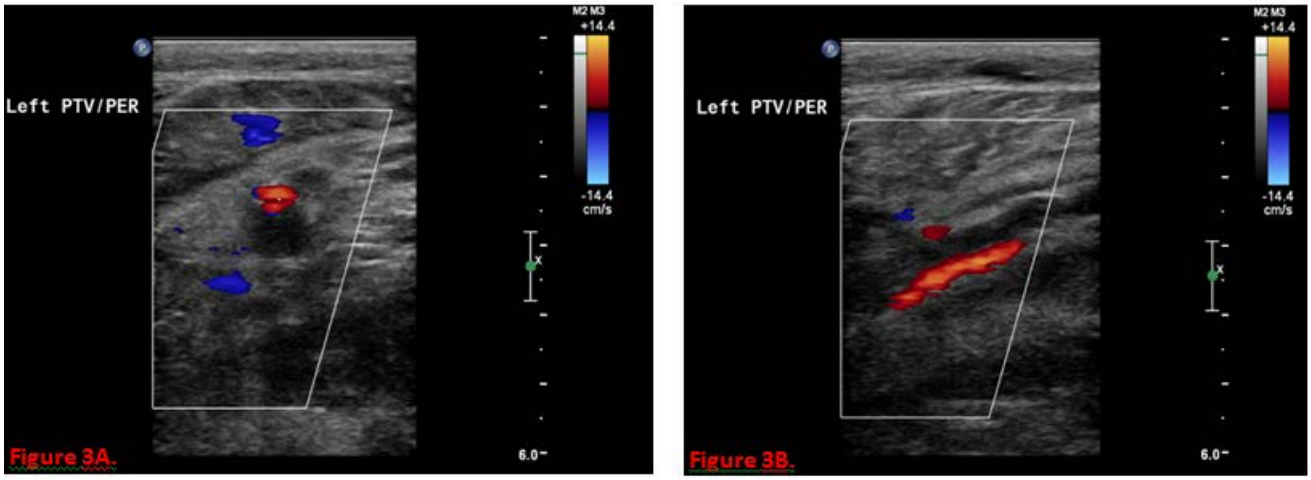

Figure 3A, B. Color doppler ultrasonography image showing occlusive thrombus extending to involve the left posterior tibial and left peroneal veins

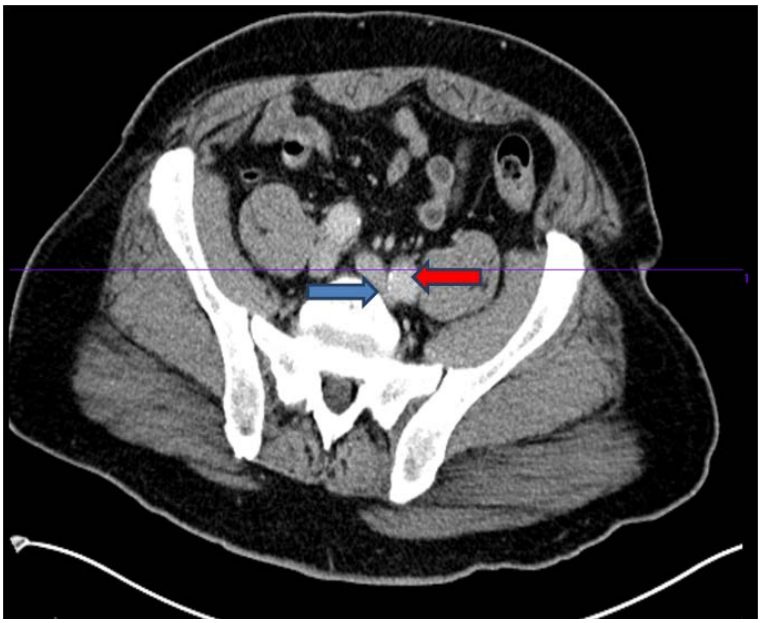

Figure 4. Enhanced axial CT of the abdomen and pelvis, demonstrating compression of the left common iliac vein (blue arrow) between the left internal iliac artery (red arrow) and the vertebral body at the S1 level

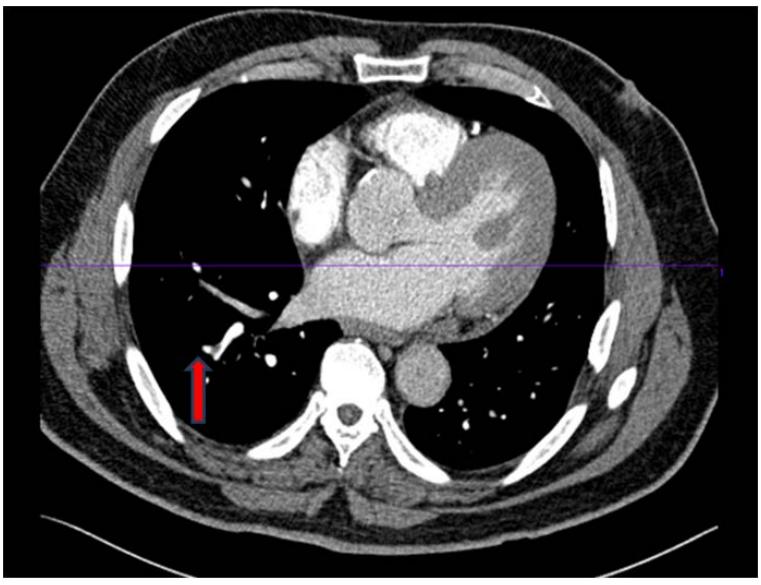

Figure 5. Enhanced computed tomography (axial view) of the chest showing small filling defect in the sub segmental branches of the right lower lobe (arrow).

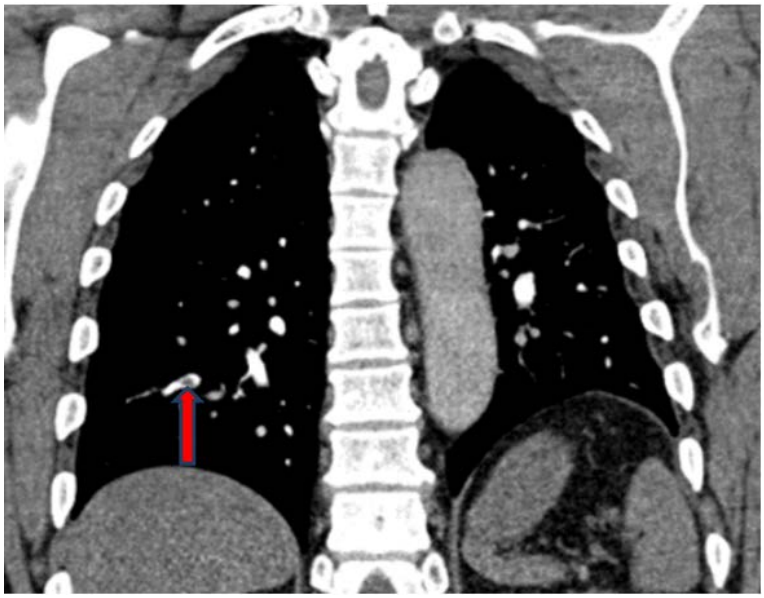

Figure 6. Enhanced computed tomography (coronal view) of the chest showing small filling defect in the sub segmental branches of the right lower lobe (arrow).

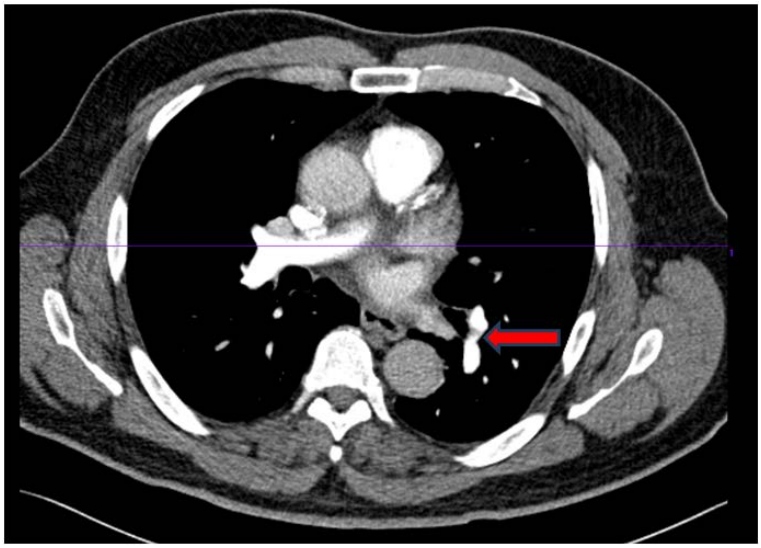

Figure 7. Enhanced axial view of the CT chest showing small filling defect in the sub segmental branches of the left lower lobe (arrow)

Duplex ultrasound of the left lower extremity showed a large thrombus in the left popliteal, posterior tibial and peroneal veins (see Figure 1, Figure 2, Figure 3A and Figure 3B). CT abdomen and pelvis with IV contrast demonstrated significant compression of the left common iliac vein as it crosses posterior to the left internal iliac artery, consistent with May-Thurner syndrome (see Figure 4). Spiral chest CT demonstrated subsegmental emboli in the bilateral lobe pulmonary arteries (see Figure 5, Figure 6 and Figure 7). Patient was started on therapeutic dose of enoxaparin twice a day. Patient was later transitioned to apixaban and was referred to an advanced vascular center to consider the need for angioplasty and stenting as well as for possible thrombolysis.

4. Discussion

May-Thurner syndrome (MTS) was first described in 1908 by Virchow, who observed that iliofemoral vein thrombosis was five times more likely to occur in the left leg than in the right leg. The syndrome was not fully understood until the mid-20th century, when May and Thurner discovered an anatomical variant in 22% of 430 cadavers where the right common iliac artery compressed the left common iliac vein against the fifth lumbar vertebra. They postulated that the chronic pulsations of the overlying right iliac artery led to development of a “spur” in the vein wall and that this spur would result in partial venous obstruction [2].

The classic clinical presentation is that of a younger female in the second or third decade of life with left lower extremity swelling, which is contrasted with our patient’s presentation as a 65-year-old male. Compression of the left common iliac vein in our patient was noted to be at the S1 level between the left internal iliac artery and the sacrum. The exact prevalence of this anatomical variant is unknown, with only a few similar cases citing this variant [3,4]. Swelling was predominantly noted up to the level of the knee in our patient,

which correlated with doppler results noting thrombi in the left peroneal, popliteal and posterior tibial veins.

Catheter-based venography is warranted if there is a sufficient level of suspicion for MTS in a patient with acute symptoms or if the patient has advanced clinical manifestations of chronic venous disease.

In the absence of DVT, patients with only mild symptoms warrant conservative management with compression stockings. For advanced nonthrombotic MTS with symptoms/signs of advanced chronic venous insufficiency, treatment is targeted toward reducing the severity of the stenotic venous lesion using angioplasty and stenting of the affected segment. Angioplasty of the venous stenotic lesion alone is not sufficient as is associated with high recurrence rates [5]. Stenting is not universally agreed upon, and recurrence rates may depend on stent type used [6]. Extending the stent into the inferior vena cava (IVC) has no negative impact [7]. Angioplasty and stenting of the compressible segment also decreases the recurrence rate of superficial reflux following ablation therapies.

If MTS is strongly suspected in a patient with venous thromboembolism (VTE), treatment begins with full therapeutic anticoagulation, if not contraindicated. Our patient was initiated on a full therapeutic dose of enoxaparin100 mg twice a day, which was converted to apixaban 5 mg twice a day. Therapeutic anticoagulation should be continued using similar dosing, monitoring and duration per VTE guidelines. The DVTs detected were considered to be provoked in nature by the external compression of the left iliac vein. Hence, a minimum of three months of full therapeutic anticoagulation is warranted [8]. The patient presented would have been subjected to indefinite anticoagulation if provocative factors were not investigated, which brings to question the possible benefits of investigating for MTS in patients with unexplained VTE. Subsequent treatment is aimed at decreasing the volume of thrombus using catheter-directed thrombolysis or pharmacomechanical thrombolysis, evaluating for intrinsic venous stenosis, and, if present, angioplasty and stenting of the diseased iliocaval segment. With successful treatment of MTS, rates of post-thrombotic syndrome are less than 10%, compared to 80-90% if no treatment provided. Following stenting, concurrent antiplatelet therapy is reasonable, provided bleeding risk is low.

Learning Points

- The prevalence of May-Thurner syndrome (MTS) is unknown for certain and is likely underestimated, largely because most individuals with this anatomic anomaly remain asymptomatic.
- The most common site of left common iliac vein compression is between the right common iliac artery and the fifth lumbar vertebra. However, the incidence of our reported anatomical variant of MTS, where the left common iliac vein is compressed by the left internal iliac artery, is underreported.
- Clinicians should have a high index of suspicion for MTS in the presence of unprovoked DVT in the left lower extremity, recurrent left sided DVT and/or signs of chronic venous hypertension.
- Angioplasty and stenting of the affected lesion and subsequent antiplatelet therapy is the definitive treatment for MTS.
- For patients with VTE in the presence of May-Thurner syndrome, anticoagulation management is similar to patients with provoked VTE. Therefore, it can be argued that in patients with an unexplained cause with VTE, investigation for May Thurner syndrome can impact duration of anticoagulation.

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