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Phlegmasia Cerulea Dolens in May Thurner Syndrome. A threat to life & limb.

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Introduction

Phlegmasia Cerulea Dolens (PCD) is an uncommon, severe type of deep venous thrombosis (DVT). The etiology is usually multifactorial and results in extensive thrombotic occlusion of major and collateral vasculature. PCD is rare but more common in patients that have May Thurner Syndrome (MTS). MTS is an anatomical abnormality in which the right iliac artery compresses the left iliac vein.¹ Emergent medical intervention is required to prevent irreversible limb ischemia and potentially fatal gangrenous necrosis.²

Case Report

A 76-year-old female who had just completed a 2-hour and then subsequent 8-hour flight, was admitted to Sky Ridge Medical Center for left lower extremity pain, swelling, and color change. She had a medical history that was significant for symptoms of one left lower extremity DVT that could not be confirmed with ultrasound two years before. For the current admission, the physical examination showed a left lower extremity with non-pitting edema, 2+ pulses, warmth, and purplish-blue discoloration. Non-invasive vascular ultrasound of the left lower extremity showed an extensive, acute-appearing deformity thrombus involving the left common iliac vein, external iliac vein, common femoral vein, and deep femoral vein. Acute thrombus as well as May Thurner Syndrome was also seen on CT-Abdomen Pelvis.

The patient was treated by interventional radiology. They performed a left common iliac vein recanalization and reconstruction, a left external iliac vein, common femoral vein, and superficial femoral vein thrombectomy with on-table hemolysis (Using heparin and tenecteplase) and left common iliac vein angioplasty and stenting. The post-procedure iliofemoral vein ultrasound demonstrated near complete resolution of the thrombus. The patient was then placed on 1mg/kg of enoxaparin BID for the next 24 hours while being observed at the hospital, and discharged with a prescription for prophylactic apixaban.

Discussion

MTS is anatomic variation in which the left common iliac vein is compressed by the right common iliac artery. The prevalence of this condition is difficult to estimate because most patients are asymptomatic. Recent studies suggest that it occurs in 2-5% of patients undergoing DVT evaluation, and is even more common in women.³

Most DVTs are treated with standard anti-coagulation and thrombectomy as needed. This approach is not suitable for May Thurner patients with proximal lower extremity DVTs, however, because it does not address the underlying cause. The compression of the left common iliac vein in May Thurner patients contributes to stasis which increases their risk of clot formation, consistent with Virchow's triad. It is critical that these patients are recognized so that their acute thrombus can be lysed, and a prophylactic endovascular stent can also be placed.

MTS causes scar formation such that angioplasty alone for DVT treatment is not effective. Gao et al studied MTS patients who did not benefit from catheter-directed thrombolysis with a steady infusion of low-dose urokinase. When these patients were subsequently treated with low-dose urokinase infusion, endovascular stent placement, and percutaneous angiography, 81% achieved significant (28% complete and 53% partial) recanalization. Patency was maintained in 67% of veins 3-24 months later.² Because the May Thurner patient in this case did not have a stent placed after her first DVT, it is not surprising that she developed another DVT on the same leg, and fortunate that her condition was recognized so a stent could be placed.

Phlegmasia Cerulea Dolens, or "painful blue edema," is an uncommon complication of DVTs that is more common in patients with May Thurner Syndrome. It threatens both life and limb with limb amputation rates of 12-50% and mortality rates of 20-40% once diagnosed.³ The most severe complication of Phlegmasia Cerulea Dolens is venous gangrene that can cause a fatal blood stream infection. This condition is particularly problematic in patients with other risk factors for DVT including those who are pregnant, have cancer, smoke, or have stasis promoting anatomic variations such as May Thurner syndrome.³ For all patients with Phlegmasia Cerulea Dolens, prompt recognition that reverses the ischemia and prevents future occlusions is critical.

References

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Figures



Fig 1: Four hours after patient presented to the ED, before thrombolytic therapy and endovascular stent placement.