Phlegmasia alba dolens presenting in plasma donor with underlying May-Thurner syndrome

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Background

Phlegmasia alba dolens is a rare but serious complication of deep vein thrombosis (DVT) involving iliofemoral or iliocaval veins, when clot burden is extensive enough to cause threat for limb ischemia, appearing as a “painful white leg”. With phlegmasia alba dolens, collateral vein supply is sufficient to prevent ischemia. However, progression into the more serious phlegmasia cerulea dolens may occur, where nearly all venous supply is occluded causing limb-threatening ischemia. Patients with increased clotting risk are at greatest risk for developing this rare manifestation, including those with May-Thurner syndrome, an anatomical variant where the right iliac artery compresses the left iliac vein, causing extrinsic venous compression resulting in slowed blood flow. While blood and plasma donation is usually a safe process, there have been multiple cases of thrombotic events in patients with increased underlying clotting risk, now including our case of May-Thurner syndrome.

Case Presentation

A 19-year-old male presented with left leg swelling, pain and discoloration, consistent with phlegmasia alba dolens. He reported a history of donating plasma twice weekly for the past year. Venous leg ultrasound revealed extensive and occlusive deep vein thrombosis throughout the distal inferior vena cava, common iliac, common femoral, femoral, popliteal and saphenous veins. He was started on heparin infusion. Patient underwent mechanical thrombectomy and venogram with interventional radiology during which he was found to have anatomy consistent with May-Thurner syndrome. He later underwent workup for hypercoagulability which was non-revealing. His DVT is thought to be provoked from his underlying May-Thurner syndrome.

a. Left leg with phlegmasia prior to thrombectomy

May-Thurner syndrome creates an inherent increased risk for DVT.

Plasma or blood donors should consider screening for underlying clotting risk factors prior to regular donation.

Conclusions

While blood and plasma donation does not typically increase clotting risk, there have been reported cases of plasma donors developing associated thrombotic events, including upper arm DVT and stroke. Some were later identified with underlying genetic hypercoagulability. Similarly, in our patient, having MTS increased his baseline risk for DVT. However, plasma donation may have further provoked this risk. One theory to be further investigated is a possible transient depletion of anti-thrombotic factors. Overall, plasma and blood donors may benefit from screening for underlying risk factors prior to regular donation.

Resources

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