Double Trouble: A Case of Secondary Evans Syndrome from Systemic Lupus Erythematosus with May-Thurner Syndrome

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Objective:

Provide a concise analysis in assessing a female patient with hypercoagulable autoimmune disorder with May-thurner syndrome, highlighting its impact on exacerbating the thrombotic condition and underscore the imperative for additional imaging workup to prevent diagnostic oversights.

Methods:

A young female with a sudden onset of swelling, intractable pain, and skin mottling of the left leg. She also had a miscarriage at 20 weeks of gestation on her 1st pregnancy. DVT screening revealed acute DVT from external iliac vein to popliteal vein of the left leg. Further investigation showed anemia and thrombocytopenia, along with positive lupus anticoagulant and anti-phospholipid antibodies. Given a positive antiglobulin test and presence of thrombocytopenia, she was diagnosed with Evans Syndrome secondary to Systemic Lupus Erythematosus. The presentation of acute DVT at the left lower extremity of a female patient prompted a CT venogram which revealed compression of the left common iliac vein by the right common iliac artery and thrombus distal to the compression area consistent with May-Thurner syndrome.

Results:

Managing a patient with acute DVT, hemolytic anemia, and thrombocytopenia in a resource-limited setting presents challenges. The more common approach in the Philippines is surgical thrombectomy with possible veno-venous bypass. A multidisciplinary team opted for an endovascular approach of the patient in a situation where blood conservation is imperative. Due to logistics problem, procurement of the endovascular thrombectomy device was made available after 1 month post symptom onset. Despite the risk of technical failure, it was prudent to try to recanalize the iliofemoral system. The procedure included endovascular thrombectomy, angioplasty of iliac and femoral veins, and deployment of a venous stent at the iliofemoral stenosis. Postoperatively, the patient received low molecular weight heparin which eventually transitioned to warfarin, with significant reduction in thigh and leg circumference within 1 day postoperatively from 58cm to 47cm. Surveillance at 1 month post-procedure showed patent stent and iliofemoral veins, with complete resolution of leg edema and healed wounds, affirming t he success of the intervention.

Conclusion:

May-Thurner syndrome should be part of a workup in a female patient presenting with acute DVT of the left lower extremity. Consider endovascular pharmacomechanical thrombectomy with venous stent placement, even after 1 month from presentation, to reduce venous obstruction and lower post-thrombotic syndrome incidence.