Spontaneous Iliac Vein Rupture due to May-Thurner Syndrome and its Staged Management

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This is the author's manuscript of the article published in final edited form as:

Ingram, M., Miladore, J., Gupta, A., Maijub, J., Wang, K., Fajardo, A., & Motaganahalli, R. (2019). Spontaneous Iliac Vein Rupture Due to May-Thurner Syndrome and Its Staged Management. Vascular and endovascular surgery, 53(4), 348-350. https://doi.org/10.1177/1538574419831488

Abstract: We present a case of a 58-year-old otherwise healthy women who presented with left lower extremity deep venous thrombosis (DVT) and was found to have pulmonary embolism along with a ruptured left internal iliac vein. Our patient was hemodynamically stable upon presentation; therefore, a staged approach was undertaken. Initially, an inferior vena cava (IVC) filter was placed and the patient was slowly advanced to therapeutic anticoagulation and subsequently discharged. She then returned two weeks after discharge for venogram, mechanical thrombectomy and stenting. At 1 year follow-up in clinic she was found to have patent stents and resolution of symptoms.

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Introduction: Robert May and Josef Thurner first described left iliac vein compression by the right iliac artery and fifth lumbar vertebrae in 1957. They hypothesized that the mechanical compression and pulsating artery eventually contribute to venous obstruction [1].This syndrome is commonly described in young to middle-aged women and especially after multiple pregnancies. Complications of May-Thurner Syndrome are often deep vein thrombosis and pulmonary embolism. While May-Thurner's syndrome presents with deep vein thrombosis with or without pulmonary embolism, spontaneous iliac vein rupture remains a very rare complication. Our case report adds to the understanding of this unusual pathology and its management using a staged treatment approach.

Case: Informed consent has been obtained from the patient (or patient's guardian) for publication of the case report and accompanying images

A 58-year-old otherwise healthy woman with no previous history of venous symptoms presented to the emergency department (ED) complaining of syncope, sudden onset lower abdominal pain associated with rapid swelling and erythematous discoloration of her left leg. Two days prior to presentation the patient had been moderately exercising when she experienced acute pain in the left lower abdomen, left hip and groin area. In the ED, computed tomography (CT) of the chest, abdomen, and pelvis with contrast demonstrated findings concerning for May-Thurner syndrome with compression of the left iliac vein and associated massive deep venous thrombosis including the left common iliac vein, left internal iliac vein, and common femoral vein, femoral vein and popliteal vein. It also demonstrated a moderate sized hematoma in the pelvis (fig 1) concerning for venous rupture and a left subsegmental pulmonary embolism. A lower extremity duplex exam showed incompressible left femoral, deep femoral, popliteal, gastrocnemius, and posterior tibial and peroneal veins along with a partially compressible left common femoral vein. She was hemodynamically stable with edema of her left lower extremity. She did not have any physical exam signs concerning for

phlegmasia. Given the findings concerning for iliac vein rupture with massive deep venous thrombosis as well as pulmonary embolism, the decision was made to insert an inferior vena cava (IVC) filter to prevent further embolic events. She was monitored in ICU for further bleeding over next 48 hours. We initiated low dose heparin anticoagulation after 24 hours of initial presentation with gradual advancement of the dose of anticoagulation to attain a full therapeutic goal at discharge. The patient remained hemodynamically stable throughout and was discharged on the 4th day on low molecular weight heparin. Her left limb swelling, and symptoms of heaviness continue to be present. Plans were made to follow up as an outpatient for a mechanical thrombectomy two weeks after initial presentation.

Fourteen days after initial presentation, the patient underwent a trans-popliteal vein access venogram, mechanical thrombectomy with 8 Fr Indigo® system (Penumbra Inc, Alameda, CA) and intravascular ultrasound guided (IVUS) left iliac venous stenting. During the procedure, significant clot burden (Fig 2) was removed from femoro-popliteal and iliac veins and completion venogram demonstrated patency of the proximal femoro-popliteal system with filling defect within the left common iliac venous system. Intravascular ultrasound (IVUS) confirmed compromised left common iliac vein lumen. Two 16 x 90 mm Wallstents (Boston Scientific, Marlborough, MA) were placed in the left iliac venous system after pre-dilation with a 6 x 100 mm balloon. Post-dilation was performed with a 12-mm balloon. A small caliber balloon was intentionally used due to underlying concern for venous rupture. At completion, IVUS and venography demonstrated a persisting filling defect at the distal end of the stent and the venous access sheath was left in place to continue local anticoagulation. On postoperative day one, venous duplex demonstrated residual thrombus within the femoral vein and total occlusion of distal deep veins. The decision was made to return to the operating room to initiate chemical thrombolysis. A thrombolytic catheter was placed and remained in place for 24 hours for low dose catheter directed thrombolysis for 12 hours. Completion venography revealed resolution of filling defects in the femoral vein with a widely patent iliofemoral venous system (Fig 3). The patient was discharged on oral anticoagulation therapy. She subsequently followed up in clinic with a duplex study demonstrating patency of the stents and left lower extremity veins with no residual thrombus. In the next 6 weeks she also underwent removal of

the inferior vena cava filter. Venography at the time of removal of IVC filter showed patent stents with no residual thrombus (Fig 4) in IVC as well as in Iliac-femoral veins . Patient had resolution of her symptoms and remains symptom free with patent stents at the end of 1 year.

Discussion: May-Thurner Syndrome (MTS) is a rare, but well documented syndrome caused by the compression of the left iliac vein by the overlying right iliac artery. This leads to stasis in the iliofemoral deep venous system which can progress to deep vein thrombosis (DVT). While the exact incidence of MTS is unknown, it is estimated to occur in 2-5% of individuals who present with a lower extremity venous disorder [2]. Relatively common complications of MTS are DVT and pulmonary embolism (PE). Spontaneous left iliac vein rupture is a rare complication of MTS. Per Hosn et al there have only been 48 reported cases of spontaneous iliac vein rupture [3]. The spectrum of presentation is variable with some patients presenting with mild symptoms and others presenting with hypovolemia and shock [4]. The dichotomy presented by controlling the bleeding from the ruptured vein while also treating the DVT and associated pulmonary embolism makes management of this condition difficult. Additionally, an open approach is not without significant morbidity and mortality. Lin et al describe a 27% perioperative mortality [5] and Tannous et al describe a similar 29% mortality [6]. Jiang et al found a 16.7% operative mortality with associated 50% postoperative morbidity rates [7]. Our patient at presentation had intra-abdominal bleeding with massive DVT and PE. We staged her treatment by addressing the most pressing issue in her case. We placed an IVC filter to reduce the risk for further pulmonary embolism as she had contraindication for immediate anticoagulation. We also desired to reduce the risk of the venous stasis syndrome by performing definitive treatment of the DVT. In their 25 year retrospective review Mohr et al found the cumulative incidence rates of venous stasis syndrome to be 7.3%, 14.3%, 19.7%, and 26.8%, at one-year, 5-year, 10-year, and 20-year, respectively [8]. Additionally, due to the low prevalence, definitive management strategies do not exist for spontaneous iliac rupture. In our case, the patient presented in a hemodynamically stable condition which allowed us to utilize a staged approach. As the patient continued to remain stable, the decision was made to proceed with low dose anticoagulation to prevent further thrombus formation while also remaining vigilant for any concern for ongoing hemorrhage and hemodynamic instability.

Conclusion: This report provides information and makes a case for staged management, balancing the management of deep venous thrombosis with risks of bleeding from a spontaneously ruptured vein. We add to the existing evidence that in a hemodynamically stable patient, a conservative approach of delayed thrombectomy and recanalization is a reasonable decision to reduce mortality and long-term morbidity associated with chronic venous insufficiency.

Funding : This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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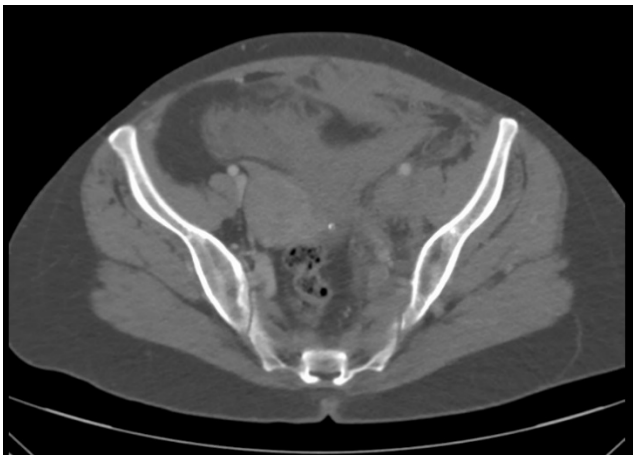

Fig 1. CT scan showing L iliac rupture with retroperitoneal hematoma

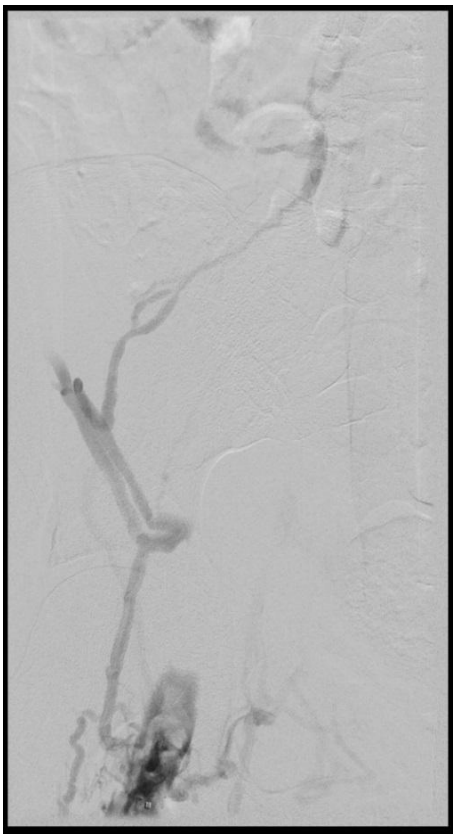

Fig 2. Femoro-popliteal vein venogram prior to intervention

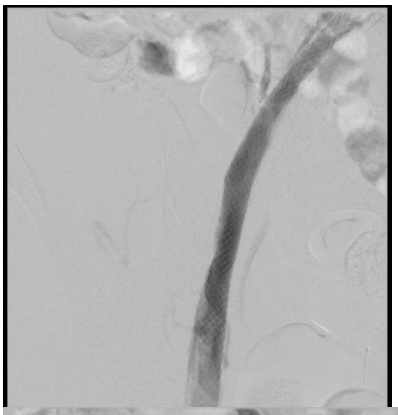

Fig 3. Completion iliac stent demonstrating resolution of thrombus

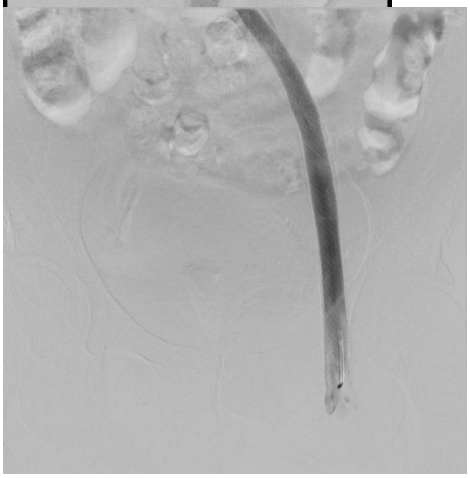

Fig 4. Angiography at 2 months follow-up demonstrating continued patency of left iliac vein