Iliofemoral Thrombosis Secondary to Iliac Compression Syndrome in Double Inferior Vena Cava

See-Wei Low, MD1; Kapildeo Lotun, MD2; Kwan Seung Lee, MD2
From the 1University of Arizona and 2Section of Vascular Medicine, Sarver Heart Center, University of Arizona, Tucson, Arizona.

ABSTRACT: Duplication of the inferior vena cava (D-IVC) joined by a pelvic connector is a rare congenital anomaly. May-Thurner syndrome (MTS), also known as iliac compression syndrome, is caused by a localized stricture at the point where the left common iliac vein is crossed by the right common iliac artery resulting in left common femoral deep venous thrombosis. The combination pathology of D-IVC and MTS is rarely described. We describe a case of a patient with iliofemoral thrombosis precipitated by MTS with D-IVC, heterozygous mutation of factor V Leiden, and describe our successful treatment approach.

VASCULAR DISEASE MANAGEMENT 2015;12(4):E54-E61
Key words: deep venous thrombosis, thrombectomy, interventional cardiology

Duplication of the inferior vena cava (D-IVC) is an uncommon congenital anomaly with an incidence of 0.3% to 3.0% in the population.1 May-Thurner syndrome (MTS), which is also known as iliac compression syndrome, is a rare condition that results in compression of the left common iliac vein by the right common iliac artery, crossing anterior to the vein.2 This narrowing of the left common iliac vein is a significant risk for developing deep venous thrombosis (DVT), especially of the left lower limb. We describe successful treatment of a very rare combination of pathology of D-IVC, heterozygous mutation of factor V Leiden and MTS in a case of extensive left lower limb DVT.

CASE PRESENTATION
A 30-year-old female presented with a 1-week history of left lower extremity swelling, pain, and erythema. The pain was continuous and dull in nature with pain score of 9/10. The pain was aggravated by movement and slightly relieved by rest and cold compression. She did not experience any shortness of breath, chest pain, or fever. On physical examination, she was hemodynamically stable and was not in distress. Her left lower extremity was warm, erythematous, swollen, and tender. Her risk factors for DVT included a significant family history of recurrent DVT and use of oral contraceptive pills for 1 year. There was no history of trauma or recent long-distance travel. Her complete
blood count and basic metabolic profile were within normal limits. However, her prothrombin and activated prothromboplastin times were 14.8 seconds and 36.9 seconds respectively, with an INR of 1.2. Her fibrinogen level was significantly elevated at 856 mg/dL.

Initial imaging with lower extremity Doppler revealed a large, occlusive DVT extending from the left tibial vein to the left common iliac vein. There was also evidence of D-IVC on the initial Doppler imaging study. A computed tomography (CT) scan confirmed D-IVC and a filling defect in the left common iliac vein and no pulmonary embolism. Screening for thrombophilia revealed a heterozygous mutation for factor V Leiden. She was started on anticoagulation therapy with intravenous heparin but continued to have significant pain and swelling of the leg with preservation of arterial pulses.

Considering the thrombus burden and increased risk of post-thrombotic syndrome, a decision to perform venous thrombectomy was made. Pelvic iliac venography was performed, which showed the D-IVC anomaly. A connection was noted between the right common iliac vein and the left inferior vena cava (IVC). A decision was made at this point to deploy IVC filters into both right and duplicate left IVC in the infrarenal positions, as there was dynamic flow between the left IVC into the right IVC at both iliac and suprarenal levels. There was also left thoracic azygos flow from the left IVC. Two OptEase filters (Cordis Corporation) were deployed, with one in the left IVC and another in the right IVC in the infrarenal position. Venography performed after the deployment of filters confirmed the appropriate deployment level. Access was then obtained at the left posterior popliteal vein.

**Figure 1.** Computed tomography of pelvis (axial plane) showed compression of the left common iliac vein between the left common iliac artery and lumbar vertebrae.

**Figure 2.** Venography showed duplicated inferior vena cava anomaly with communication above pelvic level and confluence at L1, suprarenal position.
and a 5 Fr Cragg-McNamara thrombolytic infusion catheter (Covidien Inc) was advanced into the left common iliac position. Unfractionated heparin followed by tissue plasminogen activator (tPA) infusion (2mg/hour for 6 hours followed by 1mg/hour) was administered via the Cragg-McNamara catheter in an ICU setting for 24 hours.

She was then brought back to the catheterization laboratory the next day for venography and her residual thrombus burden was assessed. Venography was performed with demarcation of the limits of residual thrombus and showed some improvement in blood flow. An 8 Fr, 30 cm therapeutic length Trellis device (Covidien Inc) was then advanced with the distal port in the left common iliac vein and proximal port in the left deep femoral vein. Trellis thrombectomy was performed for 10 minutes with a total of 10 mg of tPA administered. Thrombus material was aspirated. The therapy was once again repeated, from the left deep femoral vein to the left popliteal vein. Final images were obtained which showed marked improvement in blood flow. Her left lower extremity swelling along with symptoms of DVT significantly improved within 24 hours and she was discharged home after being started on warfarin oral anticoagulation therapy.

Three weeks later, she was brought back to the cardiac...
Venography revealed re-occlusion of the left iliac system secondary to thrombus. A decision was made to proceed with balloon angioplasty of the left iliac system. After multiple balloon dilatations, intravascular ultrasound (IVUS) was performed, which revealed compression of the left iliac vein by the right iliac artery. Two self-expanding 12 mm x 60 mm and 10 mm x 60 mm Protégé GPS self-expanding stents (Covidien Inc) were then deployed into the left common iliac venous system. There was excellent flow through the stents via both wide-open bilateral IVC and pelvic connectors. We chose not to remove the IVC filters at this juncture because of the presence of extensive clot burden. The pelvic connector between left IVC and right iliac vein was noted to be behind the aortoiliac bifurcation and was compressed. IVUS of the pelvic connector was performed, which confirmed the suspicion of compression. After performing balloon dilatation of the compression, a 12 mm x 40 mm Protégé GPS self-expanding stent was deployed which subsequently showed excellent flow in the connector and in bilateral IVC.

Two weeks later, she underwent venous Doppler,
which revealed normal flow in bilateral IVC, the iliac veins and bilateral lower extremity. She also underwent repeat venography, which confirmed patent stents with good blood flow through the left iliac system and no recurrence of thrombus formation. Both IVC filters were removed. She was continued on warfarin anticoagulation and was followed up in an outpatient clinic after 1 month without recurrence of symptoms. One year later, she became pregnant and carried the pregnancy with no complications under the care of hematology, obstetrics and gynecology, and cardiology. During her pregnancy, she was treated with enoxaparin sodium injections daily and this was later substituted with heparin in the 36th week of her pregnancy. She had an uneventful pregnancy and delivery with no recurrence of DVT. At 2 years, she has remained asymptomatic with no recurrence.

**DISCUSSION**

We highlight a case of a patient who presented with DVT in her left lower leg, who was subsequently found to have D-IVC, MTS, and a heterozygous mutation of factor V Leiden. A D-IVC arises from persistence of both right and left supracardinal veins.\(^3\) In D-IVC, a left-sided IVC ascends to the level of the renal veins to join the normal right IVC along the right side of the spine through a vascular structure, in this case through a pelvic connector that may pass either anterior or posterior to the aorta at the level of the renal vein.\(^4\) D-IVC is an uncommon congenital anomaly with an incidence of 0.3% to 3.0% in the population.\(^1\) It can cause major venous hemorrhage during vascular surgeries, thromboembolic complications, tumor extension, and diagnostic error.\(^5\) D-IVC is usually
discovered incidentally during a radiologic work-up via CT, MRI, or during abdominal surgeries. Some studies have identified D-IVC to be a risk factor for DVT in patients younger than 30 years of age with DVT by promoting venous stasis.

It is clinically important to recognize the presence of D-IVC in the therapy of DVT with IVC filters because connectors are often present between both, necessitating the accurate placement of IVC filters to prevent pulmonary embolism. The typical site of IVC filter placement is at the level of the L3 vertebral body, caudal to the renal veins. However, in some cases, suprarenal (T12-L1) placement of the IVC filters is indicated, such as in the presence of thrombus in the renal veins, with poor placement of infrarenal filter, or in pregnant patients. Indications for use of IVC filters are protection against recurrent DVT when there are contraindications to anticoagulation therapy or failure of anticoagulant therapy. The use of retrievable IVC filters as compared to permanent IVC filters lowers the risk of filter thrombosis and occlusion, filter migration, filter fracture, and vena caval thrombosis.

May-Thurner syndrome is also known as iliac vein compression syndrome and is characterized by the occurrence of left common iliac obstruction secondary to compression of the left iliac vein by the right common iliac artery. May-Thurner syndrome is seen in 22% of autopsies. However, this syndrome is often under-recognized as a cause of left iliofemoral DVT. Failure to diagnose MTS increases the patient’s risk of recurrent DVT and post-thrombotic syndrome because sole treatment using anticoagulation has been proven ineffective in patients with underlying iliofemoral venous obstruction. In young patients who have DVT with underlying MTS, it is recommended the patient be treated with thrombolysis or mechanical thrombectomy combined with angioplasty and stenting of the...
iliac vein stenosis in order to reduce the risk of post-thrombotic syndrome. The additional use of stenting or catheter-guided thrombolysis and angioplasty is to achieve long-term patency of the iliac vein, to reduce the recurrence of thrombosis.

In addition to her risk factors, the presence of heterozygosity for factor V Leiden mutation along with the intake of oral contraceptive pills contributed to her thrombotic risk. Patients with factor V Leiden mutations have an increased risk of DVT approximately 7 times that of noncarriers. This risk is further increased by a factor of 35 when taking oral contraceptive pills.

This is a rare case that illustrates successful treatment with catheter-directed thrombolytic therapy, adjunctive mechanical thrombectomy, angioplasty, and stenting as combination therapy for extensive iliofemoral thrombosis secondary to MTS in our patient with D-IVC and heterozygous mutation of factor V Leiden and highlights the special considerations necessary in successful care of this combination.

Editor’s note: Disclosure: The authors have completed and returned the ICMJE Form for Disclosure of Potential Conflicts of Interest. The authors report no disclosures related to the content herein.

Manuscript received November 12, 2014; manuscript accepted February 20, 2015.

Address for correspondence: Dr. Kwan S Lee, 2800 E Ajo Way, Tucson, AZ 85713, United States. Email: klee@shc.arizona.edu

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