Unusual case of left iliac vein compression secondary to May-Thurner syndrome and crossed fused renal ectopia

Mohamed A. Elsharawy, MD, FRCS, *Khaled M. Moghazy,* MS, MD, *Hind S. Alsaif,* SSCRad, JDR, *Mosad M. Al-Asiri,* MD.

ABSTRACT

الضغط الخارجي على الوريد الحرقفي الأيسر ضد الفقرة القطنية الخامسة بواسطة الشريان الحرقفي الأيمن متلازمة مي و ثيرنر(May and Thurner syndrome) تعتبر متغيره تشريحية معروفه ويتسبب هذا الضغط في تخثر الوريد الحرقفي الأيسر . في هذه التقرير نعرض حالة نادرة لمريض يعاني من تخثر في الوريد الحرقفي الأيسر ممتدا حتى الوريد المأبضي وقد أثبت الفحصوصات الإشعاعية أن ذلك قد سببه كل من متلازمة مي و ثيرنر(May and Thurner syndrome) مصحوبة بكليتين ملتصقتين منتقلتين في الجانب الأيسر .تمت معالجة المريض بواسطة قسطرة مباشره لإذابة التخثر بالإضافة إلى العلاج المضاد للتجلط ، وقد تحسن المريض كثيرا، أثبتت التحاليل المخبرية بعدم وجود مرض دموي متسبب في التخثر.

External compression of the left iliac vein against the fifth lumbar vertebra by the right iliac artery (May and Thurner syndrome) is a well-known anatomic variant. We identified a rare case of May-Thurner syndrome associated with crossed fused renal ectopia on the left side. The patient presented with complete thrombosis of the left common iliac vein down to the popliteal vein. He was treated with catheter directed thrombolysis followed by anticoagulant therapy.

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From the Departments of Surgery (Elsharawy), and Radiology (Moghazy, Alsaif, Al-Asiri), King Faisal University, Al-Khobar, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Mohamed A. Elsharawy, PO Box 40081, Al-Khobar 31952, Kingdom of Saudi Arabia. Fax. +966 (3) 8966728. E-mail: elsharawya@yahoo.co.uk

May-Thurner syndrome is an uncommon disease in which the left common iliac vein is compressed against the fifth lumbar vertebra. The compression is usually caused by the right common iliac artery. Its pulsation effects result in intimal wall changes of the vein with secondary iliofemoral deep venous thrombosis (DVT).[1] This anatomical variant was first described by McMurrich in 1908,[2] and explained later through detailed anatomic, and histological studies by May and Thurner in 1957 who found such compression in 22% of autopsy series.[2] We identified a rare case of iliac vein compression (May-Thurner syndrome), which was aggravated by crossed fused renal ectopia on the left side. We presented this case to show such rare association.

Case Report. A 30-year-old Syrian male presented with a 3-day history of left leg pain and swelling. His physical examination revealed markedly swollen left leg with bluish discoloration. Ultrasound showed hypoechoic thrombus within the left femoral and popliteal veins, and non-compressible lumen with absent color flow (**Figure 1**). Contrast enhanced computed tomography (CT) of the abdomen and pelvis showed left crossed fused renal ectopia. The right common iliac artery was sandwiched between fused kidneys and left common iliac vein (**Figure 2**). The left common iliac vein was compressed with complete thrombosis. He was started on unfractionated heparin to maintain a therapeutic partial thromboplastin time of 2-2.5 times the upper limit of control. A thrombolysis catheter (Pulse Spray, Angiodynamics, USA) was placed in the popliteal vein extending across the thrombus for catheter directed thrombolysis. After a bolus dose of 100,000 units Streptokinase (Kabikinase, KABI), continuous infusion was started at 100,000 U/hr. Venography was performed at 12, 24, and 36 hours, and thrombolysis catheter was advanced. At 48 hours, the left lower limb venography showed blocked left common iliac vein with irregular filling defect in the left external iliac vein (**Figure 3**). After 72 hours of streptokinase infusion, almost all the thrombus was lysed, but there was narrowing of the vein where the iliac artery crossed it. Balloon angioplasty was performed. There was no residual stenosis, hence, no stent was placed. The catheter was removed, and low molecular weight heparin and warfarin were started, and transitioned to warfarin alone once the international normalized ratio (INR) reached 2. There was a small

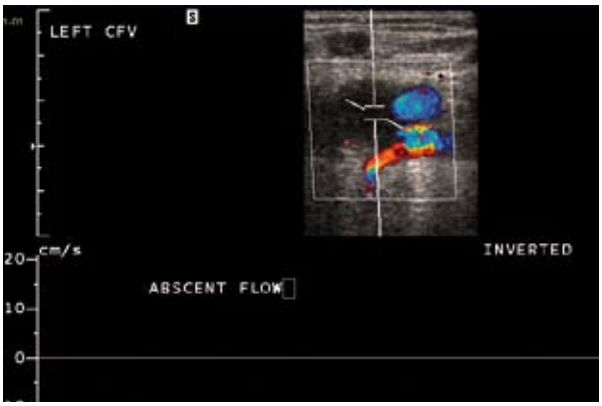

Figure 1 - Doppler study shows a hypoechoic thrombus within the left common femoral vein, with non-compressible wall and absent flow.

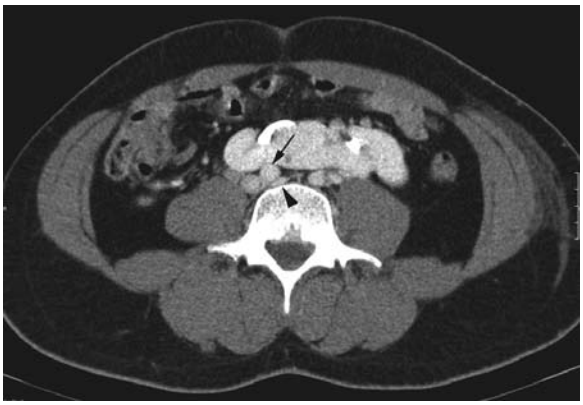

Figure 2 - Contrast enhanced CT of the abdomen showed left crossed fused renal ectopia, which may aggravate the compression of right common iliac artery (arrow) on left common iliac vein (arrowhead).

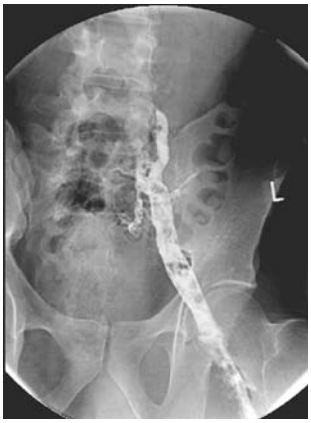

Figure 3 - Left lower limb venography showed blocked left common iliac vein with multiple irregular filling defects in the left external iliac vein.

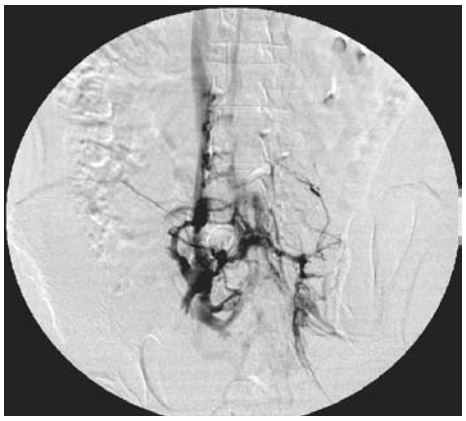

Figure 4 - Post-thrombolysis venography showed left common iliac vein had rethrombosed with extensive collaterals from left to right iliac veins.

subcutaneous hematoma at the site of catheter insertion, which was treated conservatively. There were no other complications other than discomfort at the catheter site. The swelling improved dramatically. However, check venography showed that the left iliac vein had rethrombosed, and there were extensive collaterals from left to right iliac veins (**Figure** 4). In view of the clinical improvement, no further action was carried out, and the patient was discharged on warfarin. On follow up at 6 months, there was mild leg swelling, but no pain. Warfarin was discontinued, and extensive laboratory evaluation for thrombophilia was performed. All laboratory results were normal.

Discussion. Venous stasis is a well-known cause of venous thrombosis. Extrinsic compression of the left common iliac vein between the vertebral body and the right common iliac artery, (also known as May-Thurner syndrome) is considered to cause 3 to 8 times more cases of iliofemoral deep vein thrombosis (DVT) on the left side than on the right side.[2] Approximately 50-60% of patients presenting with left-sided iliofemoral DVT have common iliac vein intraluminal webs resulting from extrinsic compression.[3] Among these abnormalities, extrinsic compression of the left common iliac vein, between the vertebral body and the right common iliac artery is the most common, and is the main cause of predominance of acute DVT in the left side, and mostly found in females. A gravid uterus,[1] tumors, aneurysms of the aorta or iliac arteries, cysts, and inflammatory processes including retroperitoneal fibrosis,[4] are other causes of symptomatic venous obstruction of the lower extremities. In addition to these known anatomic abnormalities, we found that extrinsic compression of the left common iliac vein between the vertebral body and the right common iliac artery, is aggravated by the presence of left crossed

fused ectopia. Until recently, there has been no real need to identify the presence or absence, and the cause of central venous obstruction in cases of acute iliofemoral DVT as DVT has been managed with systemic anticoagulation. Advances in catheter-directed endovascular treatment has made it important to understand the precise venous anatomy, extension of the thrombus, and presence of extrinsic causes of obstruction. Diagnosis of May-Thurner syndrome may not be easy. Doppler ultrasound is generally unable to visualize iliac vein compression. Contrast enhanced CT, has recently been suggested as a good method for detecting iliac vein compression, and identifying various extraluminal causes of venous stenosis or obstruction.[5] After catheter-directed thrombolysis, venography can accurately evaluate luminal changes, and evaluate the causes of stenosis or obstruction.[6,7] The treatment of May-Thurner syndrome has evolved over the years. Conservative treatment with compression stocking has been unsuccessful in most cases. Many surgical procedures for relief of obstruction, such as venovenous bypass, have been described with moderate success. Over the last decade, catheter directed thrombolysis has proved to be superior to anticoagulants alone.[8] In May-Thurner syndrome, some authors recommend that thrombolysis should be followed by angioplasty, and stent placement to prevent rethrombosis of the iliac vein.[6,7] However, stents were reported to have some complications such as stent thrombosis,[7,9] and migration into the right ventricle.[10] Although, the iliac vein was reported to be rethrombosed in some series, and in our patient pelvic collaterals were enough to maintain the patient asymptomatic. In such cases, additional treatment with stent placement may not be required.

In summary, a high index of suspicion of May-Thurner syndrome should be considered when a young adult presents with left sided iliofemoral vein thrombosis. Computed tomography with contrast can diagnose the compression by iliac artery or any other causes. Anatomic variations such as crossed renal ectopia may increase the compression effect of the right common iliac artery on the left common iliac vein, and may be the triggering factor in the development of deep vein thrombosis. Catheter-directed thrombolysis, and if necessary followed by angioplasty and stent is an effective, and safe treatment.

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