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Endovascular Management of May-Thurner Syndrome in a Patient with Left-Sided Superior Vena Cava: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 23
Final Diagnosis: May-Thurner syndrome
Symptoms: Claudication
Medication: —
Clinical Procedure: Venous stenting
Specialty: Interventional Radiology • Vascular Surgery

Objective: Congenital defects/diseases

Background: May-Thurner syndrome (MTS) is a condition characterized by compression of the left common iliac vein (LCFV) between the right common iliac artery (RCIA) and the lumbar vertebrae. This anatomical entrapment typically affects young women and is mostly asymptomatic. High index of suspicion is required in cases of recurrent left-sided deep vein thrombosis (DVT) and severe leg pain. We describe a case of MTS in a young male patient with a left-sided superior vena cava (LSSVC) that was successfully managed by an endovascular approach. To the best of our knowledge, the coexistence of MTS and LSSVC anomaly has not been reported previously.

Case Report: A 31-year-old man presented with a history of left-sided iliofemoral deep vein thrombosis and disabling venous claudication of 2 years' duration. Duplex ultrasound and computed tomography venogram (CTV) revealed evidence of MTS with chronic subtotal occlusion of the left common iliac vein (LCIV) with extensive venous collaterals. Venogram via the left femoral vein puncture confirmed the aforementioned findings. Retrograde recanalization of the occluded segment was attempted without success. Therefore, an antegrade approach via the right internal jugular vein was performed to facilitate recanalization. Surprisingly, venography revealed an LSSVC. The occluded CIV was successfully stented and the patient had complete resolution of his symptoms at 22-month follow-up.

Conclusions: MTS is a potentially treatable and often-overlooked pathology. In the era of expanded endovascular management of MTS, recognition of this coincidence is essential to prevent unwarranted mishaps during endovascular management when the jugular approach is used.

MeSH Keywords: Endovascular Procedures • Jugular Veins • May-Thurner Syndrome • Stents • Vena Cava, Superior • Venous Thrombosis

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Background

Iliac vein compression between the right common iliac artery (RCIA) and the lumbar spine was first described by May and Thurner in 1957 [1]. The incidence of MTS is about 20%; however, it is rarely symptomatic [2]. In patients with left lower-limb deep vein thrombosis (DVT), the incidence of MTS is about 20–50% [3]. It is believed that chronic repetitive microtrauma of the left common iliac vein (LCIV) between a static lumbar vertebra and a pulsating RCIA predisposes the patient to formation of a fibrotic spur with subsequent narrowing [3]. Such narrowing predisposes the patient to venous stasis, DVT, venous claudication, and lower extremity varicose veins [2,3].

When suspected, MTS is best diagnosed by intravenous ultrasonography [4]. Computed tomography venography (CTV) and magnetic resonance venography are also useful in establishing the diagnosis. Conventional transabdominal ultrasonography is not sufficiently sensitive for diagnosis of this condition [5].

MTS most commonly affects the left lower extremity; however, bilateral presentation is possible and is reported in 2–5% of MTS cases because of a high bifurcation of the aorta, resulting in compression of both sides [3,6]. Other anatomic variations of MTS have also been described [7].

Left-sided superior vena cava (LSSVC) is a rare congenital vascular anomaly with a prevalence of 0.3–0.5% in the general population [8]. It is the most common venous vascular anomaly in the chest, often asymptomatic, and found as an incidental finding during chest imaging or line placement [8,9].

LSSVC results from failure of the left superior cardinal vein to regress. Two possible drainage sites exist: either into the coronary sinus or directly into the left atrium. The drainage of the coronary sinus is usually into the right atrium; in such cases, there will be no shunting. However, in such a scenario, the venous intervention must be performed very carefully because unintentional perforation of the coronary sinus can be catastrophic [8,9]. Direct drainage of the LSSVC into the left atrium may result in a right-to-left shunt, which can cause paradoxical thromboembolism and air and septic embolisms [10]. Furthermore, LSSVC can be associated with other congenital heart diseases that can cause paradoxical embolization from intracardiac right-to-left, mainly via an atrial septal defect (ASD) [8,11]. In such a scenario, the recognition of coexisting MTS will be vital to prevent grave consequences. Indeed, these complications are rare, as these shunts are usually small and often clinically irrelevant [10].

To the best of our knowledge, the coexistence of LSSVC and MTS has never been previously reported. Herein, we report a case of MTS in patient with LSSVC that was successfully managed by venous stenting.

Case Report

A 31-year-old man presented with disabling venous claudication, progressive left lower-limb swelling, and varicose veins. The patient reported 2 episodes of left iliofemoral DVTs treated conservatively in the past at a district hospital; otherwise, the patient's past medical history was unremarkable. He had an average body build with a body mass index of 19.6 kg/m^2. Basic blood workup, metabolic panel, and coagulation values were within normal limits. Anticardiolipin antibodies and lupus anticoagulant were checked to rule out antiphospholipid syndrome, and their levels were normal. Protein S and protein C were also within reference ranges, and other thrombophilia tests were negative.

Clinical examination showed signs of chronic venous insufficiency on the left lower extremity, including varicose veins, edema, and hyperpigmentation of the skin without ulceration. There was no evidence of vascular anomalies in either the upper or right lower limbs.

Doppler ultrasonography revealed dilated left lower-limb superficial veins, incompetent left saphenofemoral valve, and evidence of old recanalized thrombosis of the left common femoral vein with extensive venous collaterals. Otherwise, the deep and superficial veins distal to the groin were patent and the saphenopopliteal junction was competent.

CTV revealed severe compression of the LCIV between the L5 vertebral body and the RCIA, stenosis of the left external iliac vein (LEIV), and extensive venous collaterals (Figure 1).

Management options were discussed with the patient, and he opted for invasive endovascular treatment because of his disabling symptoms. The procedure was performed in an angiographic suite (Innova™ 2100-IQ, GE Healthcare, Chicago, IL) with the patient in the supine position using a standard aseptic technique. The left CFV was accessed under ultrasound guidance, and a 6-Fr vascular access sheath (Terumo Europe NV, Leuven, Belgium) was inserted. The venogram showed narrowing of the LEIV and occlusion of the LCIV with extensive venous collaterals. The occluded segment was partially crossed with a hydrophilic guidewire (Roadrunner® PC Wire Guide, Cook Medical, Inc., Bloomington, IN) and an angled-tip multipurpose catheter (Beacon® Tip 5.0 Fr, Cook Medical, Inc., Bloomington, IN).

Entry into the inferior vena cava (IVC) was not achieved despite multiple attempts. The lesion of the LCIV was impassable, and the guidewire kept rolling backward, forming a loop, possibly due to a venous spur or scar formation in this region. We did not push the wire hard, as we felt that it might cause perforation and false passage out of the IVC (Figure 2).

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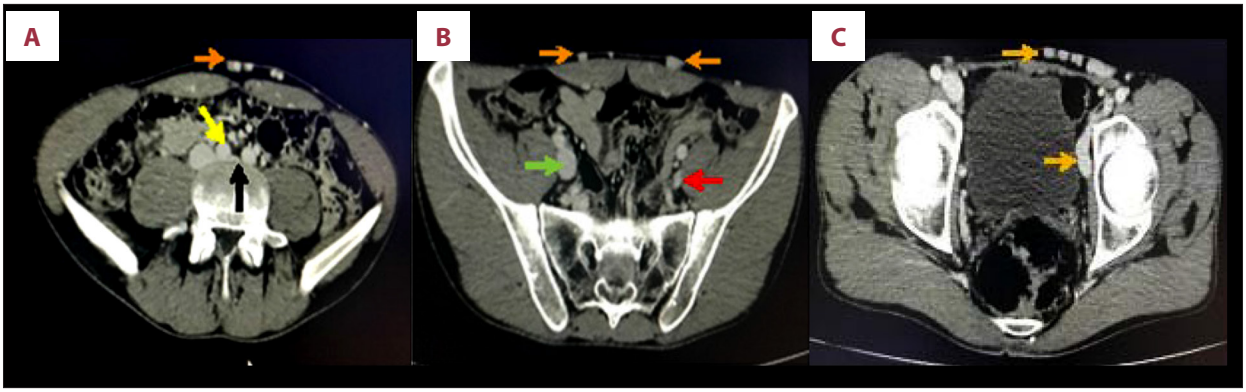

Figure 1. Pelvic CT scan in the venous phase showing (**A**) severe compression of the LCIV (black arrow) between the RCIA and the L5 vertebra, (**B**) narrowing of the LEIV (red arrow) in comparison with the REIV (green arrow), (**C**) extensive venous collaterals (small orange arrows). LCIV – left common iliac vein; LEIV – left external iliac vein; RCIA – right common iliac artery; REIV – right external iliac vein.

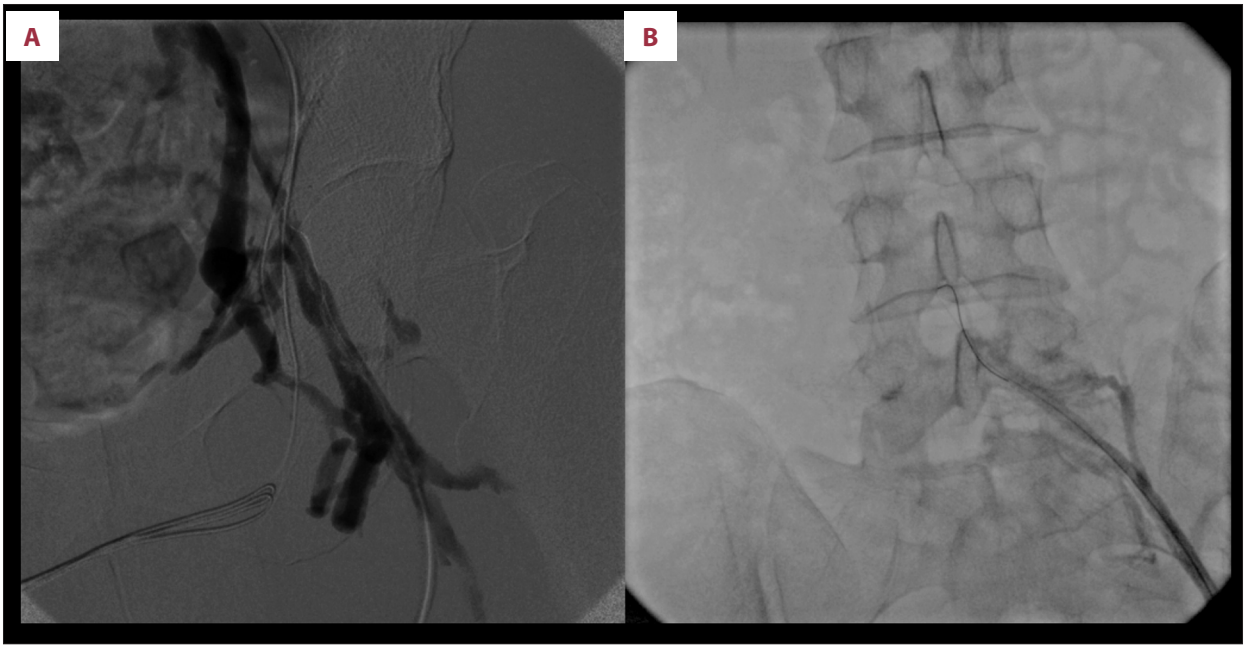

Figure 2. (**A**) Venogram through the left common femoral vein showing stenosis of the LEIV with venous collaterals. (**B**) Occlusion of the LCIV with the guidewire looping backward. LCIV – left common iliac vein; LEIV – left external iliac vein.

A decision was made to attempt crossing the lesion from the antegrade jugular approach and to insert a catheter into the distal IVC for guidance. The right internal jugular vein was accessed; surprisingly, the guide wire crossed into the left side of the chest. Venography was performed, showing an isolated LSSVC draining into the right atrium through the coronary sinus (Figure 3).

Using jugular access, we attempted to use a guidewire to cross the lesion from an antegrade approach; however, the catheter could not be engaged into the LCIV. Therefore, a guiding catheter was left inside the IVC near the ostium of the LCIV. The lesion was crossed from a retrograde approach through the LCFV, and an antegrade jugular approach was only used for marking the correct path into the IVC. After crossing the entry lesion, the occluded segment was initially dilated using an 8-mm angioplasty balloon (Armada 35 PTA Catheter, Abbott, IL), followed by successful deployment of a 16×120 mm, self-expandable Nitinol stent (Epic™, Boston Scientific, Marlborough, MA). The final venogram showed satisfactory angiographic results (Figure 4).

Cardiology consultation was conducted to exclude any possible associated anomaly with the LSSVC. All necessary investigations, including echocardiography and ECG, were normal. The patient was discharged the following day in good general

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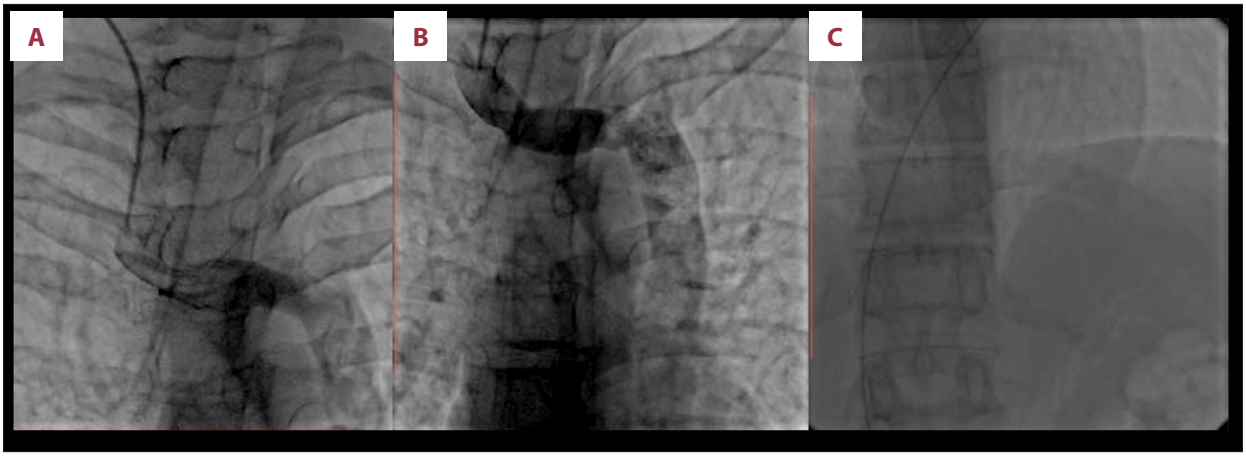

Figure 3. Venogram from the right internal jugular vein access showing (**A**) right brachiocephalic vein crossing the midline into the left side with (**B**) left-sided SVC. (**C**) Catheter crossing via the coronary sinus into the right atrium and back into the IVC. IVC – inferior vena cava; SVC – superior vena cava

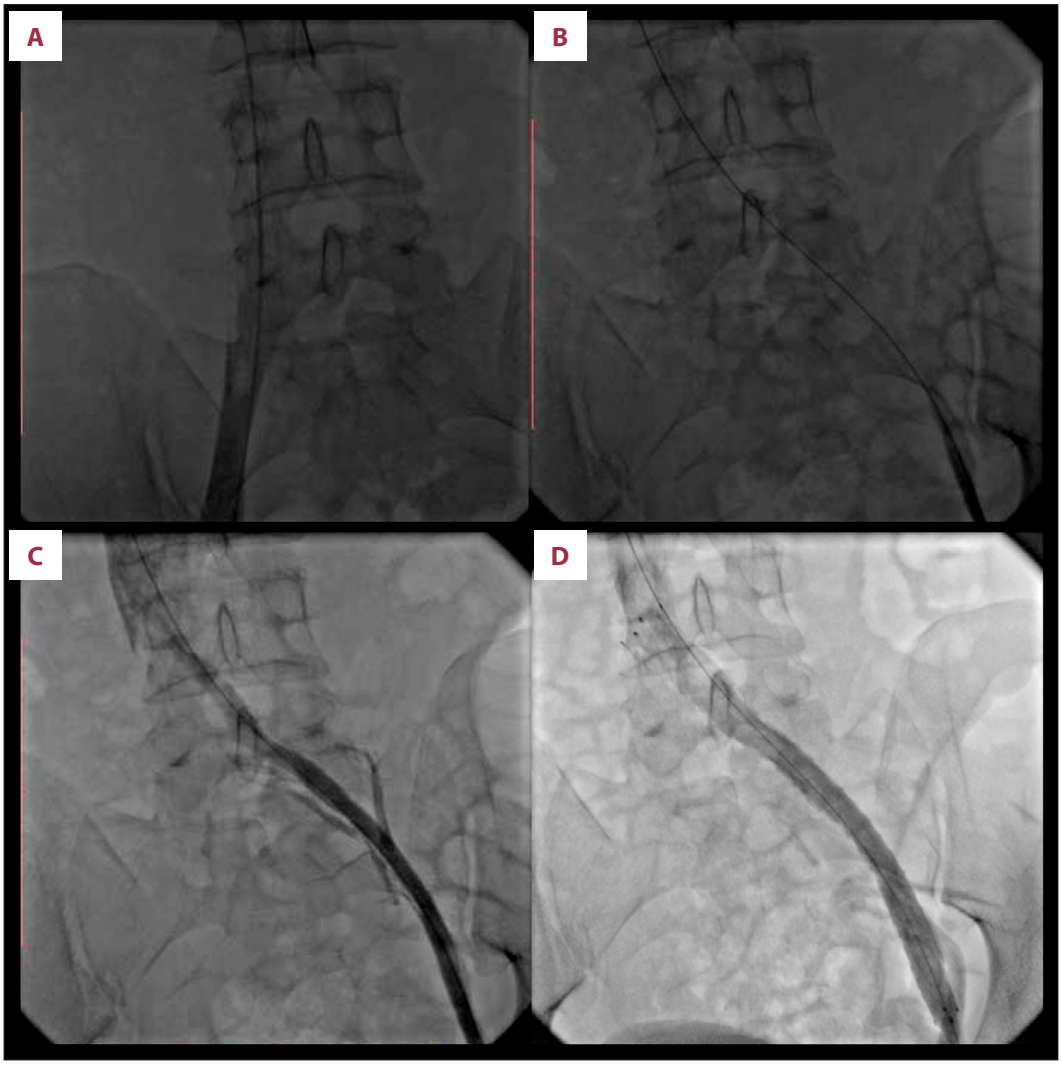

Figure 4. (**A**) Catheter inserted from the jugular approach down to the right external iliac vein. (**B**) The guidewire crossed the lesion and was successfully inserted into the IVC. (**C**) After initial balloon angioplasty. (**D**) Stent deployment. IVC – inferior vena cava.

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condition. He was given long-term oral anticoagulation therapy with warfarin. Regular follow-up in the clinic showed significant clinical improvement and Doppler ultrasonography confirmed patency of the stent at 22-month follow-up.

Discussion

Venous outflow obstruction due to MTS usually presents in the second or third decade of life and is more commonly seen in women [3,5]. Our patient was a young man who presented late to our vascular clinic with a post-thrombotic syndrome.

MTS can present acutely with iliofemoral DVT and should be managed aggressively by catheter-directed chemical thrombolysis or mechanical thrombectomy to prevent post-thrombotic syndrome [12]. Our patient had 2 episodes of DVT that were managed conservatively by his internist at another hospital. A high index of suspicion is needed for proper diagnosis of such entities. Young patients with acute DVT (especially idiopathic DVT) or chronic venous insufficiency should be referred to vascular centers for proper management.

Angioplasty followed by self-expandable iliac vein stent implantation is an efficient approach to resolve chronic symptoms of MTS. This approach has favorable technical success rates, with a 1-year patency rate of up to 94% [3,13]. Our patient was managed with venous stenting and showed significant clinical improvement with primary patency of the stent to date (22 months).

MTS is usually an isolated entity. One case of MTS associated with right-to-left shunt through a persistent foramen ovale (PFO) has been reported, in which DVT was the source of the embolus in paradoxical embolism [14]. MTS can be associated with thrombophilia, necessitating thrombophilia workup in all patients [15]. Our patient's thrombophilia workup was unremarkable, and he did not have other risk factors for his symptoms.

LSSVC can be seen in association with other congenital heart defects, including atrial septal defect, bicuspid aortic valve, coarctation of the aorta, coronary sinus ostial atresia, and cor triatriatum [8]. To the best of our knowledge, LSSVC has never been described in association with MTS. The LSSVC in our patient was not associated with any other cardiovascular anomalies.

Both MTS and LSSVC have been described in association with right-to-left shunt via PFO and ASD [8,11,13,14], and the coexistence of both anomalies in the same patient will be more relevant clinically and should not be overlooked. Furthermore, management of acute venous thrombosis in a similar sitting should not be attempted before excluding intracardiac shunting to avoid paradoxical thromboembolism.

The presence of LSSVC makes the placement of a catheter within the right side of the heart challenging. Inadvertent cannulation of the LSSVC without caution, even under fluoroscopy guidance, may result in severe complications. Possible mishaps include unusual catheter position, unintentional cannulation of coronary sinuses, and risk of cardiac perforation [8,9].

Our case highlights the importance of vigilance when attempting an antegrade transjugular approach for MTS management. Unforeseen complications can arise if its association with LSSVC is not considered.

Conclusions

MTS is a potentially treatable and often-overlooked pathology. In the era of expanded endovascular management for MTS, recognition of this coincidental finding is essential to prevent unwarranted mishaps during endovascular management when the jugular approach is used.

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Department and Institution where work was done

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Conflict of interest

None.

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