

CASE REPORT

May–Thurner syndrome: an often overlooked cause for refractory venous leg ulcers

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Key words

Chronic wound; Iliac vein compression syndrome; Iliocaval compression syndrome; May–Thurner syndrome; Venous leg ulcers

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Abstract

We report a 53-year-old female patient presenting with a refractory venous leg ulcer and unremarkable findings in the doppler Ultrasound venous mapping of the leg veins. Further comprehensive diagnostics demonstrated an underlying May–Thurner syndrome. After resolution of the primary mechanical obstruction, rapid wound healing in the following 3 weeks was documented. Iliac vein compression syndrome, commonly known as May–Thurner syndrome, is a distinguishable anatomical variant that results from an external compression over the left iliac vein exerted by the overriding adjacent right common iliac artery. It is mostly seen among young, healthy female patients and can easily be under-diagnosed. Lower extremities duplex ultrasonography remains the gold standard in diagnosing venous insufficiency, but it should not solely depend on it. Instead, clinicians should consider other possibilities, assessing the patency within the truncal veins, which in turn might contribute to the venous insufficiency along the lower limbs. An active early diagnostic approach can prevent significant overall morbidity and help patients to ease back into their daily-life activities. Therefore, it is recommended that all patients with suspected venous insufficiency and normal lower limbs duplex findings should undergo further evaluation of the truncal venous system pattern. May–Thurner syndrome, along with other causes of iliac veins compression, should be considered in the differential diagnosis in unclear persistent cases of unilateral venous symptoms.

Case report

A 53-year-old Caucasian female with a medical history of multiple sclerosis presented to our wound care unit for evaluation of a progressive chronic ulcer on her left lower leg. According to the patient, the ulcer was of 3 months duration, accompanied with moderate dull pain and swelling, exacerbated by prolonged standing. In a detailed anamnesis, the patient added that she underwent sclerotherapy for spider veins on the same affected limb 3 months earlier. Since then, she developed a non-healing wound. She applied different moist wound dressings in the period before she sought an expert consultation. With respect to her multiple sclerosis, her current medications were Prostanoids therapy, in addition to hydrocortisone, taken only at acute relapses.

[Correction added on 26 April 2017, after online publication: The author made corrections on the texts.]

Clinical examination

On clinical inspection and palpation at the first visit, there was a leg ulcer that measured approximately 5.6 cm² in diameter, with

Key Messages

- May–Thurner syndrome (MTS) is a frequently under-diagnosed cause of chronic venous leg ulcers
- young to middle-aged females presenting with unilateral persistent symptoms of CVI represent the target patient group with potential MTS
- an active approach in early diagnosis improves overall prognosis and prevents morbidity
- there exist limitations of ultrasound exploration in diagnosing truncal venous incompetence
- normal lower extremities venous duplex sonographic findings necessitate further truncal venous examination in the setting of unclear CVI cases



Figure 1 Non-healing venous leg ulcer, measuring 5.6 cm² in diameter of 3 months duration.

a pain score of 7 out of 10 points on the visual analogue scale (VAS). The ulcer was located on the left distal lower limb just below the medial malleolus. The ulcer bed was covered with fibrin as well as a small amount of necrotic tissue and was surrounded by sharply demarcated red inflamed borders (Figure 1). Besides a moderate perimalleolar oedema of the left lower leg, no other manifestations of chronic venous insufficiency (CVI) were noted. Arteria dorsalis pedis and posterior tibial pulses were palpable.

[Correction added on 26 April 2017: The lesion was mistakenly mentioned on the right leg. It should be on the left and this has been corrected in this current version.]

Differential diagnosis

A variety of pathologies can be considered in the differential diagnosis of chronic leg ulcers (1). However, from our clinical perspective and based on the patient's anamnesis of the presenting complaint, we considered amongst others venous leg ulcer, pyoderma gangrenosum or vasculitis in the differential.

Diagnostics

For the purpose of excluding inflammatory aetiologies or coexisting conditions, we run an extensive workup, including complete blood count, inflammatory markers, comprehensive metabolic panel, coagulation profile, urine and serum protein electrophoresis, anti-nuclear antibodies and anti-neutrophil cytoplasmic antibodies, which was within normal limits. On histopathological examination of a biopsy, signs of dense neutrophilic inflammatory infiltrates along with vasculitis were identified. Bacterial swab cultures found *Staphylococcus aureus*. In a lower extremities colour duplex ultrasound, no signs of chronic venous insufficiency (CVI) were found. The

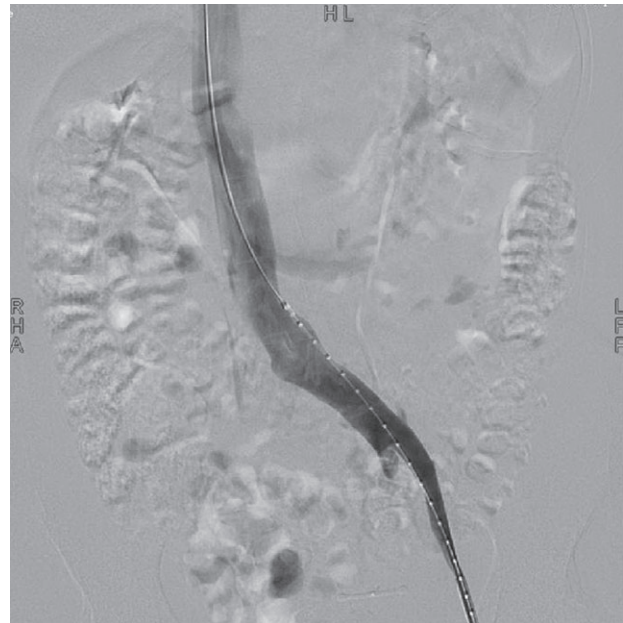


Figure 2 Contrast venogram shows the contrast flow across the confluence of the left common iliac vein into the IVC.

resting ankle brachial pressure index (ABPI) was 0.98. As part of the evaluation for any potential space-occupying lesions compromising the venous outflow, images using computerised tomography were obtained, showing no signs of masses in both the abdominal and the thorax cavities.

Therapy

The patient received prednisone initially for 4 days to suppress the painful inflammation caused by a secondary vasculitis. Non-steroidal anti-inflammatory drugs and opioids have been used to control the pain. Moreover, the local therapy regimen included moist wound care dressings, supported by consequent compression therapy consisting of daily applied short-stretch bandages.

Follow-up

During the follow-up visits, an increase in the circumference of the ulcer bed surface by almost 3 cm within 4 months after the initial presentation was noted (Figure 2). The outcome after 9 months of conventional therapy were not satisfactory, lacking significant improvement in the patient's pain and the healing progress of the ulcer. In addition to that, our patient developed a bluish discolouration of the left big toe in the process. At that stage, assessment using contrast venography for further clarification of her condition was advised, showing a severe degree of stenosis within the left common iliac vein. According to the observed radiological findings, the patient was proven to have the typical anatomical features of May–Thurner syndrome (MTS) along with a left ovarian vein reflux as part of an accompanied pelvic congestion syndrome. Therefore, an intravascular ultrasound-guided stent implantation as the treatment of choice was performed (Figures 2

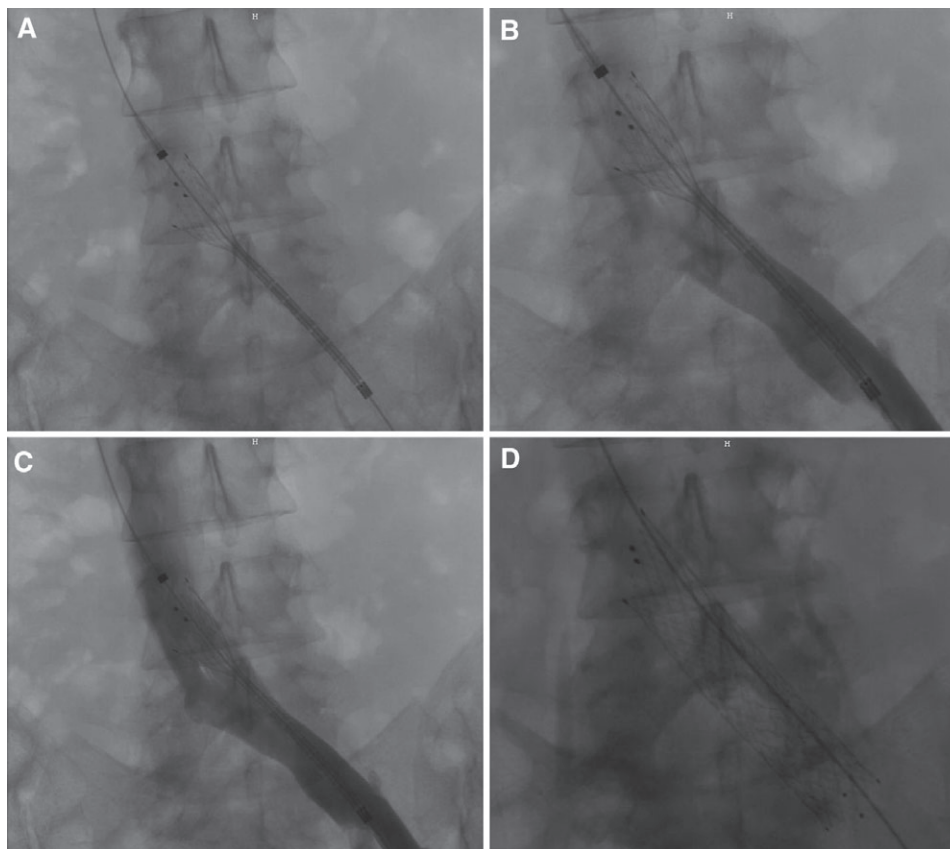


Figure 3 Images showing the preformed intravascular ultrasound (IVUS)-guided percutaneous transluminal angioplasty (PTA) with stenting implantation.

and 3) followed by coil embolisation and sclerotherapy of the left ovarian vein. As a result, venous system patency was effectively restored. No complications after the procedure were reported.

Subsequently, patient's standard care measures continued in the same manner used before the intervention. At the patient's follow-up appointment 3 weeks after the executed stent placement therapy, significant outcomes were documented. The ulcer measured 0.6 cm² in diameter; which represents an approximately 90% decrease in wound surface area. Complete reepithelialisation of the ulcer by secondary intention was reported by our team within 8 weeks.

Discussion

Non-healing leg ulcers create a significant socioeconomic burden on health systems both as providers and the patients themselves, impacting the quality of their daily life (2–5). Reported estimations of the mean cost of ulcer treatment in Germany in recent years were exceeding 9000 Euros per year (6). Currently, there are a lot of advances in the treatment strategies used; however, these different methods would not be effective for all the cases unless the underlying cause is identified and managed. In fact, these different investigations and treatment plans should be tailored to each case individually in a scheme-based approach. Ascertaining the nature of the underlying pathology plays a critical role in defining the entity of the wound in order to provide

the optimal treatment accordingly to insure a progressive healing rate and prevent further evitable complications.

The Bonner venous study carried out in Germany suggested that approximately 0.7% of the adult population suffering from venous leg ulcers (7). Our group demonstrated that in Germany, the majority of chronic leg ulcers, accounting for almost 65%, were because of an underlying CVI (8). In the European region, it has been reported that 1% of the Western Europe population is affected with leg venous ulcers, supported by similar statistics in the south of Europe (9–12). Summing up these data, we can conclude that in Europe, venous insufficiency is the most common cause of chronic leg ulcers. In daily practice, venous ulcers exhibit prolonged therapy duration and poor overall prognosis with high recurrence rate. Refractory venous ulcers need intensive care for a relatively long period of time. Different authors reported that more than 50% of the cases may actually require more than 1 year of therapy (13). Our patient follow-up continued for the full 9 months, showing no signs of healing, which indeed supports the last statement. In our case, it was evident through contrast venography that the patient suffered from a severe degree of compression of the left common iliac vein, affecting the efficiency of the venous pathway, which in turn clinically manifested as a refractory venous leg ulcer. The fact that the located stenosis within the left common iliac vein was in an area higher up in the pelvis region justifies why we were unable to detect the source of her underlying venous

incompetence at the beginning of the presentation through the lower extremities colour duplex ultrasound.

The phenomenon in which an extrinsic compression applied on the left common iliac vein by the contra-lateral overriding right iliac artery compromising the venous outflow in the lower limbs is known as MTS, also known as iliac vein compression syndrome (14). Towards the end of the 18th century, Virchow, in an attempt to explain the high incidence of left-sided deep venous thrombosis (DVT), initially referred to the proposed anatomical variant (15). In 1908, McMurrich postulated these strictures, taking a closer look into their pathology without emphasising their clinical features (16). The picture became quite clear thanks to the work of May and Thurner in 1957. They were able to describe the syndrome anatomically by conducting an anatomical dissection study on 430 cadavers, where it yielded significant results. It was found that approximately 22% of the cadavers exhibited an anatomical variant at the level of the left common iliac vein (14). The clinical features of the syndrome were later detailed by Cockett and Thomas in 1965 (17). MTS is believed to be an acquired syndrome because of the lack of supportive histological findings and the absence of any anatomical congenital signs (14,18). The left common iliac vein follows a transverse course unlike the right common iliac vein vertical ascendant path, which predisposes it to the compression effect of the overlying right common iliac artery against the lumbar spine. As a result of the compression exerted on the left common iliac vein and the chronic adjacent arterial pulsations intra-luminal changes can occur, because of the accumulation of elastin and collagen occur, leading to the formation of obstructive lesions known as ‘spurs’, hindering the venous outflow to the lower extremities (19,20). MTS is predominantly seen among young to middle-aged female patients presenting with unilateral chronic venous outflow obstruction symptoms (19,21). The actual incidence of MTS is unknown (22). Clinical presentations of the syndrome can vary from complete symptom-free cases, which represent the majority of patients, to more acute presentations of DVT or its debilitating complications. On the other hand, others might only present with symptoms and signs of chronic venous outflow obstruction, like in our patient’s case. It was estimated that almost 25% of healthy asymptomatic individuals can have >50% compression within the left iliac vein (23), which again confirms that many cases can go under-diagnosed and are incidentally detected. It is believed that the development of symptoms occurs mainly following specific provoking triggers in asymptomatic patients, for instance prolonged immobilisation, operative procedures (24) or pregnancy, causing significant haemodynamic instability in the venous flow and presenting in either acute or chronic forms. MTS can be easily under-diagnosed in the presence of other more common risk factors. It is true that duplex ultrasound remains the mainstay gold standard method in diagnosing lower limbs venous insufficiency (25–27). However, as venous incompetence is not restricted only to the lower limbs level and the fact that the iliac veins are deeply seated within the pelvic cavity, the sensitivity of the doppler ultrasound study in visualising any insufficiency within these veins above the inguinal line will be limited (28). Further investigations using other modalities like computed contrast venography or magnetic resonance

venography can assist in precisely clarifying the anatomical pattern and spotting any variants (29). Unexplained cases of unilateral venous symptoms, especially among young or middle-aged group of females, should be dealt with care within a well-integrated multidisciplinary team, aiming to alleviate the mechanical obstructed venous outflow and restoring in-line patency to the venous system.

Conclusion

Our case report highlights an important aspect in the clinical assessment of patients with CVI, particularly when it manifests as a refractory venous leg ulcer. Given the socioeconomic impact and the effects of these ulcers on patients’ quality of life, an active approach and an inclusion of unusual causes of CVI, such as MTS, in the clinical evaluation algorithm is warranted. Therefore, it is recommended that all cases of suspected CVI that demonstrated normal lower extremities venous duplex sonographic findings should undergo further comprehensive evaluation of the truncal venous system using other modalities.

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