

May Thurner Syndrome: An Important Differential Diagnosis for DVT

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Abstract

May Thurner Syndrome (MTS) is an anatomical variant in which the left common iliac vein is compressed against the fifth lumbar vertebra by the right common iliac artery resulting in lower extremity venous outflow obstruction. It is estimated to be present in about 20% of the population and predominantly seen in young females. It can present as acute or chronic DVT with symptoms of unilateral leg pain, edema, and / or varicosities. MTS related deep venous thrombosis (DVT) accounts for 2%-3% of all lower limb DVTs. In addition to standard anticoagulation, endovascular thrombolysis with angioplasty and stent placement is recommended as part of treatment. We present a case of a 43 year old female who presented with an unprovoked left DVT and diagnosis of MTS was made on CT scan.

Keywords: DVT; May Thurner syndrome; Iliac vein compression syndrome

Introduction

MTS, also known as iliac vein compression syndrome (IVCS) was first described by May and Thurner, in 1956 when they discovered the anatomical variation but as early as in 1851, Virchow noted that iliofemoral DVTs were five times more likely to occur in the left leg. MTS related DVT occurs more commonly in patients with reduced left common iliac vein diameters or severe degrees of iliac vein compression. It should be considered when a standard venous Doppler ultrasound shows that the DVT extends proximally to the iliofemoral vein indicating involvement and a possible compression of the left common iliac vein. Diagnosis often requires a CT scan or MRI to visualize the area of obstruction and the compression above it. MTS related DVT is usually recurrent and/or poorly responsive to treatment with anticoagulation alone. It therefore requires catheter-directed thrombolysis, venous angioplasty and intravascular stenting to address the underlying mechanical compression.

Case Report

A 43 year old Chinese female was admitted with a 6 day history of gradual and progressive left lower limb swelling with associated redness and warmth. There was no history of trauma to the leg, no similar episodes in the past or prolonged flight and immobilization. There was no fever, no chest pain or breathlessness. She had no significant past medical history. There was no family history of DVT. She is married with no children by choice and denied use of oral contraceptive pills. She had no history of recurrent miscarriages. She is a housewife, a lifelong nonsmoker and does not consume alcohol. On physical examination, she was not breathless at rest. Vital signs showed tachycardia at 122 beats per minute, blood pressure of 126/68 mmHg and a respiratory rate of 16 breaths per minute. The oxygen saturation (sPO₂) was 100% on room air. The cardiovascular and respiratory examinations were unremarkable. On examination of the lower limbs, the left lower limb was warm, swollen up to the thigh measuring 6 cm more than the right, shiny and tender to touch. There was no evidence of stasis dermatitis, venous ulcers or varicosities. The peripheral pulses were well felt. Investigations showed significantly elevated D Dimer level at 9.67 mg/L. Full blood count, renal panel and Chest radiograph were normal. ECG showed Sinus Tachycardia. She was given 1 dose of low molecular weight heparin (LMWH) enoxaparin which was continued at a dose of 1 mg/kg body weight 2 times a day. In view of tachycardia, CT Pulmonary Angiogram was carried out which showed

no pulmonary embolism. DVT scan (Color flow Doppler compression ultrasonography) showed complete thrombosis from the popliteal vein to the common femoral vein. However the extent of the thrombosis was not ascertained as the left external iliac vein was obscured by bowel gas (Figures 1 and 2). CT scan of the abdomen and pelvis showed diffuse venous thrombosis involving the left iliac vein down to the superficial femoral vein. The thrombus terminated at the point where the left common iliac vein crosses posterior to the right common iliac artery. In the absence of any suspicious pelvic mass, these features were suggestive of May Thurner syndrome (Figure 3). Figure 1 shows the diagrammatic representation of the anatomical variation. Protein C, Protein S, lupus anticoagulant, anti-thrombin III and auto immune markers were unremarkable. Her symptom of swelling had improved

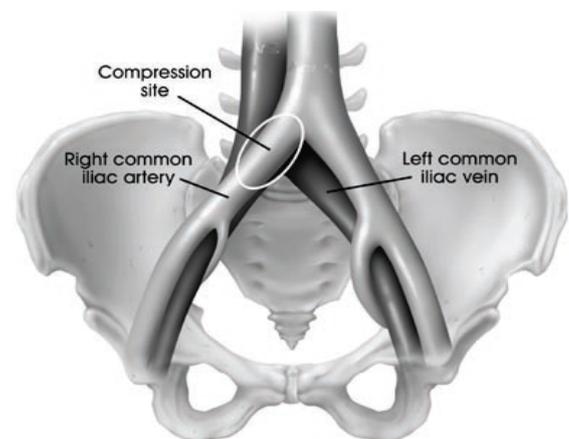


Figure 1: May-Thurner syndrome: the pulsating right iliac artery compresses the left iliac vein against the underlying vertebrae.

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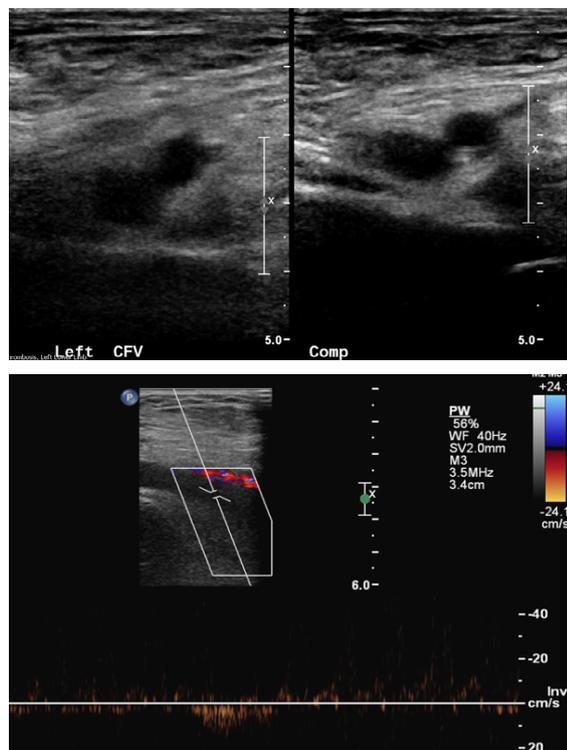


Figure 2: Ultrasound of the left common femoral vein which shows failure of compressibility on studies (upper image) and no color flow on duplex scan (lower image) confirming the diagnosis of DVT.



Figure 3: CT abdomen and pelvis of the patient showing the Right iliac artery compressing the left iliac vein and the obstruction below (arrow).

on the day following admission. She was offered catheter based thrombolysis with a possible angioplasty and intravascular stenting. However, she opted for medical management first and an intervention at a later date if symptoms recurred. The risk of post thrombotic syndrome and recurrent DVT with medical management alone was reinforced to her but she declined intervention. She was then shifted to a Factor Xa inhibitor (Rivaroxaban) for the treatment of the DVT. She was discharged on the third hospital day on Rivaroxaban to complete 6 months of treatment. On follow up, the patient remained well and again declined any intervention.

Discussion

May and Thurner were the first to provide the explanation of iliac

vein compression syndrome (IVCS) in 1956. They found that the left common iliac vein had a vascular thickening at the point where it is compressed between the fifth lumbar vertebrae and the overlying right common iliac artery (Figure 1). It is hypothesized that chronic arterial pulsation of the right common iliac artery causes deposition of elastin and collagen in left common iliac vein and continued trauma leads to local extensive intimal proliferation leading to vascular thickening which was called “venous spurs” [1]. The formation of venous spurs leads to obstruction, venous thrombosis and impaired venous return. Hence, the pathogenesis of MTS related acute or chronic DVT and chronic venous insufficiency is likely to be a combination of mechanical compression, vascular thickening due to endothelial changes and reduced iliac vein diameter.

The true incidence rate of MTS is unknown. According to autopsy studies by May and Thurner, it ranges from 22%-32% [2]. The overall prevalence of symptomatic MTS has been reported in 18-49% of patients with left sided lower limb DVT [3]. However MTS related deep venous thrombosis (DVT) accounts for only 2%-3% of all lower limb DVTs [4]. It is more common in women and 72% are aged 20-50 years old [5].

Risk factors associated with MTS related DVT are prolonged air travel, oral contraceptive use and recent pregnancy. There is also a strong association between MTS and thrombophilia. 67% of patients with MTS related DVT have some form of thrombophilia [6]. Kieran et al. showed that among 30 patients with iliac vein compression syndrome, 12 had abnormal thrombophilia screen [7]. Kolbel et al. further suggested that 78% of patients with ileofemoral DVT have one or more coagulation disorders and about 90% will have an additional risk factor [8]. Our patient was a typical middle-aged female who presented with extensive left leg DVT but she did not have any of the risk factors.

15-30% of patients with MTS are asymptomatic [1]. The most common clinical presentation is unilateral leg swelling due to acute DVT. It can also present as chronic venous insufficiency or chronic thrombosis with symptoms of venous hypertension and venous stasis namely claudication, pain, swelling, varicose veins and / or ulceration. Report of iliac vein rupture due to MTS has also been described [9].

The diagnosis of DVT is made by color duplex ultrasound as shown in our patient (Figure 2). However in MTS related DVT, this test has its limitations. Views of the iliac vein are often obscured by overlying bowel gas. In cases where the DVT is detected in the iliac vessels, the Doppler ultrasound is unable to visualize the iliac vein compression and spurs. Although Doppler ultrasound venous mapping of the lower limbs has been shown to be a reliable tool to delineate the area of DVT [10], this was not applicable in our patient as the anatomical variation is beyond the area of venous mapping.

Current noninvasive diagnostic tests include computed tomography venography (CTV), magnetic resonance venography (MRV) and Intravascular Ultrasound. Both CTV and MRV have high sensitivity and specificity in evaluating MTS related DVT allowing further identification of the compression and pelvic venous collaterals. CTV or MRV was not performed in our patient as the contrasted CT scan of the abdomen and pelvis confirmed the diagnosis of MTS (Figure 3). Intravascular ultrasound offers direct visualization of iliac vein compression, effectively identifies mural abnormalities and estimates vessel wall size. During endovascular treatment, it facilitates accurate placement of the wire across the stenosed region [11].

For the diagnosis of MTS, Contrast venography (Left common

femoral or iliac venography) is the gold standard. It not only confirms the presence of obstruction but assesses its hemodynamic significance. Direct pressure measurements are performed at the same setting. Pressure gradients of >2 mmHg measured across the compression indicates hemodynamic significance [11]. May and Thurner had advocated the use of pressure differentials to support the diagnosis of hemodynamically significant obstruction. A pressure difference between 2 iliac veins of 2 mmHg at rest or 3 mmHg after exercise is suggested to be hemodynamically significant [1]. Lastly, contrast venography helps plan therapeutic interventions and treatment can be offered at the same setting.

Management of MTS consists of standard anticoagulation for acute DVT and to prevent recurrence and long term complications, percutaneous catheter-directed pharmacothrombolysis, angioplasty and stent placement are advocated. In MTS related DVT which involves the iliofemoral vein, there is a higher occurrence of debilitating symptomatic post thrombotic syndrome and higher rates of recurrent DVT when the occlusion from the anatomical variation is not corrected. It has been noted in several studies that 6 months after anticoagulation alone, 88% of patients with iliofemoral DVT have moderate to severe venous obstruction [12]. Catheter directed thrombolysis combined with mechanical endovascular thrombectomy may be considered in patients with severe symptoms, long segment thrombosis and large clot burden [8]. Kwak et al. in their study of 16 IVCS patients with metallic stent placements following thrombectomy showed a 95 and 100% primary and secondary patency rates at 2 year follow up [13]. Open surgical thrombectomy is reserved for patients with contraindications to pharmacothrombolysis [14].

Patients with IVCS related chronic occlusion do not develop sufficient collateralization and therefore present with symptoms of venous hypertension and stasis. Treatment with endovascular stenting is recommended as studies have shown significant improvement of the symptoms. The most extensive experience has been reported by Raju et al. who described their results after the treatment of 305 cases with symptomatic chronic venous insufficiency. Primary and secondary patency rates at 24 months were 71 and 90% respectively. Complete pain relief was achieved in 71% of patients and quality of life was also significantly improved [15].

Relief of the mechanical compression of MTS is suggested in patients presenting with persistent edema even prior to the onset of DVT and venous insufficiency [16]. However, it is important to carefully consider treatment of MTS patients with minimal symptoms and "borderline" obstructions despite promising results of intravascular stenting. Literature recommends that MTS patients with non-occlusive obstruction without typical pathologic changes in flow profile (spurs) should be functionally evaluated for hemodynamic significance before stenting of the iliac vein is done [8].

In the setting of patients with MTS who have undergone thrombolysis and stent placement, the ideal duration of anticoagulation is unclear. Guidelines state that in patients who have had any form of thrombus removal (thrombolytic or thrombectomy), anticoagulant therapy is of the same intensity and duration as in comparable patients who do not undergo thrombus removal [17]. In patients who have had stent placement, anticoagulation for at least six months to prevent in-stent restenosis is recommended [5].

Our patient was diagnosed as MTS related acute proximal DVT. Her management was anticoagulation alone due to her preference. She was given Rivaroxaban (direct factor Xa inhibitor) for which clinical symptom improvement was noted. However, there is no data

specifically addressing the efficacy profile of Rivaroxaban in MTS related acute DVT.

Conclusion

May Thurner syndrome is an important anatomical risk factor for left leg proximal DVT. It remains under-recognized and should be considered in the differential diagnosis of patients with left leg DVT and or chronic leg edema. There should be a high index of suspicion of MTS in young female patients presenting with unprovoked proximal iliofemoral DVT and also in those with symptoms of chronic venous occlusion. It is important to remember that in addition to anticoagulation, treatment of MTS related DVT should include correction of the anatomical variation to prevent long term complications.

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