May-Thurner Syndrome Complicating Phlegmasia Cerulea Dolens

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ABSTRACT

May-Thurner syndrome (also known as Cockett’s syndrome or Iliac vein compression syndrome) is a venous anomaly featuring mechanical compression of the left common iliac vein by the right common iliac artery, with increased risk of development of deep vein thrombosis of the left lower limb. Phlegmasia caerulea dolens usually occurs, follow by massive venous thrombosis. May-Thurner syndrome as a cause of phlegmasia caerulea dolens is rare. We report a case of May-Thurner syndrome with deep vein thrombosis and rapid development of phlegmasia caerulea dolens, successfully treated by emergency thrombectomy, endovascular angioplasty, stenting, and systemic anticoagulation.

Keywords: May-Thurner syndrome, phlegmasia caerulea dolens, thromboembolism

INTRODUCTION

Venous thromboembolism (VTE) is a worldwide health issue, with a first-episode incidence of approximately 100 persons per 100,000 each year in the United States, and a relatively higher incidence in Caucasian populations than in Asian populations [1]. May-Thurner syndrome (MTS) is an unusual venous anomaly of the left common iliac vein characterized by compression by the right common iliac artery against the lower lumbar vertebrae, resulting in outflow obstruction and associated with a relatively high incidence of left iliofemoral VTE [2]. Phlegmasia caerulea dolens (PCD) is a life-threatening and rare scenario that is defined as massive VTE with ischemic change or venous gangrene in advanced cases [3]. We present the case of an Asian male patient with MTS manifested with acute deep vein thrombosis (DVT) with rapid development of PCD, successfully managed by emergency thrombectomy and systemic anticoagulation. The purpose of this report is to provide an opportunity for readers to increase their awareness of the clinical and radiologic characteristics of this rare condition, which could help multidisciplinary teams to perform adequate management in similar cases. The patient consented to the publication of this report.

CASE REPORT

A 61-year-old male resident of Taiwan presented with sudden-onset painful swelling and redness of the left lower limb. He had a history of well-controlled hypertension for 10 years. Old age was the only risk factor for DVT in this case. On physical examination, edematous change, tenderness, warmth, and diffuse redness with an area of cyanosis were found in the involved leg (Fig. 1). Vital signs were stable, with no signs of sepsis. Laboratory examinations were unremarkable, with the exception of an elevated D-dimer level (>10000 μg per liter). Hypercoagulable workup revealed negative findings. A bedside ultrasound...
scan revealed DVT in the left external iliac, common femoral, superficial femoral, popliteal, anterior tibial, posterior tibial, and peroneal veins (Fig. 2). However, the inferior vena cava and left common iliac vein could not be visualized via this ultrasound scan. A computed tomography (CT) scan of the pelvis and bilateral lower extremities revealed left common iliac vein compression by the right common iliac artery, indicating MTS (Fig. 3). A prominently swollen and enlarged left thigh as compared with the right thigh and DVT in the veins of the left lower extremity up to the level of the left common iliac vein were observed.

Due to the above-mentioned findings, MTS complicated by DVT and PCD was diagnosed. An emergency surgical thrombectomy was performed, and copious fresh thrombi in the left common iliac vein, left external iliac vein, left common femoral vein and left superficial femoral vein were removed. Balloon angioplasty of the left common iliac vein was then preformed. Concurrent systemic anticoagulation therapy with intravenous heparin infusion was administered for possible residual thrombi. After 6 days of hospitalization, the patient recovered uneventfully, although some residual soft tissue edema remained in the left lower extremity. Outpatient anticoagulation treatment with oral enoxaparin/warfarin for 3 months was continued.

**Figure 1.** Swelling, pain, warmth, and diffuse redness with an area of cyanosis (arrows) were found in the involved leg, indicating phlegmasia cerulean dolens.

**Figure 2.** Color Doppler ultrasound scan disclosed persistent absence of blood flow in the a. left external iliac (arrow), b. left common femoral (arrow), c. left superficial femoral (arrow), and d. popliteal veins (arrow).
in order to maintain the international normalized ratio in the range of 2.0–3.0. Subsequently, endovascular stenting of the left common iliac vein for MTS was performed 3 months later, and outpatient anticoagulation treatment was administered for another 3 months. The patient was seen 6 months after the initial episode, and was free from any symptoms of MTS and PCD. A follow-up ultrasound scan showed no residual thrombi, with improvement of venous flow in the left lower extremity.

DISCUSSION

VTE most commonly manifests with pulmonary embolism and DVT, the most common location of DVT being the lower extremities. DVT development in the proximal lower extremity above the popliteal vein level is associated with a 50% risk of pulmonary embolism without treatment, which is a life-threatening condition [4]. Risk factors of VTE include congestive heart failure, central venous catheters, chemotherapy, major general surgery, major trauma, hormone therapy, malignant neoplasm, respiratory failure, oral contraceptives, pregnancy, postpartum status, previous venous thromboembolism, hematologic disease, increasing age, and immobility due to sitting, such as in long-distance automobile travel [5]. Clinical reports have concluded that episodes of travel-related VTE affect people with the aforementioned risk factors, such as a history of VTE most commonly, and cases in which there are no predisposing factors are rare [6]. Age is another risk factor for the development of VTE, with a watershed of age 40, above which there is a significantly increased risk as compared with younger populations [5].

MTS is a venous anomaly resulting in compression of the left common iliac vein by the right common iliac artery and the lumbar spine with or without DVT [7]. Pulsatile compression and repetitive irritation at this location results in fibrotic change of the venous wall with venous spur formation, which causes stenosis of the vessel lumen [7]. Although this syndrome is unusual, its true prevalence is likely underestimated owing to the chronic nature of MTS, the clinical symptoms and signs of which include chronic leg pain, limb edema, recurrent stasis ulcers, skin color changes and postphlebitic syndrome. MTS has a propensity to occur in female patients of a young age, i.e., in their second to fourth decade of life, with a history of prolonged immobility or during pregnancy.

PCD is a rare and severe complication of DVT, featuring venous outflow obstruction, limb swelling, pain, bluish discoloration of skin, and even venous gangrene in advanced cases, which can lead to compartment syndrome and limb amputation, with an ultimate amputation rate as high as 50% in survivors [3]. Risk factors of PCD include hypercoagulable syndrome, malignancy, MTS, heart failure, intravenous drug abuse, trauma, and ulcerative colitis [8]. An immediate diagnosis of PCD can be made clinically according to the clinical triad of limb swelling, pain, and bluish discoloration. Generally, sonography is useful as an imaging diagnostic tool that is available on demand, without the need for venography, CT or magnetic resonance venography. Sonography is effective in terms of demonstrating venous thrombus in the clinical scenario of PCD [9]. However, visualization of the iliac vessels by color Doppler ultrasound is technically difficult, especially in
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cases of a large amount of bowel gas or in obese patients with much subcutaneous fat [10]. In our case, the original cause of PCD and DVT was MTS, which required CT investigation in order to demonstrate the pelvic veins and to guide further thrombectomy and endovascular interventions.

There exists no standard treatment strategy for MTS, and options vary depending on clinical presentation. Potential treatments include traditional open repair, bypass surgery, balloon angioplasty and stenting. Surgical indications of MTS are a difference of greater than 2 mmHg in the mean resting venous pressure between the bilateral common femoral veins; claudication with a triple venous system pressure of the ill extremity during exercise; or a difference in pullback pressure from the inferior vena cava to the external iliac vein [11]. No prospective large-scale clinical randomized comparative studied for the aforementioned treatment option have been conducted, although the guidelines of the Society of Vascular Surgery and the Society of Interventional Radiology both state that stenting of the iliac vein is recommended in the context of external compression [12]. For young and otherwise healthy patients, endovascular intervention for MTS by methods of balloon angioplasty or stenting is perhaps a less invasive and more graceful method than surgery. An agreed standard treatment strategy or guidelines for PCD are not available. Routinely, systemic anticoagulation with intravenous heparin is considered an essential component of treatment for PCD to prevent further blood clot aggregation, in addition to potential pulmonary thromboembolism. Surgical thrombectomy may be performed, depending on the status of disease progression. In cases of MTS, due to concurrent mechanical iliac vein compression, patients are at high risk of recurrence of DVT and post-thrombotic syndrome. Surgery or endovascular intervention is recommended in some cases of MTS, as previously described [12]. Suwanabol and colleagues advocated endovascular stenting as a mainstay for the treatment of patients with MTS owing to mechanical obstruction in this venous anomaly [8].

In conclusion, PCD is a rare condition with a high mortality rate that should be considered in patients presenting with marked limb swelling, pain, and bluish discoloration. PCD caused by MTS is even more rare, and challenging to manage. A correct diagnosis and prompt treatment are key to improving the prognosis and avoiding further limb amputation.

REFERENCES