

Unusual Presentation of May-Thurner Syndrome: A Case Report and Review of Literature

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Abstract

Iliac vein compression syndrome (IVCS) that presents with unclear etiology is known also as May-Thurner syndrome. The syndrome is due to the compression of the left common iliac vein occurring between the right common iliac artery and superior vertebrae leading to a rare presentation of deep vein thrombosis (DVT). It is uncommon for patients with an established diagnosis of IVCS to present with an obstruction of venous outflow or pulmonary emboli without the occurrence thrombosis in the deep venous system. A patient should be treated promptly to prevent life threatening consequences associated with the prior. Our case study presents a 46-year-old African American female without history of smoking or any other IVCS risk factors. We diagnosed her with a pulmonary emboli and a compressed left common iliac vein occurring without any symptoms in her lower extremities. The patient was successfully treated with catheter directed thrombolysis into the left iliac vein followed by therapeutic anticoagulation protocols.

Keywords: Iliac Vein Compression Syndrome (IVCS); Deep Vein Thrombosis (DVT); Pulmonary Embolism (PE)

Introduction

The most two common presentations of venous thrombosis are deep vein thrombosis (DVT) of the lower extremity and pulmonary embolism (PE) of the respiratory system. Venous thrombosis can be attributed to several causes that are further classified into significant, moderate, or poorly associated risk factors (Table 1). Risk factors such as surgery, prolonged bed rest, pregnancy, oral contraceptives and smoking occurred in more than 80% of patients with an established diagnosis of venous thrombosis [1,2].

Significant
Extensive Trauma
Spinal Cord Injury (SCI)
Major Surgery
Moderate
Arthroscopic Surgery
Central Venous Lines
Chemotherapy

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Congestive Heart or Respiratory Failure
Hormone Replacement Therapy
Neoplasia or Clotting Factors
Abnormalities
Hormonal contraception (Birth Control)
Prolonged Immobilization
Pregnancy/Postpartum
Previous Venous Thromboembolism
Thrombophilia
Poorly
Bed Rest More than Four Days
Old Age
Morbid Obesity
Laparoscopic Surgery
Varicose Veins
Pregnancy/Antepartum

Table 1: Risk factors associated with venous thromboembolism categorized into one of three rankings; significant, moderate, or poorly.

May-Thurner syndrome may also be referred to as ilio caval compression syndrome, cockett syndrome, or iliac vein compression syndrome. The incidence of May-Thurner syndrome ranges from 18 - 49% among patients with left-sided lower extremity DVT. It commonly seen in women between the ages of 20 and 50 [3]. Iliac vein compression syndrome (IVCS) is the result of compression occurring to the left common iliac vein between the right common iliac artery and overlying vertebrae. External compression of the vein causes changes intraluminal spacing resulting in a disturbance to blood flow potentially leading to deep vein thrombosis (DVT). Moreover, the prior may instead lead to venous hypertension without thrombosis in the left lower extremity, similar to radiologically confirmed findings in our case [4,5]. DVT typically occurs in the calf of thigh muscle and is attributed to the formation of a blood clot in a deep vein. DVT can partly or completely block venous blood flow, causing chronic pain and localized swelling. Most patients with DVT present with leg pain, fatigue, swelling, redness and visible veins [6].

May-Thurner syndrome most commonly presents as deep vein thrombosis. However, other patients with an established diagnosis of iliac vein compression syndrome may present with an obstruction of the venous outflow absent of any disturbance to the deep venous system [7]. IVCS must be recognized and treated as early as possible to prevent or reduce any irreversible changes in the patient. This syndrome was diagnosed more frequently after the introduction of catheter-directed endovascular for treatment of DVT. It is also considered as a common anatomic pattern in normal subjects, moreover which has been associated with unprovoked left iliofemoral DVT or chronic venous insufficiency [8]. However, visualization of a blood clot in the upper part of the pelvis may be difficult to detect using ultrasonography alone. If DVT is highly suspected further testing would be required using contrast venography, magnetic resonance imaging, or intravascular ultrasound imaging [9]. It must be noted that May-Thurner Syndrome is more common in those with reduced left common iliac vein diameters and/or severe degrees of iliac vein compression [10].

Pulmonary embolism (PE) is a blockage of the main artery of the lung or one of its branches by a blood clot that has travelled from elsewhere in the body through the bloodstream. PE most commonly results from an embolus of DVT that breaks off and migrates to the lung, a

process termed venous thromboembolic. The obstruction of the blood flow through the lungs resulting in changes of pressure in the right ventricle of the heart commonly lead to the symptoms and signs of PE [11]. In our case, we report the presence of PE in the absence of a preceding detectable thrombosis in the deep venous system, previously described.

Case Report

A 46-year-old African American female, with a negative history of smoking or any other risk factors associated with DVT or PE, presented in our emergency room with sudden onset of syncope and respiratory distress. These symptoms were not relieved by inhaled oxygen therapy using a 40% face mask. Afterwards, she demonstrated a poor response to Bilevel Positive Airway Pressure (BPAP) to reduce her respiratory effort. Furthermore, the patient was intubated after she experienced a rapid desaturation in blood oxygen levels. Desaturation in blood oxygen levels were detected by pulse oximeter and arterial blood gases that revealed profound hypoxemia and respiratory alkalosis.

After stabilization of the patient, a complete physical examination was determined to be unremarkable. Overall she was found to be hemodynamically stable. Her vital signs were taken and were as follows: blood pressure: 120/80, pulse Rate: 115 (beats per minute-bpm) and temperature: 37.7 Celsius. During the evaluation her respiratory rate remained at 26 breaths/min. This was concerning to us and lead us to transfer into medical intensive care unit (ICU).

Additionally, the laboratory values taken at this time were all within normal limits, except D-Dimer levels at 1970 ng/mg. Her EKG showed a sinus rhythm with a tachycardia of 115 bpm. It was at this time, we preformed a bilateral lower extremity doppler but it failed to show any evidence of blood clot(s). Cat Scan (CT) abdomen-pelvis angiography was then obtained after elevated D-Dimer levels generated clinical suspicion. It revealed thrombus in the left common iliac vein without any evidence of a space-occupying lesion. As well, images revealed a moderate sized left main trunk PE in our patient. As time progressed, a second echocardiography was conducted that revealed right ventricular dilatation and wall hypokinesis. Echocardiography results further showed a raised pulmonary artery pressure of 48mmHg with normal systolic function of the right ventricle. Venography, showed a compression of the left common iliac vein between the right common iliac artery and the overlying vertebrae.

Therapeutic intervention was administered in the form of endovascular therapy and catheter directed thrombolysis. This procedure was then followed with endovascular stenting of the left common iliac vein, as well as several other branches. An excellent angiographic response was seen after angioplasty and subsequent stenting of the left common, external veins, and left common femoral vein.

The patient was later switched to Lovenox 80 mg administered subcutaneously every 12 hours in combination with oral Coumadin until a therapeutic INR of 2.5 was achieved. Finally, the patient was successfully extubated before she was discharged on an oral Coumadin regimen with follow up of INR every 3 days until stable.

Discussion

In 1957 May and Thurner examined 430 cadavers and documented the decrease in venous flow that resulted from an intimal change in the left common iliac vein (LCIV), later called May-Thurner syndrome. It manifests with lower extremity edema, varicosities, venous stasis, venous claudicating, pain or venous ulceration. The main diagnostic study is venography, which shows compressions of the iliac vein occurring with a spur. Intravenous ultrasound has also been used in diagnosis because of increased sensitivity compared to venography protocols. After a diagnosis, therapy involves anticoagulation or surgical correction of the vein compression by either by a vein patch repair, venous crossover bypass graft, or an IVC filter used in our case study [12].

Incidences of thrombi formation increases due missed diagnoses of venous compression such as in May-Thurner syndrome [13]. In a patient with identifiable risk factors, the diagnostic workup is commonly terminated upon the diagnosis of DVT. However, this allows the

anatomic etiology which results in a variety of hazards including a recurrence, distant spread to pulmonary emboli, chronic venous stasis, and iliac vein rupture to occur unrecognized.

Other cases have been reported by Levent Oğuzkurt, *et al.* 2007 [16]: the patient was a 14-year-old girl who presented with a left lower extremity swelling accompanied by an illness lasting six months. CT abdomen revealed severe compression of the right common iliac artery, and Doppler studies detected a lower extremity DVT. Loukas, *et al.* 2008 [17] also described our syndrome but in 35-year-old Caucasian female with no prior illness or risk factors. Cerqouzzi, *et al.* 2012 [18] described iliac vein compression syndrome in a 35-year-old white female athlete who was complaining of hip pain for 3 years leading to an accidental diagnosis during a MRI study. The course of therapy in each of the prior cases was very similar to our case. Moreover, these authors stress the fact that the main key factor for improving mortality and morbidity is an early detection based mainly on clinical suspicion followed by imaging and other diagnostic modalities.

Additionally, color duplex alone is a weak diagnostic modality for our syndrome, since it lacks sensitivity to demonstrate non-occlusive thrombosis, venous spurs and intraluminal defects in the common iliac veins. CT abdomen-pelvis conducted with contrast can confirm any extrinsic compression by a space occupying lesion(s). Magnetic resonance venography of pelvic veins is the imaging technique of choice to visualize the compressed vasculature and collaterals. Acute iliofemoral DVT is identified then treated with thrombolysis followed by anticoagulation regimens. Those patients with residual compression and chronic leg manifestations should be treated with percutaneous transluminal angioplasty and stenting to prevent vessel recoil [15].

Furthermore, thrombolytic infusion should be continued for an additional 24 to 48 hours. Then an intravascular stent should be deployed in the area of iliac vein compression. Repeated imaging should be obtained to verify that the stent is positioned across the entire area of the compressed vein. After the stent placement or IVC Filter a regimen of systemic long-term anticoagulation is recommended for at least 6 months [14].

Conclusion

A specific vascular malformation, such as May-Thurner syndrome, may come to clinical attention when examining a suspicious asymptomatic, as well as symptomatic patient. An early diagnosis leading to an early treatment is required for successful intervention in reducing any long-term health consequences. We believe that further work and studies are required to assess the impact of anatomical variation, as well as different risk factors leading to venous thromboembolism in order to better diagnosis patients. We demonstrated that sometimes further testing based on suspicion is required to reveal a diagnosis that otherwise would of been missed. The successful diagnosing and treatment of atypical cases, such as ours, is essential in sustaining and improving the quality of living for all patients.

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