

Painful Blue Leg: The Darkside of May-Thurner Syndrome

Ağrılı Mavi Bacak: May-Thurner Sendromunun Karanlık Yüzü

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ABSTRACT

Phlegmasia Cerulae Dolens PCD is a rare consequence of Deep Vein Thrombosis (DVT), only 2 to 5% of patients with PCD were found to have May-Thurner Syndrome (MTS). Despite its rarity, Phlegmasia Cerulae Dolens is a life-threatening form of acute DVT that necessitates early detection and prompt treatment to save lives and limbs. May-Thurner Syndrome is characterized by an anatomical variation in which the right common iliac artery compresses the left common iliac vein, causing left-sided DVT. PCD causes severe swelling of the lower extremities, pain, and cyanosis, leading to venous gangrene and amputation. We present a case of a 67-year-old lady who presented to the Emergency Department, presented with PCD, and was diagnosed with MTS. The patient then underwent successful endovascular treatment. This study aims to emphasize the importance of early detection and intervention while dealing with acute DVT.

Keywords: Phlegmasia Cerulae Dolens, May-Thurner Syndrome, Deep vein thrombosis

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ÖZET

Phlegmasia Cerulae Dolens PCD, Derin Ven Trombozunun (DVT) nadir bir sonucudur, PCD'li hastaların sadece %2 ila 5'inde May-Thurner Sendromu (MTS) olduğu bulunmuştur. Nadir görülmesine rağmen Phlegmasia Cerulae Dolens, hayatları ve uzuvları kurtarmak için erken teşhis ve hızlı tedavi gerektiren hayatı tehdit eden bir akut DVT formudur. May-Thurner Sendromu, sağ ana iliak arterin sol ana iliak veni sıkıştırarak sol taraflı DVT'ye neden olduğu anatomik bir varyasyon ile karakterizedir. PCD, alt ekstremitelerde şiddetli şişmeye, ağrıya ve morarmaya neden olarak venöz kangrene ve amputasyona yol açar. Acil Servise başvuran, PCD ile başvuran ve MTS tanısı alan 67 yaşında bir bayan olguyu sunuyoruz. Hastaya daha sonra başarılı endovasküler tedavi uygulandı. Bu çalışma, akut DVT ile uğraşırken erken teşhis ve müdahalenin önemini vurgulamayı amaçlamaktadır.

Anahtar Sözcükler: Phlegmasia Cerulea Dolens, May-Thurner Sendromu, Derin ven trombozu

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INTRODUCTION

Phlegmasia Cerulæ Dolens (PCD) is a rare complication of Deep Vein Thrombosis (DVT); only 2% to 5% of patients presented with PCD diagnosed with May-Thurner Syndrome (MTS). MTS is caused by an anatomical variation in which the right common iliac artery compresses the left common iliac vein, causing left-sided DVT in the patient. We discuss a case of successful endovascular therapy in MTS with PCD(1).

CASE REPORT

A 67-year-old women with underlying Hypertension, Diabetes Mellitus, Dyslipidaemia, Left Knee, and Hip osteoarthritis presented to the Emergency Department with left lower limb pain and swelling for one week associated with cyanosis limb. On examination, the left lower limb was edematous and cyanosed. (Figure 1). All pulses were palpable, and bedside Doppler auscultation revealed biphasic signal at DPA and PTA, while popliteal and femoral were triphasic. The patient could still move her limb, and the sensation was reduced compared to the right lower limb. After 4 hours of admission, the patient complaining of worsening left lower limb pain.

We noted that the compartment became tense during clinical examination, and we could not palpate the DPA/PTA pulses. However, the popliteal and femoral pulses were able to feel. Doppler signals over the DPA/PTA were feeble, while popliteal showed biphasic and femoral triphasic. USG and CT Venogram of the left lower limbs showed thrombosis of the popliteal vein extending into the left external iliac vein, confirming the diagnosis of Phlegmasia Cerulæ Dolens (PCD) and May-Thurner syndrome (Figure 2). We started her on low molecular weight heparin and morphine.

A discussion was made with the vascular surgeon and planned for catheter-directed thrombolysis. Our interventional radiologist inserted a prophylactic IVC filter before proceeded with catheter-directed thrombolysis. Venogram showed multiple filling defects along left common iliac vein till left popliteal vein. The region of interest was traversed using a microwire and microcatheter (Figure 3), and a total of 15mg rTPA bolus was given during the procedure, and then 35mg rTPA infusion was given for the next 14 hours. No overt complications. A few hours after the treatment, her left lower limb had returned to normal. All the pulses were palpable with good doppler signals (Figure 4). The venogram on the first day after the intervention revealed effective catheter-directed thrombolysis. (Figures 5,6). During review at Surgical Outpatient clinic two weeks post-intervention, the left lower limb was normal. She decided not for stent placement yet and agreed to stent placement if she develops a recurrent problem or becomes symptomatic later.



Figure 1

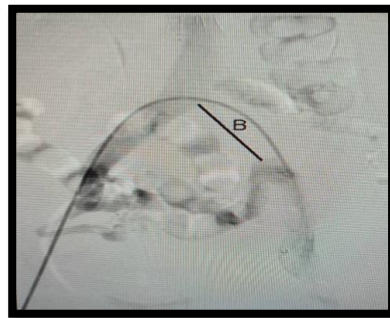


Figure 2: Noted no contrast opacification seen over the left common iliac vein with collaterals seen from the left internal iliac vein into the right common iliac vein. Also, no contrast opacification distally.



Figure 3: rtPA bolus was given with subsequent opacification of previously thrombosed vessels.



Figure 4



Figure 5 and 6: Venogram done day one post-intervention, noted complete opacification of the venous system. Successful catheter guided thrombolysis..

DISCUSSION

May-Thurner syndrome is an anatomical variant in which the right common iliac artery compresses the left common iliac vein. Mechanical compression causes intima hyperplasia and disrupts the smooth laminar flow, resulting in blood stasis and thrombosis(1). It is a rare aetiology of the symptomatic lower extremity venous disorder which accounts for only 2-3% of all lower limb DVT; hence it requires a high index of clinical suspicion (2). Because most MTS patients are asymptomatic, the exact prevalence of MTS is unknown, and the majority of it likely goes unnoticed (1).PCD, or phlegmasia cerulea dolens, is a rare and dangerous complication of iliofemoral vein thrombosis. It is a late manifestation of non-collateralized venous obstruction, which prevents adequate arterial flow, therefore causing massive fluid sequestration increased compartment pressures and ultimately leading to arterial compromised that is rapidly causing gangrene and limb loss within hours. phlegmesia alba dolens is a thrombosis that affects only the primary deep vein and spares the collateral veins. At this stage, the leg appears white, swollen, and very painful. There is no arterial compromised yet. Once it involved the collateral vein, it is called phlegmasia cerulea dolens (PCD). This painful blue limb is at significant risk of developing venous gangrene and necessitating amputation.

There is no gold standard treatment for PCD due to its rarity; however, several therapeutic options have been recommended to enhance the outcome. In PCD, 80% of survivors will develop post-thrombotic syndrome due to persistent chronic venous stasis (1). As a result, traditional DVT treatment is insufficient for PCD, and we must address the significant burden of venous thrombus. The aim of treatment for MTS includes removing the thrombus to restore blood flow and addressing the venous compression to ensure prolonged venous patency. The goal is to avoid or reduce post-thrombotic syndrome symptoms and varicosities caused by valve damage and iliac venous outflow restriction. In general, absolute bed rest, affected limb elevation, fluid resuscitation, and intravenous heparin should all be part of the initial treatment plan. However, systemic anticoagulation alone is not adequate to treat DVT with MTS, failing to address long-term sequelae. The systemic anticoagulant can only prevent the thrombus from further progression; it cannot eliminate the existing thrombus (4).

Patients diagnosed with iliofemoral thrombosis and MTS should be treated aggressively. The options to remove thrombosis would be systemic thrombolysis, catheter-directed thrombolysis (CDT), percutaneous mechanical thrombectomy, pharmaco-mechanical catheter-directed thrombolysis (PCDT), and surgical thrombectomy. Subsequently, stent placement across the common iliac vein stenosis to eliminate the compression effect by the iliac artery. The endovascular treatment, including thrombectomy followed by venoplasty and stenting, provides immediate relief of symptoms with long-term patency rates and decreased incidence of the post-thrombotic syndrome as compared to conservative anticoagulant therapy(2).

The American College of Chest Physicians (ACCP) recommends catheter-directed thrombolysis for proximal deep venous thrombosis since it has no systemic effect and reduces the risk of bleeding and PE (1,4). The CDT is associated with the complete resolution of thrombi in up to 90% of patients with PCD (1). It works by introducing a catheter into the occluding thrombus subcutaneously to give a high concentration of thrombolytic drug proximal to the thrombus, lowering the danger of systemic bleeding.

The addition of the PCDT's mechanical action to the CDT enhances the effect of the localized thrombolytic, resulting in faster thrombus clearance, which is critical in PCD to prevent the gangrenes stage (4).

Open surgical thrombectomy is the other therapy option. The main benefit is that the occlusion is relieved immediately, although it may offer a barrier for patients with co-morbidities because it needs to be done under general anaesthesia. The surgical approach was also associated with a higher risk of thrombus recurrence and mortality when compared to interventional catheters.(4). Furthermore, open thrombectomy is less effective than thrombolysis, as it cannot address the clot in small veins involved in the progression to venous gangrene. Placing an ileocecal stent in patients with acute iliocecal thrombosis or its complicated form (PCD) in the context of May-Thurner Syndrome should be considered to reduce the likelihood of thrombus recurrence and hence the risk of developing post-thrombotic syndrome. (7).

CONCLUSION

Phlegmasia cerulae dolens is a medical emergency requiring early and immediate intervention to prevent post-thrombotic syndrome. Systemic anticoagulation alone is insufficient to treat PCD; however, catheter-directed thrombolysis is an effective option against PCD with underlying MTS followed by vascular stent placement.

Conflict of interest

No conflict of interest was declared by the authors.

REFERENCES

1. Suwanabol PA, Tefera G, Schwarze ML. Syndrome Associated with the Deep Veins: Phlegmasia Cerulae Dolens, May-Thurner Syndrome, and Nutcracker Syndrome. Perspectives in Vascular Surgery and Endovascular Therapy. 2010; 22(4) 223-230
2. Siddharth Agarwal, Christopher Sosnofsky, Jamie Saum RN, Brian Taylor, Sandeep M. Patel (2021) Out of the Blue Acute May-Thurner Syndrome. J. Clinical Cardiology and Cardiovascular Interventions, 4(1)
3. R. A. Al-Hakim et al.: Management of Phlegmasia Cerulea Dolens with Percutaneous. Cardiovascular and Interventional Radiological Society of Europe (CIRSE) 2020
4. Elsaid AS et al. The ugly face of deep vein thrombosis: Phlegmasia Cerulae Dolens-Case report. International Journal of Surgery Case Reports. 2019. 107-110
5. O'Connor CT et al. A case of phlegmasia cerulae dolens as a result of May-Thurner syndrome. QJM: An International Journal of Medicine. 2020, 419-420
6. Madhavan A et al., May-Thurner syndrome as the cause of phlegmasia cerulea dolens, Am J Emerg Med (2015),
7. M.J. Seager, A. Busuttill, B. Dharmarajah, A.H. Davies, Editor's choice—a systematic review of endovenous stenting in chronic venous disease secondary to iliac vein obstruction, Eur. J. Vasc. Endovasc. Surg. 51 (1) (2016)100–120.