

Case report

A Deadly Cerulean Limb- Phlegmasia Cerulea Dolens complicated with Pulmonary Embolism

ABSTRACT

Aims: Asses the presentation of phlegmasia cerulea dolens and differentiate venous etiologies of ischemic limb with arterial thrombus

Background: Phlegmasia cerulea dolens (PCD) is a rare condition that is responsible for mortality up to 20-40%, mainly due to pulmonary embolism. Medical therapy involves administration of anticoagulants such as IV Heparin during acute setting should be initiated as soon as possible, followed by oral factor Xa inhibitors ie; rivaroxaban up to 6 months. Surgical therapy such as catheter-directed thrombolytics however remain a mainstay

Case Description: A case reported in a 51 years-old woman with underlying metastatic endometrial carcinoma, presented with sudden onset of pain and swelling of left leg. Initial bedside doppler show patent arterial perfusion over the affected leg. It was later developed into bluish discoloration and bedside doppler show poor perfusion of arteries. Subsequently, she succumbed to illness less than 24 hours from first presentation due to suspected pulmonary embolism.

Discussion and conclusion: Patients with increased risk of developing deep vein thrombosis (DVT) such as those with underlying malignancy have shown to develop a more extreme case of the disease. Phlegmasia cerulea dolens (PCD) is a rare manifestation of DVT over lower extremities that cause critical limb ischemia. Incidence of PCD is unknown however, it is estimated from a case series about 10-20% of patients underwent thrombolysis for extensive DVT, developed into phlegmasia. Common risk factors that contribute to developing PCD are malignancy, surgery and trauma.

Keywords: Phlegmasia cerulea dolens, Deep vein thrombosis, Limb ischemia, malignancy

1. INTRODUCTION

Phlegmasia cerulea dolens (PCD) is an extreme form of deep venous thrombosis (DVT). It is characterized by an acute history of excruciating pain, profound limb swelling, and skin discoloration [1]. DVT is a common venous thromboembolic (VTE) disorder, reporting an incidence rate of 1.6 per 1000 annually [2], and PCD is a consequence of massive acute venous thrombosis. The incidence is also more common in females than in males and is reported to be higher in the fifth and sixth decades of life [4]. PCD was first described by Gregoire in 1938 to differentiate massive venous thrombosis causing ischemic changes from phlegmasia alba dolens, which characterized venous thrombosis without ischemia [4].

In PCD, it is manifested with ischemia-related extensive venous thrombosis [1]. As venous flow becomes obstructed, venous hypertension increases interstitial oedema and extensive fluid sequestration into the affected limb. High compartment pressure eventually causes the arterial system to collapse, leading to acute ischemia and venous gangrene [3]. Left-sided lower extremities are more common with a ratio of 3:1 or 4:1 [4]. This may be explained by an anatomical

variant in which the right common iliac artery overrides and compresses the left common iliac vein against the lumbar spine, causing thrombosis in the left common iliac vein [5].

Factors that contributed to the development of DVT and subsequently PCD included malignancy (20–40%) [4], with higher cases of thrombosis reported in gynaecology malignancy-related patients [6]. Other less common causes are advanced age, with more prominent cases reported in the fifth and sixth decades of life [4]. Use of contraception, post-surgery, and trauma are among other factors. Administration of anticoagulants [7,8] and surgical intervention such as catheter-directed thrombolytics remain a mainstay therapy [8]. The condition, if left untreated, leads to amputation of the gangrenous limb and death, mainly due to the development of pulmonary embolism. In this study, we reported a PCD case of a 51-year-old woman with underlying pelvic malignancy who presented with a one-day history of left lower limb swelling and pain, mimicking acute limb ischemia.

2. PRESENTATION OF CASE

A 51-year-old woman with underlying metastatic cervical adenocarcinoma and receiving palliative chemotherapy presented with a sudden onset of pain and swelling of the left lower limb. She was referred to the surgical team by an emergency physician for acute limb ischemia on the same day upon arrival in the emergency department. Upon reviewing, she appeared breathless and in pain. Her left lower limb was swollen with bluish discoloration, and she was unable to move the toes and ankle. The left distal pulses were well-felt up to the femoral artery. This raised a suspicion of venous ischemia instead of arterial ischemia. The ultrasound doppler revealed a long segment of thrombus from the left common femoral vein up to the external iliac vein. The electrocardiogram showed an S-wave in lead I, a Q-wave, and an inverted T-wave in lead III. Her latest computed tomography (CT) of the thorax and abdomen, which was done one month ago, showed a huge pelvic mass with multiple lung metastases. She was treated with extensive DVT, resulting in PCD and pulmonary embolism secondary to an advanced cervical adenocarcinoma. Anticoagulant was commenced immediately once the diagnosis was made. However, she deteriorated fast to the point where she became more breathless, and the bluish limb extended to the contralateral lower limb was observed. She succumbed to death within 24 hours of presentation, despite the prompt initiation of anticoagulant therapy. CT Angiography was unfortunately not done in view of the patient deteriorating fast.



FIG 1. PHLEGMASIA CERULEA DOLENS DURING FIRST HOUR OF PRESENTATION AND SUBSEQUENT HOUR

3. DISCUSSION

PCD is a limb [9] and life-threatening condition [1]. Venous obstruction and intravascular hypertension cause sequestration of fluid, increasing the compartmental pressure. The arterial system will eventually collapse, thus, resulting in tissue ischemia and limb gangrene. Other differential diagnoses, such as acute cellulitis, peripheral artery disease, and acute arterial occlusion might be considered.

Pulmonary embolism, which contributes up to 10%–15% of hospital deaths with relation to the occurrence of DVT [1], is the biggest factor in mortality in PCD as well.

Ultrasound is the main imaging modality that facilitates the diagnosis of PCD. The inability to compress the vein during ultrasonography is pathognomonic for a thrombus in the vein [9]. Catheter venography and arteriography were mentioned as the gold standard in one case reported for their ability to be therapeutic [9]. However, a case reported in a healthy male patient who presented with a sudden onset of leg swelling shows CT-Angio is superior as it can detect any pelvic mass causing venous compression [10].

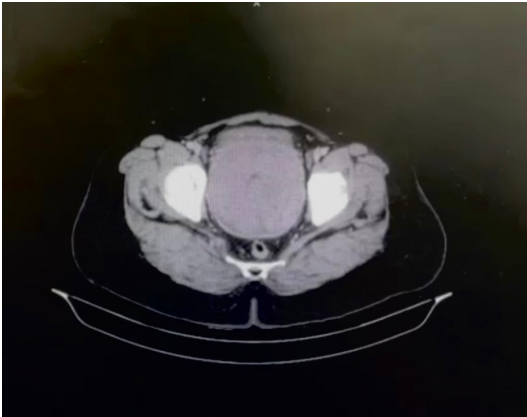


Fig 2. CT-Abdomen one month prior to presentation show huge pelvic mass secondary to metastatic cervical carcinoma.

Early and fast recognition are essential for survival outcomes, as immediate intervention can be initiated. Systemic anticoagulant combined with catheter-directed deep vein thrombolysis has been the treatment for PCD. Fluid resuscitation is essential to replace losses of fluid sequestration in the lower limb [11].

A clinical challenge may arise to differentiate limb ischemia caused by arterial or venous occlusion. PCD of a limb is distinguished by its extensive swelling, out-of-proportion pain, and skin discoloration, while in patients with ischemic limb causes by arterial occlusion, they presented with pulselessness followed by pain, limb paralysis, parasthesia, pallor, and poikilothermia. Venous occlusion caused by thrombosis is detected through ultrasound, while contrast enhanced CT is a recommended imaging method to locate multiple thrombosis that are often seen in patients with peripheral artery disease. However, challenging as it may appear to differentiate both conditions, a patient with a known case of vascular disease indicates the ischemic limb is caused by an arterial thrombus, such as a patient with previous cardiovascular disease who specifies an atherosclerotic or stent- or graft related thrombus. Trauma precipitates an arterial thrombus, and patients with aortic dissection may indicate a similar arterial origin [12]. Nevertheless, both conditions benefit from immediate intervention with both medical and surgical interventions such as intravenous heparin and thromboembolectomy, respectively.

A case reported in a patient with iliac vein compression and deep vein thrombosis showed that open venous thrombectomy promotes fast limb decongestion and contributes to good morbidity and overall mortality for the patient [11]. However, it is worth noting the apparent disadvantages of thrombectomy, such as recurrent thrombosis [11], and the need to perform the procedure under general anaesthesia. In this case, the immediate initiation of anticoagulants is sufficient to prevent venous propagation.

As previously reported, mortality in venous thrombosis is mostly caused by the development of pulmonary embolism. The prognosis remains poor in this case due to the fact that the patient may have already developed massive pulmonary embolism prior to the arrival of the emergency department.

4. CONCLUSION

Common risk factors that contribute to developing PCD are malignancy, surgery and trauma. Though rare and deadly, a high clinical suspicion and prompt diagnosis of PCD are crucial for patients with advanced pelvic malignancy presented with an ischemic limb.

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As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

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As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

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