

# Phlegmasia cerulea dolens in a long distance driver

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## Abstract

Phlegmasia cerulea dolens (PCDs) is a rare and serious complication of massive deep venous thrombosis of the lower extremities, which has a high mortality rate. It is characterized by the sudden onset of severe leg pain, massive edema, cyanosis, venous gangrene, compartment syndrome and arterial compromise, often followed by pulmonary embolism and death. We report one case of a long distance driver who presented with PCDs, complicated by fatal pulmonary embolism.

**Key words:** Compartment syndrome, deep vein thrombosis, phlegmasia cerulea dolens

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## Introduction

Deep vein thrombosis (DVT) of the lower extremities is an important cause of morbidity and mortality, particularly in hospitalized patients. Its pathogenesis is explained by the Virchow's triad of hypercoagulability, blood stasis, and endothelial injury.<sup>[1]</sup> Risk factors for thrombosis are identified in over 80% of patients with DVT, and may be either acquired or inherited.<sup>[2]</sup>

Phlegmasia cerulea dolens (PCD) is a rare ischemia-associated DVT, which may lead to limb loss and even death.<sup>[3]</sup> This condition is characterized by massive venous thrombosis and obstruction of the venous drainage of an extremity, resulting in massive venous congestion and fluid sequestration, venous gangrene, compartment syndrome, and arterial compromise.<sup>[3]</sup> Its optimal treatment remains controversial and can include

anticoagulation, thrombolysis, thrombectomy, and fasciotomy.<sup>[4]</sup>

## Case Report

A 35-year-old man presented to our emergency department with a 3-day history of painful swelling of his left lower limb. He was a long distance driver who regularly drives over 900 km/day. He denied any previous history of DVT, recent trauma, or hospitalization. His family history was negative for coagulopathy.

On examination, he was of normal body habitus, but tachypneic (respiratory rate 36 breaths/min) and tachycardic (pulse rate 100 beats/min), with a blood pressure of 150/110 mmHg. His left leg was remarkable for diffuse swelling and bluish discoloration [Figure 1]. The leg was tender, cold, and with weak distal arterial pulses. Auscultation of the chest revealed decreased breath sounds posteriorly on the right side. The remainder of the examination was unremarkable.

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**Figure 1:** The affected leg with swelling, pallor and discoloration

The admission laboratory workup showed white blood cells (WBC) of  $31 \times 10^9/L$ , hemoglobin of 16.6 g/dl, hematocrit of 50.5%, mean cell volume (MCV) of 77.8 L and a platelet count of  $329 \times 10^9/L$ . Other laboratory results showed a serum creatinine of 217  $\mu\text{mol/L}$ , urea of 12.9 mmol/L, and potassium of 5.8 mmol/L (all elevated above normal) and sodium 126 mmol/L. Chest X-ray revealed a right pleural effusion. An urgent Doppler ultrasound demonstrated DVT of all of the left lower extremity veins, including the left external iliac and femoral veins. Three days prior to admission, his laboratory values revealed the following: WBC  $12.77 \times 10^9/L$ , hemoglobin 12.9 g/dl, hematocrit 40.4%, MCV 81 L, platelet  $254 \times 10^9/L$ , serum urea 7.3 mmol/L, creatinine 78  $\mu\text{mol/L}$ , sodium 138 mmol/L and potassium 4.3 mmol/L. The diagnosis of PCD was made on the basis of the above clinical and ultrasound findings. The patient's PCD was complicated by compartment syndrome, as his left lower limb was tensely distended, cold, and poorly perfused. The right lung findings were consistent with a coexistent pulmonary embolism.

In-patient intravenous fluids, systemic anticoagulation with intravenous heparin, and intravenous antibiotics were immediately initiated, and the patient underwent an emergency lower leg fasciotomy, which initially improved perfusion to his limb. Eight hours after fasciotomy, gangrenous changes were noted in his left foot. The limb was elevated, and intravenous heparin anticoagulation was continued. Despite the above measures, his general condition worsened, and the patient died 24 hours after admission from a suspected pulmonary embolism.

## Discussion

PCD is a rare complication of lower extremity proximal DVT that is characterized by total, or near-total thrombotic occlusion of the venous drainage of the extremity.<sup>[3]</sup> The occlusion leads to massive extravasation of fluid into the interstitial space, which in turn causes interstitial edema and, in severe cases, hypovolemic

shock.<sup>[5]</sup> As a result of fluid extravasation, the vascular hydrostatic pressure is reduced while interstitial pressure increases. A significant drop in arterial hydrostatic pressure and/or an increase in interstitial pressure may lead to vessel collapse.<sup>[3]</sup> In our patient, evidence for intravascular hypovolemia were the development of hemoconcentration and derangement of his renal function tests within the course of illness.

Patients with PCD initially present with severe leg pain and swelling secondary to venous occlusion, which then progress to tissue ischemia and gangrene. Other findings include cyanosis, arterial hypotension, cutaneous blebs or bullae, and absent peripheral pulses.<sup>[3]</sup> Factors that may precipitate PCD include malignancy, hypercoagulable states, anatomic variability, previous DVT, femoral vein catheterization, surgery, heart failure, pregnancy, and prolonged immobility.<sup>[2]</sup> As with our patient, more than 48 hours of immobility in the preceding month is one of the important contributors to DVT.<sup>[6]</sup> Nevertheless, multiple precipitating factors may coexist in one patient. The diagnosis of PCD is established clinically by the presence of four cardinal signs: Edema, purplish discoloration, pain, and severe venous outflow obstruction.<sup>[7]</sup> Radiological investigations that could be performed include contrast venography, duplex ultrasonography, and magnetic resonance venography.<sup>[4]</sup>

The management of PCD varies with its clinical severity. Conservative medical treatment with bed rest, leg elevation, fluid resuscitation, and intravenous heparin anticoagulation is an option for mild and nongangrenous PCD.<sup>[8]</sup> Because large volumes of fluid may be lost into the tissues of the affected lower extremity,<sup>[9]</sup> vigorous fluid resuscitation is necessary to replace these losses and systemic anticoagulation helps prevent propagation of the clot and consequent thromboembolism.<sup>[7,8]</sup> However, conservative management is associated with a high failure rate,<sup>[10]</sup> and other treatment modalities such as venous thrombectomy, surgical fasciotomy and thrombolysis must be considered early in the management of PCD.<sup>[8,10-12]</sup>

Compared to standard anticoagulation therapy, thrombolysis effectively dissolves venous thrombi and consequently minimizes postphlebotic sequelae.<sup>[12,13]</sup> Its main limitation is failure to reach an optimal concentration of the thrombolytic agent at the thrombus site, hence catheter-directed thrombolysis (CDT) is preferred.<sup>[12]</sup> With CDT, a catheter multiple lateral wall openings are introduced and advanced through the thrombotic segment(s) of the affected veins, for delivery of the thrombolytic agent directly into the clots. However, the clinical advantage of this directed approach over standard anticoagulation has recently been questioned.<sup>[14]</sup> In this recent study that involved 90,618 DVT patients, in-hospital mortality was not significantly different

between patients treated with CDT versus standard anticoagulation. In addition, CDT was associated with higher rates of adverse events (blood transfusion, pulmonary embolism, intracranial hemorrhage, and need for vena cava filter placement). Nonetheless, CDT was not an available treatment option for our patient.

Venous thrombectomy is advised in severe PCD cases with venous gangrene, as well as when conservative treatment fails.<sup>[8]</sup> The clinical and hemodynamic effects of thrombectomy are significantly superior to conservative management, especially in patients with extensive ilio-femoral thrombosis that is treated within 3 days of presentation. Thrombectomy results in instant venous decompression, but should be reserved for patients with contraindications to thrombolysis.<sup>[11]</sup> Its main disadvantage is the high risk of recurrent thrombosis and postthrombotic syndrome.<sup>[15]</sup> Fasciotomy is recommended when there is threatening limb ischemia, to relieve compartment pressure.<sup>[11]</sup> In our patient, fasciotomy was done to relieve the compartment syndrome.

This case report highlights a rare but catastrophic complication of the common clinical syndrome of DVT. Regardless of the mode of treatment, mortality associated with PCD remains high.<sup>[14]</sup> Our patient died of massive pulmonary embolism within a few hours after admission.

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#### Conflicts of interest

There are no conflicts of interest.

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