

Rapid Deterioration and Death Caused by Bilateral Phlegmasia Cerulea Dolens

Mejgon Joya, BS; Patricia Khatib, MD; Erin L. Simon, DO

A 58-year-old woman with a history of deep vein thrombosis and pulmonary embolism presented for evaluation of significant back and leg pain and difficulty ambulating.

Phlegmasia cerulea dolens (PCD), a life-threatening complication of deep venous thrombosis (DVT), is characterized by massive iliofemoral thrombus that extends to the collateral veins, leading to fluid sequestration and elevated compartment pressures that ultimately compromise arterial flow. Phlegmasia cerulea dolens can rapidly progress to compartment syndrome and gangrene.^{1,2} The affected limbs of patients with PCD can be hypoxic and appear purple in color due to substantial lack of blood flow, with diminished or absent pulses. Risk factors for PCD include malignancy, hypercoagulable states, venous stasis, contraceptive agents, inferior vena cava (IVC) filter, aneurysm, history of DVT, trauma, heparin-induced thrombocytopenia, femoral vein catheterization, antiphospholipid syndrome, or pregnancy.³⁻⁶ Failure to treat PCD early and aggressively carries an amputation rate of up to 50% and a mortality rate of up to 40%.⁴

We present the case of a patient with

PCD, whose condition rapidly deteriorated despite prompt diagnosis and treatment.

Case

A 58-year-old woman presented to the ED with a 1-day history of back and leg pain and difficulty walking. When asked about the severity of her pain, she rated her leg pain at 10 on a scale of 0 to 10. The patient's history was significant for DVT and pulmonary embolism (PE), for which a Greenfield IVC had been placed and for which she was on prophylactic warfarin therapy. The patient stated that she had been taken off warfarin several weeks prior to presentation in preparation for an elective colonoscopy and dental procedure, but had restarted the warfarin therapy 2 days prior to presentation. She had no history of diabetes mellitus or renal disease.

Initial vital signs at presentation were: blood pressure, 120/91 mm Hg; heart rate, 110 beats/min; respiratory rate, 24 breaths/min; and temperature, 96.6°F. Oxygen saturation was 100% on a nonrebreather mask.

Ms Joya is a summer research fellow; department of emergency medicine research, Cleveland Clinic Akron General, Ohio. **Dr Khatib** is an attending, department of emergency medicine, Cleveland Clinic Akron General, Ohio. **Dr Simon** is an associate professor, Northeast Ohio Medical University; and emergency medicine research director, department of emergency medicine, Cleveland Clinic Akron General, Ohio.

Authors' Disclosure Statement: The authors report no actual or potential conflict of interest in relation to this article.

DOI: 10.12788/emed.2018.0091

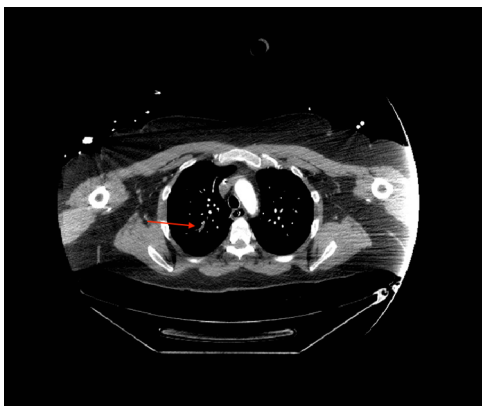


Figure 1. Computed tomography angiogram of the patient's chest demonstrating a large subsegmental pulmonary embolism (red arrow).

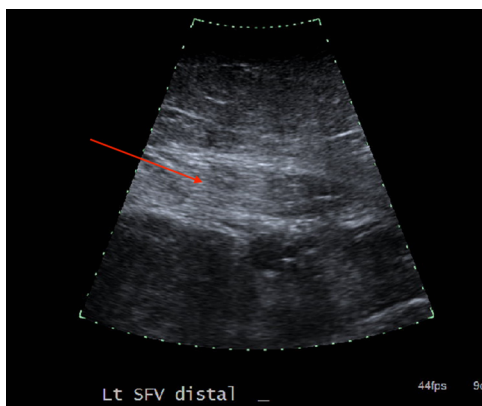


Figure 2. Ultrasound image of the patient's left lower leg demonstrating an extensive blood clot in the superficial femoral vein (red arrow).

On examination, the patient was alert and oriented to person, time, and place, but appeared dyspneic. An electrocardiogram revealed sinus tachycardia. On physical examination, lung sounds were clear to auscultation bilaterally with good air movement, and the abdomen was soft and nontender with normal bowel sounds. The dorsalis pedis and posterior tibial pulses were absent bilaterally, lower extremity capillary refill was 3 seconds, and the legs appeared mildly erythematous and cool to touch. No speech or neurological deficits were present.

Laboratory evaluation was remarkable for metabolic acidosis, venous pH, 7.11; bicarbonate, 11.7; partial pressure of carbon dioxide, 37.6; lactic acid, 8.8 mEq/L leu-

kocytosis, 24,900 u/L; glucose, 296 mg/dL; creatinine, 2.41 mg/dL; and international normalized ratio, 1.36.

Before additional laboratory studies and imaging could be obtained, the patient developed altered mental status, hypotension, and paralysis of the lower extremities. She was orally intubated for airway protection and was given a total of 4 L of normal saline intravenously (IV) for hypotension and acidosis; sodium bicarbonate for metabolic acidosis; norepinephrine for hypotension; fentanyl for pain; and ondansetron for nausea. A central line and arterial line were placed for administering medication and hemodynamic monitoring.

Computed tomography (CT) angiography of the chest, abdomen, and pelvis demonstrated multiple subsegmental bilateral PE with no arterial pathology (**Figure 1**). Beside ultrasound revealed extensive bilateral DVTs involving the superficial and common femoral veins (**Figure 2**). The patient's bilateral DVTs, arterial compromise, and leg cyanosis led to the diagnosis of PCD.

Critical care and vascular surgery services were consulted, and the patient was admitted to the intensive care unit. Since the patient was too unstable to undergo thrombectomy, she was given IV tissue plasminogen activator. Despite aggressive pharmacological treatment, the patient's condition continued to deteriorate. On hospital day 2, the patient's family changed the patient's code status to do-not-resuscitate/comfort-care only; she died shortly thereafter.

Discussion

This case illustrates the severity and complications of PCD and the rapidity with which this condition can deteriorate. At the time of ED presentation, the patient had already developed bilateral PCD, metabolic acidosis, and bilateral PE. Unfortunately, due to decreased venous return, decreased cardiac output, and severe shock, she quickly became unstable and

progressed rapidly to multisystem organ failure leading to death.

Risk Factors

A prior patient history DVT and an IVC filter are both significant risk factors for the progression of DVT to PCD;^{3,6} however, in this case, IVC filter failed to prevent emboli from reaching the lungs. Extensive thrombi led to severely decreased venous return and cardiac output, causing life-threatening shock, ischemia, and metabolic acidosis. A lactic acid level taken on hospital day 2 was elevated at 19 mEq/L, demonstrating the severity, morbidity, and progression of PCD.

Signs and Symptoms

The three cardinal signs that lead to a clinical diagnosis of PCD are edema, pain, and violaceous discoloration or skin mottling.³ Although most commonly found in the lower extremity, PCD can occur in any limb due to occlusion of venous outflow.⁷ Unfortunately, a clinical diagnosis of PCD is not often made until the venous occlusion becomes severe enough to impair arterial flow and cause venous gangrene, tissue ischemia, shock, and death.⁸

Although IVC filters are designed to prevent life-threatening PE, there are risk factors associated with their use. Whether placed recently or decades prior, urgent investigation, such as immediate CT scan, should be undertaken in patients presenting with DVT-like symptoms who have a history of an IVC filter, to ensure the filter has not shifted from its original placement and is not occluding the IVC.

Conclusion

Phlegmasia cerulea dolens is an uncommon vascular emergency, but one that has a high-morbidity and high-mortality rate. This case demonstrates the importance of early diagnosis, aggressive treatment, and the severe complications that can develop in PCD.

There are cases in the literature where patients diagnosed with PCD had a suc-

cessful outcome with pharmacological or surgical intervention such as thrombectomy. Treatment for PCD is most effective when instituted early in onset. As seen in our patient, the tendency for rapid deterioration in PCD can limit potentially life-saving therapeutic options, decreasing the chances of a successful outcome. Emergency physicians, therefore, must be aware of the high-mortality rate associated with this disorder and the possibility of rapid progression from stable to critical condition.

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