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BILATERAL PHLEGMASIA CERULEA DOLENS AFTER WARFARIN REVERSAL FOR ACUTE RECTAL BLEEDING: A CASE REPORT

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Abstract—Background: Deep vein thrombosis (DVT) is a common disease that is diagnosed in approximately 1 in 1000 adults annually. Extensive DVT can lead to life- or limb-threatening diagnoses such as phlegmasia cerulea dolens (PCD), phlegmasia alba dolens, and venous gangrene. PCD, also known as massive iliofemoral venous thrombosis, is rare, and a severe complication of DVT. **Case Report:** We report a case of a 94-year-old bedridden woman with past medical history of dementia, hypertension, pulmonary embolism, DVT, and atrial fibrillation. The patient was admitted to the hospital for bright red blood per rectum and an elevated international normalized ratio (INR) of 5.7. On admission, her dose of warfarin was suspended and she was given 4 units of fresh frozen plasma as well as 10 mg of i.v. vitamin K. She was discharged home with an INR normalized to 1.3 and cessation of her rectal bleeding. At discharge, she was not restarted on warfarin, nor was any bridging therapy used. The patient returned to the Emergency Department a week later for worsening pain and bluish discoloration of her bilateral lower extremities. An ultrasound (US) examination showed that she had developed bilateral PCD, after INR reversal. **Why Should an Emergency Physician Be Aware of This?:** Emergency physicians commonly care for patients who present with acute DVT or treat patients on anticoagulant therapy who require cessation of medications or administration of prothrombotic agents to reverse bleeding. Cases of extensive clot burden leading to PCD have been reported in the literature, however, reports of bilateral PCD secondary to cessation of warfarin have been scarce. PCD should be considered carefully as one of the complications in warfarin reversal, as it

requires immediate attention and surgical intervention to prevent limb loss. © 2017 Elsevier Inc. All rights reserved.

Keywords—complications; DVT; anticoagulants; venous stasis; prothrombotic agents

INTRODUCTION

It is estimated that there are over 600,000 venous thromboembolic events each year in the United States (1,2). In the case of deep vein thrombosis (DVT), some of its more severe complications, such as phlegmasia cerulea dolens (PCD), phlegmasia alba dolens, and venous gangrene, have been noted to be potentially life threatening. PCD initially starts with a large thrombus formation in an extremity. Venous congestion, venous hypertension, and subsequently, arterial spasms as a result of overwhelming edema causes the extremity to become white, painful, and swollen (3). At this stage, the disease is referred to as phlegmasia cerulea alba. As the clot continues to expand into the collateral venous circulation, additional venous congestion creates elevated hydrostatic pressures and causes arterial insufficiency, leading to a violaceous discoloration (3). It is at this point that phlegmasia cerulea alba then develops into PCD. With continued rise in pressures, irreversible tissue loss results in venous gangrene due to inadequate perfusion,

providing a low likelihood of limb viability (4). Our case report details the case of a 94-year-old woman who developed bilateral PCD after international normalized ratio (INR) reversal for rectal bleeding.

CASE REPORT

A 94-year-old bedridden woman with past medical history of dementia, hypertension, pulmonary embolism, DVT, and atrial fibrillation (on warfarin) presented to the Emergency Department (ED) with lower-extremity pain. The patient had been admitted to the hospital approximately a week prior for bright red blood per rectum and a supratherapeutic INR of 5.7. During the prior admission, her warfarin dose was suspended and she was given 4 units of fresh frozen plasma as well as 10 mg of i.v. vitamin K. She was subsequently discharged home when her bleeding stopped and her INR normalized to 1.3 (notably subtherapeutic for prevention of DVT, pulmonary embolism, and atrial fibrillation). At discharge, she was not restarted on warfarin, nor was any bridging therapy used. The patient returned to the ED for worsening pain and bluish discoloration of her lower extremities, which was noted shortly after discharge. On examination, she had bilateral, 2+ lower-extremity pitting edema with violaceous discoloration and diminished peripheral pulses (Figure 1).

A duplex venous ultrasound revealed extensive bilateral DVTs involving the bilateral external iliac veins and extended distally to include the bilateral popliteal and calf veins (Figure 2). Clinical findings were concerning for phlegmasia cerulea dolens. The patient was placed on i.v. heparin. Vascular Surgery and Interventional Radiology were consulted for emergent thrombectomy. The patient was brought to the interventional suite for mechanical thrombectomy and suffered a cardiac arrest shortly into the procedure. She was successfully resuscitated and transferred to the Intensive Care Unit, where she suffered several more episodes of cardiac arrest. After a prolonged discussion with her family, the patient was placed under a “Do Not Resuscitate” order and expired the following day.

DISCUSSION

PCD is one of the most severe complications of DVT. It carries a mortality rate of 40% and an amputation rate of 50% (5). Due to its rare occurrence, the incidence of this disease is still unknown (3). The patient presented in this case report had extensive bilateral venous thrombosis that involved her bilateral external iliac veins and extended distally into her bilateral popliteal and calf veins. If left untreated, patients with PCD may develop thrombi that extend into the capillaries, causing venous



Figure 1. A 94-year-old woman presented with a 1-week history of bilateral lower leg pain. Lower extremity examination revealed cold, violaceous skin changes, decreased peripheral pulses, and pitting edema concerning for phlegmasia cerulea dolens.

gangrene, compartment syndrome, arterial compromise, and ultimately, limb loss (6). Patients with PCD often have associated pulmonary emboli as well as underlying malignancy (7). The cardinal signs of edema, pain, violaceous discoloration, and extensive DVT are needed for the diagnosis (8). Cyanosis is the pathologic finding and usually progresses from distal to proximal portions of the extremity.

In this case, the patient's status for prolonged immobility coupled with a recent INR reversal with fresh frozen plasma and vitamin K likely precipitated the extensive number of clots in her lower extremities, which led to the development of PCD. At the request of her family, which was first informed of the associated risks involved, the patient was brought to the interventional suite for mechanical thrombectomy but had the procedure suspended early due to cardiac arrest. The incidence of PCD is extremely rare. Most cases reported in the literature have been with regard to unilateral PCD. Only a handful of cases of bilateral PCD have been reported (9–15). This report re-emphasizes the extreme rarity of bilateral PCD and its heightened risk of mortality when compared with deep venous thrombosis. Severe complications of DVT, such as PCD, should also be carefully

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