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A rare case of phlegmasia cerulea dolens complicated with iliac vein perforation

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ABSTRACT

Venous thromboembolism (VTE) is a major healthcare problem in the world. It includes pulmonary embolism and deep vein thrombosis (DVT) that result in significant morbidity and mortality. Phlegmasia cerulea dolens (PCD) is a rare but potentially life-threatening severe form of acute DVT, which is characterized by marked swelling of the extremities with pain and cyanosis, and further leads to arterial ischemia due to venous hypertension and compartment syndrome, and ultimately cause gangrene with high amputation and mortality rates. However, there was rare case reported about PCD complicated with venous perforation. Herein, we report a scarce case of left iliofemoral DVT suffering from iliac vein perforation with extraperitoneal hematoma. Although this case treated by iliac vein stenting with cover stent and ultrasound-assisted thrombolysis (EKOS). Patient still finally received below the knee amputation due to reperfusion injury over distal foot despite great improved venous blood flow. Our case should be the first case of PCD complicated with venous perforation. This rare case reminds physicians that venous perforation is a possible complication of PCD and may lead to poor outcome. Early hemostasis by stenting and performing thrombectomy treatment as soon as possible is suggested to save the life and limb.

Keywords: deep vein thrombosis; phlegmasia cerulea dolens; ultrasound-assisted thrombolysis; venous perforation

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Introduction

Venous thromboembolism (VTE) is a major healthcare problem, and its consequent complication including pulmonary embolism (PE) and deep vein thrombosis (DVT) that result in significant mortality, and morbidity. Phlegmasia cerulea dolens (PCD) is a rare but potentially life-threatening severe form of acute DVT, which is characterized by marked swelling of the extremities with pain and cyanosis, and further leads to arterial ischemia due to venous hypertension and compartment syndrome, and ultimately cause gangrene with high amputation and mortality rates [1-2]. However, there was rare case reported about PCD complicated with venous perforation and the treatment strategy was unknown in the literature. Herein, we report an extremely rare case of PCD complicated with iliac vein perforation and also report our treatment strategy and further patient's outcome.

Figure 1. Left lower extremity swelling and distal foot cyanosis (Phlegmasia cerulea dolens complicated with compartment syndrome)

Case presentation

A 50-year-old woman presented to the emergency department (ED) due to progressive swelling, painful sensation, and cyanotic change over the left foot (Figure 1A & 1B). Profound shock (59/25 mmHg) was also noted at ED. Initial laboratory data showed severe anemia (hemoglobin 9.3 g/dL), thrombocytopenia (42000/ul), elevated D-Dimer (73 mg/L), and metabolic acidosis. For unknown reason of dropped hemoglobin, abdominal CT was performed which showed thrombosis in the left femoral, iliac veins and inferior vena cava. In addition, extraperitoneal hemotoma was also noted in the pelvis (Figure 2). Emergent angiography by the radiologist was arranged but no overt evidence of active arterial bleeding. They also tried to find if bleeding in the venous system but failed due to left iliac and femoral vein thrombosis. The patient was then admitted

to the intensive care unit due to unstable hemodynamic status with vasopressor use. During the hospitalization, left lower extremity cyanosis with tense skin and bullae was found and fasciotomy was emergently performed due to compartment syndrome. Cardiologist was then consulted for PCD & endovascular therapy was suggested. Initial venography showed left iliofemoral DVT with venous perforation over left external iliac vein, which may related to the high pressure of venous thrombosis. Viabahn stent was used to seal the perforation and angioplasty was performed in combination with ultrasound-assisted catheter directed thrombolysis (CDT by EKOS device). After several days of urokinase infusion, patient blood flow became greatly improved and apixaban was given for further anticoagulation treatment. However, despite successfully regained blood flow of left foot. Left distal foot suffered from reperfusion injury with some gangrene change. Initial wound care was suggested instead of amputation by the plastic surgeon but the patient and family refused long-term wound care. The patient finally received below-knee amputation.

Figure 2. Computed tomography showed Left iliofemoral deep vein thrombosis with extraperitoneal hematoma (Yellow arrow: extraperitoneal hematoma)

Discussion

PCD is a rare but fulminant life threatening condition, which is caused by acute massive venous thrombosis [1-2]. Patients may suffer from severe leg pain, swelling, cyanosis, compartment syndrome, venous gangrene, and arterial compromise. Release of inflammatory mediators can cause a vasodilatory state, which may develop circulatory collapse and hypovolemic shock [3]. The pathophysiology of PCD involves extensive venous obstruction leading to increased interstitial tissue pressure, arrest of capillary blood flow, tissue ischemia

and further gangrene formation, which can cause limb loss and even mortality. Reported mortality rates range from 20% to 41% and amputation rates range from 12% to 50% [4]. Initial management should include absolute bed rest, leg elevation, fluid resuscitation, and the intravenous heparinization. Thrombus removal such as surgical thrombectomy or endovascular therapy using CDT or percutaneous mechanical thrombectomy (PMT) should be performed as soon as possible to restore venous outflow. However, disease outcome are strongly depend on the initial disease severity of PCD and the physician's experience [5-6]. Early, quick detection and effective treatment may save such patients limbs and life. In addition, early fasciotomy to relief pressure due to acute compartment syndrome is also needed for limb saving.

PCD is a relatively rare and severe form of DVT and there was no case of PCD complicated with venous perforation reported in the literature. Our case should be the first case of PCD complicated with external iliac vein perforation and extraperitoneal hematoma episode. The mechanism of iliac vein perforation was considered to be related to severe venous hypertension and compartment syndrome. Because PCD is already a life-threatening emergency, venous perforation may cause further higher mortality in this patient group. In our case, the cause of amputation may be related to the delayed reperfusion of left venous system which caused tissue damage and distal foot gangrene formation.

Conclusion

Our case is an extremely rare case of PCD complicated with venous perforation. This case reminds physicians that venous perforation is a possible complication of PCD and may lead to poor outcome. Early hemostasis and performing thrombectomy treatment as soon as possible is suggested to not only save the life but also save the limb.

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