

中文題目：罕見嚴重靜脈血栓引起的疼痛性大腿青腫及靜脈破裂

英文題目：A rare case of phlegmasia cerulea dolens complicated with iliac vein 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 complication 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] However, there was rare case reported about PCD complicated with venous perforation. Herein, we report a rare case of left iliofemoral DVT suffering from acute compartment syndrome of left leg. However, computed tomography revealed left iliofemoral DVT & possible left iliac vein perforation with extraperitoneal hematoma. Progressive left leg cyanosis and bullae formation was noted. PCD was diagnosed and emergent fasciotomy was performed. After unstable hemodynamic improved, this patient was treated by iliac vein viabahn stenting for venous perforation & further catheter-directed thrombolysis for left iliofemoral DVT.

Case presentation

A 50-year-old woman presented to the emergency department (ED) due to dizziness, dyspnea and progressive swelling, painful sensation over the left foot. Profound shock (59/25 mmHg) was also noted at ED. Initial lab 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 thromboses in the left femoral, iliac veins and inferior vena cava. In addition, extraperitoneal hemotoma was also noted in the pelvis. Emergent trans-arterial embolization by the radiologist was arranged but no overt evidence of active arterial bleeding. She 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 EKOS enhanced CDT treatment. 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 gangrene change. The patient still finally received below-knee amputation.



Figure 1. Deep vein thrombosis with extraperitoneal hematoma



Figure 2. left lower extremity cyanosis with compartment syndrome

Discussion

PCD is a fulminant, limb and life threatening condition, which is caused by acute massive venous thrombosis. [2] Patients may present with sudden and 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 hypovolaemic shock. [3]

The pathophysiology of PCD involves extensive venous, obstruction leading to increased interstitial tissue pressure, arrest of capillary blood flow, tissue ischemia and ultimately gangrene, which can cause limb loss and even death. Reported mortality rates range from 20% to 41% and reported amputation rates among survivors 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, the treatment outcome seems to strongly depend on the initial grading severity of PCD and the physician's experience. [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.

Although PCD is a rare and severe form of DVT, 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. Physicians should keep in mind this complication and treat PCD patients as soon as possible to avoid the amputation or even mortality risk.

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