

A painful massive cyanotic edema of the left leg

Alessandra Marchetti, Guglielmo Guarona, Fabrizio Elia

Emergency Medicine, San Giovanni Bosco Hospital, Torino, Italy



A 70-year-old woman presented to our Emergency Department because of acute pain and swelling in her left lower limb. She suffered from depressive syndrome and hypertension, and she had a previous history of breast and uterine cancer. She denied fever and trauma. Her medication included olmesartan/hydrochlorothiazide and unspecified antidepressants. Physical examination revealed blood pressure of 90/60 mmHg, heart rate of 78 bpm, peripheral oxygen saturation of 94% in room ambient, and unilateral edema of the left lower limb with bluish skin discoloration, decreased peripheral sensitivity, and weak peripheral pulses. Laboratory findings showed White Blood Cell count (WBC) $5.53 \times 10^9/L$, Hemoglobin (Hb) 12.5 g/dL, Platelet count (PLT) 137,000/ μL , International Normalized Ratio (INR) 1.22, glucose 449 mg/dL, creatinine 1.19 mg/dL (nv 0.5-0.96 mg/dL), C-reactive protein 0.26 mg/dL (nv <1.0 mg/dL), D-dimer 28,920 ng/mL (nv <500 ng/mL), fibrinogen 182 mg/dL (nv 200-400 mg/dL).

Question

Given these results, which is the possible diagnosis?

1. Cellulitis
2. Lymphedema
3. Phlegmasia Cerulea Dolens
4. Acute arterial occlusion

Correspondence: Alessandra Marchetti, Emergency Medicine, San Giovanni Bosco Hospital, Torino, Italy.
Tel.: +39-3206055080.
E-mail: alessandra.marchetti93@gmail.com

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Answer

The correct answer is Phlegmasia Cerulea Dolens (PCD). PCD is a rare, life-threatening massive deep vein thrombosis of the limbs, with a higher incidence rate in the lower extremities (90%), particularly the left leg. The iliofemoral segment is almost always involved and occluded in the lower extremities due to the anatom-

ical relationship between the right iliac artery overlying the left iliac vein.¹ PCD is characterized by complete obstruction of the venous outflow and consequent increased compartment pressure with limb ischemia and venous gangrene.² Its most pathognomonic feature is the presence of cyanosis. Paraesthesia and motor weakness can be present if edema causes severe arterial compromise and compartment syndrome.

PCD is more common in women than men. The highest incidence is in the fifth and sixth decades of life. It is a real medical emergency with a high incidence of mortality and limb loss that requires immediate evaluation and treatment.³ The risk of limb amputation and the estimated mortality are 20-50% and 20-40%, respectively.⁴⁻⁶ Given the involvement of the iliofemoral segment, PCD is associated with significant post-thrombotic morbidity and high recurrence rates if not treated adequately.

The etiology remains unknown in approximately 10% of cases. The leading causes are malignancy (20-40%), followed by hypercoagulable disorders, venous stasis or insufficiency, use of hormonal therapy or oral contraceptives, prolonged immobilization, May-Thurner syndrome (left iliac vein compression between the overlying right iliac artery and the lumbar spine), spinal surgery, trauma, pregnancy, IVC filter placement, central venous catheterization, and others (*i.e.*, inflammatory bowel disease, heart failure).²⁻⁸ The American Society of Vascular Surgery suggests removing the thrombus directly (grade 1A) or using medicine to remove the thrombus (grade 2C), if available.⁷ The evidence regarding PCD management is insufficient, and a universal consensus on a standard procedure has not yet been reached. Consequently, personnel from each center administrates different treatments based on their experience and available resources.²

In this case report, a Computed Tomography (CT) scan of the chest, abdomen, and lower limbs was performed and showed massive venous thrombosis of the left iliac vein and hypoplastic inferior vena cava; pulmonary embolism was excluded. Unfractionated heparin was started, and vasopressors were administered because of ongoing hemodynamic instability. The patient underwent pharmaco-mechanical catheter-directed thrombolysis, and a self-expanding stent was placed in the common iliac vein.

Unfortunately, the procedure was complicated by hemorrhagic shock and disseminated intravascular coagulation. The patient passed away shortly after admission.

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