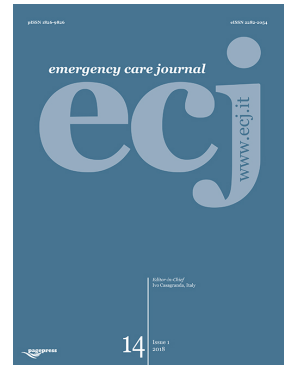


Emergency Care Journal

Official Journal of the [Academy of Emergency Medicine and Care \(AcEMC\)](#)



eISSN 2282-2054

<https://www.pagepressjournals.org/index.php/ecj/index>

Publisher's Disclaimer. E-publishing ahead of print is increasingly important for the rapid dissemination of science. The **Early Access** service lets users access peer-reviewed articles well before print / regular issue publication, significantly reducing the time it takes for critical findings to reach the research community. These articles are searchable and citable by their DOI (Digital Object Identifier).

The **Emergency Care Journal** is, therefore, e-publishing PDF files of an early version of manuscripts that undergone a regular peer review and have been accepted for publication, but have not been through the typesetting, pagination and proofreading processes, which may lead to differences between this version and the final one. The final version of the manuscript will then appear on a regular issue of the journal.

E-publishing of this PDF file has been approved by the authors.

Emerg Care J 2024 [Online ahead of print]

To cite this Article:

Marchetti A, Guarona G, Elia F. **A painful massive cyanotic edema of the left leg.** *Emerg Care J* doi: 10.4081/ecj.2024.12877

©The Author(s), 2024
Licensee [PAGEPress](#), Italy

Note: The publisher is not responsible for the content or functionality of any supporting information supplied by the authors. Any queries should be directed to the corresponding author for the article.

All claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article or claim that may be made by its manufacturer is not guaranteed or endorsed by the publisher.



A painful massive cyanotic edema of the left leg

Alessandra Marchetti, Guglielmo Guarona, Fabrizio Elia

Emergency Medicine, San Giovanni Bosco Hospital, Torino, Italy

Correspondence: Alessandra Marchetti, Emergency Medicine, San Giovanni Bosco Hospital, Torino, Italy.

Tel.: +39-3206055080.

E-mail: alessandra.marchetti93@gmail.com

Key words: acute arterial occlusion, cellulitis, lymphedema, deep vein thrombosis.

Contributions: all the authors made a substantive intellectual contribution. All the authors have read and approved the final version of the manuscript and agreed to be held accountable for all aspects of the work.

Conflict of interest: the authors declare no potential conflict of interest.

Funding: none.

Availability of data and materials: all data underlying the findings are fully available upon reasonable request to the corresponding author.

Ethics approval and consent to participate: the article contains no personal medical information, and the patient is not identifiable. The photos are original and there are no copyright constraints.

Informed consent: the patient provided consent for access to medical records at the time of admission.



Descriptive legend

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 a unilateral edema of the entire 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

Answer. The right 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 anatomical 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 main 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 administers 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 pharmacomechanical 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.

References

1. Gardella L, Faulk J. Phlegmasia Alba and Cerulea Dolens. 2022. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK563137/>
2. Said A, Sahlieh A, Sayed L. A comparative analysis of the efficacy and safety of therapeutic interventions in phlegmasia cerulea dolens. *Phlebology* 2021;36:392-400.

3. Chinsakchai K, Ten Duis K, Moll FL, de Borst GJ. Trends in management of phlegmasia cerulea dolens. *Vasc Endovascular Surg* 2011;45:5-14.
4. Oguzkurt L, Tercan F, Ozkan U. Manual aspiration thrombectomy with stent placement: rapid and effective treatment for phlegmasia cerulea dolens with impending venous gangrene. *Cardiovasc Intervent Radiol* 2008;31:205-8.
5. Oguzkurt L, Ozkan U, Demirturk OS, Gur S. Endovascular treatment of phlegmasia cerulea dolens with impending venous gangrene: manual aspiration thrombectomy as the first-line thrombus removal method. *Cardiovasc Intervent Radiol* 2011;34:1214-21.
6. Elsaid AS, Alqattan AS, Elashaal E, et al. The ugly face of deep vein thrombosis: Phlegmasia cerulea dolens – case report. *Int J Surg Case Rep* 2019;59:107-10.
7. Meissner MH, Gloviczki P, Comerota AJ, et al. Early thrombus removal strategies for acute deep venous thrombosis: clinical practice guidelines of the Society for Vascular Surgery and the American Venous Forum. *J Vasc Surg* 2012;55:1449–146.
8. Mumoli N, Invernizzi C, Luschi R, et al. Phlegmasia cerulea dolens. *Circulation* 2012;125:1056-7.

Submitted: 29 July 2024

Accepted: 29 August 2024

Early access: 26 September 2024