Phlegmasia Cerulea Dolens

Nicola Mumoli, MD; Claudio Invernizzi, MD; Riccardo Luschi, MD; Giovanni Carmignani, MD; Alberto Camaiti, MD; Marco Cei, MD

An 83-year-old man with a recent history of thyroid carcinoma presented with sudden pain and swelling of the left leg with bluish discoloration and livedo reticularis throughout. Two days before admission, swelling of the left lower extremity developed, and the diagnosis of partial left femoral vein thrombosis was confirmed by venous duplex ultrasonography and compression ultrasound. Anticoagulation with low-molecular-weight heparin and oral warfarin was initiated, and the patient was discharged home receiving maintenance anticoagulant therapy and wearing compression stockings.

On arrival, the patient’s arterial blood gas analysis and 12-lead ECG were normal. His blood pressure was 127/78 mm Hg, pulse rate was 84 bpm, and respiratory rate was 16 breaths per minute. His international normalized ratio was 1.2 (normal range, 0.8–1.2) and D-dimer level was 2530 μg/L (normal range, <190 μg/L). On physical examination, the left leg was markedly swollen, violaceous, painful, tender, and slightly warm up to the inguinal regions (Figure 1). Compression ultrasound was performed again, and an occlusive thrombus was visualized from the left common iliac vein to the calf veins (Figure 2). A diagnosis of phlegmasia cerulea dolens was made, and to prevent potential irreversible venous gangrene and subsequent limb loss, the patient underwent catheter-directed thrombolysis combined with thrombectomy and showed subsequent symptomatic improvement. Seven days later, during oral anticoagulation therapy, the limb discoloration was entirely resolved (Figure 3), and recanalization of the limb vessels was apparent on power Doppler (Figure 4).

Phlegmasia cerulea dolens is a rare syndrome caused by diffuse venous thrombosis that is characterized by sudden

Figure 1. The left leg is swollen, violaceous, and tender up to the inguinal regions.

Figure 2. Compression ultrasound scan of the left common femoral vein shows occlusive thrombus that proved to be incompressible.

Figure 3. Seven days later, discoloration and swelling of the left leg were entirely resolved.

From the Departments of Internal Medicine (N.M., R.L., G.C., A.C., M.C.) and Vascular Surgery (C.I.), Ospedale Civile Livorno, Livorno, Italy. Correspondence to Nicola Mumoli, MD, Department of Internal Medicine, Ospedale Civile Livorno, Viale Alfieri 36, 57100, Livorno, Italy. E-mail n.mumoli@usl6.toscana.it (Circulation. 2012;125:1056-1057.) © 2012 American Heart Association, Inc. Circulation is available at http://circ.ahajournals.org DOI: 10.1161/CIRCULATIONAHA.111.051912
pain, swelling, purple ecchymosis, and arterial ischemia with loss of distal pulses. The so-called blue phlebitis is an often fatal form of venous thrombosis and frequently results in shock, venous gangrene, pulmonary embolism, and death. To make the correct diagnosis, 4 cardinal signs are necessary: edema, violaceous discoloration, pain, and severe venous outflow obstruction. Risk factors include malignancy, femoral vein catheterization, heparin-induced thrombocytopenia, antiphospholipid syndrome, surgery, heart failure, and pregnancy. Currently, although randomized trials are not still available and guidelines for treatment are still not clearly documented, 3 therapeutic options are advocated alone or in combination: anticoagulants, thrombolytic therapy, and venous thrombectomy. This case demonstrates that a rapid, aggressive, multimodal approach to phlegmasia cerulea dolens can be also used in the elderly population and can result in successful limb salvage. Finally, prospective studies are needed to establish the best treatment options in these patients; however, pending long-term follow-up, a highly individualized approach is recommended to treat these patients.

Disclosures

None.

References

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