

Case Report

Phlegmasia Cerulea Dolens Report of a Case with Unusual Presentation

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Abstract

Phlegmasia cerulea dolens is a rare entity secondary to severe Deep Vein Thrombosis (DVT), usually in the lower extremities, and associated with rapidly progressive pain and marked swelling that may compromise the perfusion of the limb. This situation may lead to venous gangrene and the limb's amputation secondary to a compartment syndrome.

We present the case of a patient with a surgical history of lumbar spine surgery three months before the start of the described symptoms. She referred a clinical description of rapidly progressive edema and the appearance of cerulean colour in her left lower limb.

With diagnostic aids, acute DVT was confirmed caused by extrinsic compression of the left iliofemoral axis secondary to chronic retroperitoneal hematoma.

Keywords: Phlegmasia; Venous gangrene; Venous thrombosis

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Background

The term phlegmasia is linked to the classical description of Trousseau's syndrome, due to the clear association of its presentation with oncological diseases [1,2]. Common causes of phlegmasia include benign pathological situations such as thrombophilia, sepsis, post-surgical conditions, heart failure and interventional venous accesses in the femoral vein. It occurs slightly more frequently in women than in men, in a ratio of 4:3 and with a preference for the left lower limb. It is commonly explained by the existence of venous compression, physiological or otherwise, of the right common iliac artery on the common left iliac vein.

It can occur at any age, although its peak incidence is set between the 5th and 6th decade of life. Its classical clinical presentation has been described as a sudden onset of pain in about 75% of cases, followed by edema and a violaceous skin discoloration. The physical examination shows painful edema of the limb and a bluish colour. In addition, the patient may present some degree of shock, absence of pulse in the lower extremity and coldness of the limb. If the condition is allowed to evolve, bullae, paresthesias, motor paralysis, renal failure, venous gangrene and even death may occur.

This disease has a risk of amputation between 12 and 25% of described cases, and a mortality rate close to 25%, where about 30% is due to a pulmonary thromboembolism. The diagnosis is fundamentally clinical and requires only confirmation with an ultrasound study [3].

Presentation of the Case

Female patient, 63 years old, without known allergies, or significant pathological history, who was being medically treated for chronic low back pain, which worsens and presents pain irradiated to the postero-external surface of the LI extremity, and paresthesias of left metamer S1 distribution. Lumbar MRI shows lumbar spondylosis, left L5-S1 herniated disc on a lateral recess syndrome. There is also segmental instability.

Since the patient was not improving with conservative treatment, she underwent surgery, performing a L5-S1 left hemilaminectomy, facetectomy, foraminotomy and discectomy with hard-root decompression. Transpedicular arthrodesis L5-S1 with distraction maneuvers - reduction with titanium system (compatible resonant) Romeo by Spineart and arthrodesis on transverse processes with autologous bone graft and B Gel. The patient recovery is satisfactory after a prophylactic doses treatment with LMWH until full mobility is recovered (three weeks).

During the third postoperative month and while walking, she suddenly notices progressive edema of the entire left lower limb, initially painlessly, but rapidly progressive. Given these symptoms, she decides to turn to the emergency room at the "Gómez Ulla" Military Hospital in Madrid, Spain. She had a marked swelling of the lower limb which obliged the medical personnel to remove her clothes with scissors before attending her. During the physical examination,

the patient is found with stable vital signs, without cardiac arrhythmia, no dyspnea nor chest pain. Arterial pulses were present and symmetric in both lower extremities, there was lower limb edema with an important difference in diameter compared to the contralateral limb and a cerulean discoloration of the entire limb (Figure 1). With a diagnosis of severe Deep Vein Thrombosis (DVT) associated to phlegmasia, anticoagulation with LMWH and complete rest in the Trendelenburg position were indicated.



Figure 1: Clinical finding. Notice the edema and changes of perfusion of the skin on the left leg.

An abdominal-pelvic angio-CT was performed, which confirmed the presence of DVT of the left iliofemoral axis and chronic compressive hematoma on left iliac venous axis whose most probable cause was the venous shearing caused by the osteosynthesis material (Figures 2 and 3).



Figure 2: Angio-CT. Notice the presence of the tip of the osteosynthesis screw protruding into the retroperitoneal space.

The patient evolves favorably, having resolved the edema, the pain and the perfusion of the limb, being discharged with oral anticoagulation and elastic compression stockings.

After three months of anticoagulation, a vascular venous echodoppler was performed, finding repermeabilization of the left iliac venous axis and minimal post-thrombotic changes (thickening of the wall and discrete reduction of its caliber when compared to the right iliac venous axis). The total anticoagulation time was six months.

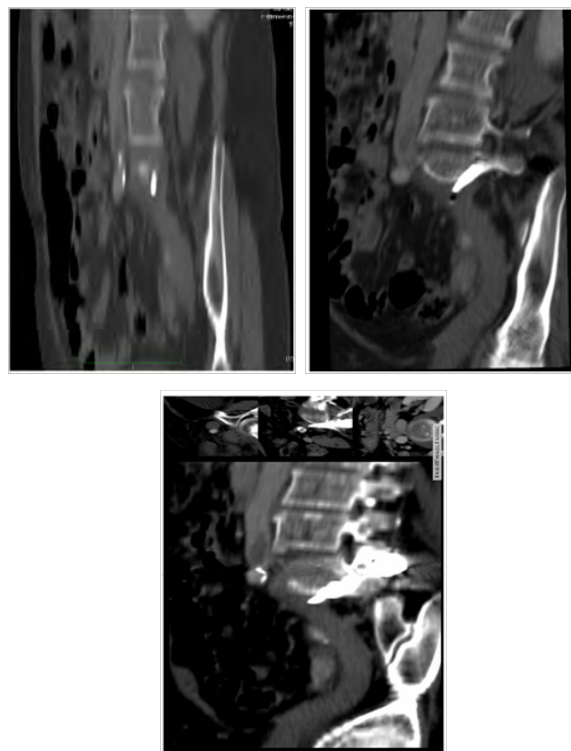


Figure 3: Reconstruction of the Angio-CT. Notice the presence of compressive hematoma on the left iliac venous axis and associated venous thrombosis.

Discussion

There are publications in the literature that describe the behavior and the natural history of both phlegmasias, dawn and cerulean, involving the concept of “venous gangrene” [2], clearly indicating that it is a serious entity with a reserved prognosis [3]; and that despite the technological advances in endovascular surgery that would suggest the ideal management of this type of pathology [4-6], there is no consensus regarding its management and that it is not free of complications [7]. Some authors report good results with conservative management [8], others with surgery and that offers a range of therapeutic options [9].

The case presented was medically managed with good results, but we are aware that a kind of severity score should be established, because undoubtedly, in cases of greater complexity and aggressiveness than we have described, more daring actions such as mechanical thromboaspiration will be required, with or without the implantation of a cava tempor filter, the use of fasciotomies, venous stenting, etc.

Conflict of Interests

The authors declare that they have no conflicts of interest.

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