Phlegmasia cerulea dolens in the Emergency Department: A case report

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ABSTRACT Background: Phlegmasia cerulea dolens is a rare form of deep venous thrombosis (DVT) characterised by massive thrombosis of the veins in a lower extremity. Presumptive diagnosis is made on clinical grounds and confirmed with a point-of-care ultrasound (POCUS). Diagnostic failure can lead to limb loss and even death. Case: We describe a case of a 38-year-old male who presented to the emergency department (ED) with painful cyanotic swelling of his right lower extremity. Clinical exam and POCUS confirmed the diagnosis of phlegmasia cerulea dolens. This patient was successfully treated with anticoagulation and endovascular thrombolytic therapy. Conclusion: This report emphasises the importance of early diagnosis of the disease with POCUS, which can confirm the diagnosis of phlegmasia within minutes of clinical suspicion. Additionally, it describes the successful therapy with anticoagulation and endovascular thrombolysis.

KEYWORDS Phlegmasia cerulea dolens, DVT, POCUS, Cellulitis, gangrene, phlegmasia alba, venous thrombosis, amputation, thrombectomy

Introduction

The incidence of DVT is increasing in developed nations, with a calculated incidence of about 1 per 1000 adults [1,2,10]. Significant outcomes of venous thrombosis include pulmonary embolism (PE), recurrence, post-thrombotic syndrome, phlegmasia, limb loss and even death.

Phlegmasia is a rare, but severe form of DVT. It results from massive thrombosis that occurs within the deep veins of a lower limb causing ischemia and possible limb loss. The disease encompasses a spectrum beginning from phlegmasia alba dolens (PAD) to phlegmasia cerulea dolens (PCD). In the early stages of the disease (PAD), the significant burden of vein thrombosis occludes the deep veins of the limb, causing arterial ischemia, and slowing the venous drainage that remains somewhat patent through the spared collateral veins. This results in a milky white appearance of the limb. PCD occurs at a later stage due to the increased venous outflow pressures and occlusion of the collateral veins. This results in severe swelling, cyanosis and compartment syndrome. The final stage of the spectrum is venous gangrene [3].

Clinical symptoms include sudden onset of pain, swelling, cyanotic discolouration of the affected limb due to venous congestion. In more advanced cases, gangrene can be seen. Early diagnosis is critical as PCD is often reversible when treated promptly [2]. Clinical suspicion of PCD is confirmed with POCUS [4], a fast, affordable and noninvasive diagnostic imaging modality readily available in nearly all emergency departments.

We present a rare case of phlegmasia cerulea dolens, which was quickly suspected in the ED on clinical grounds and confirmed with POCUS. This led to the prompt establishment of therapy and successful recovery of the patient.

Case report

A 38-year-old male presented to the emergency department complaining of painful swelling with purplish discoloration of his right lower extremity with associated weakness and numbness (Fig. 1). Except for being an active smoker (8 pack-year smoking history), he had no other significant past medical history.
Categorically, he denied any history of coagulation disorder or recent trauma. On physical exam, the right lower extremity was cyanotic and swollen. Both bilateral posterior tibial and dorsalis pedis artery pulses were palpable. PCD was clinically suspected, and the patient underwent a POCUS (Fig. 2), with a 2-point simplified compression test. The results showed a lack of compressibility of the right common femoral vein, right greater saphenous vein and right popliteal vein. Subcutaneous (SC) Fondaparinux was immediately administered, and a Doppler ultrasound of the right lower limb was requested as per protocol. The Radiology Department again confirmed the diagnosis of Doppler ultrasound. The patient was admitted to the hospital, and endovascular thrombolytic therapy was administered. The patient subsequently improved uneventfully.

Discussion

Phlegmasia cerulea dolens (PCD) is a rare, but extreme form of DVT that results in massive thrombosis of the deep venous system in a lower limb. Its incidence is not known. Risk factors include cancer, hypercoagulability, previous surgery, immobilisation, male gender or tobacco use.

The early form of phlegmasia, PAD, is characterised by thrombosis of the major deep veins and arterial ischemia. It has a milky white appearance. The late form of phlegmasia, PCD, is due to increased venous outflow pressures causing occlusion of the deep and collateral veins, which result in a cyanotic and swollen appearance. If untreated, this can progress to the point of becoming an irreversible condition in the form of gangrene. Further complications include fluid sequestration and circulatory shock.

Clinical findings and physical exam are key in the process of early diagnosis. However, it can be quickly diagnosed with POCUS [4, 5] and if needed, confirmed with duplex ultrasonography. Imaging characteristics of DVT on ultrasound include non-compressible, enlarged and hyperechogenic veins showing lack of flow on duplex ultrasound. The advantages of POCUS include the portability, affordability, noninvasive modality and availability in almost every ED.

Contrast-enhanced computed tomography (CT) is rarely used. However, if performed, it can show the extent of the thrombus, especially in the pelvis where ultrasound is less sensitive. For example, it can show the proximal extent of the thrombus, which can even reach the common iliac vein or go beyond the iliac bifurcation (Fig. 3).

Treatment of phlegmasia should be started as soon as possible. Medical treatment includes anticoagulation with an elevation of the affected limb. Recent literature suggests that subcutaneous low-molecular-weight heparins such as Enoxaparin [6] or Fondaparinux [7] are safe and effective in the treatment of PCD. New oral anticoagulants, Factor Xa inhibitors (rivaroxaban, apixaban, edoxaban) and oral direct thrombin inhibitors (dabigatran) can be prescribed in DVT, but are not suitable for PCD [8].

Surgical treatment of PCD involves endovascular intervention or open surgical thrombectomy. Endovascular targeted thrombolytic therapy is the intervention of choice and has been proven to be effective, and safe (Fig. 4) [9]. In some cases, thrombolytic therapy can be combined with endovascular mechanical thrombectomy, depending on the extent of the thrombus burden and local experience. Absolute contraindications for endovascular thrombolysis include recent head trauma, recent cerebrovascular accident (less than two months), severe hypertension, allergy to thrombolytic agents or active bleeding in a noncompressible space.

Open surgical thrombectomy is an alternative for patients who generally have contraindications for an endovascular approach.

Conclusion

This case report emphasizes the need for early clinical suspicion of phlegmasia cerulea dolens which can be quickly diagnosed with POCUS to expedite limb-saving or even life-saving treatment.
Figure 3A: Point of care ultrasound (POCUS) showing occlusion of the right common femoral vein (CFV) due to an acute thrombus. Coronal contrast-enhanced CT (of a different patient) showing complete thrombosis of the right common iliac vein (RCIV), with some extension into the iliac bifurcation. The left common iliac vein (LCIV) remains patent. Figure 3B: Axial contrast-enhanced CT at the level of the hips showing complete thrombosis of the right common femoral vein (V). Compare with the ipsilateral patent and usually enhancing right femoral artery (A) and contralateral left common femoral vein (V') and left femoral artery (A'). Figure 3C: Axial contrast-enhanced CT at the level of the upper femoral diaphyses is again showing the thrombus in the right femoral vein (V) with overlying subcutaneous inflammation. The left femoral vein (V') is patent and contrast-enhancing.

Take home messages
1. Phlegmasia cerulea dolens is an extreme form of DVT that causes painful cyanotic swelling of a limb due to massive DVT. It can ultimately lead to irreversible gangrene, limb loss and even death.
2. Early suspicion of the disease is critical and can be quickly confirmed with Point of Care Ultrasound, performed in the ED.
3. Early diagnosis is critical to establish a prompt treatment.
4. Treatment should be started as soon as the diagnosis of PCD is made. Anticoagulation (unfractionated and low-molecular-weight heparin) and endovascular thrombolysis are the mainstays of treatment.

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References