

## Phlegmasia Cerulea Dolens in a Patient with Newly Diagnosed Rheumatic Disorder

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**Article History**

Received: 03.09.2018

Accepted: 16.09.2018

Published: 30.09.2018

**DOI:**

10.36347/sjmcr.2018.v06i09.013



**Abstract:** Phlegmasia cerulea dolens (PCD) is a rare and critical complication of deep vein thrombosis (DVT) that results from extensive thrombotic occlusion of the major and collateral veins of an extremity, which back up into the arterial vasculature causing limb cyanosis then eventual gangrene and limb death. The prompt recognition of this condition is crucial to prevent progression of clot burden resulting in surgical intervention. Doppler ultrasound investigation of both venous and arterial vasculature is used to determine DVT. Any cause of DVT, both provoked or unprovoked can lead to PCD. Once identified, anticoagulation must be started immediately. A common rheumatologic condition such as Systemic Lupus Erythematosus (SLE) is known to cause coagulopathies and thrombosis, but currently PCD being a presenting symptom of SLE has not yet been documented in literature. We describe a case where PCD is the presenting manifestation of SLE and highlight the need for rapid recognition to prevent irreparable limb damage.

**Keywords:** Phlegmasia Cerulea Dolens, Vascular Surgery, Rheumatology, Deep Vein Thrombosis, Systemic Lupus Erythematosus, Anticoagulation, Antinuclear antibody.

### INTRODUCTION

Phlegmasia cerulea dolens (PCD) coined from Greek terminology translates to a painful, blue inflammation of the extremities. It is a rare and critical complication of deep vein thrombosis (DVT) that results from extensive thrombotic occlusion of the major and collateral veins of the extremity. This thrombotic occlusion compresses the arterial vasculature leading to limb cyanosis, gangrene and death. Common presenting symptoms include acute onset of limb tenderness, skin discoloration, edema, sensory loss, and cyanosis and in the most severe cases, wet gangrene [1]. There should be a high degree of suspicion of PCD in patients presenting with these symptoms and diagnostic Doppler ultrasonography is crucial to detect thrombosis. Once the diagnosis is confirmed, anticoagulation with intravenous heparin must be started immediately. Further management may include surgical intervention, such as catheter-directed thrombolysis, mechanical thrombectomy, or limb amputation [2]. This case depicts a 57-year-old male with no past medical history presenting with PCD who required emergent lower limb amputation due to irreversible ischemic damage [3]. Common causes of unprovoked DVT were ruled out, and investigation into etiology revealed vasculitis in the amputated lower extremity. The patient was later diagnosed with

systemic lupus erythematosus (SLE) presenting as acute PCD.

### CASE DISCUSSION

A 57-year-old male with no diagnosed past medical history presented to the emergency room after being unable to ambulate for 24 hours due to severe pain in his left lower extremity. In the emergency room, the patient's left lower extremity appeared swollen, discolored with diminished pulses and paresthesia. The patient denied risk factors for a provoked DVT, including recent travel, prolonged immobility, trauma or known hypercoagulable state. The patient had not seen a physician in almost a decade, felt to be in good health, and knew of no medical disorders in his immediate family.

The patient revealed one week prior to presentation an abrupt episode of a cramping sensation in his left calf that woke him up from sleep. The patient did not seek medical attention because the cramping episode resolved. He noticed his left lower extremity progressively began to appear discolored with a reddish hue appearing in the subsequent days. Twenty-four hours before presentation, his left lower extremity transformed from red to black discoloration with

mottled edematous skin and bulla formation up to the proximal left thigh.

On physical examination the patient had blistering with fluid extravasation, pulselessness, large motor and sensory deficits, superficial gangrene and necrosis of the distal toes. Due to the clinical presentation, stat venous and arterial Dopplers were ordered. Additionally, vascular surgery and cardiology teams were consulted. Bilateral venous Doppler ultrasound was remarkable for acute DVT involving the left common femoral, sapheno-femoral junction, and superficial femoral veins. Due to the presence of wet gangrene and irreversible damage, above the knee amputation (AKA) was recommended. Guillotine AKA was performed immediately to prevent impending sepsis, prior to further investigation into the etiology. The patient tolerated the procedure well and after an extended hospital stay he was safely discharged home. The patient was informed that he would require lifelong anticoagulation to decrease his risk of DVT and potential future recurrence of PCD.

During the patient's admission, further evaluation included computed tomography (CT) scan with intravenous contrast as well as CT abdominal aortogram with runoff were obtained. CT abdomen concluded a filling defect (Image A), which was subsequently followed by a CT aortogram showing occluded distal arteries (Image B). These findings in conjunction with the venous Doppler confirmed PCD. CT abdomen was performed to rule any apparent malignancies as the culprit for acute thrombosis, given

this is the most common etiology of PCD. The rare iliac compression syndrome known as May-Thurner syndrome was also not visualized on CT.

An extensive hypercoagulable workup was concurrently performed (Table 1). Antiphospholipid syndrome, factor V Leiden, prothrombin mutation, homocysteinemia were ruled out as causes of unprovoked DVT. Continued diagnostic workup during admission found the patient to be ANA positive with a titer >1:160, which has a specificity of 96% for diagnosing SLE [4]. Pathology of the amputated lower extremity revealed evidence of occlusive thrombi in major blood vessels with an infiltration of neutrophils and lymphocytes within the tunica media, indicative of vasculitis.

The incisiveness of medical attention is extremely crucial for PCD, as in this case, leading to surgical treatment. In PCD, amputation rates range between 12%-50%. Vasculitis prevalence in SLE is reported to be between 11%-36% [5]. Reports also state that males with SLE represent 4%-22% of all SLE patients, and tend to have higher frequencies of renal disease, skin manifestations, cytopenias, serositis, neurologic involvement, thrombosis, cardiovascular disease, hypertension and vasculitis than women [6]. Although not present in this patient, some literature reports that 22.4% of lupus patients are associated with antiphospholipid syndrome (APS) [7]. Unfortunately, there is minimal literature available discussing SLE patients and thrombosis in the absence of APS [8].

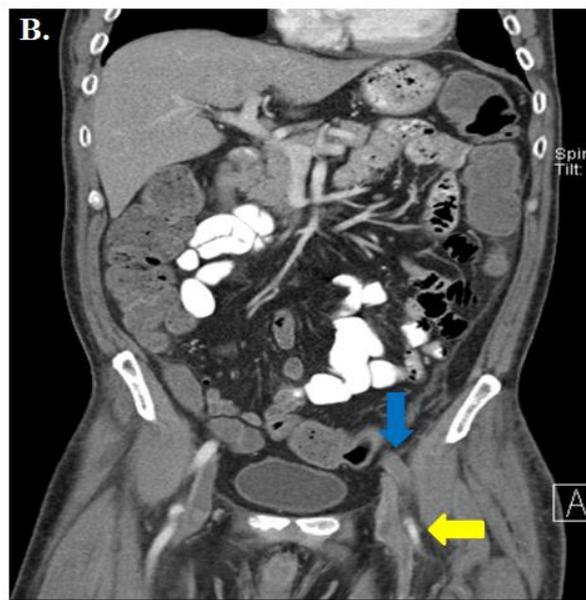
**Table-1: Hematologic Workup for Hypercoagulable State**

Special Chemistries	Results	Special Chemistries	Results
Anti-Thrombin III Activity	WNL	ANA	Positive; Titer 1:160
Protein C Activity	WNL	Anti-SSA	Positive
Protein S Activity	WNL	Anti-SSB	Negative
Lupus Inhibitor	WNL	Cardiolipin Antibody	Negative
Homocysteine	WNL	Beta-2-glycoprotein	Negative
MTHFR Genotyping	Negative	Scleroderma 70	Negative
C3 Complement	WNL	RNP Antibody	Negative
C4 Complement	WNL	Factor V Leiden	Negative
Prothrombin Mutation	Normal genotype	Occult Blood, Stool	Negative

WNL: Within normal limits; MTHFR: methylene tetrahydrofolate reductase; ANA: antinuclear antibodies; Anti-SSA: anti-Sjogren's Syndrome A; Anti-SSB: anti-Sjogren's Syndrome B; RPN: ribonucleoprotein



**Image A:** CT abdomen and pelvis with intravenous contrast showing filling defect involving the left external iliac artery indicating thrombus



**Image B:** CT abdominal aortogram with runoff to the femoral arteries bilaterally, shows non-occlusive thrombus within the left external iliac artery proximally (blue arrow) with occlusive thrombus extending into the mid and distal external iliac artery (yellow arrow)

## CONCLUSION

In conclusion, phlegmasia cerulea dolens is a severe, end stage form of deep vein thrombosis that needs prompt recognition to avoid grave prognosis such as limb amputation, shock, and/or death. More importantly, the etiology must be determined to clearly manage the patient as 50% of patients with PCD have an occult malignancy [9]. This case highlights the presentation of a life threatening thrombosis caused by a common rheumatologic disorder requiring prompt recognition and treatment to prevent irreversible damage.

## ACKNOWLEDGEMENTS

The authors are thankful to Dr. Kevin Carter, Department of Radiology at McLaren Oakland Hospital, for his time and expertise.

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