

PERCUTANEOUS MECHANICAL THROMBECTOMY IN PHLEGMASIA CERULEA DOLENS: CASE REPORT AND LITERATURE REVIEW

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Abstract

Introduction: Phlegmasia cerulea dolens is a potentially life-threatening complication of deep venous thrombosis, causing marked swelling and sudden severe pain in the limb, associated with cyanosis, edema and compartment syndrome that together compromise arterial supply. There is no consensus on its treatment.

Case Report: A 36-year-old woman, with a history of cosmetic surgery 8 days before admission (abdominal liposuction), was admitted to the emergency department with edema, cyanosis, severe pain, decreased temperature and tenderness of the left lower limb. At physical exam, no distal pulses on the left lower limb were found. Angio-CT was performed, showing occlusion of left femoral vein, external and common iliac veins.

The patient started treatment with enoxaparin (80 mg, subcutaneous, bid) and percutaneous mechanical thrombectomy (PMT) of the left iliac vein sector was performed, followed by balloon angioplasty and stenting of the left iliac vein sector. It was also deployed a temporary filter in the inferior vena cava. Thrombophilic workup was negative.

The patient presented thorough clinical remission after the procedure (Villalta score 0). Two years after surgery, the patient is asymptomatic, and the Doppler ultrasound is unremarkable concerning morphologic changes throughout the left iliac vein sector.

Conclusion: The treatment of phlegmasia cerulea dolens is challenging due to its severity and poor prognosis. Minimally invasive procedures, such as PMT can be an alternative to open surgery. It can also avoid the use of thrombolytics in patients with relative / absolute contraindications to its use.

Keywords: Phlegmasia; deep venous thrombosis; compartment syndrome

INTRODUCTION

Deep venous thrombosis (DVT) and acute pulmonary embolism (PE) are the two most frequent manifestations of venous thromboembolism (VTE).^{1,2} Although rare, phlegmasia cerulea dolens (PCD) is a potentially life-threatening complication of acute DVT.^{2,3}

Patients usually present with marked swelling and sudden severe pain in the limb, associated with cyanosis, edema and compartment syndrome that together compromise arterial supply.^{1,2} Delay in treatment may result in amputation or even death^{1,3}; however, there is no consensus

on its treatment.²

CASE REPORT

A 36-year-old woman, with a history of cosmetic surgery 8 days before admission (abdominal liposuction), was admitted to the emergency department with edema, cyanosis, severe pain, decreased temperature and tenderness of the left lower limb (figure 1) with two hours onset. The patient also presented with hypotension (89/63 mmHg) and tachycardia (117 beats per minute). At physical examination presented hypoesthesia and no distal pulses



Figure 1

Initial presentation of phlegmasia cerulea dolens - edema and cyanosis.

on the left lower limb.

D-dimers' value was 13902 ng/mL (cut off value < 243 ng/mL). DVT of the left femoral vein was confirmed by Doppler ultrasound. Angio-CT was performed, excluding PE and showing occlusion of left femoral, external and common iliac veins (figure 2).

The patient was started on enoxaparin (80 mg, subcutaneous, bid) and percutaneous mechanical thrombectomy of the left iliac vein was performed (Aspirex®, Straub Medical, Wangs, Switzerland), via mid-thigh left femoral vein, followed by balloon angioplasty and stenting (sinus-Venous®, Optimed, Ettlingen, Germany) of the left iliac vein sector (figure 3), as an iliac venous stenosis was detected on completion phlebography. It was also deployed a temporary filter in the inferior vena cava (figure 4) (Capturex®, Straub Medical, Wangs, Switzerland), through the right jugular vein during the procedure.

Thrombophilic factors such as factor V Leiden mutation (R506Q), prothrombin gene mutation (G20210A), methylenetetrahydrofolate reductase (MTHFR) 677T and MTHFR 1298C, Protein S deficiency, Protein C deficiency, Antithrombin deficiency, activated protein C resistance, lupus anticoagulant, factor VIII deficiency, anticardiolipin (aCL) antibody immunoglobulin (IgG and IgM) and anti-b2 glycoprotein-I antibodies (IgG and IgG) were all either negative or within normal range.

After the procedure, the patient started using com-

pression hosiery (23-32 mmHg) and changed the anticoagulation therapy to oral acenocumarol (target INR 2-3) for 3 months, then to apixaban (5 mg, bid) for 12 months. After this period, an apixaban dose of 2,5 mg, bid was maintained.

At the end of the procedure, clinical remission was total. After 2 years follow-up, the patient has no symptoms or signs of venous hypertension. Doppler ultrasound showed no evidence of structural or endoluminal changes in left iliac vein sector stents.

DISCUSSION

PCD is the most severe presentation of DVT, causing an obstruction of venous drainage of the lower limb. The most common clinical presentation is sudden severe pain, swelling, cyanosis, edema and compartment syndrome that in combination compromise arterial supply and may cause the neurologic impairment of the limb. Circulatory collapse and shock frequently ensue in this clinical setting. (1) Furthermore, in the lower extremities, left-sided involvement is 3-4 times more common than right-sided. (1) In our case report, symptoms were described to the left lower limb. An early diagnosis was performed due to high clinical awareness.

Development of DVT is multifactorial.⁴ Malignancy is the most common etiology and represents 20-40% of the cases.¹ Other associated risk factors include thrombophilias, surgery, trauma, pregnancy and venous stenosis, such as left iliac venous compression syndrome (IVCS), also known as May-Thurner Syndrome (MTS).⁴ Nevertheless, 10% of patients have idiopathic PCD.¹ Our patient performed a surgery 8 days before symptom onset. Testing for thrombophilia may not be consensual in this setting. However due to the patient' young age and eventual implication in future decisions to keep on full anticoagulation we thought this could add valuable information. Malignancy and pregnancy were excluded. During phlebography, an

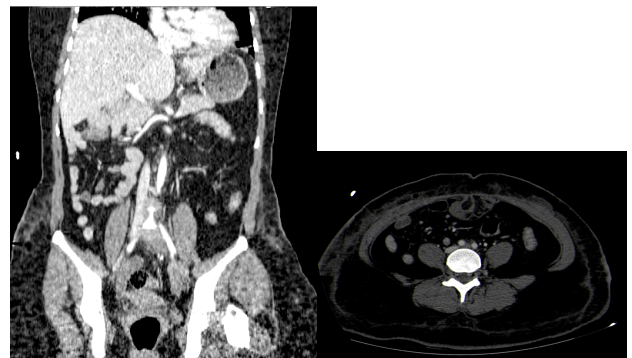


Figure 2

Computed tomographic angiography showing occlusion of left iliac vein (A and B).

iliac venous stenosis was detected. Surgery, obesity and MTS were probably the triggering factors to PCD.

The goal of treatment is to prevent propagation of thrombus formation and maintain the patency of the venous circulation, resulting in life and limb salvage.³ Anticoagulation, surgical thrombectomy with or without fasciotomy, catheter-directed thrombolysis, percutaneous transluminal angioplasty with or without stenting, or a combination of these are treatment options described in the literature.³

In this case, initial management included absolute



Figure 4

Temporary filter introduced in the inferior vena cava, showing fragments of the clot.

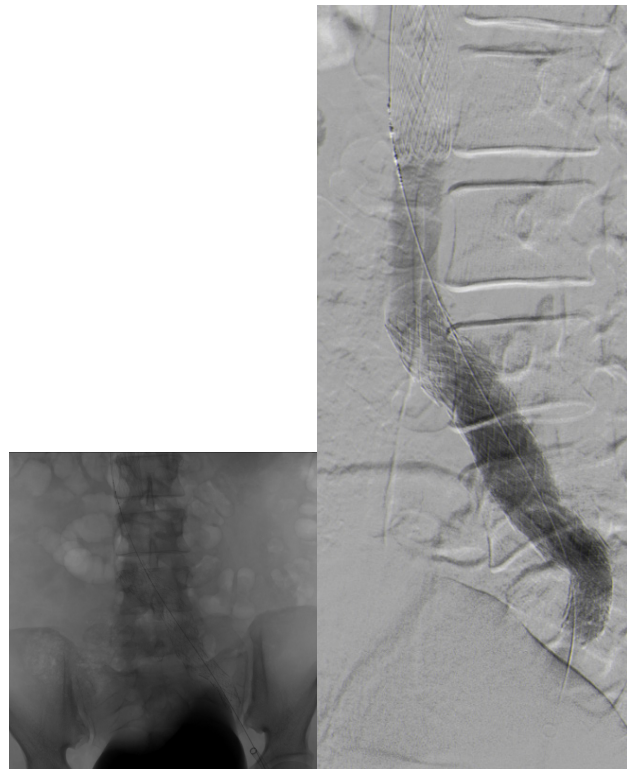


Figure 3

Phlebography at the end of the procedure, showing stenting (A) and patency of the left iliac vein sector (B), still with temporary filter in place.

bed rest, leg elevation, fluid resuscitation and low-molecular-weight heparin (LMWH) (enoxaparin, 80 mg, subcutaneous, bid). LMWH is a safe and effective treatment of proximal DVT and PE. Still, there is no recently published evidence that supports the use of these drugs in PCD and can lead to a poor outcome if used isolated.^{1,3}

Beyond initial management and due to the severity of patient's symptoms along with the dismal evolution, a percutaneous mechanical thrombectomy (PMT) of the left iliac vein sector (Aspirex®, Straub Medical, Wangs, Switzerland), a temporary filter in the inferior vena cava (IVC) (Capturex®, Straub Medical, Wangs, Switzerland) and percutaneous transluminal balloon angioplasty and stenting of the left iliac vein sector (sinus-Venous®, Optimed, Ettlingen, Germany) were performed. PMT devices are capable of macerating thrombus, and aspirating devices can remove the macerated thrombus fragments. So, it can increase the surface area of residual thrombus and create a central flow channel within an occluded vein, which might improve the efficiency of applied thrombolysis. The main objective was to achieve venous outflow more quickly due to sensory loss.

The risk of PE may be increased by thrombolytic therapy due to clot fragmentation and manipulation by venous catheters³. In this case, an IVC filter was introduced to avoid an eventual PE and due to the type of IVC filter system used (Capturex®, Straub Medical, Wangs, Switzerland), immediate removal by a single catheter de-

vice at the end of the procedure was possible. The debris captured by the IVC filter are shown in figure 4.

After the PMT, an IVCS was detected and percutaneous transluminal balloon angioplasty, with stent deployment, was performed. Compared to anticoagulation alone, endovascular angioplasty and stenting may decrease the risk of recurrent thrombosis; still, its benefits are not well established.⁴

Unfortunately, there is no clear-cut indication for the type, dose or duration of antithrombotic management after endovascular stenting for IVCS up to the time of this publication.⁴ Due to the conspicuous presentation of this case, two years after the event, our decision is to keep this patient under anticoagulation until new data becomes available.

CONCLUSION

The treatment of PCD is challenging due to its severity and prognosis. Once identified it must be considered for more aggressive management. The goal of treatment is to prevent propagation of thrombus formation and preserve the patency of the venous circulation, avoiding the development of a compartment syndrome.

Minimally invasive procedures, such as PMT can be an alternative to open surgery. It can also avoid the use of thrombolytics in patients with relative / absolute contraindications to its use.

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