Cellulitis of Leg: An Unusual Case of Phlegmasia Cerulea Dolens

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Abstract: Phlegmasia cerulea dolens (painful blue edema) is an uncommon severe form of deep venous thrombosis which results from extensive thrombotic occlusion (blockage by a thrombus) of the major and the collateral veins of an extremity. We presented here a case of cerulea dolens in 23 years female who is ANC of 8 months came with swelling and tenderness in lower limb. Patient is managed conservatively with injectables and anti thrombotic agents.

Keywords: deep venous thrombosis, venous embolism

1. Introduction

Phlegmasia ceruleol dolens is a severe form of deep venous thrombosis characterized by severe venous outflow obstruction, marked limb swelling, pain, bluish discoloration, and even venous gangrene if the condition is untreated. Triggering factors include malignancy, femoral vein catheterization, heparin-induced thrombocytopenia, antiphospholipid syndrome, surgery, heart failure, and pregnancy.

2. Case Report

23 years old female came with chief complaint of swelling in left lower limb since 5 days. Patient also complaint of discharge of serous fluid from the same leg since 5 days. Patient is a case of ANC of 8 months and complaining of decrease foetal movement since 1 day. There is continuous presence of fever since 1 day. There is no history of DM/Hypertension/TB/Congenital heart disease/any medicine or drugs intake. Patient is primigravida without any history of abortion. Patient is vitally stable with a pulse of 86bpm and BP of 110/70mm of hg. Systemic examination is normal. On examination of left lower limb it is edematous with skin discoloration, blebs are present and pin prick also present. No evidence of crepitus.

3. Investigations

CBC:14.1/7000/249000,PT/INR:15/1.07,Na/K:129/4.17
USG OBS: suggestive of 32 weeks gestation with IUFD
USG DOPPLER: suggestive of mild subcutaneous edema, NO evidence of DVT, no thrombus/embolus. Release incision was taken and tissue sent for gas gangrene examination. DIC profile of patient was D-DIMER: 3388 (raised), Sr FIBRINOGEN: 520 (raised), APTT: 37.9 (raised), PT/INR:12.4/1.13, BUN/Creat: 42/2.4. Treatment for DVT initiated. Induction for IUFD done and patient delivered vaginally macerated baby. Patient becomes vitally unstable and kept on ventilator with ionotropic supports. On next day the condition of leg worsens, blackening increases up to thigh, extremely cold, femoral and other peripheral pulses present. Repeat USG Doppler done which did not suggestive of DVT, embolism, flow on arteries was normal.
Patient was not maintaining vitals even on ionotropic supports. Patient was died on the next day.

4. Discussion

Overall clinical features of phlegmasia cerulea dolens are difficult to diagnose. Such cases, 40-60% also have capillary involvement, which results in irreversible venous gangrene that involves the skin, subcutaneous tissue, or muscle. Under these conditions, the hydrostatic pressure in arterial and venous capillaries exceeds the oncotic pressure, causing fluid sequestration in the interstitium. Venous pressure may increase rapidly, as much as 16- to 17-fold within 6 hours. Malignancy, Hypercoagulable syndrome, Surgery, Trauma, ulcerative colitis, Inferior vena cava filter insertion, May-Thurner syndrome (compression of the left iliac vein by the right iliac artery).

Pregnancy has often been associated especially during the third trimester, when the uterus is large enough to compress the left common iliac vein against the pelvic rim (ie, milk leg syndrome). Finally, 10% of patients with phlegmasia have no apparent risk factors. Supportive evidence is provided by ultrasonography and diagnostic venography. It helps to diagnose DVT and its progression along with monitoring for generation of new emboli. Contrast enhanced venography and magnetic resonance venography can also be used but all are associated with complications or disadvantages related individually with them. The development of a standard treatment for phlegmasia cerulea dolens is ongoing. Intervention is usually required in order to prevent gangrene. Treatment consists of a multimodal approach, including systemic anticoagulation, catheter-directed thrombolysis, systemic thrombolysis, surgical thrombectomy, and AngioJet thrombectomy. The advantages of thrombolysis over the other methods are wider distribution of the thrombolytic agent with blood flow and the resultant beneficial effect on both large and small venous channels. This patient was not vitally stable to undergoes and invasive intervention and tolerate such long duration procedures.

Treatment: Medical management is the cornerstone of treatment of cerulea dolens. As soon as the diagnosis is suspected intravenous Heparin should be initiated at a dose of 80-100U/Kg as bolus followed by 15-18U/Kg/Hour as maintenance. Activated partial thromboplastin time should be monitored and kept under therapeutic range. Low-molecular-weight heparins (LMWHs) have been found to be safe, effective, and convenient for use. The oral factor Xa inhibitors rivaroxaban and apixaban are approved for treatment of deep venous thrombosis (DVT) and can potentially be used for treatment of phlegmasia. If patients improve with elevation of limb and anticoagulation and do not progress to critical limb ischemia, compression therapy with stockings or elastic bandages can help to decrease edema and swelling.

Surgically can be managed as endovascular intervention. Catheter-directed thrombolytic therapy is the mainstay of therapy for the extensive DVT that is usually associated with phlegmasia. It is minimally invasive, effective, and safe. The patient is kept on heparin drip. A combination of pharmacomechanical thrombolysis using devices that macerate and aspirate the clot (eg, AngioJet or Trellis) or balloon angioplasty to break down the thrombus is performed. Venous occlusive lesions or areas of stenosis can be treated with stenting after dissolution of the acute thrombus. Patient should undergoes check angiography after endovascular procedure. Endovascular iliocaval reconstruction has been described for the treatment of iliocaval thrombosis.

Catheter-directed thrombolysis is contraindicated in certain patients. Absolute contraindications include the following: Active bleeding, Recent neurologic surgery (cranial or spinal), Trauma or cerebrovascular accident less than 2 months previously, Brain tumor, Severe hypertension, Coagulopathy, Allergy to thrombolytic agents.

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Open surgery:
Open surgical thrombectomy is an alternative treatment for patients who cannot undergo thrombolysis and who require thrombus removal. Transabdominal cavotomy and thrombectomy can also be performed. This approach permits better control of the cava above the thrombus and thus provides protection against PE, but it carries a higher morbidity. Procedures that have been performed in an effort to decrease the rethrombosis rate include cross-pubic venu-to-vein reconstruction with polytetrafluoroethylene (PTFE) or the great saphenous vein (GSV) in conjunction with an arteriovenous fistula between the femoral artery and the GSV. These adjuvant procedures may be especially beneficial in cases that involve proximal iliofemoral vein constriction, damage, or external compression. Surgical thrombectomy cannot open the small venules that are affected in venous gangrene and is therefore thought to be less effective than thrombolysis in clearance of acute thrombus. For patients with acute compartment syndrome, a four-compartment fasciotomy and decompression are required to prevent muscle necrosis. Finally, if all efforts fail, an amputation is required. It is best to delay the procedure as long as possible so as to reduce edema, allow venous channels to recanalize, and allow necrotic tissue to demarcate.

5. Conclusion
Phlegmasia cerulea dolens is very fatal condition with an underlying DVT. If treated early in suspicion the chances of cure increases. The most common cause of mortality is pulmonary insufficiency. Medical and surgical management are equally important as the treatment of cerulea dolens and should be instituted as soon as possible.

References
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