CASE REPORT

PHLEGMASIA CERULEA DOLENS – A RARE LIFE THREATENING CONDITION

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ABSTRACT: Phlegmasia cerulean 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. Etiological factors include malignancy, femoral vein catheterization, heparin – induced thrombocytopenia, anti - phospholipid syndrome, surgery, heart failure, and pregnancy.

KEY WORDS: Phlegmasia cerulean dolens, deep vein thrombosis.

INTRODUCTION: More than 600,000 cases of venous thromboembolism are estimated to occur each year in the United States. Pulmonary embolism (PE) complicates approximately 50% of cases of untreated proximal deep venous thrombosis (DVT) and contributes to 10 – 15% of all hospital deaths. Less frequent manifestations of venous thrombosis include phlegmasia alba dolens, phlegmasia cerulean dolens (PCD), and venous gangrene. All three manifestations result from acute massive venous thrombosis and obstruction of the venous drainage of an extremity.

CASE REPORT: A 35 years old male patient with 5 year treated type 2 diabetes mellitus, Presented with massive swelling of left leg, progress over the period of 2 days, agonising pain, cyanosis and many blisters on medial aspect of left leg. He is alcoholic, non smoker, ambulated. On general examination patient was afebrile, pulse – 80/min, regular, all peripheral publication well felt, except – left tibial and left dorsalis pedis (because of massive swelling). Had massive oedema of left leg with many blisters on medial aspect of leg & cyanosis, but no e/o cellulitis. Other systemic examination were normal. Patient was clinically diagnosed with deep vein thrombosis of left leg and confirmed by Doppler ultrasound (fig 1). He was treated with Limb elevation, immobility and low molecular weight heparin (LMWH) for 7 days then switched over to oral anticoagulants (warfanin), with this treatment swelling reduced in size(Fig 2).

DISCUSSION: This entity was first described by Fabricus Hildanus in the 16th century. In 1938, Gregoire made an outstanding description of the condition and used the term "phlegmasia cerulean dolens" to differentiate ischemia – associated massive venous thrombosis from phlegmasia alba dolens, which describes fulminant venous thrombosis without ischemia (1). The exact incidence of this condition is not reported. Precipitating factors include malignancy, which is the most common cause, femoral vein catheterization, heparin – induced thrombocytopenia, anti-phospholipid antibody syndrome, surgery, heart failure, and pregnancy (2). Pregnancy has often been associated with phlegmasia alba dolens, especially during the third trimester when the uterus is large enough to compress the left common iliac vein against the pelvic rim (i.e., milk leg syndrome). Finally, 10% of patients with phlegmasia have no apparent risk factors as in our case.

Phlegmasia dolens is a rare form of massive venous thrombosis of the lower extremities that is associated with a high degree of morbidity, including venous gangrene, compartment type syndrome, and arterial compromise. (3)The development of a standard treatment for phlegmasia cerulean dolens is ongoing. Intervention is usually required in order to prevent gangrene. (4) For phelegmasia alba dolens and mild non gangrenous forms of phlegmasia cerulean dolens (PCD), conservative medical treatment, such as steep limb elevation, anticoagulation with intravenous administration of heparin anticoagulation is to decrease the risk of proximal clot propagation or thromboembolism. Heparin does not directly affect limb swelling. The best nonsurgical method to decrease edema is steep leg elevation. Thrombolysis seems to be another attractive alternative in the management of PCD and venous gangrene. In 1970, Paquet was the first to use thrombolysis for the treatment of PDC. (6) Some authors propose catheter – directed thrombolysis directly into the vein with high doses of urokinase or tissue plasminogen activator (t – PA). Other authors support the method of intra – arterial low – dose thrombolysis via the common femoral artery, reasoning that the arterial route delivers the thrombolytic agent to the arterial capillaries and, subsequently, to the venules. The intra – arterial approach seems to be more effective in cases with venous gangrene. Systemic thrombolysis has also been used. (7)

Surgical thrombectomy performed through a femoral venotomy allows instant decompression of the venous hypertension.

Fasciotomy alone or in conjunction with thrombectomy, or thrombolysis reduces compartmental because of the prolonged wound healing and the risk of infection.

Concomitant administration of heparin and long – term anticoagulation are mandatory. Regardless, thrombectomy in patients with PCD is associated with a high rate of rethrombosis. Surgical thrombectomy cannot open the small venules that are affected in venous gangrene, and it does not prevent valvular incompetence or postphlebitic syndrome. The incidence of postphlebitic syndrome may be as high as 94% among survivors.

SUMMARY: Phlegmasia cerulean dolens (blue, painful leg) is an uncommon manifestation of deep – vein thrombosis and results from massive ischemia. Despite all of the therapeutic modalities described above, phlegmasia cerulean is a life threatening conditions with overall mortality rates of 20 – 40%. Pulmonary embolism is responsible for 30% of the deaths reported from PCD. Overall, amputation rates of 12 – 50% have been reported among survivors.

Image of phlegmasia cerulea dolens (Before Treatment)

Image of phlegmasia cerulea dolens (After Treatment)

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