Phlegmasia Cerulea Dolens: A Life- and Limb-threatening Disease

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Abstract

Phlegmasia cerulea dolens (PCD) is a rare syndrome caused by venous thrombosis and characterised by a triad of limb oedema, cyanosis and pain. It requires early recognition as delay of treatment can cause gangrene, limb amputation and in extreme cases, death. A 67year-old Chinese lady, with underlying hypertension, diabetes mellitus and dyslipidaemia presented to the emergency department with a 2 days history of pain, oedema and bluish discoloration over the entire left leg. She had a history of fall 6 months prior and since then she used a walking stick for mobilization. This patient underwent ultrasound doppler left lower limb, which showed features suggestive of long-segment left lower limb deep vein thrombosis. A diagnosis of PCD was made. Subsequently, she went for a CT angiogram and venography of the left lower limb which confirmed thrombosis of the left calf vein extending to the long segment of the left common iliac vein. She was commenced on intravenous heparin infusion and then underwent inferior vena cava filter insertion and catheter directed thrombolysis. Repeat venogram showed successful catheter directed thrombolysis of the left lower limb deep venous thrombosis (DVT). Treatment should be initiated as soon as the diagnosis of PCD is suspected. Currently, guidelines for treatment are lacking however 3 therapeutic options are advocated alone or in combination: anticoagulants, thrombolytic therapy, and venous thrombectomy. An early recognition of PCD and appropriate decision regarding the treatment is essential to preserve the limb.

Keywords: Phlegmasia cerulea dolens, venous thrombosis, thrombolysis

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