

DIAGNOSIS

Our differential diagnosis comprised: phlegmasia cerulea dolens (PCD), arterial insufficiency and compartment syndrome. The factors which led to our final diagnosis of PCD included: the recently diagnosed DVT and pulmonary emboli, a sub-therapeutic INR, the low creatinine kinase level (usually markedly elevated in compartment syndrome), and the absence of recent leg trauma. The patient's clinical presentation of a cool, blue, intensely painful and swollen leg with absent distal pulses also supported our PCD diagnosis. We did not measure compartment pressures.

Our patient was immediately transferred to a tertiary care hospital where he underwent catheter-guided venous urokinase thrombolytic therapy and ultrasound-accelerated thrombolysis. This resulted in a dramatic symptomatic and clinical improvement, although subsequently the patient's first, third and fourth toes on the left foot became necrotic necessitating partial amputations. The underlying cause of the patient's PCD was also investigated and no evidence of malignancy or other disease was found on computed tomographic scans of the thorax and abdomen.

PCD is a severe form of venous thrombosis with clotting of the deep and superficial veins. Venous pressure can increase to such an extent that arterial perfusion may be impaired, as was the case in our patient. The elevated venous pressure coupled with oedema and compromised arterial flow may lead to gangrene which may in turn produce shock and death. PCD is a highly morbid condition and amputation rates of 12 to 50% have been described. Mortality rates range from 20 to 40%. Modern treatment may result in better outcomes.

PCD patients usually present with sudden severe limb (usually leg) pain, cyanosis (cerulea) and oedema. Prompt diagnosis and treatment is necessary to save the patient's limb and life. Many cases are preceded by the less severe phlegmasia alba dolens, characterised by pain, oedema and blanching (rather than cyanosis).

PCD treatment consists of: systemic anticoagulation, thrombectomy and/or intravenous thrombolysis. Immediate referral to an interventional radiologist and a vascular surgeon with expertise in PCD treatment is necessary. Once the patient is stabilised, further analysis should be performed to elucidate the aetiology of the PCD. Malignancy is the most common cause of PCD (20 to 40%).

REFERENCES

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