Severe outcome in rare upper extremity phlegmasia cerulea dolens

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CASE

A 53-year-old woman with hypertension and COPD arrived at the emergency department (ED) by EMS after she had developed respiratory distress. She was intubated in route to the hospital and deteriorated into asystole requiring CPR for 20 minutes. Return of spontaneous circulation was achieved in the ED, and the patient was admitted to the medical intensive care unit (MICU) for further management. There was a purpuric discoloration on the patient's right upper arm, extending proximally from the fingers to the dorsum of the right forearm (Figure 1); pulses were palpable. The patient was afebrile, hemodynamically stable, and on mechanical ventilation. Labs revealed a WBC of 29 K/μL, sodium 149 mmol/L, Cr 2.8 mg/dL, elevated liver enzymes, and creatinine kinase 3000 U/L. Lactic acid was 8.8 mmol/L; arterial blood gas results included pH 7.2 and $PaCO_2$ 55 mm Hg. Computed tomography of the head revealed mild, diffuse cerebral edema, and hypertonic saline was started. Upon re-evaluation of the forearm, it was noted that the discoloration had worsened significantly. Ultrasound venous duplex revealed a deep venous thrombosis in the radial vein and superficial thrombosis in the cephalic vein from the hand to the axilla level as well as thrombosis in the basilic vein from the mid upper arm to the hand. X-ray of the arm showed no fractures, no osseous abnormalities, nor gas formation. Therapeutic anticoagulation was started, and surgery was consulted for possible compartment syndrome. At the time of surgical evaluation, the patient had no peripheral pulses in the affected extremity; she was taken to the operating room for emergent fasciotomy of the forearm and hand. Her postoperative course was complicated by significant

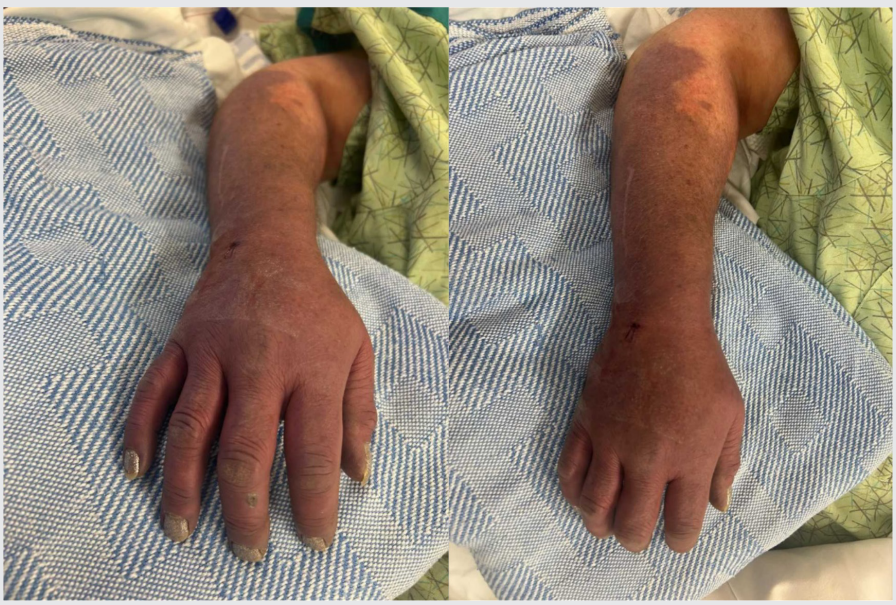

Figure 1. Right upper extremity with extensive purpuric discoloration of the skin, extending proximally to the level of the radial head.

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bleeding from the surgical site. Ultimately, the patient developed multisystem organ failure, and her family decided to transition to comfort care.

Discussion

Phlegmasia cerulea dolens (PCD) involving the upper extremity is a rare but critical condition characterized by severe venous obstruction and leads to significant swelling, pain, and bluish discoloration of the affected limb.[1] Timely diagnosis and treatment are crucial to prevent severe complications, such as venous gangrene. Spontaneous involvement of the upper extremity in PCD is extremely rare, with only a few cases documented.[1] The prognosis for upper extremity PCD is often poor, with high morbidity and mortality rates. Factors such as underlying malignancies, coagulopathies, and the presence of concurrent lower extremity DVT can exacerbate the condition, leading to severe outcomes, such as irreversible venous gangrene and limb loss.[2] The rising venous pressure predisposes these patients to compartment syndrome; therefore, early diagnosis and intervention are crucial in managing PCD.[3] Common therapeutic approaches include anticoagulation therapy and extremity elevation. However, more invasive procedures like catheter-directed thrombolysis or surgical thrombectomy may be necessary in severe cases or in patients with contraindications to anticoagulants like heparin-induced thrombocytopenia.[4] This case shows the characteristic presentation of PCD with rapid progression to compartment syndrome despite anticoagulation therapy. Once diagnosis is suspected, clinicians must consider a multidisciplinary approach and perform frequent physical examinations and neurovascular checks of the affected extremity. Frequent reevaluation for clinical progression and the need for more aggressive measures is imperative in the management of these patients. In this case, the patient was unresponsive on mechanical ventilation and unable to report her symptoms, leading to a delay in recognizing the severity and rapid progression of her condition, resulting in a poor outcome.

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