SYMMETRICAL PERIPHERAL GANGRENE OF BILATERAL HANDS AND UNILATERAL FOOT IN SETTING OF SEPTIC SHOCK REQUIRING PROLONGED HIGH DOSE VASOPRESSORS

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Symmetrical peripheral gangrene (SPG) is characterized by progressive ischemic damage in two or more extremities, without large vessel obstruction. This syndrome has been reported in several etiologies such as infections, disseminated intravascular coagulation (DIC), and low cardiac output states. It is also known as purpura fulminans (PF). It carries a risk of limb amputation leading long term morbidity and higher rates of mortality due to prolonged hospitalisation. SPG is the sign of progressive disseminated intravascular coagulation (DIC). The main pathogenesis theory, is microthrombosis associated with disturbed pro coagulant – anticoagulant balance, leading to tissue necrosis. The treatment thus involves aggressive treatment of root cause and theoretically involves heparin-based anticoagulation.

Introduction:

Case Presentation:
A 63-year-old female with history of diabetes mellitus, hypertension, hypothyroidism and ischemic heart disease on a regular treatment was admitted at outside hospital with fever and abdominal pain for 1 week. Social history included no smoking, alcohol or illicit drug use and no relevant family history was reported. On presentation, she was examined and investigated to be diagnosed with acute pyelonephritis with septic shock leading to acute kidney and liver injury and Disseminated intravascular coagulation. She was started with hydration and broad spectrum antibiotics later targeted according to urine culture which grew e. coli sensitive to carbapenems + glycopeptides and polymyxins (colistin, meropenem and teicoplanin) along with invasive mechanical ventilation in view of acute type 2 respiratory failure and high inotropic and vasopressor support.i.e ( IV noradrenaline 30ug/min and IV dopamine 15ug/kg/min ).

Four days after vasopressor administration, a bluish discolouration was noted on the patient’s upper and lower extremities with prolonged capillary refill time and cold extremities but dopperable pulses in extremities. Two days later patient developed thrombotic thrombocytopenic purpura, for which patient received 5 cycles of plasmapheresis, dalteparinand multiple transfusions.

On continued evaluation, with clinical improvement, inotropes and vasopressors were tapered and negative autoimmune studies and negative culture reports were noted; but with progressive symmetrical peripheral gangrene, patient underwent right upper limb above elbow amputation on day 15th of hospital admission.

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On day 17th of admission, patient developed hyponatremic seizure leading to hypoxic brain damage requiring prolonged ventilation and antiepileptics and hence was tracheostomised and shifted to Sushrut Hospital and research centre for further management.

On clinical evaluation and vascular surgery opinion, patient was found to have dry gangrene with non-palpable but dopplerable pulse in left upper limb limiting to index, ring, middle and little finger. Whereas, well felt line of demarcation, more extensive blackish discoloration was observed in left lower limb 10 cm below tibial tuberosity. After Doppler study to rule out any signs of deep vein thrombosis, patient underwent left hand index, middle, little and ring finger amputation with ALP flap & right leg trans-metacarpal amputation with free ALT flaps in two settings one week apart followed by vacuum-assisted dressing and broad spectrum antibiotic coverage. Patient’s clinical status continued to deteriorate until she lapsed into asystole cardiac arrest and was pronounced dead 47 days after the onset of her initial symptoms.

**Discussion:**
Symmetrical peripheral gangrene (SPG) is uncommon clinical presentation of acute ischemic changes in two or more limbs without distal arterial obstructions and absence of invasive bacterial proliferation. Mostly fingers and toes are affected but rarely gangrene has been noticed in nose, ear lobes and scrotum. Although a wide array of infective and non-infective etiological factors has been linked with SPG, the common risk factors associated with dry gangrene includes diabetes mellitus, atherosclerosis, and long-term smoking.

It has been described that DIC associated SPG is primary driven with sepsis, low-flow states, vasospastic conditions, myeloproliferative disorders, and hyperviscosity syndromes. The condition is aggravated by hypothermia, vasopressor infusion, immunosuppression, malignancy, diabetes mellitus, and renal failure. Symmetrical peripheral gangrene carries a mortality rate as high as 35% to 40% and an equally high morbidity rate; the literature reports an amputation rate upwards of 70%.

Delayed diagnosis and treatment of sepsis and prolonged vasopressors and inotropes can lead to peripheral gangrene as was first reported in 1973. Thereby, suggested first-line measures for treatment of SPG when identified early include discontinuation of vasopressors, reversal of DIC by cautious anticoagulation, and aggressive treatment of shock and sepsis. Adjuvant therapy with tissue plasminogen activator, plasmapheresis, sympathetic blockade, and aspirin has been recognized to contribute to favourable outcome.

The only definitive treatment that has been established for gangrene is amputation of the necrotic digits after development of a clear line of demarcation.

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