Bilateral symmetrical digital gangrene of upper and lower limbs due to purpura fulminans caused by *Streptococcus pyogenes*: A rare entity

Sir,

We report an unusual case of 72-year-old female with acute gastroenteritis due to *Streptococcus pyogenes*, leading to low cardiac output state and acute kidney injury, who developed bilateral symmetrical peripheral gangrene (SPG) of upper and lower limbs [Figures 1 and 2]. All peripheral pulses were palpable and color Doppler of upper and lower limb vessels indicated normal flow. Investigations revealed hemoglobin 10.9 g/dL, platelet count 44,000 cells/mm^3, PT INR 1.72, low antithrombin-lll levels and deranged liver function. Plasma was positive for fibrin degradation products and D-dimers. She was shifted to Intensive Care Unit suspecting septic shock and resuscitated with intravenous (IV) fluids and dopamine infusion. In view of persisting hypotension and desaturation, she was intubated and mechanically ventilated. Unfractionated heparin was started to treat digital gangrene, attributing it to the pro-thrombotic state of sepsis. She developed bleeding per vaginally and orally in course of 2 days while her PT-INR rose to 2.8, platelet count decreased to 11,000 cells/mm^3 and fibrinogen levels were 65 mg/dL, indicative of consumptive coagulopathy. This was treated with transfusion of six units FFP, four units each of cryoprecipitate and platelet, and bleeding was controlled within 2 days.

After 72 h, blood culture was positive for *S. pyogenes*, sensitive to ceftriaxone, amoxicillin/ampicillin

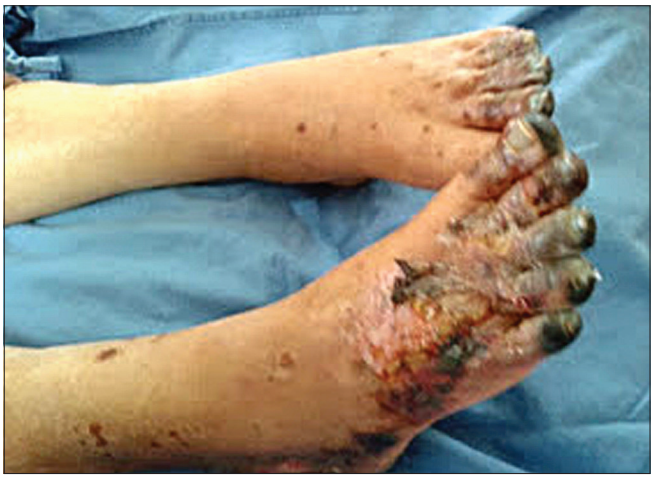

Figure 1: Digital gangrene of both lower limbs, involving forefoot on the right side

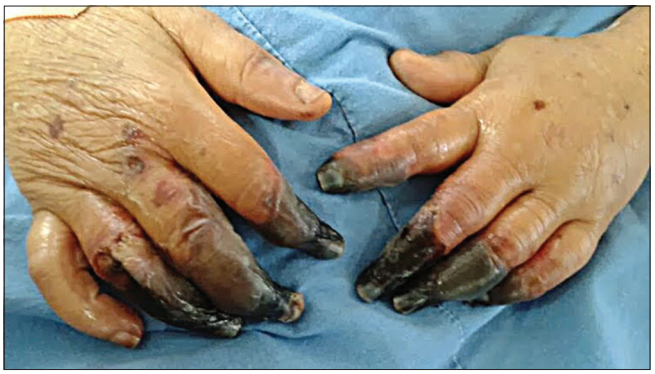

Figure 2: Digital gangrene of both upper limbs

and tetracycline/doxycycline, which was started immediately. The diagnosis of pupura fulminans (PF) due to *S. pyogenes* leading to SPG was made. Her general condition improved within 72 h with no further progression of gangrenous areas, which were well delineated in a week. She was discharged against medical advice on day 11 due to the economic constraint.

Pupura fulminans is an acute life-threatening complication of disseminated intravascular coagulation (DIC) manifested by rapidly progressive cutaneous hemorrhage and necrosis leading to SPG.[1] Clinical features include tissue necrosis, intact peripheral pulse, elevated serum lactate, association with vasospastic disorders and multi-organ failure. This is attributed to the pro-thrombotic state of DIC, which leads to widespread microvascular thrombosis.[2] Common pathogens responsible for PF include *Meningococci, Pneumococci, Staphylococci, Klebsiella* and *Enterococci*.[3] *S. pyogenes* the causative organism in our case has not been reported till now in the literature.

Awareness of this clinical condition will help in the prompt recognition, early administration of treatment to prevent the spread of gangrenous areas and avoidance of expensive investigations like angiography or unwanted surgical procedure in the critically ill patient. Aggressive resuscitation using IV fluids, vasopressor infusions, correction of acid-base and electrolyte imbalance and mechanical ventilation can stop the spread of gangrenous area along with rapid recovery. Mortality associated with SPG can be reduced by early recognition and administration of culture sensitive antibiotics. Although heparinization is associated with thrombotic (peripheral gangrene) as well as bleeding tendencies (consumptive coagulopathy), posing therapeutic dilemma, it remains the mainstay of treatment to control thrombosis.[4] Amputation of gangrenous areas is deferred till the general condition of the patient improves, and the gangrenous area is well-demarcated.[3] Protection of gangrenous extremities with antiseptic dressing, halting the spread and demarcation by topical nitroglycerine paste, phentolamine, epoprostenol or sympathetic blockade can be tried.[2] Novel modalities of treatment include plasmapheresis, papaverine, streptokinase, hyperbaric oxygen therapy, dextran-40, epsilon aminocaproic acid, ketanserin, and hirudin.[1,3,5-7]

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