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Symmetrical Peripheral Gangrene Due to Disseminated Intravascular Coagulation

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Critical Situations: Dermatology in the Acute Care Setting

REPORT OF A CASE

A 50-year-old white man developed fevers, chills, shortness of breath, and a cough that produced green sputum. Five days later, he was prostrate and was admitted to his local hospital with fulminant septic shock and associated disseminated intravascular coagulation, renal failure, and adult respiratory distress syndrome. *Staphylococcus aureus* was cultured from samples of his sputum and blood. He required ventilator support, fluids, and inotropic support to maintain arterial blood pressure, and he underwent hemodialysis for renal failure. He received intravenous antimicrobial therapy, initially with erythromycin, ceftriaxone, and gentamicin, later with piperacillin sodium–tazobactam sodium, and then with vancomycin hydrochloride.

Twenty-four hours after admission, his fingers and toes became cyanotic. This condition progressed rapidly to gangrene involving the hands, feet, tip of the nose, ears, and posterior scalp area. Hydrocortisone (100 mg every 6 hours) was administered intravenously for 6 days and then slowly tapered. The patient's general condition stabilized, but the gangrene persisted.

After 3 months, he was transferred to Mayo Clinic, Rochester, Minn, for consideration of amputation of the gangrenous areas. On physical examination, symmetri-

cal well-demarcated dry gangrene involving the hands (to the wrists), feet, legs (to the level of the knees), tip of the nose, ears, and posterior scalp area was noted (**Figure**).

THERAPEUTIC CHALLENGE

In a case of gangrene involving all extremities, it was important to salvage as much of each limb as possible. Initially, therefore, a nonsurgical approach was used to minimize the degree of amputation. Gentle debridement of the proximal portions of the gangrenous areas revealed pink, viable underlying tissue in places of apparent gangrene.

Radiography of the hands and feet showed no evidence of osteomyelitis, although marked soft tissue swelling was observed. Magnetic resonance imaging showed multiple infarcts bilaterally in the talus, calcaneus, multiple tarsal bones, small regions of the distal aspect of the tibia, and fingers.

After 1 month, the nonviable tissue was well demarcated, and bilateral below-the-knee amputations and finger amputations were performed.

One year later, after intensive rehabilitation, the patient was able to ambulate with his bilateral lower extremity prostheses. After multiple corrective plastic pro-



Gangrene symmetrically affecting acral areas. A, feet; B, hands.

cedures on his hands, he was able to pinch objects between his "fingers."

COMMENT

Symmetrical peripheral gangrene is a rare but well-documented condition that was first described by Hutchinson¹ in 1891 as the sudden onset of acral gangrene occurring in a symmetrical distribution. In some cases, the initial skin lesions are hemorrhagic and then progress to gangrene, so-called purpura fulminans. Up to 85% of patients with symmetrical peripheral gangrene have associated disseminated intravascular coagulation, and approximately 50% of the patients who survive require amputation of a limb.² In the patient described herein, the symmetrical peripheral gangrene was associated with disseminated intravascular coagulation resulting from infection.

The pathogenesis of symmetrical peripheral gangrene is not well understood. Any condition that critically diminishes the blood, nutrient, or oxygen supply to acral areas over a prolonged period can lead to this condition. Possible pathogenic mechanisms include vascular obstruction of any cause: intraluminal (such as that related to hypercoagulable states³), intramural (such as that related to vasoconstrictive drugs⁴ or collagen vascular disease⁵), and external (such as that related to extracorporeal shockwave lithotripsy⁶), in addition to conditions that produce a low cardiac output.⁷

Disseminated intravascular coagulation, often due to infection,^{8,9} is the condition most often associated with symmetrical peripheral gangrene,² as seen in the case reported herein. It is hypothesized that occlusion of the microvasculature by microthrombi results in hypoperfusion and tissue ischemia, leading to infarction and gangrene.

A mortality rate of 35% has been reported for symmetrical peripheral gangrene associated with disseminated intravascular coagulation, with 48% of reported survivors requiring amputation of at least 1 lower extremity.² Johansen and Hansen¹⁰ reported that 3 of 10 patients died after the onset of gangrene.

Because symmetrical peripheral gangrene is rare, reports of therapeutic alternatives are largely anecdotal and not based on controlled clinical trials. Rarely has treatment been shown to prevent progression or to reverse incipient gangrene. Management of underlying causes, such as disseminated intravascular coagulation, seems appropriate. If a precipitating factor, such as dopamine infusion,¹¹ can be identified, it should be corrected if possible. Treatment success has been reported for individual patients who received epoprostenol and tissue plasminogen activator infusion⁸; sympathetic blockade¹⁰; the combination of plasmapheresis, leukapheresis, and antibiotics¹²; or aspirin.³ It has been suggested that anticoagulation

might be of benefit in preventing the progression of gangrene.¹³ However, Johansen and Hansen¹⁰ reported that treatment with aspirin, heparin, and streptokinase seemed to have no effect. The addition of oral corticosteroids did not help either.

Amputation of the gangrenous areas may be inevitable, but initially a nonsurgical approach to management is preferred to allow time for the patient's condition to stabilize and to allow the gangrene to become demarcated.¹⁴ The degree of gangrene may be less extensive than initially expected, as in our patient.¹⁰ Bone scans may be helpful in determining the appropriate level for amputation.¹⁵

Thus, in this rare and devastating condition, correction of underlying disease, meticulous supportive management, and a cautious approach before proceeding to amputation are suggested to minimize morbidity and mortality.

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