Symmetrical Peripheral Gangrene and Disseminated Intravascular Coagulation Associated With Pneumococcal Sepsis

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To the Editor:
Symmetrical peripheral gangrene (SPG) is a rare but devastating complication of septicemia. Most cases of SPG are associated with disseminated intravascular coagulation (DIC).

We present the case of a 35-year-old woman who had undergone splenectomy due to abdominal trauma at 3 years of age; she was referred to hospital with fever and severe prostration with onset 6 days previously in the form of fever, myalgia, and dry cough. Tachycardia, tachypnea, hypotension, and fever were all evident in the initial evaluation. Closer physical examination revealed nothing except for the presence of bibasal crackles during the cardiopulmonary auscultation. Initial complementary tests clearly showed the presence of leukocytosis with left shift (leukocytes: 11 600/µL [range: 4000-10 000/µL], neutrophils: 91% [40%-75%], band forms: 46% [1%-3%]), mixed acidosis, sinus tachycardia, and bilateral pleural effusion with an air bronchogram in the retrocardiac area. Serum biochemistry, coagulation study, biochemistry, and urine sediment, as well as cerebrospinal fluid (CSF) analysis were normal. Due to her hemodynamic state and the initial diagnosis of sepsis caused by encapsulated bacterial respiratory infection, the patient was transferred to the intensive care unit (ICU), where she was stabilized with a volume infusion and perfusion of vasoactive amines and empirical treatment with cefotaxime 2 g every 6 hours and intravenous azithromycin 500 mg/d.

Six hours after admission to the ICU the patient was re-evaluated clinically and petechial skin lesions were found on the acral zones of the extremities, coalescing to form ecchymotic plaques. Peripheral pulses were palpable. A second complete blood count and coagulation study performed at this time were consistent with DIC (platelets: 20 000/µL [150 000-400 000/µL], prothrombin time: 20 s [10-12.5 s], partial thromboplastin time: 82.5 s [20-40 s], fibrin degradation products: 650 µg/mL [< 8 µg/mL], D-dimer: 8947 ng/mL [< 500 ng/mL]). Red cell concentrates and platelets, as well as fresh plasma, were administered. The patient's urine tested positive for Streptococcus pneumoniae antigen and the same agent was isolated in the CSF culture. Antibiotic susceptibility testing revealed the organism was sensitive to penicillin and treatment was initiated with doses of 4 million units every 6 hours. The patient progressed well clinically over the following days, with the skin lesions healing except on several fingers on both hands where necrosis and dry gangrene with mummification occurred (Figure). When the necrotic areas had been outlined, 15 weeks later, these areas were amputated and reconstruction was completed with flaps.

SPG is an uncommon but well documented syndrome first described by Hutchinson in 1891. It consists of symmetrical gangrene in acral regions with no evidence of large-vessel occlusion or vasculitis. The lesions begin in the form of erythema or purpural lesions that develop
into gangrene within 24-48 hours. Hemorrhagic blisters are common as are proximal purpural zones that do not always develop into necrosis. SPG has been linked to many underlying medical processes, and is most prevalent in serious infections in patients with certain risk factors. The most commonly implicated microorganisms are meningococci, pneumococci, streptococci, and staphylococci. SPG associated with pneumococcal sepsis principally affects splenectomized patients and is considered to be an extremely serious condition with high rates of associated morbidity and mortality.

Up to 85% of cases of SPG are linked to DIC. Although it has been proposed that DIC leads to ischemia and posterior gangrene of the acral zones through the formation of intravascular clots in the microcirculation, other authors have related SPG to an initial spastic rather than thrombotic process in the vessels. Other factors potentially present in septic patients (severe hypotension, endothelial damage, microembolism, and the use of inotropic drugs) could also play a role in the pathophysiology of this entity.

There is no specific treatment for SPG. Treatment of the underlying cause and DIC is of vital importance. Isolated cases of SPG have been successfully treated through sympathetic blockade, leukapheresis and plasmapheresis, acetylsalicylic acid, or a combination of anticoagulants and vasodilators. Early amputation is contraindicated, as secondary infection of the necrotic tissue is uncommon and delimitation of the ischemic lesions occurs with time. Initially, treatment is based on protection of the extremities and nursing care, with debridement, skin grafts, and partial or total amputation performed later. Surgery must be followed by rehabilitation of the patient with physiotherapy in order to conserve the highest possible level of functionality.

We wish to use this case to stress that, in the clinical setting of sepsis, SPG is considered a valuable skin marker of DIC and can therefore be seen as a sign of a very poor prognosis in this group of patients.

References