CASE REPOSITORY

Necrotizing Sweet Syndrome of the Upper Extremity After Elective Hand Surgery

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Sweet syndrome, or acute febrile neutrophilic dermatosis, is a systemic disease process mainly characterized by hyperpyrexia and skin lesions. A newly described entity, necrotizing Sweet syndrome, is a severe and locally aggressive dermatological condition that clinically and histopathologically resembles a necrotizing soft tissue infection. It is characterized by pathergy, a nonspecific inflammatory response to cutaneous trauma resulting in a propagation of the disease. In contrast to a necrotizing infection, this condition responds to systemic steroids. A high clinical suspicion is required in order to distinguish a necrotizing polymicrobial infection from noninfectious necrotizing Sweet syndrome. We present a case following elective hand surgery. *(J Hand Surg Am. 2017;■(■):1.e1-e6. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Dermatosis, necrotizing, neutrophilic, pathergy, Sweet syndrome.

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Sweet syndrome, or acute febrile neutrophilic dermatosis, is a systemic disease process characterized by hyperpyrexia, peripheral blood and skin neutrophilia, and edematous skin lesions. A newly described entity, necrotizing Sweet syndrome (NSS) or acute necrotizing neutrophilic dermatosis, is a severe systemic variant characterized by locally aggressive skin lesions that may be mistaken for a necrotizing soft tissue infection.[1] This condition, like many neutrophilic dermatoses, is characterized by pathergy, a nonspecific inflammatory response to skin trauma that typically manifests as erythematous to violaceous papules, plaques, pustules, or ulceration. We present a case of NSS following elective hand surgery.

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CASE REPORT

The patient was a healthy 47-year-old right-handed man with Dupuytren contracture. His past surgical history was notable for a left Achilles tendon repair, complicated by a hematoma and small ulcer that took several months to heal, and an uncomplicated left-hand ganglion cyst excision surgery. The family history was notable for a female cousin who developed pyoderma gangrenosum after a C-section surgery. The patient underwent a right partial palmar fasciectomy and presented on postoperative day 2 with fever (100.5° F), swelling, palmar erythema, and pain and was treated with local wound care and oral antibiotics with no improvement. The patient had persistent fever and elevated white blood cell count (19,000 mcL) 24 hours later and was taken to the operating room for debridement and cultures. There was noted to be fat necrosis, but no evidence of bacteria on Gram stain (Fig. 1). A computed tomography scan demonstrated phlegmonous changes and palmar soft tissue swelling. He was also noted to have a pustule at the prior site of a peripheral intravenous catheter in his left antecubital fossa. Subsequent surgical debridement demonstrated further fat necrosis without foul odor. The Gram stain was negative and cultures did not grow any organisms.

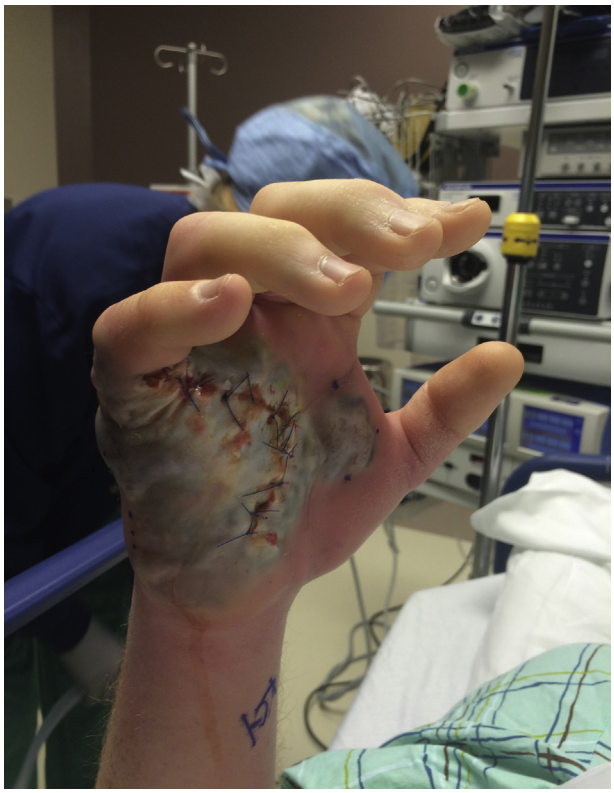

FIGURE 1: Clinical image, 3 days following fasciectomy, demonstrates significant drainage from the surgical wound with expanding skin necrosis, prior to debridement.

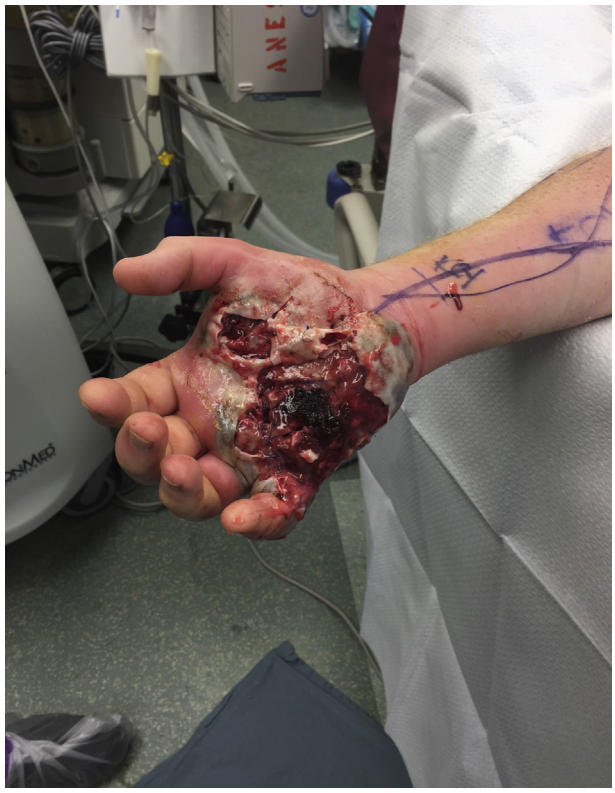

FIGURE 2: Clinical appearance following initial operative irrigation and debridement depicts full-thickness loss of dermis.

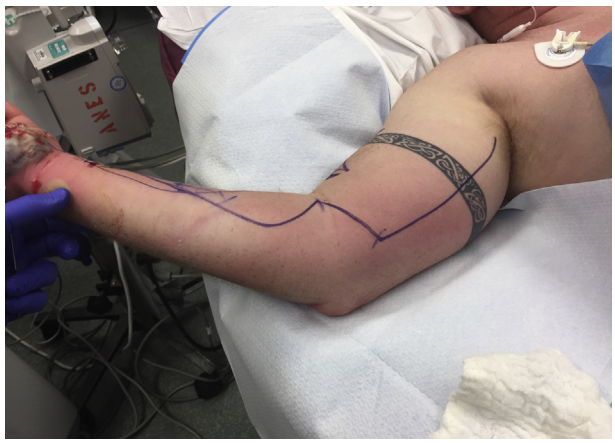

FIGURE 3: Clinical image depicts proximal extension of erythema. Planned surgical incision is outlined in the event fasciotomy is indicated.

He was transferred to a tertiary care center where he was admitted to the intensive care unit, placed on broad-spectrum antibiotics, and underwent repeated surgical debridements of nonviable soft tissue and purulent exudative material with the working diagnosis of a necrotizing infection (Figs. 2, 3). He underwent right upper extremity fasciotomies after cloudy fluid was noted to be traveling proximally along fascial planes. Intraoperative bacterial, mycobacterial, and fungal cultures remained negative throughout his stay, with no organisms ever identified (Fig. 4). He continued to exhibit signs suggestive of systemic inflammation despite intravenous antibiotic treatment and serial debridements. He had temperatures as high as 104.4° F, white blood cell of 54,000 cells/mL, erythrocyte sedimentation rate of 52, and C-reactive protein of 317 mg/L (Fig. 5). His inflammatory markers, fevers, and leukocytosis all seemed to worsen immediately after each debridement. Consults were obtained from dermatology and rheumatology because he was not improving following standard surgical debridement.

Pathological specimens demonstrated necrosis and massive infiltration of neutrophils involving the dermis and superficial and deep subcutaneous tissues with no evidence of organisms on specialized stains, including Grocott-Gomori methenamine silver and Brown-Brenn stains (Fig. 6). Given the patient's fevers, rising leukocytosis (with neutrophilia), worsening acute-phase reactants, negative cultures, and apparent pathergy, dermatology was concerned that this may be a rare case of NSS or necrotizing neutrophilic dermatosis.