

Sweet heart: A case of pregnancy-associated acute febrile neutrophilic dermatosis with myopericarditis

Wesley Yung-Hsu Yu, BS,^a Erica Manriquez, BS,^a Tina Bhutani, MD,^b R. Krishna Chaganti, MD,^c
Beth S. Ruben, MD,^{b,d} Brian S. Schwartz, MD,^e and Lindy P. Fox, MD^b
San Francisco, California

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Sweet syndrome (acute febrile neutrophilic dermatosis) is an inflammatory disorder characterized by the abrupt onset of fever, erythematous papules, or plaques showing a neutrophilic infiltrate on histopathology and rapid response to treatment with systemic steroids. The disease may present with extracutaneous manifestations that may be life threatening.¹ Although the pathogenesis is poorly understood, Sweet syndrome has been associated with drug exposure, malignancy, autoimmune diseases, infection, and pregnancy.²⁻⁴ Here we report a case of pregnancy-associated Sweet syndrome complicated by myopericarditis requiring surgical therapy for cardiac tamponade.

CASE REPORT

A 32-year-old pregnant woman at 27 weeks of gestation had fever, leukocytosis, and papulopustular eruption. She was admitted 5 weeks prior for diabetes mellitus associated with acute-on-chronic renal failure requiring hemodialysis and remained hospitalized at the time of her eruption. She also had diabetic retinopathy and bilateral above-the-knee amputations, recurrent venous thromboses requiring a Greenfield filter, daily cephalexin suppression for chronic urinary tract infection, chronic *Staphylococcus aureus* osteomyelitis of the right hand, and active methamphetamine use.

Examination of her skin found more than 40 solitary, erythematous, deep-seated, 0.5- to 1-cm discrete and grouped papules concentrated over the dorsal surface of her hands but also scattered over the upper extremities, chest, abdomen, and eyelids. Several lesions were erythematous papules with pustular

centers (Fig 1). To evaluate her leukocytosis and rash, blood cultures were drawn and skin biopsies were performed. She was empirically treated with vancomycin, meropenem, acyclovir, and amphotericin. No improvement was observed on antimicrobial therapy.

Her laboratory results showed a white blood cell count of 20,000/mm³ (neutrophils 18,700/mm³ [1,800-6,800]; lymphocytes, 1,470/mm³ [1,000-3,400]; monocytes, 1,850/mm³ [200-800]; eosinophils, 230/mm³ [0-400]; basophils, 20/mm³ [0-100]). Antinuclear and anti-double-stranded DNA antibody titers were below threshold values. Results of an extensive infection workup, including blood cultures; skin biopsy cultures for bacteria, fungi, and mycobacteria; herpes simplex virus and varicella zoster virus culture and direct fluorescent antibody; as well as enterovirus polymerase chain reaction of pustule scrapings, were negative. Two sequential skin biopsies from the right forearm showed similar features, including a dense dermal neutrophilic infiltrate with leukocytoclasia, marked papillary dermal edema, and formation of neutrophilic intraepidermal pustules. No microorganisms were identified via staining (Fite, Grocott's Methenamine Silver, Periodic acid-Schiff-diacetate, Brown-Brenn, immunoperoxidase studies for herpes simplex virus I/II and varicella zoster virus; Fig 2). The usual findings for coxsackievirus infection—ballooning and reticular degeneration—were not observed. Results of age-appropriate screening for malignancy and immunofixation electrophoresis were normal.

Three days after presentation, the patient had chest pain, a pericardial rub, and diffuse ST-elevations, and troponin level was elevated at 0.94 μg/L (normal,

School of Medicine,^a Department of Dermatology,^b Division of Rheumatology,^c Department of Pathology,^d and Division of Infectious Diseases,^e University of California, San Francisco.

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Correspondence to: Lindy P. Fox, MD, 1701 Divisadero St, Box 0316, San Francisco, CA 94143. E-mail: foxli@derm.ucsf.edu.

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Fig 1. Erythematous deep-seated papules were scattered over the patient's face, arms, chest, and hands. Note involvement of the eyelids and solitary pustules surmounting papules.

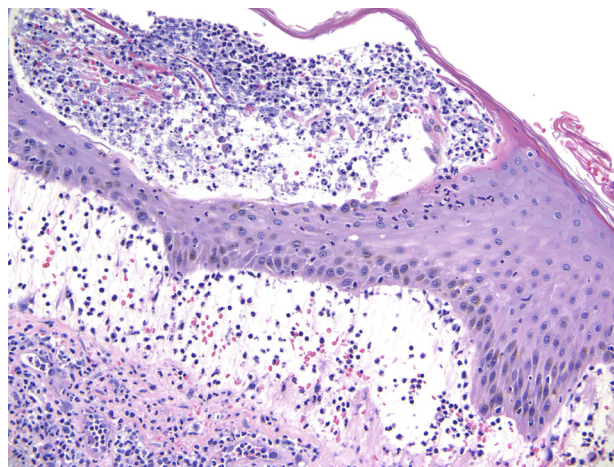


Fig 2. Histopathologic features. A punch biopsy of the right forearm shows a dense diffuse superficial and deep infiltrate with an intraepidermal pustule and marked subepidermal edema, bordering on vesiculation, with leukocytoclasia evident in the edematous papillary dermis. (Original magnification: $\times 200$.)

<0.05). Echocardiogram found a small pericardial effusion. Several days later she had hypotension, and a repeat echocardiogram found increased pericardial fluid with right atrial collapse. She was taken for urgent pericardial window with removal of 350 mL of bloody fluid resulting in immediate hemodynamic improvement. She also underwent a cesarean section. Cultures from pericardial tissue were without growth. Given the lack of any laboratory support for a diagnosis of infection and no improvement with

broad antimicrobial coverage, the decision was made to treat the patient for Sweet syndrome complicated by myopericarditis. She was prescribed prednisone, 1 mg/kg/d for 5 days, followed by a prolonged taper. The skin lesions and pericarditis resolved, and she was discharged 2 weeks after starting prednisone.

DISCUSSION

Sweet syndrome classically presents with tender, erythematous, mammillated plaques. The eruption

often favors the head, neck, and upper extremities.² Vesicles or pustules have been described; however, they tend to be grouped on erythematous plaques.⁵ Scattered discrete pustules as predominantly seen in this case are uncommon, which made it difficult to exclude disseminated infection such as *Candida*, herpesvirus, or coxsackie virus on examination alone. There was no evidence of vasculitis on biopsy, differentiating this case from neutrophilic dermatosis of the dorsal hands.⁶

Systemic manifestations of Sweet syndrome typically include fever, leukocytosis, arthralgias/arthritis, myalgias and, less commonly, pulmonary or renal involvement. Hepatic, central nervous system and gastrointestinal involvement are unusual.² Cardiac sequelae are extremely rare.^{7,8} Cases of acquired cutis laxa leading to cardiac complications have been observed months to years after the initial presentation of Sweet syndrome in the pediatric population.⁹⁻¹¹ There is only one previous case report, to our knowledge, of myopericarditis in an adult with Sweet syndrome.¹² That patient presented with pericarditis and congestive heart failure. She was found to have Sweet syndrome on biopsy of a lower extremity nodule, and there were no other skin findings. She died suddenly of cardiac arrest. On autopsy, marked neutrophilic infiltration of the pericardium and myocardium was seen. The underlying disease thought to be inducing Sweet syndrome was myelodysplastic syndrome.

Pregnancy is a rare cause of Sweet syndrome. It typically occurs in the first or second trimester and often affects the head, neck, and upper extremities.³ The disease may recur with additional pregnancies but has not been reported to increase infant morbidity or mortality.³ Systemic steroids are the treatment of choice, although in many cases, cutaneous lesions resolve spontaneously. Other chronic conditions in our patient, including diabetes, chronic kidney disease, and methamphetamine use, have not been associated with Sweet syndrome to

our knowledge. A complete review of our patient's medications found no known associations with Sweet syndrome, except for furosemide, which she had taken several years before onset and continued to take at discharge without recurrence of skin lesions.

The atypical cutaneous lesions in combination with the rarity of cardiac involvement in Sweet Syndrome made this case diagnostically challenging.

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