Consultant360 Multidisciplinary Medical Information Network

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Sweet Syndrome

Volume 59 - Issue 6 - June 2019

Authors:
Jaclyn Rivington, MD; Patrick Twohig, MD, MPH; David Soma, MD; and Jennifer Hanrahan, DO, MA
Department of Internal Medicine, Case Western Reserve University, MetroHealth Medical Center, Cleveland, Ohio

Citation:
Rivington J, Twohig P, Soma D, Hanrahan J. Sweet syndrome. *Consultant.* 2019;59(6):187-189.

A 60-year-old woman presented with a 4-day history of right-sided anginal chest pain, palpitations, vaginal pain, and dysuria. Her medical history included coronary artery disease, type 2 diabetes, hypertension, essential thrombocytosis (*JAK2*-positive), tobacco use, and hepatitis C.

At presentation, she was febrile (temperature, 39.9°C), tachycardic, and hypoxic (oxygen saturation, 80% on room air). She had tender right labial swelling without induration, fluctuance, or crepitus. The remainder of the physical examination findings were benign.

Laboratory tests revealed leukocytosis (white blood cell count, 17,700/μL with an absolute count of 22 bands), a lactate level of 2.0 mg/dL, and a D-dimer level of 707 ng/mL.

Computed tomography scans of the abdomen and pelvis showed a complex 4 × 3-cm fluid collection involving the right labial fold. She was started on broad-spectrum antibiotics and underwent incision and drainage. Intraoperative cultures were negative for bacterial pathogens.

The fever persisted, and she developed acute respiratory distress syndrome. Her labial lesion became increasingly necrotic with extension to the levator ani and urethra. Surgical pathology results showed necrotic tissue but were otherwise nonspecific.

Despite multiple courses of antimicrobial therapy, her clinical condition worsened, ultimately requiring mechanical ventilation. On her 16th hospital day, she developed new ulcerative skin

requiring mechanical ventilation. On her 10th hospital day, she developed new ulcerative skin lesions on her neck and right wrist (**Figure**). She became increasingly hypotensive and was transitioned to comfort care; she died one day after skin biopsies revealed neutrophilic dermatosis consistent with histiocytoid Sweet syndrome (H-SS).

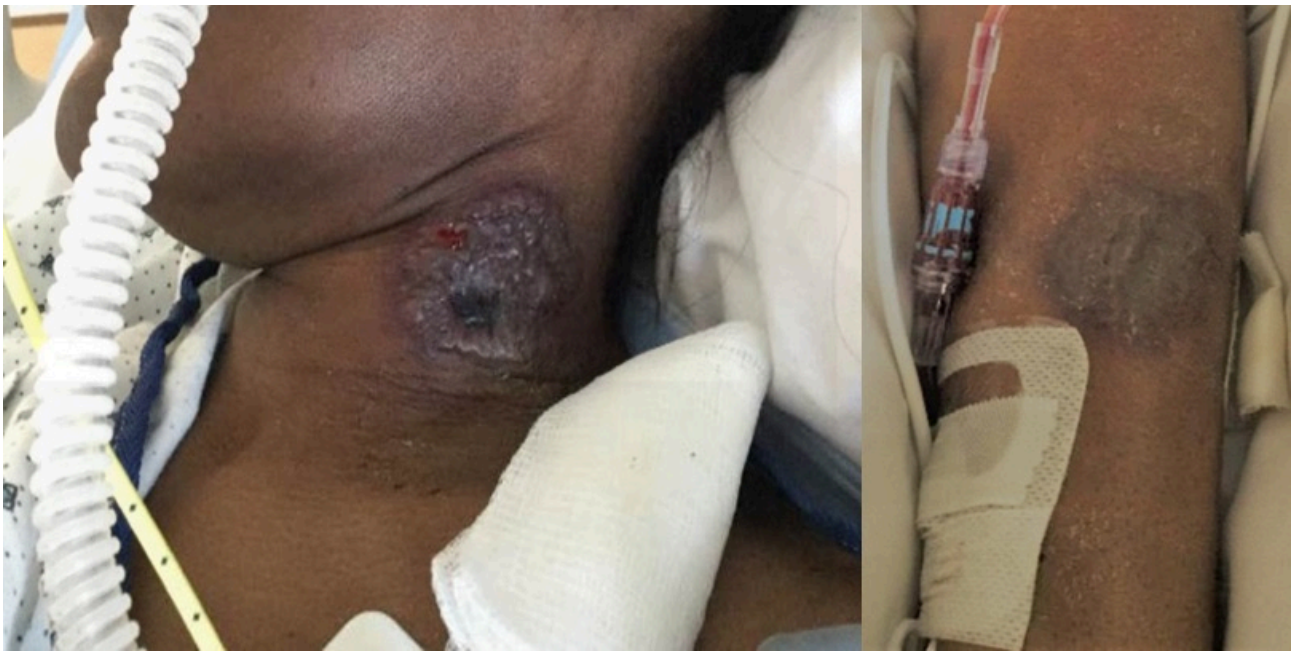

Figure. *New ulcerative skin lesions developed on the patient's neck and right wrist on day 16 of hospitalization.*

Discussion. Sweet syndrome (SS), or acute febrile neutrophilic dermatosis, is an uncommon inflammatory disorder characterized by painful skin nodules, often with fever and leukocytosis.[1] The diagnosis is confirmed with biopsy showing dense neutrophilic infiltrates without vasculitis.[2] There are 3 subtypes—classical, malignancy-associated, and drug-induced—which are further subclassified by histology.[3] H-SS is a subset of the classical subtype.[3,4] Only 100 cases of H-SS have been previously reported.[5] Our patient's case appears to be the first reported case of H-SS presenting with a labial abscess.

We utilized a large health care database to further characterize the relationship between commonly associated diseases and SS. Retrospective analysis was performed using the IBM Explorys database (1999-2018), a pooled, deidentified clinical database of more than 50 million unique patients in the United States. At the time of analysis, there were 54,714,160 patients in the database, of whom 740 had Sweet syndrome. Odds ratios (OR) were calculated to determine the relationship between SS and other diseases or population subsets using SNOMED and ICD codes. Results are listed in the **Table**.

Table. Odds Ratios of Having Different Diseases/Characteristics Among Patients With Sweet Syndrome in the Explorys Database

Comorbidity/Characteristic	Odds Ratio	95% Confidence Interval
Essential thrombocytosis	19.1	(12.2-29.7)
Acute myeloid leukemia	151.6	(110.2-208.6)
Myelodysplastic syndrome	131.6	(101.1-171.4)
Systemic lupus erythematous	26.2	(19.6-34.9)
Inflammatory bowel disease	11.7	(8.5-16.1)
Chronic myeloid leukemia	0	0
Female	3.5	(2.99-4.1)
Male	1	(0.93-1.26)
Labial abscess	0	0

Our patient was not on any medications associated with drug-induced SS. Hematologic malignancies, especially myelodysplastic syndrome (OR, 131.6), have been associated with an increased risk of SS.[4,5] This was confirmed by our database search (**Table**). Although our patient had no history of cancer, she did have essential thrombocytosis (ET), which increases the risk of SS (OR, 19.1). Classical SS is commonly seen after infection. We suspect that the labial abscess was the sentinel event causing her SS, and that her ET increased her predisposition for H-SS.

Our patient's case has similarities to those in the literature and in our population database analysis. She was female (OR, 3.5), she had an underlying myeloproliferative disorder (ET; OR, 19.5), and her skin lesions are common in H-SS, seen as asymmetric, tender, red-violaceous nodules on the face, neck, and arms.[1,3,5]

Conversely, there were several unique features of our patient's case. Our patient initially presented with labial ulcerations and chest pain. SS typically presents with constitutional symptoms.[1] Her initial clinical picture was consistent with sepsis secondary to her labial abscess, since no characteristic H-SS lesions were present. Most patients survive long enough to be treated with corticosteroids or immunosuppressants, which often results in resolution of symptoms.[1] However, our patient progressively decompensated and died prior to initiation of therapy. This severity of H-SS has not been previously documented.

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