Sweet outcome for a rare inflammatory condition

Salah Abdel Jalil, Owais Abdul-Kafi, James Smith, Ala’ Abdel Jalil

ABSTRACT

Sweet syndrome is a rare inflammatory condition that was first described by Douglas Sweet in 1964 as an acute febrile neutrophilic dermatosis.1 It can be associated with infections, inflammatory conditions, pregnancy, drugs, and malignancy. It is usually divided into three subtypes based on etiology: classical (idiopathic); malignancy-associated; and drug-induced.

We describe a patient with classical Sweet syndrome who had a dramatic response to corticosteroids. Our patient met the major criteria for diagnosis (positive histopathology and an abrupt onset of a painful rash), along with 4 minor criteria (fever, preceding upper respiratory tract infection, dramatic response to steroids, and leukocytosis).

Presentation

A 47-year-old Caucasian female with an unremarkable past medical history presented to the emergency room with a chief complaint of fever and chills for 2 days. On the morning of her presentation, she developed a non-pruritic, tender rash over her extremities that started as papules and plaques and rapidly progressed into pseudovesicular lesions. She also had cough and malaise over the preceding three weeks. She denied any antibiotic or non-steroidal drug exposure before her illness. She also denied anorexia, weight loss, abdominal pain, blood in her stool, or changes in bowel habits.

Although she was allergic to penicillin, this rash was not similar to the macular rash she developed after penicillin. She smoked cigarettes, and admitted to occasional marijuana and cocaine use.

Assessment

On physical exam, she had a temperature of 38°C, blood pressure of 162/87 mmHg, pulse rate of 120 beats per minute, and was in no apparent distress. Skin exam revealed tender, erythematous papules and plaques (Figure 1), along with pseudovesicular lesions with raised edges and central pallor resembling ‘targetoid-lesions’ of different stages (Figure 2). Some lesions developed eschar formation over the upper and lower extremities. The rash notably spared the trunk, palms and soles. She did not have any lymphadenopathy or organomegally. The rest of her physical exam was unremarkable.

She was admitted for further evaluation. Laboratory tests revealed a white blood cell count of 12,400/Ul with 80% neutrophils and evidence of mild iron deficiency anaemia.

Chest x-ray was negative for pneumonia. Collagen vascular disease antibodies, HIV, cryoglobulins, and serum immunoglobulin levels were all within the normal limits. Hepatitis C antibody was positive, but PCR and genotype testing were negative and liver enzymes were within normal limits. Occult malignancy work-up with a mammogram and upper and lower endoscopy was negative, and she did not have manifestations of a lymphoproliferative disease. Hematologic malignancy was felt less likely, so bone marrow biopsy was deferred. A skin biopsy was obtained, which demonstrated dense neutrophilic dermatosis without evidence of leukocytoclastic vasculitis, suggestive of Sweet syndrome, (Figure 3).

Management

The patient was started on prednisone 40 mg daily and had a dramatic improvement in her skin lesions within the first few days (Figure 4). The precipitating aetiology of this patient’s Sweet syndrome was felt to be a preceding upper respiratory tract infection.
Figure 1: Lower extremities image shows erythematous papules and plaques bilaterally.
Figure 2: Bullous lesions on lateral aspect of right hand and forearm.

Figure 3: Hematoxylin & eosin stain microscopy (100X), showing dense neutrophilic infiltration of dermis, without evidence of leukocytoclastic vasculitis.
Sweet syndrome is characterised by an abrupt onset of painful skin papules, plaques, or nodules. Fever and leukocytosis usually accompany the cutaneous lesions. Sweet syndrome has been observed in association with different conditions and has been classified into classical, malignancy-associated, and drug-induced.

Classical (idiopathic) Sweet syndrome is most frequently preceded by a gastrointestinal, upper respiratory tract, or other infection. It might also be associated with inflammatory conditions like inflammatory bowel disease, pregnancy, and, less frequently, other autoimmune diseases.

Malignancy-associated Sweet syndrome, which constitutes 20–25% of cases, is more common in older patients, and is most often associated with a haematological malignancy, especially acute myelogenous leukaemia (AML) and the myeloproliferative disorders. The rash could precede, accompany, or follow the malignancy diagnosis. Drug-induced Sweet syndrome has a temporal relation with certain medications, including non-steroidal anti-inflammatory medications, colony-stimulating factors, antibiotics, contraceptives, and other medications. The skin lesions usually resolve after the offending medication is discontinued.

According to the screening procedure originally suggested by Su and Liu in 1986, and modified by Von Den Driesch in 1994, the diagnosis of classical Sweet syndrome requires both major criteria: a) an abrupt onset of painful erythematous skin papules or nodules; and b) histopathologic evidence of dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis. In addition, the diagnosis requires meeting at least two of the four minor criteria: a) fever >38°C; b) association with an underlying haematologic or solid malignancy, pregnancy, inflammatory condition (eg, IBD), or preceding upper respiratory or gastrointestinal infection; c) excellent response to treatment with corticosteroids or potassium iodide; and d) at least three of the four abnormal laboratory values (ESR >20mm/hour, elevated CRP, WBC count >8,000/Ul, neutrophils >70% of WBC count).

Characteristic skin lesions of Sweet syndrome are in the form of tender erythematous papules or nodules which can develop into erythematous plaques with a characteristic papillomatous surface. Bullous Sweet syndrome and subcutaneous Sweet syndrome are less common manifestations. A skin biopsy should be performed whenever possible. A punch biopsy is usually obtained for histologic examination, and microbial stains should also be performed to rule out an infectious aetiology.

Systemic corticosteroids therapy is the first-line treatment for Sweet syndrome, and usually results in dramatic clinical improvement. Colchicine, Dapsone, and...
CLINICAL CORRESPONDENCE

potassium iodide are additional effective therapies. Symptoms often begin to improve within 48 hours and skin lesions usually resolve within one to two weeks. Relapse may occur after tapering or discontinuation of glucocorticoids, and may be more likely to occur in patients with malignancy-associated disease.  

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Author information:
Salah Abdel Jalil, Resident in general surgery department, Najah National University Hospital, Aseera Street, Nablus , West Bank-Palestine; Owais Abdul-Kafi, University of Missouri School of Medicine, One Hospital Drive, MA204, DC018.00, Columbia, MO 65212, US; Ala’ A Abdel Jalil, Division of Gastroenterology & Hepatology, CE 405 – DC 043.00, University of Missouri-Columbia; James Smith, Hematology & Oncology, McLeod Regional Medical Center, 506 E. Cheves St. Florence, SC 29506, US.

Corresponding author:
Salah Abdel Jalil, Resident in general surgery department, Najah National University Hospital, Aseera street, Nablus , West Bank-Palestine.
salah.a.jalil@outlook.com

URL:

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