Case in Point

Sweet Syndrome: The Uncommon Painful Rash

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After 4 visits to the emergency department, this patient was admitted to the hospital, diagnosed, treated, and eventually responded well to treatment for Sweet syndrome.

A 60-year-old woman presented to the hospital with a 7-day history of a rash that developed predominantly over her hands and upper extremities. She stated she had worked for several hours in her garden at home. Later that evening she noticed pruritis of her upper extremities. The next morning she developed small vesicles on her fingers, fingertips, and upper extremities. She reported no erythema, fevers, bug bites, new chemical exposures, recent travel, or recent illness.

The next day she was evaluated and diagnosed with chigger bites. Two days later she returned to the emergency department (ED) for worsening and painful vesicles on her hands. She was diagnosed with scabies. Two days later she returned to the ED with additional tense, painful vesicles and pustules on her hands and with left-sided arthralgias. She was admitted to the hospital for further evaluation.

The examination revealed a confluence of papules and vesicles on the patient's hands and forearms (Figures 1 and 2), and her forearms were tender to palpation. She had limited motion due to pain and edema of the hands. She was afebrile and remained so throughout her hospitalization. Laboratory results revealed a slight elevation of alkaline phosphatase, serum creatinine, and significantly elevated C-reactive protein (CRP). Based on the biopsy results, a diagnosis of Sweet syndrome was made.

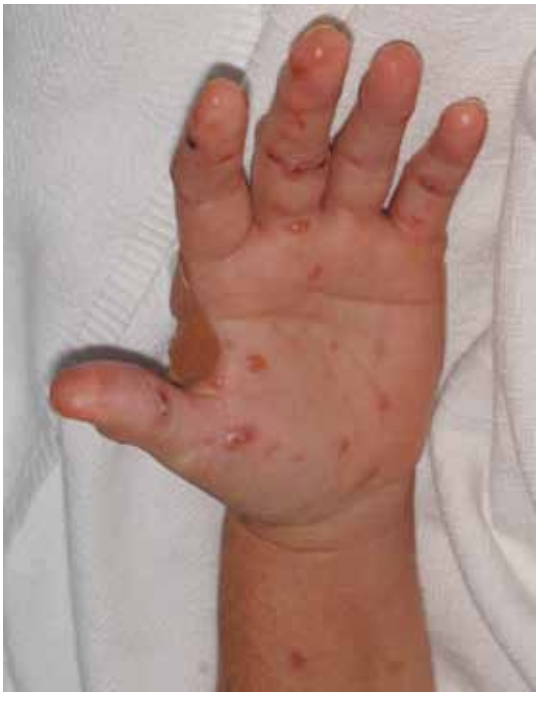

Figure 1. Note edematous hand and tense vesicles on fingertips, necrotic blisters on third digit.

Sweet syndrome, also known as acute febrile neutrophilic dermatosis, is an uncommon dermatosis described as an abrupt onset eruption of red-to-purple papules and nodules that coalesce into plaques in the setting of fever and peripheral neutrophilia. It may be associated with an underlying malignancy, inflammatory bowel disease, pregnancy, and medications, so its diagnosis should warrant further investigation. Its wide variety of presentations make it a diagnostic challenge. Sweet syndrome is the prototypical neutrophilic dermatosis first described in a small female population.[1]

A relatively uncommon disease, Sweet syndrome seems to have a female predominance.[1-3] Although a majority of cases are idiopathic, it can occur after a mild respiratory infection or be associated with an underlying malignancy, most commonly with acute myelogenous leu-

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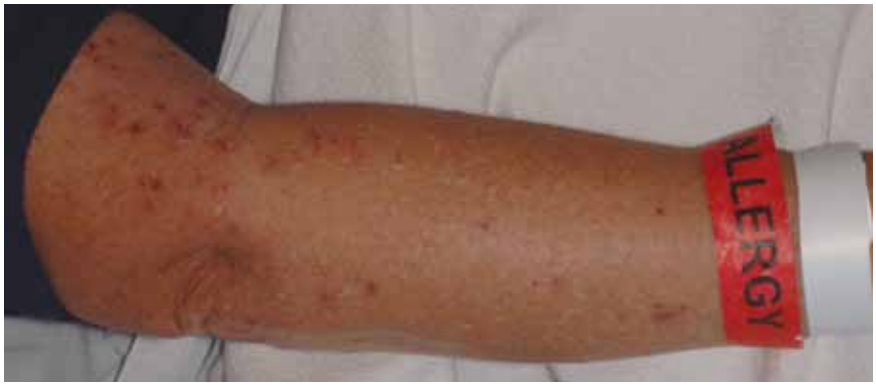

Figure 2. Similar lesions located on forearm.

kemia, followed by solid tumors of the genitourinary and gastrointestinal tract.[1-4] The cutaneous manifestations include eruption with papules that can enlarge to form plaques. Pustulation and vesicles can also occur. These lesions are asymmetric and predominantly found on the upper extremities, face, and neck and are often described as feeling tender. The patient may report hypersensitivity within the affected area. Although Sweet syndrome predominantly affects the skin, extracutaneous involvement of the musculoskeletal system, gastrointestinal tract, heart, and kidneys have been reported.[1-3]

The diagnosis of Sweet syndrome is made by fulfilling both of the following major criteria: (1) an abrupt onset of plaques or nodules that are tender; and (2) histology of the lesions that include predominantly neutrophilic infiltration of the dermis in the absence of vasculitis.[1,5]

In addition, 4 signs are considered minor criteria, and 2 are required for diagnosis. These include: (1) preceding infection of the respiratory or gastrointestinal tract, or underlying cancer; (2) fever or general malaise; (3) good response to systemic corticosteroids; and (4) abnormalities of laboratory values, which can include elevated erythrocyte sedimentation rate (ESR), CRP, segmented neutrophils in a peripheral blood smear, or leukocytosis. Three of the 4 laboratory abnormalities are required.[5]

Although the patient fulfilled the major criteria, she met only 1 minor criterion. A CRP test was obtained; however, an ESR was not obtained during hospital admission. Since the patient responded well to steroids with significant reduction in lesions and other symptoms, it was very strongly suggestive of Sweet syndrome.

Treatment of Sweet syndrome consists of daily systemic steroids starting at 1 mg/kg and tapered over 4 to 6 weeks. Treatment generally results in complete resolution of lesions. Extracutaneous symptoms also respond well to steroid therapy. However, if an underlying malignancy is present, the symptoms may not respond to steroids, and treatment should be focused on the malignancy.

Other treatments include indomethacin, colchicine, potassium iodide, dapsone, and intravenous immunoglobulin, which can be used in the long-term management of the disorder. The proposed mechanism of action is thought to inhibit neutrophil chemotaxis. Several other drugs have also been used with good success.

The patient responded well to steroids. A serum workup revealed abnormalities in CRP, alkaline phosphatase, creatinine, and arthralgias that have been described with this disorder. Biopsy pathology showed classic histological features of Sweet syndrome. After hospitalization, the patient completed a prolonged steroid taper, and her lesions resolved 1 week after her hospital stay. Given Sweet syndrome has an association with a variety of malignancies, the patient is currently undergoing the necessary screening examinations, including a mammography, colonoscopy, and an evaluation of her blood work. ●

Author disclosures

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