

A Case of Cellulitis-Like Sweet Syndrome

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Abstract

Observation: Sweet syndrome (SS) is a rare dermatological disease characterized by fever, leucocytosis, well-demarcated, erythematous papules and plaques which show dense neutrophilic infiltration. It can be idiopathic or associated with infections, malignancies, drug hypersensitivity, sarcoidosis, and autoimmune diseases. Lesions are generally on upper extremities and face. Leg lesions are rare. It is generally treated with systemic or local corticosteroids.

Herein, we present a case of SS located in the lower extremity that mimics cellulitis because of its rarity.

Introduction

Sweet's syndrome (SS), which is characterized by its clinical and histological features, is also known as acute febrile neutrophilic dermatosis, and was first described in 1964 by *Robert Douglas Sweet* [1]. Although the disease is seen in both men and women, it is most common in women between the ages of 40-60 years. SS presents with a combination of skin lesions, sudden high fever, and neutrophilia in peripheral blood and tissue. It is thought to be a reaction to a variety of antigenic stimuli such as infections and neoplasms. Lesions usually occur on the face and upper extremities and are very rare in lower limbs [2, 3, 4].

Here, we present a rare case of a cellulitis-like SS that settled in the lower extremity.

Case Report

57-year-old female patient presented to the Infectious Diseases Clinic of our hospital with redness

on the front of her right leg, along with swelling, weakness, muscle and joint pain, and high fever. It was elucidated during her clinical history that she had a visit to another hospital with the same complaints 10 days ago, and was treated with



Figure 1. Lesion in the patient's right leg

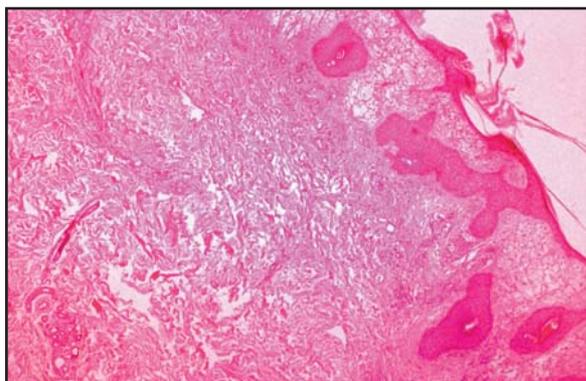


Figure 2 . The band-like edema in the superficial dermis underneath the atrophic epidermis (Hematoxylin-eosin stain, x40)

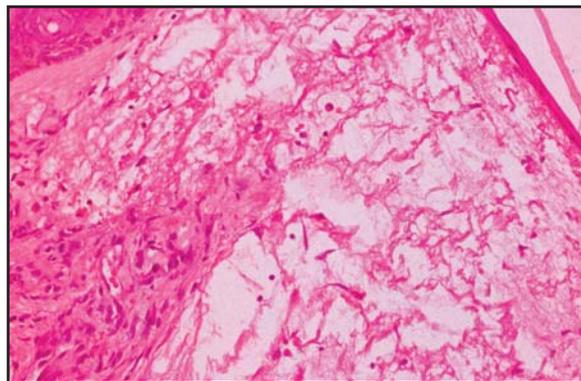


Figure 3 . The higher magnification view of the edema and mild perivascular infiltration of leukocytes, and neutrophils in the superficial dermis (Hematoxylin-eosin stain, x200)

3x1g of ampicillin/sulbactam parenterally for five days with a diagnosis of cellulitis. As her symptoms did not subside, she came to our hospital's Infectious Diseases Clinic. The dermatologic examination revealed erythema and edema on the right leg (**Figure 1**). Upon evaluation of the patient's medical history, we found out that the patient had type 2 diabetes mellitus for seven years. She had been on medication by oral antidiabetic agents (gliclazide and metformin) since 2009. Her family history was unremarkable. The results of the laboratory examination were as follows: white blood cell count was 5.26 K/uL; C-reactive protein (CRP) was 52.9 mg/L; neutrophils were 2.85 K/uL; sedimentation rate was 55 mm/h; fasting blood glucose was 129 mg/dL; and glycated hemoglobin A1C was 6.3 H. Her total urinalysis, blood smear, liver, and kidney function tests were within the normal limits. No pathology was detected on the chest radiography or abdominal ultrasonography. The patient was diagnosed as cellulitis and treated with 4x2 g ampicillin/sulbactam and 2x400 mg ciprofloxacin intravenously while at the Infectious Diseases Clinic. No resolution of clinical lesions was observed following one week of treatment, so a dermatology consultation was ordered and a skin punch biopsy procedure was performed. Histopathologically, a thick, band-like edema that contained sparse lymphocytes and erythrocytes was detected in the superficial dermis just underneath the atrophic epidermis showing mild orthokeratosis (**Figure 2**). Slightly dilated capillaries with swollen endothelium as well as sparse polymorphonuclear leukocytes, erythrocytes, and lymphocytes around the vessels were seen in the superficial dermis under the edematous area (**Figure 3**). In addition, sparse histiocytes and lymphocytes were observed between the adipocytes in subcutaneous adipose tissue. It was noted in the pathology report that the histopathologic findings might be indicative of late stage SS. After evaluating clinical and laboratory findings in

conjunction with histopathological features, the patient was diagnosed as SS and a treatment of 40 mg/day of systemic methylprednisolone was started. A follow-up exam after weeklong treatment revealed that the lesions regressed significantly.

Discussion

SS (acute febrile neutrophilic dermatosis) is a sudden onset of neutrophilic dermatosis accompanied by fever and peripheral neutrophilia and is characterized by sensitive red-purple papules, plaques, and nodules [1, 2, 3]. Although infections, vaccines, malignancies, hypocalcemia, and drugs have been suspected in the etiology of disease, because the cause of SS cannot be determined in more than half of all cases, they are described as idiopathic [4, 5, 6, 7]. In recent years, it has been proposed that excessive secretion of cytokines, particularly IL-1 (interleukin-1), and an abnormal immune response to those cytokines might be among the mechanisms that initiate pathogenesis [2, 5, 6]. Since the etiology of SS could not be determined in our case, it was considered as idiopathic.

In SS, the skin findings are often disease-specific and are therefore clinically diagnostic. The characteristic skin lesions include painful, sharp-edged erythematous plaques which are often seen on the face, neck, upper chest, and upper extremities. Eczematous, pustular, vesicular, and crusted lesions may also occur. Asymmetrically distributed single or multiple lesions may occur, but the shape and size of lesions are variable. Although annular or arcuate lesions are often observed, small acneiform lesions may also develop.

Typical plaques vary in size from a few millimeter to one centimeter and usually heal without scarring after a few weeks or months [1, 2, 8]. Atypical lesions such as blisters, ulcers, and oral mucosal involvement can be observed in SS cases associated with malignancies. Atypical clinical presentations are observed most frequently in patients with hematological malignancies. These patients commonly relapse and tend to suffer chronically from the disease [2, 7, 9]. In our case, the lesion was observed on the leg, which is rarely seen. There was no oral mucosal involvement.

Systemic symptoms such as fever, headache, arthralgia, myalgia, and malaise accompany SS. Additionally, conjunctivitis, episcleritis, conjunctival hemorrhage, and ocular congestion are observed in some cases [2, 6]. Our patient's systemic symptoms were fever, weakness, arthralgia, and myalgia.

Leukocytosis, neutrophilia, high sedimentation rate, and elevated CRP are important laboratory findings in SS. The neutrophil ratio in peripheral blood is usually above 70% and leukocytosis are typical features, but they may not be present in all patients. Moreover, the presence of neutrophil dominated mixed dermal infiltrate along with subepidermal edema constitutes a crucial finding in the histopathological examination [1, 3, 5]. Although our patient did not have leukocytosis, she did have a high sedimentation rate and elevated CRP, besides the histopathological findings were compatible with late stage SS.

The standard treatment of SS is systemic corticosteroids. Long-term low-dose therapy may be required due to frequent recurrence. Potassium iodide, colchicine, dapsone, doxycycline, clofazimine, chlorambucil, cyclosporine, nonsteroidal anti-inflammatory drugs, and cyclophosphamide are among other treatment options [2, 9]. Our case was treated with systemic corticosteroid and the lesions receded rapidly after the treatment.

SS is diagnosed according to the major and minor criteria described by Su and Liu, which were later modified by von den Driesch [3, 10]. Major diagnostic criterias include sudden onset, sensitive, erythematous plaques or nodules along with neutrophil rich dermal infiltrate histopathologically. Meanwhile, the minor criterias include fever over 38 °C, increase of inflammatory markers in the serum (erythrocyte sedimentation rate > 20 mm / h, elevated CRP,

leukocytes > 8000mm³, and neutrophils > 70%), response to systemic corticosteroids or potassium iodine therapy, concomitant infection, inflammatory disease, malignancy, or pregnancy. The presence of two major and at least two minor criterias is required for a SS diagnosis [2, 3, 5]. In our case, the diagnosis of SS was made due to the presence of two major and three minor criterias.

In conclusion, we believe that in patients who are thought to have cellulitis clinically but do not respond to systemic antibiotic therapy, rare dermatoses such as SS should be considered in the differential diagnosis.

Conflict of Interest

No conflict of interest was declared by the authors.

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