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Case Report

A dermatologic emergency; Sweet's syndrome

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ABSTRACT

Background: Sweet's syndrome (SS), also known as acute febrile neutrophilic dermatosis, is a rare condition characterized by recurrent erythematous skin lesions. Skin lesions appear as papules, nodules and plaques located on the upper extremity, trunk, neck and face.
Case report: A 72-year-old male patient presented to the emergency department with a 10-day history of generalized rash, generalized muscle and joint pain and high fever. He had a history of upper respiratory tract infection. He presented with painful erythematous plaques on both lower and upper extremities and the trunk as well as serohemorrhagic bullous lesions on the feet. The laboratory results revealed WBC count of $20.6 \times 10^3/mm^3$ (76.9% neutrophils), CRP (c-reactive protein) of 33 mg/L and erythrocyte sedimentation of 110/h. The patient was referred to a dermatologist with prediagnosis of SS due to the presence of typical painful skin lesions, high fever and neutrophilic leukocytosis. A systemic corticosteroid therapy was initiated. The diagnosis for SS was confirmed after the skin biopsy revealed the presence of dense dermal infiltrate of neutrophils and leukocytoclasis in the upper dermis. The patient responded rapidly to corticosteroids and the skin lesions improved.
Conclusion: We reported this case as it was a rare life-threatening dermatosis diagnosed in the emergency department, which is generally difficult to diagnose therein, and the skin lesions appeared on the lower extremities.

Sweet's syndrome (SS), or acute neutrophilic dermatosis, is a rare inflammatory condition characterized by the sudden onset of a rash [1]. Sweet's syndrome was first described by Robert Douglas Sweet [2]. More than 425 cases have been reported since [3]. It is characterized by tender erythematous lesions accompanied by high fever that mostly occur on the face, neck, trunk and upper extremities [4]. Our patient presented with idiopathic Sweet's syndrome and was regarded to be of significance due to the rare presentation of predominant skin lesions on the lower extremities and the diagnosis in the emergency department.

A 72-year-old male patient presented to the emergency department with complaints of erythematous and bullous lesions over body, high fever, fatigue, muscle and joint pain with a 10-day history. It was noted that he had a history of upper respiratory tract infection. On admission, the GCS score was 15 and the vital signs were as follows: blood pressure = 128/76 mmHg, pulse = 129/min, fever = 38.3 °C and SO_2 = 96. Physical examination revealed painful erythematous maculopapular plaques of various diameters on both lower and upper extremities (Fig. 1). Serous and hemorrhagic bullous lesions were observed on the feet (Fig. 2). The laboratory test results were as follows: WBC count = $20.6 \times 10^3/mm^3$ (76.9% neutrophil), CRP (c-reactive protein) =33 mg/L, erythrocyte sedimentation = 110/h, serum creatine = 1.1 mg/dL, ALT = 15 U/L, AST = 15 U/L and GGT = 17 U/L. The patient was referred to a dermatologist and then underwent a skin biopsy upon the diagnosis of vasculitis, SS and pemphigus. A corticosteroid and antihistamine therapy was initiated. The biopsy revealed the presence of dense dermal infiltrate of neutrophils and leukocytoclasis in the upper dermis. The patient was diagnosed with classical Sweet's syndrome as he presented with both major criteria and three minor criteria. The lesions had disappeared upon the 10-day follow-up.

Sweet's syndrome presents in three clinical settings which can be categorised as classical (or idiopathic), malignancy-associated and drug-induced [1]. Idiopathic SS is characterized by a constellation of clinical symptoms which include high fever and tender erythematous skin lesions (nodules, papules, plaques). This SS type typically presents with elevated neutrophil count and diffuse inflammatory infiltrate of neutrophils in the upper dermis [5]. The diagnostic criteria for Sweet's Syndrome were first proposed by Su and Liu in 1986 and subsequently modified by Von den Driesch in 1994 (Table 1). Both major criteria and two of the four minor criteria are required to confirm the diagnosis of classical (idiopathic) SS [2,6]. Classical SS is often associated with upper respiratory tract infections, gastrointestinal infections, pregnancy and inflammatory bowel diseases. It occurs more frequently in female patients with a higher rate of recurrence. It mostly presents with fever and is usually accompanied by general symptoms like headache, myalgia and fatigue. Fever mostly precedes the appearance of lesions by several days to weeks. Skin lesions of Sweet's syndrome are typically tender. They often present as red-purple papules and nodules that

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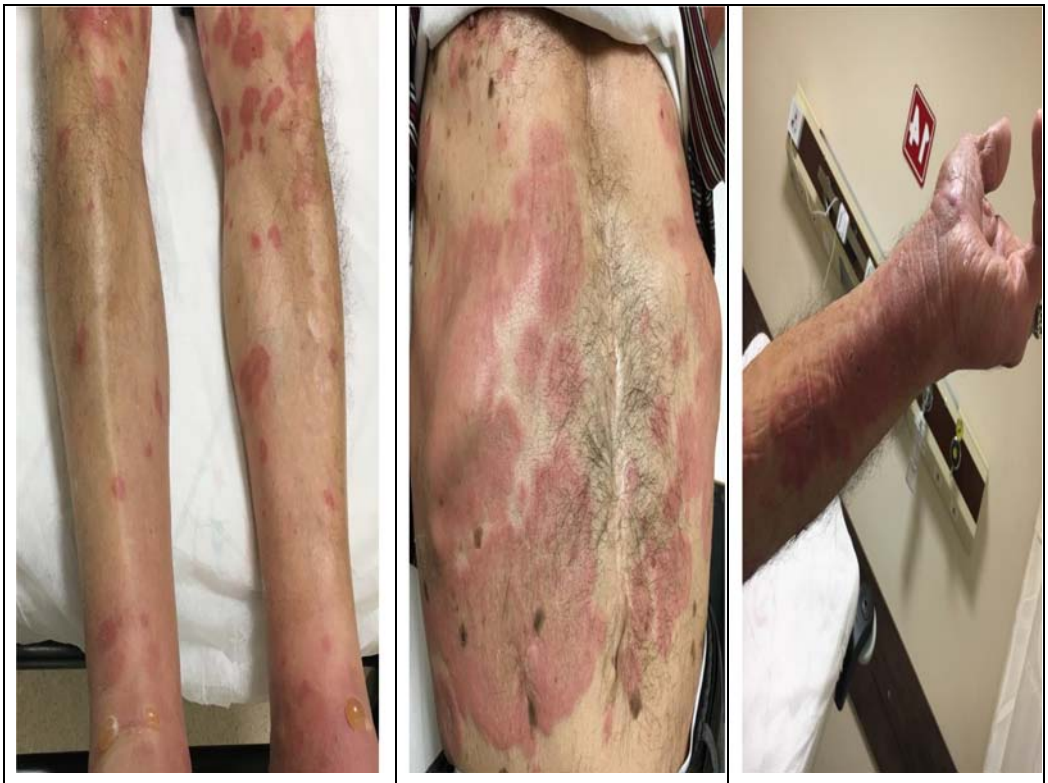
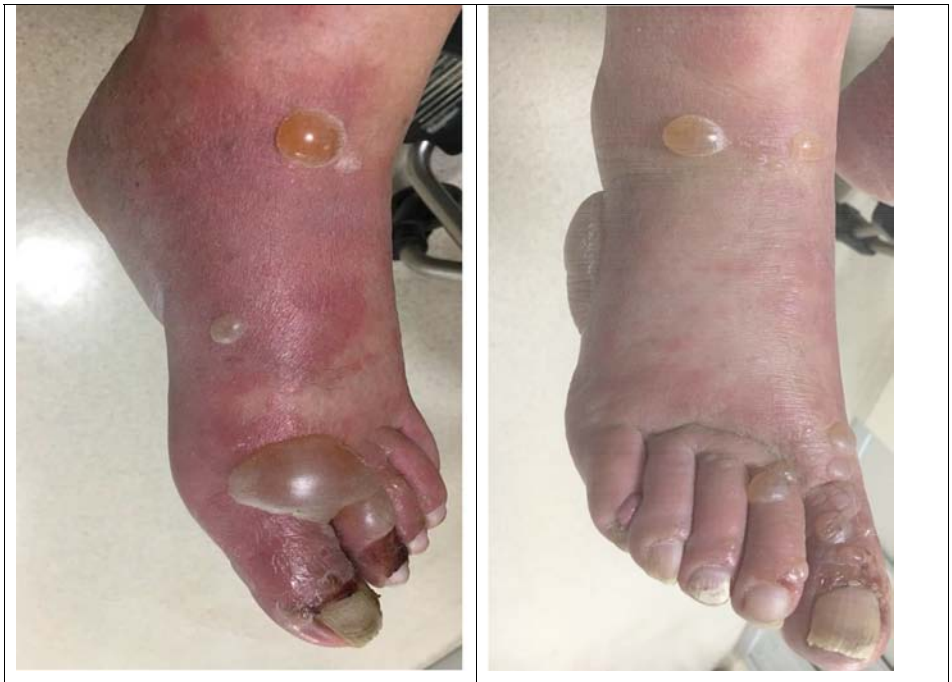


Photopgraph of the lower extremities	Photopgraph of the trunk	Photopgraph of the left upper extremity

Fig. 1. Erythematous maculopapular plaques of different diameters demonstrated on patient's body.



Photopgraph of the left foot	Photopgraph of the right foot

Fig. 2. Serous and perforated hemorrhagic bullous lesions demonstrated on the patient's feet.

Table 1
The diagnosis of classical (idiopathic) Sweet's syndrome.

Major criteria
Sudden onset of painful erythematous nodules or plaques Dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis
Minor criteria Fever >38 °C Association with an underlying;
• Hematologic/visceral malignancy, • inflammatory disease/pregnancy, • or preceded by an upper respiratory/gastrointestinal tract infection/vaccination
Abnormal laboratory values at presentation (three of four):
• ESR[a] >20 mm/h. • Positive[b] CRP • WBC[c] >8000 • >70% neutrophils
Excellent response to treatment with systemic corticosteroids/potassium iodide

*The presence of both major criteria, and two of the four minor criteria is required in order to establish the diagnosis of classical Sweet's syndrome.
[a] ESR: erythrocyte sedimentation rate.
[b] CRP: c-reactive protein.
[c] WBC: leukocytes.

tend to coalesce to form plaques. Some lesions present as vesicles and bullea as a result of the edema in the upper dermis [7]. Common laboratory results include elevated peripheral leukocyte and neutrophil counts and elevated sedimentation rate [2]. The pathological diagnostic criteria for Sweet's syndrome include a diffuse infiltrate of mature neutrophils in the upper dermis in addition to the edema associated with polymorphonuclear leukocytes. SS may precede the initial diagnosis of cancer or may accompany the disease. It is mostly associated with hematological malignancies. Several medications have been associated with drug-induced Sweet's syndrome, but it is most commonly observed in patients that use granulocyte-colony stimulating factor. Systemic corticosteroids are considered the gold standard for treatment. They are effective in improving the symptoms and skin lesions associated with dermatoses, thus providing rapid treatment. Other first-line agents are potassium iodide and colchicine.

Although SS is a benign skin disorder which is rare in the literature, the diagnosis of patients in the emergency department may be helpful in the early diagnosis and treatment of life-threatening malignancies associated with dermatoses.

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Declaration of Competing Interest

None declared.

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