



CASE REPORT

Sweet's Syndrome Limited on the Palms and Soles: A Case Report

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Sweet's syndrome was first described as a reactive dermatosis characterized by sudden onset of fever, leukocytosis, and erythematous plaques infiltrated with neutrophils. Therefore, Sweet's syndrome is also known as acute febrile neutrophilic dermatosis. However, subsequently, it became clear that fever and neutrophilia in Sweet's syndrome vary depending on the case, and several other characteristics have been described. The lesions in Sweet's syndrome are typically observed not only in the limbs but also in the face, neck, and upper trunk. A 28-year-old female without a specific medical history presented in a hospital following the complaint of painful erythematous patches and pustules on her palms and soles. She had no previous history of palmoplantar pustulosis and other infections or malignancies. A skin biopsy showed diffuse dermal infiltration of neutrophils. Laboratory tests showed increased neutrophil count and erythrocyte sedimentation rate. After systemic corticosteroid administration was initiated, the lesions gradually disappeared. The patient was subsequently diagnosed with Sweet's syndrome according to histology, clinical feature, and response to treatment. However, there have been few reports of Sweet's syndrome confined to the individuals' palms and soles. According to the literatures, although the dorsum of the hand is frequently affected, the palmoplantar involve-

ment as in our case appears to be rare. (**Ann Dermatol 33(5) 459~462, 2021**)

-Keywords-

Foot, Hand, Skin diseases

INTRODUCTION

Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, was first described in 1964 by Sweet¹. The syndrome can be subdivided into the following groups: idiopathic, malignancy associated, and drug-induced². Cutaneous manifestation is characterized by tender erythematous plaques, nodules, or papules. Prodromes such as fever, malaise, or arthralgia are also observed. Histologic finding reveals diffuse infiltration of neutrophils in the dermis. The lesions in Sweet's syndrome are typically observed in the face, neck, upper trunk, and limbs. A few cases of Sweet's syndrome in the literature have been localized on the individuals' palms and soles. To the best of our knowledge, this is the first case of Sweet's syndrome in Korea with the lesions only observed on the patient's palmoplantar area.

CASE REPORT

A 28-year-old female presented with a 1-month history of multiple bullae, pustules, and erythematous patches on her palms and soles (Fig. 1). There was no previous history of skin disease or family history of psoriasis. She also had no other infection or medication history. During the first month prior to the hospital visit, the skin lesions did not spread beyond the palmoplantar area. However, the number of lesions had increased.

On examination, multiple raised erythematous patches covered with pustules and bullae confined to both her

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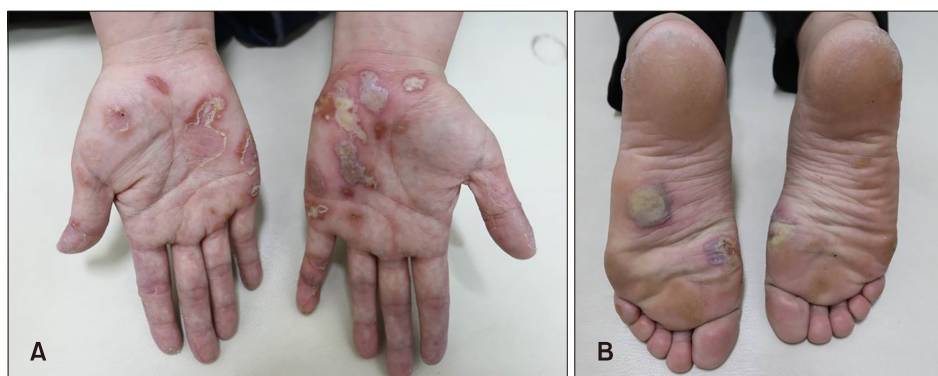


Fig. 1. (A, B) Multiple scattered bullae, pustules, and erythematous plaques with erosion on both palms and soles.

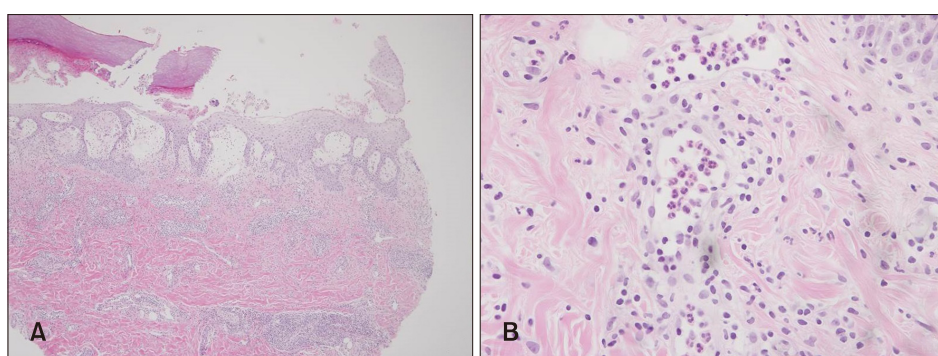


Fig. 2. Subcorneal blister formation, edema of the papillary dermis, and diffuse dermal infiltration of neutrophils and lymphohistiocytes are observed (A: H&E, ×40; B: H&E, ×400).

palms and soles were found, accompanied by severe pain. She did not complain of itching. The blisters were relatively hard on palpation. Patient's mucous membranes were unaffected, and further general examination was unremarkable.

Hematologic investigations showed an elevated white blood cell count, specifically neutrophils. Patient's erythrocyte sedimentation rate increased to 53 mm/h, and her C-reactive protein level was 9.1 mg/dl. Other laboratory results, including serum biochemistry, immunoglobulins, antistreptolysin O titers, viral titers, and potassium hydroxide preparation for fungal infection and fungus culture, were normal or negative.

A biopsy was performed on the palmar lesion. The histology showed subcorneal blister formation and spongiosis in the epidermis, edema of the papillary dermis, diffuse interstitial infiltration of neutrophils, and superficial and deep perivascular infiltration of lymphohistiocytes and neutrophils without vasculitis (Fig. 2). The immunofluorescence findings were negative.

These findings were consistent with the clinical diagnosis of Sweet's syndrome. The patient initially received oral prednisolone at a dose of 20 mg/day, which led to an improvement of skin symptoms. However, while tapering the dosage of prednisolone, there were two relapses. Again, the patient took prednisolone at a dose of 20 mg/day; sub-

sequently, the dosage was carefully reduced by 5 mg monthly to 7.5 mg/day, and the patient's skin condition has well maintained with improved condition since then. We received the patient's consent form about publishing all photographic materials.

DISCUSSION

Sweet's syndrome (acute febrile neutrophilic dermatosis) was first described in 1964¹. Sweet's syndrome is characterized by fever, leukocytosis, and tender erythematous plaques, which could be observed in any parts of the body. On histology, a dermal infiltrate of neutrophils was observed, several of which showed nuclear fragmentation³.

Sweet's syndrome can be classified into idiopathic (50% of patients), malignancy-associated (up to 35% of patients), and drug-induced⁴. The idiopathic type or classic type typically develops few days after an upper respiratory tract infection, and it is characterized by the sudden onset of tender red-colored skin plaques and nodules. The surface of the plaques can have vesicular or transparent appearance due to severe edema of the upper portion of the dermis.

There are various atypical variants of Sweet's syndrome. Patients with inflammatory bowel syndrome may develop

a pustular variant characterized by pustules overlying erythematous plaques. A second variant presents a subcutaneous manifestation, characterized by erythematous and tender dermal nodules over the extremities, and a third variant develops bullous or pustular lesions. New histopathologic variants, such as histiocytoid, neutrophilic panniculitis, and Sweet's syndrome-associated leukemia cutis, have recently gained special attention^{5,6}.

The diagnostic criteria suggested in 1986 have been generally accepted⁷. To establish the diagnosis of Sweet's syndrome, two major criteria comprising characteristic skin lesions and a largely neutrophilic infiltration of the dermis without leukocytoclastic vasculitis should be observed in addition to two of the following four minor criteria: (1) pyrexia; (2) association among an underlying hematologic or visceral malignancy, inflammatory disease, or pregnancy or preceded by an upper respiratory or gastrointestinal infection or vaccination; (3) elevated erythrocyte sedimentation rate and C-reactive protein levels and presence of leukocytosis or neutrophilia; and (4) excellent response to systemic steroid treatment or potassium iodide.

Multiple lesions appear predominantly, but isolated lesions have also been reported. The lesions in Sweet's syndrome are typically observed not only in the limbs but also in the face, neck, and upper trunk. Plaques are usually several centimeters in size, but larger lesions up to 20 cm in width have been described. Discrete pustules may occur close to more typical lesions. The lesion may involve the mucous membrane. Rarely, systemic involvement is possibly observed in the lungs, liver, kidneys, and central nervous system. Histopathologic examination shows a dense dermal inflammatory infiltration mainly comprising neutrophils, with no features of leukocytoclastic vasculitis. Sweet's syndrome is generally accepted as a reactive dermatosis. It is hypothesized that the pathogenesis of Sweet's syndrome is caused by helper T cells through the production of cytokines, such as interferon-gamma, interleukin-2, and interleukin-1, and granulocyte colony-stimulating factor, which are found to be remarkably elevated in the serum of patients with Sweet's syndrome^{8,9}. Another suggested mechanism of the disease is the inappropriate function of neutrophils, including inadequate lysosomal enzyme activity, lowered oxidative burst, and both activated and inactivated neutrophil chemotaxis¹⁰.

Systemic corticosteroids or potassium iodide are the treatment of choice for Sweet's syndrome. Most of the patients have a complete response to treatment. Dapsone is used as a second-line treatment in patients who experienced recurrence with systemic corticosteroids.

In this case, we initially suspected palmoplantar pustulosis. However, we diagnosed this case as Sweet's syndrome for

the following reasons: (1) sudden onset of painful skin lesions without itching; (2) diffuse dermal neutrophilic infiltration in the histopathological examination; (3) increased erythrocyte sedimentation rate and C-reactive protein levels and presence of leukocytosis; (4) prompt response to systemic steroid and completes treatment without recurrence; and (5) exclusion of other diseases through immunofluorescence and fungal study. Subcorneal blister and spongiosis are not typical histological findings of Sweet's syndrome but are often observed¹¹. The patient's clinical manifestation was larger bulla and pustule, than the microvesicle. Moreover, the blisters were relatively hard on palpation. For these reasons, dyshidrotic eczema could be ruled out. In addition, the id reaction was differentiated by excluding fungal infection through potassium hydroxide preparation, fungus culture, and biopsy. Autoimmune bullous diseases were ruled out on the basis of biopsy findings, clinical manifestations, and negative immunofluorescence findings. In our case report, the patient had no previous histories of palmoplantar pustulosis and other infections or malignancies.

Sweet's syndrome can occur anywhere in the body, specifically on the face, neck, trunk, and extremities. However, there have been few reports of skin lesions confined to the palms and soles. Consistent with our case, some reported cases showed lesion recurrence after steroid withdrawal^{12,13}. More studies will be subsequently required to confirm our findings, but considering the palmoplantar involvement in Sweet's syndrome, a more careful tapering of steroid doses is required.

If erythematous plaques with pustules confined to both palms and soles are observed, clinicians will initially suspect palmoplantar pustulosis. Based on our case report, clinicians should be aware of the lesions in Sweet's syndrome that are possibly confined to both palms and soles. For suspected patients, additional history taking, laboratory tests, and biopsy are required.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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REFERENCES

1. Sweet RD. An acute febrile neutrophilic dermatosis. *Br J Dermatol* 1964;76:349-356.
2. Cohen PR, Kurzrock R. Sweet's syndrome: a neutrophilic dermatosis classically associated with acute onset and fever. *Clin Dermatol* 2000;18:265-282.
3. Kemmett D, Hunter JA. Sweet's syndrome: a clinicopathologic review of twenty-nine cases. *J Am Acad Dermatol* 1990;23(3 Pt 1):503-507.
4. Rochet NM, Chavan RN, Cappel MA, Wada DA, Gibson LE. Sweet syndrome: clinical presentation, associations, and response to treatment in 77 patients. *J Am Acad Dermatol* 2013;69:557-564.
5. Marzano AV, Ishak RS, Saibeni S, Crosti C, Meroni PL, Cugno M. Autoinflammatory skin disorders in inflammatory bowel diseases, pyoderma gangrenosum and Sweet's syndrome: a comprehensive review and disease classification criteria. *Clin Rev Allergy Immunol* 2013;45:202-210.
6. Neoh CY, Tan AW, Ng SK. Sweet's syndrome: a spectrum of unusual clinical presentations and associations. *Br J Dermatol* 2007;156:480-485.
7. Su WP, Liu HN. Diagnostic criteria for Sweet's syndrome. *Cutis* 1986;37:167-174.
8. Giasuddin AS, El-Orfi AH, Ziu MM, El-Barnawi NY. Sweet's syndrome: is the pathogenesis mediated by helper T cell type 1 cytokines? *J Am Acad Dermatol* 1998;39:940-943.
9. Kawakami T, Ohashi S, Kawa Y, Takahama H, Ito M, Soma Y, et al. Elevated serum granulocyte colony-stimulating factor levels in patients with active phase of sweet syndrome and patients with active behcet disease: implication in neutrophil apoptosis dysfunction. *Arch Dermatol* 2004;140:570-574.
10. von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). *J Am Acad Dermatol* 1994;31:535-556; quiz 557-560.
11. Sitjas D, Puig L, Cuatrecasas M, De Moragas JM. Acute febrile neutrophilic dermatosis (Sweet's syndrome). *Int J Dermatol* 1993;32:261-268.
12. Keefe M, Wakeel RA, Kerr RE. Sweet's syndrome, plantar pustulosis and vulval pustules. *Clin Exp Dermatol* 1988;13:344-346.
13. Sommer S, Wilkinson SM, Merchant WJ, Goulden V. Sweet's syndrome presenting as palmoplantar pustulosis. *J Am Acad Dermatol* 2000;42(2 Pt 2):332-334.