

Case Report

A case of Sweet's syndrome with marked facial swelling and fluid collection

一例有明顯面部腫脹並積液的史維特症候群

JE Seol 薛禎恩, SH Park 朴蘇熙, DH Kim 金道亨, JN Kang 姜情暖, H Kim 金孝鎮

A 43-year-old woman presented with a one-day history of painful erythematous patches with swelling on face, accompanied by fever of 38.4°C. Laboratory tests showed leukocytosis with neutrophil predominance and increased erythrocyte sedimentation rate and C-reactive protein level. Histopathological examination revealed dense neutrophil infiltration in dermis. She was diagnosed with Sweet syndrome and treated with systemic corticosteroids, after which she showed clinical improvement. As the corticosteroids were being tapered, however, facial swelling developed with focal fluid collection. Incision and drainage were performed, and cyclosporine, colchicine, and dapsone were administered. After 16 weeks, she showed marked clinical improvement.

一名四十三歲的女性，因持續一天的臉部腫脹和伴隨的疼痛紅斑及攝氏 38.4 度發熱而求診。化驗所檢查顯示白血球增多，當中以嗜中性粒細胞為主；另外，紅細胞沉降率和C反應蛋白水平升高。組織病理學檢查發現病灶真皮層有嗜中性粒細胞浸潤。她被診斷為患有史維特症候群，施以全身性皮質類固醇治療後，皮膚情況得到臨床改善。但隨著皮質類固醇逐漸減量，面部卻又再次腫脹並有局部積液。在積液處進行了切口及引流手術，並改用環孢菌素、秋水仙鹼和氨苯砒等藥物治療。16 週後，患者的臨床症狀有顯著改善。

Keywords: Sweet syndrome

關鍵詞：史維特症候群

**Department of Dermatology, Busan Paik Hospital,
Busan, Korea**

JE Seol, MD
SH Park, MD
DH Kim, MD
JN Kang, MD
H Kim, MD, PhD

Correspondence to: Dr. H Kim

Department of Dermatology, Busan Paik Hospital, College of Medicine, Inje University, 75, Bokji-ro, BusanJin-gu, Busan, Korea (postal code: 47392)

Introduction

Sweet's syndrome (SS) commonly manifests as painful erythematous papules and nodules on the face, neck, and upper extremities,¹ accompanied by fever, malaise, and leukocytosis.² The exact cause of SS is unknown.^{3,4} Systemic corticosteroids are the gold standard therapy for SS.^{5,6} We report an unusual case of SS with development of focal fluid collection.

Case report

A 43-year-old woman presented with a painful erythematous patch with marked swelling on the face. She also complained of a febrile sensation, malaise, and headache, which began abruptly the day before. She had had a history of upper respiratory infection one week earlier, but there was no history of trauma or medication use.

Physical examination revealed tender erythematous swollen patches on her face, especially on the forehead and both cheeks (Figure 1). Body temperature was 38.4°C. The laboratory evaluation showed neutrophilia (18,309/mm²), an increased C-reactive protein level (CRP; 28.89 mg/dL) and increased



Figure 1. Erythematous swollen patches were shown on the face.

erythrocyte sedimentation rate (ESR; 116 mm/h). Diabetes mellitus was diagnosed incidentally at admission by a routine screening blood test. All other tests were within normal limits, including liver and renal function tests.

Histopathological examination of a forehead specimen showed mild papillary dermal oedema and dense inflammatory cell infiltration, consisting mainly of neutrophils and histiocytes (Figure 2).

Based on the clinical, laboratory, and histopathological findings, we made a provisional diagnosis of SS, ruling out cellulitis. Methylprednisolone 16 mg/day, dexamethasone 10 mg/day, and cefazodone 2 g/day were administered together at first. Gram staining and tissue cultures performed to exclude infectious causes were negative. Therefore, infectious causes were excluded and cefazodone was stopped on the fifth day. Both the clinical features and laboratory parameters improved (neutrophils 6,347/mm², CRP level 3.63 mg/dL). The patient was discharged seven days after admission on a decreased dose of systemic corticosteroid (methylprednisolone 4 mg/day) (Figure 3).

However, two weeks after discharge from our hospital, the skin lesions worsened, especially on the forehead (Figure 4a). The methylprednisolone dose was increased to 16 mg/day, and cyclosporine 200 mg/day was added, but there was no clinical improvement (Figures 4b & 4c). Ultrasonography showed diffuse fluid collections in her forehead. Yellowish mucoid fluid was drained from these lesions on four occasions. Gram staining and fluid cultures were negative, and cytological analysis revealed increased neutrophils (>30/mm²). Colchicine 1.2 mg/day and dapsone 50 mg/day were added. After 10 weeks of treatment, the skin lesions had improved markedly (Figure 4d). Maintenance treatment with colchicine 0.6 mg/day and

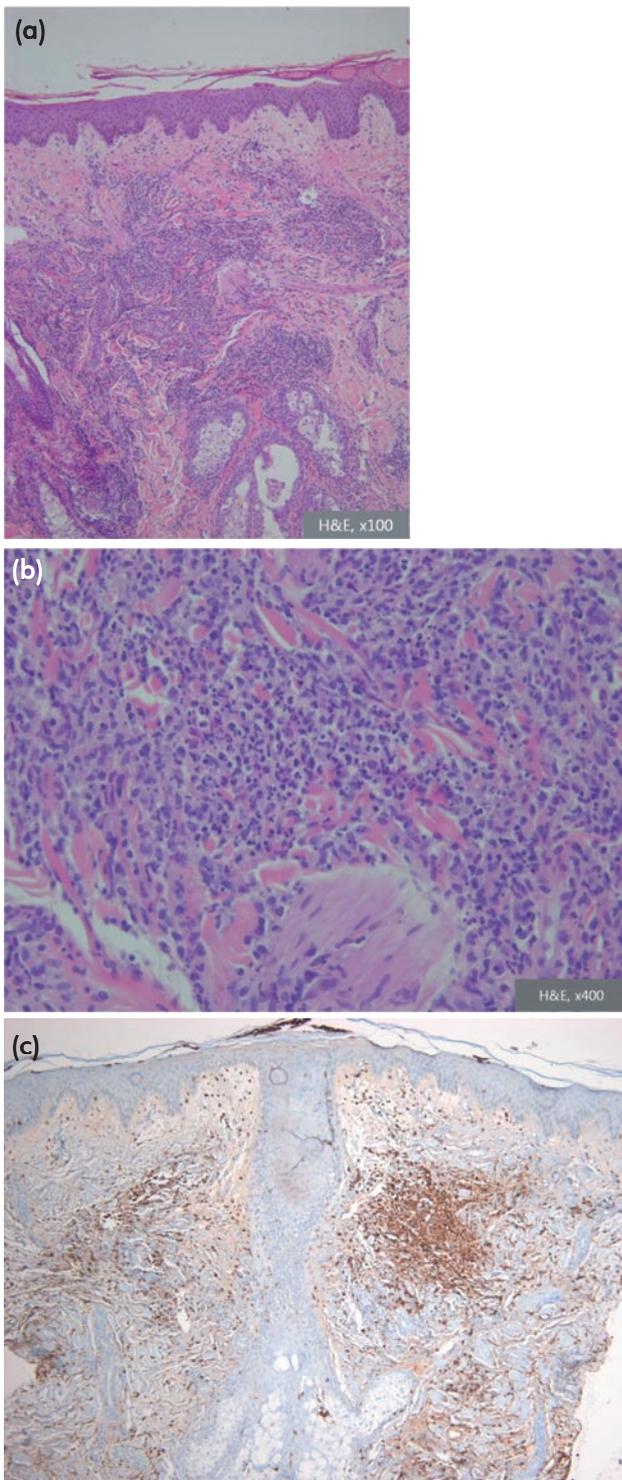


Figure 2. (a) Histopathological examination revealed mild papillary dermal oedema and dense inflammatory cells infiltration (H&E, x100). (b) High-powered view of specimen showed inflammatory cells mainly consisted of neutrophils and histiocytes (H&E, x400). (c) Myeloperoxidase staining showed positive result (MPO, x100).

dapsone 50 mg/day was given for three months, and there was marked improvement by the end of treatment (Figure 4e).

Discussion

Sweet's syndrome, also known as acute febrile neutrophilic dermatosis, is characterised by painful skin lesions, pyrexia, neutrophilia, elevated acute phase reactants, and a rapid response to corticosteroids.² The exact cause of SS is unclear, but infections, autoimmune diseases, malignant diseases and drugs are thought to be associated with SS.⁷ It has been suggested that circulating antibodies, cytokines, and immune complexes contribute to its pathogenesis.⁷ Underlying malignancy, especially haematological malignancy, should be ruled out in cases of SS with severe manifestations.⁷ Our patient had severe skin lesions, marked facial swelling, and



Figure 3. Skin lesions improved after one week of systemic corticosteroid treatment.

focal fluid collection on the forehead, but there was no evidence of haematological malignancy on laboratory evaluation.

There are two major and four minor criteria for SS.⁸ The major criteria are: (1) sudden onset of painful erythematous plaques or nodules, (2) dermal oedema and neutrophilic infiltration of the dermis without signs of vasculitis histopathologically.⁸ The four minor criteria are (1) associated underlying disease (upper respiratory tract or gastrointestinal infection, autoimmune disorder, visceral or haematological malignancy or pregnancy), (2) fever ($>38^{\circ}\text{C}$), (3) abnormal laboratory findings initially (ESR >20 mm/h, positive CRP, leukocytosis >8000 , neutrophils $>70\%$), and (4) an immediate response to systemic therapy with corticosteroids or potassium iodide.⁸ Both major criteria and at least two minor criteria should be met to diagnose SS.⁸ Our patient had skin lesions consisting of warm, erythematous, swollen patches and focal fluid collection resembling cellulitis, but the histopathological findings that were consistent with SS, and all of the minor criteria were met. In addition, tissue and fluid cultures revealed no evidence of infection.

First-line treatment of SS includes systemic corticosteroids, potassium iodide, and colchicine. Of these, corticosteroids are the main treatment and are often administered at a dose of 1 mg/kg/day prednisone and tapered to 10 mg/day within four to six weeks.⁹ Second-line systemic treatment includes indomethacin, clofazimine, cyclosporine, and dapsone. All of these agents can be used as monotherapy or combination therapy either as the first-line therapy or after failure of first-line therapy.⁹ Our patient responded rapidly to systemic corticosteroid therapy. However, the early tapering of the systemic corticosteroids due to the incidental diagnosis of diabetic mellitus might have contributed to the worsening of the skin lesions and severe inflammation might have resulted in the focal neutrophil-rich fluid collection.

Several cases of SS with atypical clinical features, such as cellulitis-like lesion, zoster-like lesion, rapidly expanded erythematous lesion with haemorrhage, and multiple tense pustules and abscess lesion, have been reported,¹⁰⁻¹² but SS manifesting as swollen erythematous patches and subsequent focal fluid collection has not been reported. Here, we reported a case of SS with unusual clinical features.



Figure 4. (a) Skin lesions aggravated, especially in forehead, two weeks after discharge. (b,c) Exacerbation of skin lesions with focal fluid collection on the forehead despite an increased dose of systemic corticosteroid and addition of cyclosporin. (d) Skin lesions significantly improved with administration of dapsone, colchicine and fluid drainage on four occasions. (e) After three months of maintenance treatment with dapsone and colchicine, skin lesions resolved completely.

References

1. Anzalone CL, Cohen PR. Acute febrile neutrophilic dermatosis (Sweet's syndrome). *Curr Opin Hematol* 2013;20:26-35.
2. Cohen PR. Sweet's syndrome: a comprehensive review of an acute febrile neutrophilic dermatosis. *Orphanet J Rare Dis* 2007;26:34.
3. Marzano AV, Cugno M, Trevisan V, Lazzari R, Fanoni D, Berti E, et al. Inflammatory cells, cytokines and matrix metalloproteinases in amicrobial pustulosis of the folds and other neutrophilic dermatoses. *Int J Immunopathol Pharmacol* 2011;24:451-60.
4. Wojcik AS, Nishimori FS, Santamaria JR. Sweet's syndrome: a study of 23 cases. *An Bras Dermatol* 2011; 86:265-71.
5. Cohen PR. Neutrophilic dermatoses: a review of current treatment options. *Am J Clin Dermatol* 2009;10:301-12.
6. Schadt CR, Callen JP. Management of neutrophilic dermatoses. *Dermatol Ther* 2012;25:158-72.
7. Villarreal-Villarreal CD, Ocampo-Candiani J, Villarreal-Martinez A. Sweet Syndrome: A Review and Update. *Actas Dermosifiliogr* 2016;107:369-78.
8. Su WP, Liu HN. Diagnostic criteria for Sweet's syndrome. *Cutis* 1986;37:167-74.
9. Cohen P, Honigsmann H, Kurzrock R. Acute febrile neutrophilic dermatosis, In: Freedberg IM, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffell DJ, Wolff K, editors. *Fitzpatrick's dermatology in general medicine*. 8th ed. New York: McGraw-Hill, 2012:362-70.
10. Surovy AM, Pelivani N, Hegyi I, Buettiker U, Beltraminelli H, Borradori L. Giant cellulitis-like Sweet Syndrome, a new variant of neutrophilic dermatosis. *JAMA dermatol* 2013;149:79-83.
11. Kroshinsky D, Alloo A, Rothschild B, Cummins J, Tan J, Montecino R, et al. Necrotizing Sweet syndrome: a new variant of neutrophilic dermatosis mimicking necrotizing fasciitis. *J Am Acad Dermatol* 2012;67:645-54.
12. Yoon TY, Ahn GB, Yang TH, Shin HS. Sweet's syndrome with abscess-like lesions in patient with acute myelogenous leukemia. *J dermatol* 2000;27:794-7.