CASE REPORT OPEN ACCESS

A case of simultaneously developed sweet's syndrome and systemic lupus erythematosus in a man

Hiroyuki Hounoki, Maiko Okumura, Koichiro Shinoda, Reina Ogawa, Hirofumi Taki, Kazuyuki Tobe, Kyoko Shimizu, Teruhiko Makino, Tadamichi Shimizu

ABSTRACT

Introduction: Sweet's syndrome is characterized by the development of tender, erythematous plaques, fever, arthralgia and leukocytosis. Sweet's syndrome is reported to be associated with underlying infections, malignant neoplasms, pregnancy, drugs, and autoimmune diseases. However, Sweet's syndrome associated with systemic lupus erythematosus has been rarely reported. Although both Sweet's syndrome and systemic lupus erythematosus predominantly occur in women, we are reporting a case of simultaneously developed Sweet's syndrome and systemic lupus erythematosus in a man. Case Report: A 48-year-old previously healthy man was admitted for intermittent fevers, arthralgias, and eruption. Laboratory investigations revealed elevated C-reactive protein, lymphocytopenia, and low complements with elevated immune complex. Antinuclear antibodies, anti-DNA antibodies were positive. Systemic lupus erythematosus was diagnosed based on the presence of arthritis, lymphopenia, anti-DNA antibodies and antinuclear antibodies. A biopsy specimen from an erythematous papule from the right sole showed a slight lichenification and an infiltration of neutrophils and lymphocytes in dermis without leukocytoclastic vasculitis. Thus, this case also met the modified criteria for Sweet's syndrome proposed by von den Driesch. Prednisolone was begun at a dose of 20 mg/day, resulting in diminishing skin eruption and resolution of arthralgia. Conclusion: Sweet's syndrome is categorized into classical, drug-induced, and malignancy-associated form. Only one case of concurrence of classical Sweet's syndrome and systemic lupus erythematosus has been reported in a man. This is the second case of coincidence of classical Sweet's syndrome and systemic lupus erythematosus in an adult man.

Keywords: Sweet's syndrome, Systemic lupus erythematosus, Erythematous plaques, Arthralgia

****

Hounoki H, Okumura M, Shinoda K, Ogawa R, Taki H, Tobe K, Shimizu K, Makino T, Shimizu T. A case of simultaneously developed sweet's syndrome and systemic lupus erythematosus in a man. International Journal of Case Reports and Images 2012;3(9):36–38.

****

doi:10.5348/ijcri-2012-09-180-CR-10

Hiroyuki Hounoki[1], Maiko Okumura[2], Koichiro Shinoda[1], Reina Ogawa[2], Hirofumi Taki[1], Kazuyuki Tobe[1], Kyoko Shimizu[3], Teruhiko Makino[3], Tadamichi Shimizu[3]
Affiliations: [1]First Department of Internal Medicine, University of Toyama, 2630 Sugitani, Toyama, Toyama 930-0194, Japan; [2]Department of Dermatology, University of Toyama, 2630 Sugitani, Toyama, Toyama 930-0194, Japan.
Corresponding Author: Hirofumi Taki, First Department of Internal Medicine, University of Toyama, 2630 Sugitani, Toyama, Toyama 930-0194, Japan; Ph: +81 76 434 7287; Fax: +81 76 434 5025; Email: htaki-tym@umin.ac.jp

Received: 17 July 2011
Accepted: 11 April 2012
Published: 01 September 2012

INTRODUCTION

Sweet's syndrome, or acute febrile neutrophilic dermatosis, was first described by Sweet RD in 1964 [1]. This syndrome is characterized by the development of tender, erythematous plaques, fever, arthralgia and leukocytosis [2, 3]. The syndrome is reported to be

associated with underlying infections, malignant neoplasms, pregnancy and drugs [2, 3]. In addition, Sweet's syndrome is also associated with autoimmune diseases [2, 3]. However, Sweet's syndrome associated with systemic lupus erythematosus (SLE) has been rarely reported [4–6]. Here, a case of simultaneously developed Sweet's syndrome and SLE in a man is reported.

CASE REPORT

A 48-year-old previously healthy man was admitted for intermittent fevers, arthralgias, and eruptions. He had never taken any meditations before occurrence of the symptoms. Laboratory investigations were C-reactive protein 2.31 mg/dL (normal range: <0.3 mg/dL), white blood cells 4300/mm^3 with lymphocytes 830/mm^3, CH50 18 U/mL (normal range: 30–46 U/mL), C3 37.0 mg/dL (normal range: 71–129 mg/dL), C4 5.7 mg/dL (normal range: 12–34 mg/dL) and immune complex 165.0 μg/mL (normal range: <3.0 μg/mL). Viral markers for hepatitis were negative. Antinuclear antibodies (ANA), anti-DNA antibodies were positive at 1:40 speckled pattern and 19 U/mL (normal range: <10 U/mL), respectively. Anti-SS-A antibodies, anti-SS-B antibodies, antineutrophil cytoplasmic antibodies against proteinase 3 and myeloperoxidase, anti-RNP antibodies, anti-Sm antibodies, anti-Jo-1 antibodies, anti-cardiolipin antibodies and lupus anticoaglant were all negative. Screening for malignancy by means of gallium scintigraphy, chest and abdominal computed tomography, and gastrointestinal fiberscope indicated no evidence of malignancy. Leukocytosis, which is a predominant feature of Sweet's syndrome, was not always observed in this case [1]. Rather, lymphopenia in addition to positive ANA and anti-DNA antibodies with hypocomplimentemia and high levels of immune complex in the serum were observed in this case. SLE was diagnosed based on the presence of arthritis, lymphopenia, ANA and anti-DNA antibodies according to the revised classification criteria of SLE.

A biopsy specimen from an erythematous papule from the right sole (Figure 1) showed a slight lichenification and an infiltration of neutrophils and lymphocytes in dermis without leukocytoclastic vasculitis (Figure 2A–B). Furthermore, immunofluorescence was positive for complement deposition at dermal-epidermal junction, indication a positive for lupus band test. This finding further helps to confirm the diagnosis of SLE. Oral administration of prednisolone was begun at a dose of 20 mg/day, resulting in diminishing skin eruptions and a resolution of arthralgia. Thus, this case met the modified criteria for Sweet's syndrome proposed by von den Driesch [1].

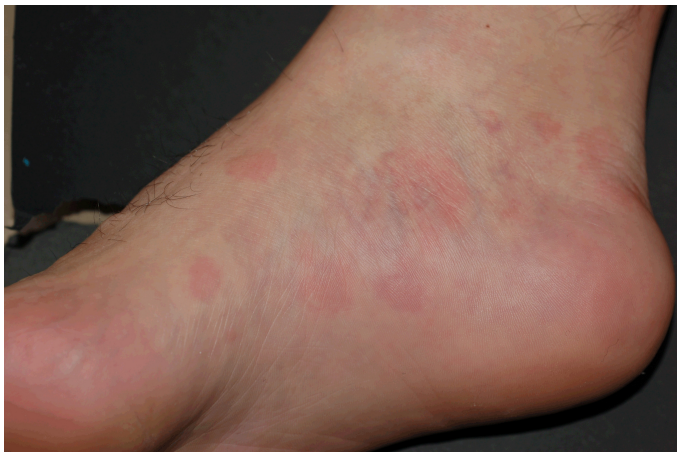

Figure 1: Erythematous plaques on the right sole.

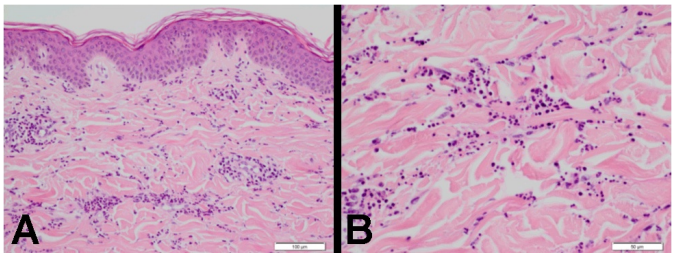

Figure 2: Hematoxylin-eosin stained histopathological images. Skin lesion showed a slight lichenification and an infiltration of neutrophils and lymphocytes in dermis without leukocytoclastic vasculitis in (A) low-power (original magnification X40), and (B) high-power (original magnification X400) magnification.

DISCUSSION

Sweet's syndrome is reported to be associated with underlying infections, malignant neoplasms, pregnancy, drugs, and autoimmune diseases [2, 3]. Thus, Sweet's syndrome is categorized into classical, drug-induced, and malignancy-associated form. We diagnosed this case as a classical Sweet's syndrome.

The characteristic histopathologic features of Sweet's syndrome are diffuse infiltrate of predominantly neutrophils with leukocytoclasia, papillary dermal edema, swollen endothelial cells and an absence of leukocytoclastic vasculitis. The pathological findings of this patient were consistent with Sweet's syndrome. In addition, immunofluoresence of immunoglobulin and complement deposition at the basement membrane of the skin was positive along with serum hypocomplementemia and high immune complex levels, suggesting a coexistence of SLE.

Concurrence of lupus including drug-induced lupus and subacute cutaneous lupus with Sweet's syndrome had been reported previously in adults [7, 8]. However, only three cases of concurrence of classical Sweet's syndrome and SLE have been reported, in whom two are women and one is a man [4–6]. Although both classical Sweet's syndrome and SLE predominantly occur in women, this is the second case of coincidence of classical Sweet's syndrome and SLE in an adult man.

CONCLUSION

Sweet's syndrome associated with SLE has been

rarely reported. This is a second case of concurrence of classical Sweet's syndrome and SLE observed in a male patient.

****

Author Contributions

Hiroyuki Hounoki – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Maiko Okumura – Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published
Koichiro Shinoda – Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published
Reina Ogawa – Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published
Hirofumi Taki – Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Kazuyuki Tobe – Final approval of the version to be published
Kyoko Shimizu – Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published
Teruhiko Makino – Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published
Tadamichi Shimizu – Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). J Am Acad Dermatol 1994;31:535–6.
2. Sweet RD. An acute febrile neutrophilic dermatitis. Br J Dermatol 1964;76:349–56.
3. Cohen PR. Sweet's syndrome-a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis 2007;2:34.
4. Hou TY, Chang D-M, Gao H-W, Chen C-H, Chen H-C, Lai J-H. Sweet's syndrome as an initial presentation in systemic lupus erythematosus: a case report and review of the literature. Lupus 2005;14:399–402.
5. Gollol-Raju N, Bravin M, Crittenden D. Sweet's syndrome and systemic lupus erythematosus. Lupus 2009;18:377–8.
6. Fernandes NF, Castelo-Soccio L, Kim EJ, Werth VP. Sweet syndrome associated with new-onset systemic lupus erythematosus in a 25-year-old man. Arch Dermatol 2009;145:608–9.
7. Goette GK. Sweet's syndrome in subacute cutaneous lupus erythematosus. Arch Dermatol 1985;121:789–91.
8. Sequeria W, Polisky RB, Alrenga DP. Neutrophilic dermatitis (Sweet's syndrome); association with a hydralazine-induced lupus syndrome. Am J Med 1986;81:558–60.