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# Sweet's Syndrome: A Rare Case Report

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## ABSTRACT

Sweet's Syndrome, also known as acute febrile neutrophilic dermatosis is a rare disease of unknown etiology, but has been associated with autoimmune processes, malignancies, infections, GI disorders and drug reactions. It is more prevalent in women than men, except in case of malignancy associated origin. Clinical features of this disorder include acute onset of painful plaques on skin, neutrophilic leukocytosis, fever, dermal infiltrations by neutrophils. Usually, sweet's syndrome is presented in three main clinical settings which includes: the classical (idiopathic), malignancy-associated, and drug induced. Classical Sweet's Syndrome (CSS) is usually seen in women between the age of 30 to 50 years. It is frequently preceded by an upper respiratory illness and could be linked to pregnancy, inflammatory bowel disease, and both. Recurrence of the dermatosis is usually seen in one third of the patients experiencing classical sweet's syndrome. Malignancy associated Sweet's Syndrome (MASS) is usually present in patients with confirmed cancer and manifested as a paraneoplastic syndrome. Also occurs in those whose solid tumor or hematologic dyscrasia associated with sweet's syndrome was previously undetected. Drug induced Sweet's Syndrome (DISS) occurs in patients being treated with granulocyte-colony stimulating factor. Various other drugs including antibiotics like Minocycline, Trimethoprim Sulfamethoxazole, nitrofurantoin, antiepileptics like Carbamazepine, Diazepam, NSAIDS, Diuretics and Retinoids can also cause DISS. Diagnosis of sweet syndrome is usually done by two major criteria (histopathology and acute cutaneous lesion), and four minor criteria's (infection, malignancy, medications, fever, and extra cutaneous manifestations like leukocytosis and corticosteroid response. Systemic corticosteroids are the golden standard of treatment for Sweet's Syndrome. Systemic corticosteroids (0.5mg to 1mg/kg/day) for 4-6 weeks improves the systemic symptoms and reduce the skin lesions. For treating localized lesions, topical application of high potency corticosteroids or intralesional corticosteroids are effective. Other first line agents including potassium iodide and colchicine are also used. Second line agents include Indomethacin, Cyclosporine, Dapsone and Clofazimine.

***Keywords:*** Sweet's Syndrome, Corticosteroids, Malignancy, Leukocytosis

## INTRODUCTION

Sweet's syndrome also known as (acute febrile neutrophilic dermatosis) a rare disorder was first described in 1964 by Robert Douglas Sweet. [1] It is mainly characterized by fever, acute onset of typical skin lesions, neutrophilic leukocytosis, acute onset of painful plaques and histological findings of dense neutrophilic infiltrations on the skin. [2] Histological features include neutrophilic infiltrate which is located on the superficial layer of the dermis and papillary edema.

Usually, Sweet's syndrome is presented in three main clinical settings out of which [1]. the classical one or the para-infectious sweet's syndrome; represented as a hypersensitivity reaction that occurs before an infection, [2]. Malignancy associated sweet's syndrome (para-neoplastic), usually occurs in association with acute myelogenous leukemia in children. [3]. An adverse drug reaction which occurs less frequently, connected with an underlying disease condition (drug induced sweet's syndrome). Medications associated with drug induced Sweet's Syndrome include Antibiotics (Minocycline, Trimethoprim Sulfamethoxazole, Nitrofurantoin, Nor-floxacin), Antiepileptics (Carbamazepine, Diazepam), Colony stimulating factors (Granulocyte colony stimulating factors), NSAIDS (Celecoxib, Diclofenac), Antineoplastics (Bortezomib, Lenalidomide), Retinoids (All trans retinoic acid).[3] Usually the confirmatory diagnostic test for sweet's syndrome is the skin biopsy. Oedema with infiltrate of mature neutrophils with neutrophil fragmentation in the upper epidermal region are the hallmarks of the histological examination. Upper extremities, neck and the face are the common sites where skin lesions occur. Systemic corticosteroids are the golden standard of treatment for Sweet's Syndrome. Systemic corticosteroids (0.5mg to 1mg/kg/day) for 4-6 weeks improves the systemic symptoms and reduce the skin lesions.

## CASE REPORT

A 61 years old female patient was admitted under General medicine department in a tertiary care hospital presenting with the complaints of fever for 6 days (on and off) and macuopapular nodules and rashes all over the skin for 3 days. The patient had medical history of Dyslipidemia and Hypothyroidism. On examination the patient was conscious and febrile (Temperature: $99^0$F). Physical examination showed erythematous papules and oedematous plaques all over the skin, non tender urticated plaques over the forearm, few atypical lesions and mild pruritis. The patient had a medical History of Hypothyroidism, Dyslipidaemia and was treated with T. Levothyroxine 50mcg,T. Rosuvastatin 5mg for the respective disease condition. Among the laboratory tests performed, neutrophils, ESR, CRP, rheumatoid factor, serum LDH and Ferritin were found to be elevated. Sweet's syndrome was suspected and skin biopsy was indicated for histopathological study. The histology described epidermis with focal mild acanthosis, parakeratosis, and spongiosis. Reticular dermis showed perivascular and interstitial neutrophilic infiltration. Areas of oedema with leukocytoclasis were also noted. Some of the sections showed perivascular lymphohistiocytic infiltration along with occasional eosinophils. Mild neutrophilic exocytosis is also present. The patient was treated under the expert guidance of a Dermatologist at the tertiary care hospital. The respective condition was managed with Tab. Prednisone (Omnacortil) 80mg, Inj. Ceftriaxone (Monocef) 1gm, Cap Doxycycline (Doxy) 100mg, Inj. Pantoprazole (Pantop) 40mg for 7days, together with Tab Ivermectin 12mg, Tab. Levocetirizine 5mg was given for managing allergic symptoms and Dermadew Cream, Mometasone cream, Mupirocin ointment for and Liquid cream (after bath) was given for local application to the skin blisters. On the $7^{th}$ day she was discharged with stable condition, skin lesions also partially healed. Oral Corticosteroid Tab. prednisolone 30mg OD for 5 days, followed by Tab. prednisolone 20mg OD for 1 week.

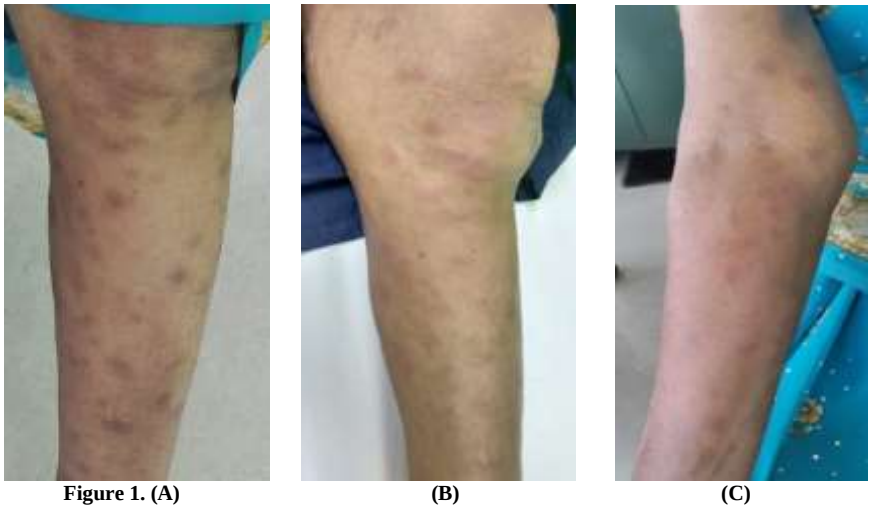

**Figure 1. (A)** **(B)** **(C)**

A : Blister and crusted ulcers in the left leg. B : Blisters in the right leg. C : Blisters and papules on right forearm and arms.

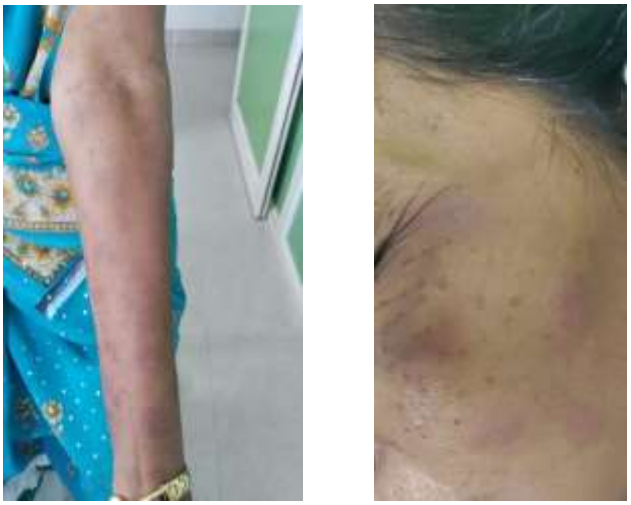

**Figure 2. (A)** **(B)**

A: Papules and erythematous nodules in left arm and forearms.
B: Blisters on the face.

## DISCUSSION

Sweet's Syndrome is a rare condition described as 'acute febrile neutrophilic dermatosis'. [4] Sweet's Syndrome can occur in three clinical types : classic/idiopathic, paraneoplastic, and drug induced.[5] Classical or para infectious Sweet's syndrome is usually characterized by high fever (higher than $38^0$C), neutrophilia and leucocytosis. Cutaneous manifestations like skin lesions with erythematous papules or nodules are common with this condition. Usually the confirmatory diagnostic test for sweet's syndrome is the skin biopsy. [6] Edema with infiltrate of mature neutrophils with neutrophil fragmentation in the upper epidermal region are the hallmarks of the histological examination. Upper extremities, neck and the face are the common sites where skin lesions occur. [7] Systemic symptoms that coexist with these symptoms are arthralgias, myalgias and headache. [8] The most commonly observed laboratory findings are neutrophilia and elevated ESR. The diagnostic criteria of Sweet's Syndrome include two major criteria and two minor criteria. [9] Among the major criteria; 1. Sudden onset of painful erythematous plaques or nodules, 2. Dense neutrophilic infiltrate. Minor criteria include fever $>38^0$C, abnormal laboratory test with elevated ESR, CRP, leucocytes and neutrophils, association with any underlying

malignancy, haematological or inflammatory disease, or pregnancy, also preceded by an upper respiratory tract infection or gastrointestinal infection. In our patient, the minor and major diagnostic criteria were prominent. Due to various associated pathologies, screening for sweet's syndrome is proposed. This includes measurement of blood pressure, laboratory tests including hemogram, ESR, LDH, and uric acid. [10] Also detection for HIV, Anti-Nuclear Antibodies (ANA) and serum immunoglobulin levels are tested. A numerous variety of medications have also contributed to the development of Sweet's Syndrome, which includes antibiotics, antineoplastics agents, antiepileptics, and colony stimulating factors. [11] In our patient the underlying cause of the disease was not clear. The skin manifestation of Sweet's syndrome can persist for weeks and months if not treated. The main stay of treatment for this disease is the use of corticosteroids. [12] The initial dose is 1mg/kg/day and tapered over 4-6 weeks of time period. In our case there was a good initial response to the use of corticosteroids, therefore progressive reduction of dose was done with clinical monitoring of the patient. [13] In cases of drug induced Sweet's syndrome, subsequent withdrawal of the drug results in resolution of the disease. [14] The patient must be followed up on regular basis monitored for recurrence of lesions as the recurrence rate of occurrence of skin lesions are high in these cases. Thus, follow up is important for investigating development of new skin lesions. [15] In our case the patient was regularly followed up after the discharge.

## CONCLUSION

The sweet's syndrome is an uncommon condition of unknown pathogenesis. In our patient the exact aetiology of developing Sweet's syndrome was not clearly established. But there are several mechanisms that contribute to the development of this disease. The mechanisms include genetic susceptibility, dermal dendrocytes, cytokines, circulating autoantibodies, and immune complexes. If not treated, the clinical manifestations may persist for weeks and months. The mainstay of treatment for sweet's syndrome is corticosteroids. However, first line agents like potassium iodide and colchicine and second line agents like dapsone, cyclosporine, indomethacin and clofazimine can be used.

### *Declaration by Authors*

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**Conflict of Interest:** The authors have the required patient consent form, on which the patients have agreed to participate in the study and be represented in the corresponding publication. Although the patients are aware that the writers would take precautions to take their names secret, anonymity cannot be guaranteed.

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