Sweet's syndrome: A Therapeutic challenge

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Abstract
Sweet syndrome (acute febrile neutrophilic dermatosis) is not a common disease, etiology of which is unknown. It is an inflammatory neutrophilic dermatosis characterized by non-itchy, tender, erythematous papules and plaques most commonly distributed on the arms, upper body, head and neck. It commonly affects adults in between 30- 60 years with female predominance with 4:1 sex ratio. The disease is associated with fever, peripheral leukocytosis and wide variety of diseases. It shows excellent response to systemic corticosteroids. A 28 year old female presented with multiple red elevated lesion over face, back, upper and lower limbs, which were associated with pain and fever. She gave history of sore throat before appearance of lesions. On cutaneous examination erythematous papules and plaques, plaques were studded with pseudovesicles were seen on face, back, upper and lower limbs. They were tender and associated with fever. Patient underwent clinical and systemic examination followed by routine and histopathological investigations. Complete blood picture showed neutrophilia (94%) and histopathological examination showed dense neutrophilic infiltrate with oedema in dermis. The patient was prescribed topical steroids, oral Dapsone and corticosteroids, but patient did not show any improvement. Later on patient was put on Dexamethasone pulse therapy along with oral Azathioprine for 18months. Lesions resolved and no recurrence was observed in months.

Keywords: Sweet syndrome, Neutrophilic dermatosis, Recurrent.

Introduction
Sweet syndrome is also known as acute febrile neutrophilic dermatosis and Gomm- button disease. The first case was described by Dr. Robert Douglas Sweet in 1964.[1] It is an uncommon disease of unknown etiology, characterized by tender, non-pruritic, erythematous papules or plaques, which may enlarge or coalesce to form plaques commonly distributed on the arms, upper body, head and neck. Its exact incidence is not known. It primarily affects adults, in age group of 30 to 60 years with a 4:1 female dominance.[2] It has been associated with autoimmune processes, malignancies, infections, drug reactions, and gastrointestinal disorders e.g. inflammatory bowel disease. Histopathological examination reveals dense dermal neutrophilic infiltrate with oedema.[3] It responds very well to systemic corticosteroids.

Case Report
A 28 year old female presented with multiple red elevated lesions over face, back, upper and lower limbs, which were acute in onset and gradually progessive. They were associated with pain and fever. She gave history of sore throat before appearance of lesions. On cutaneous examination erythematous papules measuring 0.3 to 1cm in diameter and plaques measuring 2-6 cm in diameter were present over abovementioned sites which were studded with pseudovesicles (Fig. 1,2,3). Lesions were tender and indurated. Patient underwent clinical and systemic examination followed by routine and histopathological investigations. Complete blood picture showed neutrophilia (94%), haemoglobin and total leucocyte count was 12.3g/dL and 21000/*u*L respectively. ESR was raised. Her lipid, liver, renal and urine profile was normal. 4mm punch biopsy was done under local anaesthesia from left upper limb which revealed dense dermal neutrophilic infiltrate with oedema. At first patient was prescribed topical steroids, oral Dapsone (100mg/day), corticosteroids (1mg/kg/day) and other symptomatic medications, but patient did not show evident improvement. Then patient was put on Dexamethasone pulse (DP) therapy which not only showed total remission but also averted recurrence along with oral azathioprine in dose of 50mg twice daily also given. DP therapy was given for 18months, but Azathioprine was continued for another 6months and patient is followed up for 2 years.

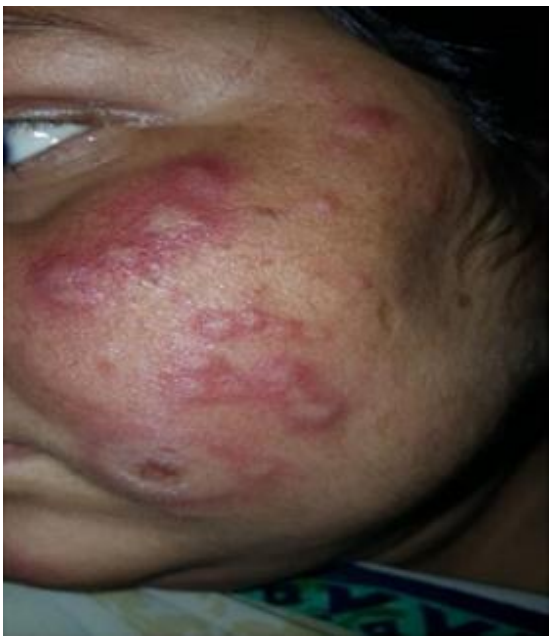

Fig. 1: Showing papules and ulceration in the plaque with erythematous base over cheek.

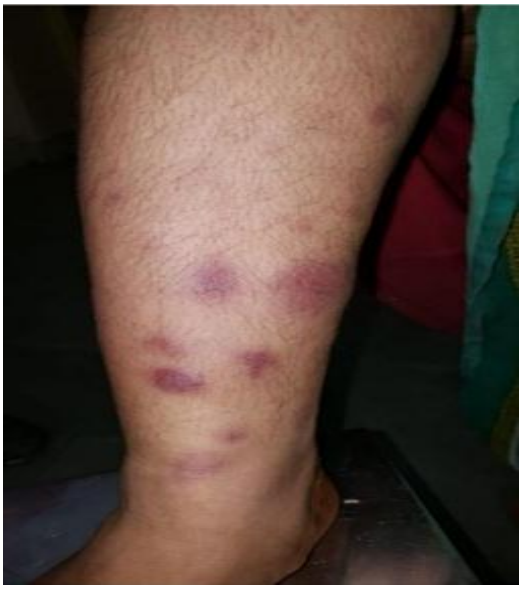

Fig. 2: Showing erythematous macules and plaques over leg.

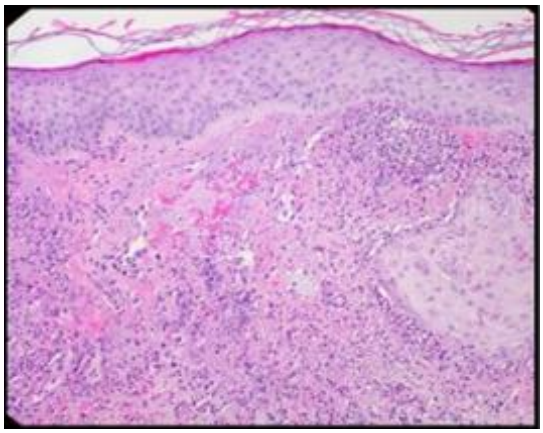

Fig. 3: Histopathological slide revealing massive neutrophilic infiltration.

Discussion

Sweet syndrome is also known as acute febrile neutrophilic dermatosis is an uncommon entity. It was first described by Dr. Robert Douglas Sweet in 1964. The classical features are fever, neutrophilia, cutaneous eruptions consisting of erythematous papules and plaques, and a dermal non-vasculitic neutrophilic infiltration on skin biopsy.[5,6] These plaques are painful but are not associated with pruritus. Other skin presentation such as pustules, vesicles, purpura, ulcers and hemorrhagic lesions have been described[5]. It is basically divided into three categories: Idiopathic or classical Sweet's syndrome, Drug induced and malignancy associated Sweet's syndrome.[8] The diagnostic criteria for Sweet's syndrome established by Su and Liu in 1986, later modified by Von Den Driesch in 1994[4]. All major and two minor criteria should be fulfilled for its definitive diagnosis. Lesions are self-limiting within six to eight weeks.[9] Prednisone should be started at a dose of 40–60 mg per day, with gradual tapering off over four to six weeks, is the conventional treatment for Sweet's syndrome.[6,9] Sudden stoppage or quick tapering off, of steroids leads to relapse. In recurrent disease, In relapses other described drugs are colchicine, potassium iodide, dapsone, doxycycline, indomethacin, clofazimine, isotretinoin and cyclosporine.[9]

Table1: Diagnostic criteria for Sweet syndrome[4]

a. Major criteria	• Sudden onset of typical lesions • Histopathological findings showing neutrophilic dermal infiltrates with the absence of leukocytoclastic vasculitis
b. Minor criteria	• Fever (>38C), recent infections • Arthralgia, conjunctivitis or associated malignancy • Leucocytosis with neutrophilia >70% on peripheral smear, elevated ESR and CRP. • 4. Excellent response to systemic corticosteroids or Potassium iodide

Conclusion

Sweet syndrome being one of the most common neutrophilic dermatoses has been easily treated with conventional therapy like Dapsone, oral steroids and other immunosuppresants. Our case did not respond to these treatment options and had persistent relapses, hence we had to employ DP therapy. This is the first case reported to our best knowledge of sweets syndrome successfully treated with DP therapy in remission with 3 years of follow up.

Conflict of Interest: None.

Reference

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