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Sweet Syndrome - Report of a Case and Review of Literature'

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1. Introduction

Sweet syndrome is a rare inflammatory skin disorder, also known as acute febrile neutrophilic dermatosis or Gomm–Button disease which commonly presents with tender, erythematous plaques, papules and nodules mostly distributed over the arms, upper body, face and neck [(1).] It is mainly associated with fever and peripheral leucocytosis. It was first described by Robert Douglas Sweet in 1964 [(2).] Females are affected more than males. It is characterised by neutrophilic infiltration of papillary dermis, where an underlying chance of unknown malignancy can be present [(1).] Here we report a case of sweet syndrome who presented to our OPD

2. Case Report

35 - year - old female came with complaints of sudden onset of itchy erythematous plaques over face, chest, forearm, dorsal aspect of feet for past 10 days. She gives history of fever 2 days prior associated with diarrhoea and vomiting. She was treated outside with siddha medication for dermatophytosis, which she stopped 1 day prior coming to our OPD. No history of loss of sensation or any pre - existing lesions. She gives history of taking fertility treatment for 12 years. She has underwent hysterectomy for multiple fibroid uterus 1 year before.

On examination well - defined erythematous plaques with tenderness distributed symmetrically over the face, chest, back, forearm, dorsal aspect of feet.

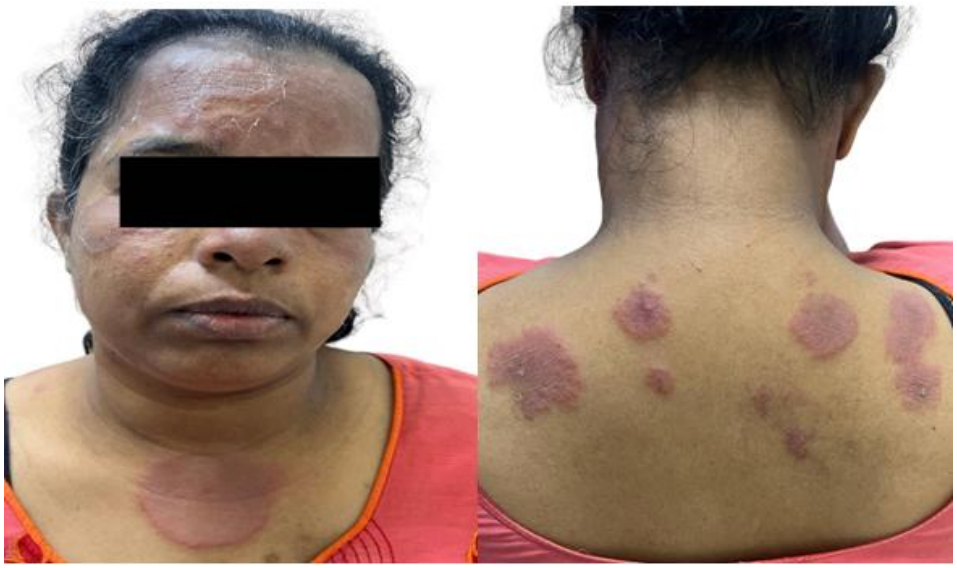

Figure 1 & Figure 2: well - defined erythematous plaques distributed symmetrically over the face, chest (figure 1) and back (figure 2)

To view in terms of the diagnostic criteria of sweet syndrome, lab investigations were done which showed decreased haemoglobin, elevated leukocyte count: 10200cells/microL and ESR of 126mm. For treatment basis her LFT was done which was found to be normal. USG abdomen and pelvis was done which showed a 9x10cm large pelvic mass. To rule out any associated malignancy MRI - pelvis was done which showed bulky uterus with multiple (>10) uterine fibroids, a large 4.6x 5cm large anterior

subserosal fibroid having a mass effect on dome of urinary bladder.

To confirm the diagnosis skin biopsy was done.

Histopathological report revealed epidermis showing mild acanthosis, hyperkeratosis and spongiosis. Dermis shows dense neutrophilic infiltrate with perivascular and peri appendageal mixed inflammatory cell infiltrate.

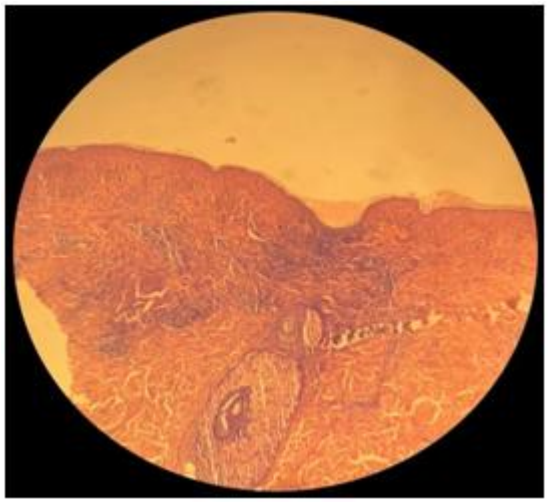

Figure 3: 10X view
Epidermis showing mild acanthosis, hyperkeratosis and spongiosis. Dermis shows dense neutrophilic infiltrate with perivascular and peri appendageal mixed inflammatory cell infiltrate

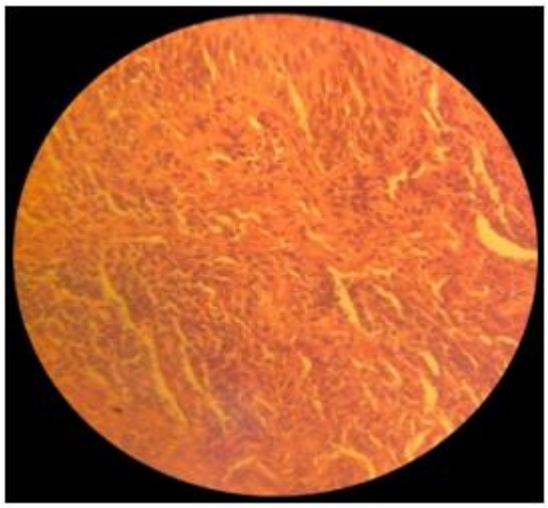

Figure 4: 40X view:
Dermis showing dense neutrophilic infiltrate with perivascular and peri appendageal mixed inflammatory cell infiltrate

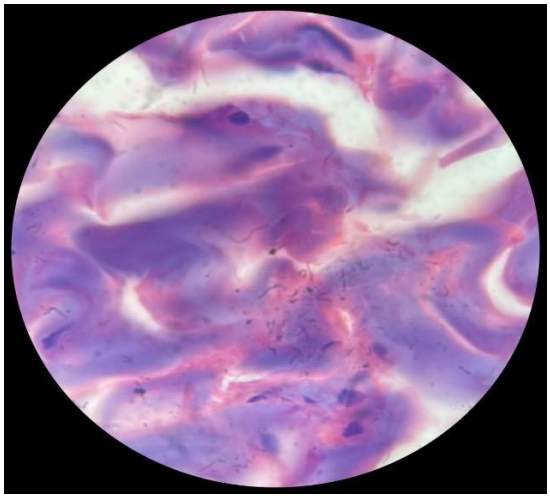

Figure 5: Oil immersion
showing multilobulated nucleus with cytoplasm of neutrophils present in dermis.

Correlating clinical, laboratory and histological findings we came to a diagnosis of sweet syndrome. On treatment basis patient was started on Oral Dapsone, Oral Prednisolone, potent topical steroids and antihistamines. Patient started to improve symptomatically and lesion tends to resolve gradually over a period of 4 weeks.

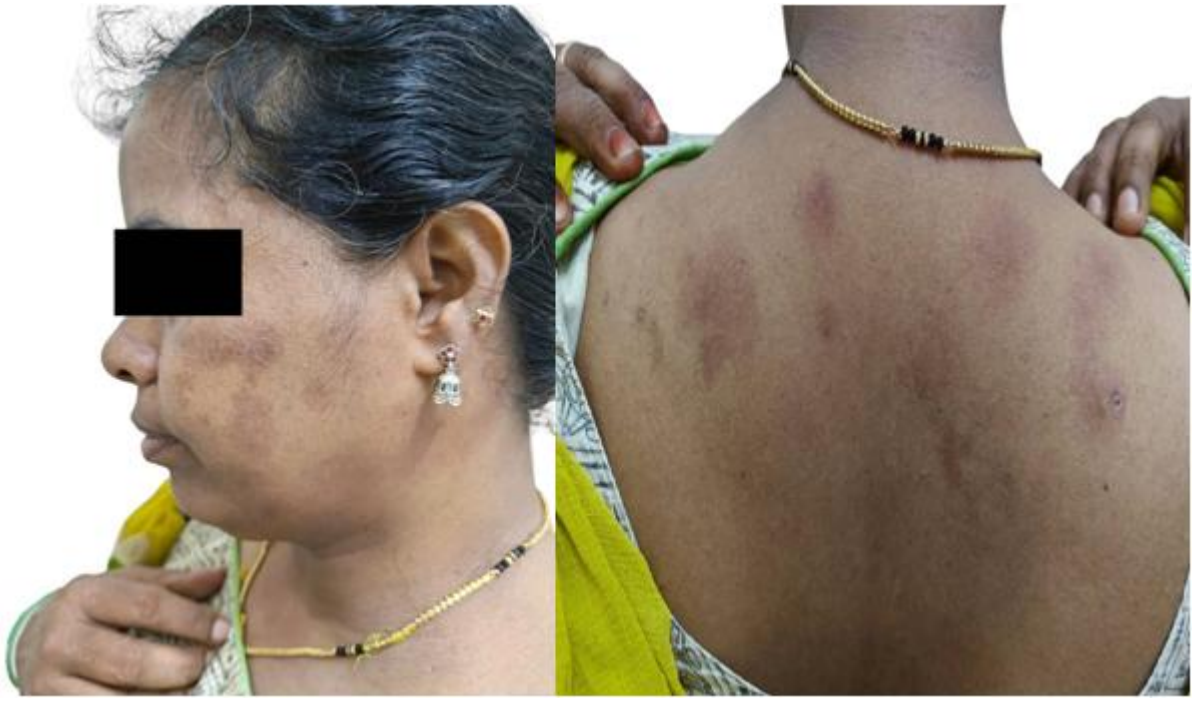

Figure 6 & 7: Erythematous plaques over body resolving after 4 weeks of treatment

3. Discussion

Sweet syndrome also known as acute febrile neutrophilic dermatosis, which presents as multiple, tender, erythematous nodules and plaques, most frequently located on upper extremities, back, neck and face [(1).] Pseudo vesicles may also present.

Sweet syndrome is classified into 3 types.

1) Classical Sweet syndrome
2) Malignancy associated Sweet syndrome
3) Drug induced Sweet syndrome

Classical Sweet Syndrome

Most commonly affects women of ages between 30 and 60yrs [(3).] It is preceded by fever, upper respiratory or gastrointestinal tract infections. A diagnostic criteria is present for the diagnosis of sweet syndrome which is shown below in table 1.

Table 1 shows diagnostic crietria of sweet syndrome.

Table 1

Major criteria	**Minor criteria**
1) Sudden onset of tender erythematous plaques and nodules 2) Histopathological findings shows dense neutrophilic infiltrate without any evidence of leukocytoclastic vasculitis	1) Fever>38C 2) Associated malignancy, inflammatory disorders with prior respiratory and gastrointestinal infections. 3) Shows good response to systemic corticosteroids 4) Abnormal lab values showing • ESR>20mm • Leukocytes>8000 • Neutrophils>70% • - Increased C - Reactive protein

The presence of both major and 2 of the 4 minor criteria is required for the diagnosis of sweet syndrome ([4).]

Malignancy Associated Sweet Syndrome:

It is mostly associated with acute myelogenous leukemia[(5).] Other hematological malignancy includes Myelodysplasia, Multiple myeloma, Monoclonal gammopathy, lymphomas and carcinomas of gastro intestinal tract, breast and genitourinary organs. Incidence is equal among males and females.

Drug - Induced Sweet Syndrome:

It is a rare subtype. The most common drug resposible for it is granulocyte - colony stimulating factor [(6, 7)] and cytokines [(8).] Patient will give history of fever or neoplastic disorder, which will be difficult in differentiating DISS from other SS. Some of the culprit drugs causing DISS are listed below in table 2.

Table 2 shows some drugs responsible for drug - induced sweet syndrome.

Table 2

• Antibiotics	Trimethoprim - sulfamethoxazole Norfloxacin Ofloxacin Quinupristin/dalfopristin
• Colony stimulating factors [6, 7]	Granulocyte - colony stimulating factor Granulocyte - macrophage - colony stimulating factor Pegfilgrastim
• Antiepileptics	Carbemazepine Diazepam
• Antihypertensives	Hydralazine
• Antipsychotics	Clozapine
• Antineoplastics	Bortezomib Imatinib mesylate
• Antithyroid hormone drugs	Propylthiouracil
• Retinoids	All - trans retinoic acid
• Contraceptives	ethinyl estradiol Levonorgestrel
• Diuretics • Nonsteroidal anti - inflammatory drugs	Furosemide Diclofenac

Extracutaneous manifestations of sweet syndrome includes,

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1) Joint involvement: non erosive inflammatory arthritis ([9, 10]) and arthralgias.
2) Ocular involvement: Conjunctivitis, Scleritis, Episcleritis, Uveitis, Keratitis and choroiditis
3) Others: Aseptic meningitis, sterile osteomyelitis, myocarditis, pleural effusion.

The Sweet syndrome must be ruled out from other conditions like

- cellulitis,
- Hypersensitivity drug reactions
- Pyoderma gangrenosum
- Erythema nodosum
- Erythema multiforme
- Behcet's Disease
- Leukemia cutis
- Leukocytoclastic vasculitis

Sweet Syndrome is highly responsive to systemic corticosteroids like prednisolone with a daily dose of 40 mg to 60 mg [9, 10]. In case of recurrence following tapering of steroids, steroid sparing agents like colchicine, Dapsone ([11]), Methotrexate, chlorambucil, Isotretinoin, cyclosporineare used with good outcome. Most cases resolves and some persists, with variable prognosis depending on the underlying cause. Generally, with appropriate treatment and diagnosis, lesions resolve without any scarring.

4. Conclusion

Sweet syndrome aka Acute neutrophilic dermatosis can present typically and atypically and diagnosis is quite challenging. Biopsy is always done to confirm the diagnosis and it is important to rule out any underlying haematological malignancy. Shows good response to Systemic corticosteroids and Dapsone.

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