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CLASSICAL SWEET SYNDROME: A CLINICAL AND PATHOLOGICAL CORRELATION

Dermatology

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ABSTRACT

Sweet syndrome is a rare neutrophilic dermatosis presenting with fever, leukocytosis, and tender erythematous plaques. We report a case of a 34-year-old woman with recurrent febrile rash on the face and upper limbs. Histopathology confirmed classical Sweet syndrome with dense neutrophilic infiltration and no vasculitis. The patient showed marked improvement with systemic corticosteroids and indomethacin. This case underscores the importance of early recognition and improving the quality of life.

KEYWORDS

Classical sweet syndrome, Corticosteroids, Indomethacin

INTRODUCTION:

Sweet syndrome is a rare inflammatory skin condition disease characterized by a constellation of clinical symptoms and physical findings, which include fever, blood, and tissue neutrophilia, leading to the development of tender, erythematous inflammatory skin lesions, histopathologically characterized by neutrophilic infiltrate without evidence of leukocytoclastic vasculitis with an average age of onset is 30–60 years, having a female predominance. A skin biopsy and the discovery of diffuse neutrophilic dermal aggregations in the absence of vasculitis play a crucial role in the diagnosis, which is established worldwide using a diagnostic approach utilizing major and minor criteria.

CASE REPORT:

A women aged 34 years old presented to Skin & V.D OPD due to a 2-year history of abrupt onset of painful erythematous maculopapular rash over her face, neck, bilateral upper limbs which were non-pruritic, later on coalesced to form plaques with an uneven surface. The rash was associated with abrupt onset on fever which was persistent with rash and generalised malaise. She reported no history of recent upper respiratory infection, gastrointestinal infection, or vaccination with no history of past haematologic or visceral malignancy, smoking, or alcohol abuse with a negative urine pregnancy test.

On physical examination, multiple tender erythematous nodules over face(*Figure 1a*), bilateral upper extremities *(Figure 1b,c,d)* of varying diameter were seen. Some of the lesions were found to be crusted. Eyes & oral mucosa were intact. Her vital signs revealed a temperature of 101.7° F and a heart rate of 98 beats per minute.

Laboratory investigations showed white cell count of 17,090 /uL with 88% neutrophils, haemoglobin of 7.90 g/dL, erythrocyte sedimentation rate of 60 mm/hour, creatinine of 0.73 mg/dL and normal liver function tests. Blood cultures remained negative for any growth, viral markers were non-reactive, slit skin smear for acid fast bacilli was negative, C3 & C4, rheumatoid factor, circulating antineutrophil cytoplasmic antibody, perinuclear antineutrophil cytoplasmic antibody and antinuclear antibody tests were negative.

A computed tomography scan and ultrasound of the abdomen did not reveal any obvious pathology. A skin biopsy of a nodule on her right upper limb revealed a hyperkeratotic and neutrophilic infiltration in epidermis and neutrophilic microabscess in the dermis associated with neutrophilic panniculitis with no granulomas in the sections.*(Figure 2)*. There was no evidence of vasculitis present. On the basis of clinical features and histopathological features, patient was diagnosed as classical sweet syndrome.

Patient was started on prednisolone 40mg and indomethacin 150 mg daily for 2 weeks and resolution in lesions which were present on face(*Figure 3a*) and bilateral upper extremities(*Figure 3b*) were gradually seen.

DISCUSSION:

Dr Robert Douglas Sweet first described and used the term acute febrile neutrophilic dermatosis, back in 1964 based on his observations in 8 women[1]. Therefore, the name "Sweet's syndrome" is established as the eponym for acute febrile neutrophilic dermatosis and used worldwide. It is an uncommon disorder characterized by abrupt onset of fever, which can also be accompanied by other symptoms like arthralgia, headaches, elevated white blood cell count, and a painful red papular skin rash, with a neutrophilic infiltrate of the upper dermis on histology without signs of vasculitis. It is generally classified into 3 categories based on the underlying etiology and clinical scenario, including classical (idiopathic) Sweet's syndrome, malignancy-associated Sweet's syndrome (diagnostic criteria is outlined in *Table 1*), and drug-induced Sweet's syndrome(the diagnostic criteria is outlined in *Table 2*).

Table 1: Diagnosis Requires Two Major Criteria And Two Of Four Minor Criteria.

Classical & Malignancy associated Sweet Syndrome
Major Criteria: 1. Abrupt onset of painful, erythematous plaques/nodules. 2. Dense neutrophilic infiltrate without leukocytoclastic vasculitis on histopathology. Minor Criteria: 3. Pyrexia (>38˚C). 4. Preceded by upper respiratory or gastrointestinal infection, vaccination, associated with pregnancy or inflammatory disease. 5. Responds to treatment with corticosteroids or potassium iodide. 6. Abnormal labs (3 of 4): ESR > 20 mm/hr, positive C-reactive protein, WBC > 8,000, neutrophils > 70 %.

Table 2: Diagnostic Criteria For Drug-induced Sweet Syndrome (Diagnosis requires all 1-5 criteria)

1. Abrupt onset of painful erythematous plaques or nodules. 2. Histopathological evidence of dense neutrophilic infiltration without evidence of vasculitis. 3. Pyrexia, >38°C. 4. Temporal relationship between drug ingestion and clinical presentation, or temporally related recurrence after oral challenge. 5. Temporally related resolution of lesions after drug withdrawal or treatment with systemic corticosteroids.

The most prevalent kind of Sweet's syndrome, as described in this case study, is the classical type, which is typically linked to an infectious process involving the upper respiratory or gastrointestinal tract (GIT) like Crohn's disease, ulcerative colitis and other inflammatory conditions as underlying etiologies like Systemic lupus erythematosus, Dermatomyositis. Clinical features in Malignancy-associated Sweet's syndrome may appear with, prior to, or after to the underlying cancer where skin lesions may become ulcerated or bullous and mimic pyoderma gangrenosum. The most common malignancies associated with Sweet's syndrome are haematological malignancies,

most commonly being acute myelogenous leukaemia. Solid tumours with carcinomas of the genitourinary tract, breast, and GIT have been reported as well.

Drug-induced Sweet syndrome has also been documented which are all-trans retinoic acid, granulocyte colony-stimulating factor, sulfa drugs, tetracycline, minocycline, nonsteroidal anti-inflammatory drugs, carbamazepine, oral contraceptives, hydralazine, clindamycin, fluoroquinolones[3]. Recently a case of niraparib PARP-Inhibitor induced Sweet Syndrome in gynaecologic cancer has been reported by Nora Badiner[3].

The pathogenesis of Sweet's disorder is obscure as of date and it is accepted to be multifactorial. Extreme hypersensitivity is accepted to be the fundamental prompting component driving the pathogenesis. Recent studies have driven to the understanding that adjacent to the innate immunity , the adaptive immunity too plays a critical part, proven by the raised levels of IL-1α, IL-1β, IL-2, and interferon-γ and TNF-α which are Type 1 T helper cells (Th1)-related cytokines which leads to enactment and recruitment of neutrophils.

Treatment of sweet syndrome comprises of local and systemic therapy. Local Therapy is used for localised lesions where topical or intralesional corticosteroids can be used either alone or along with other therapy. Systemic therapy comprises of corticosteroids which are the mainstay of the treatment ; started with 1 mg/ kg/day of oral prednisolone and then tapered to 10 mg/day in 3 to 5 days when a desired clinical response is observed. Other commonly used 1st line consists of potassium iodide(KI) which is administered as enteric-coated tablets (900 mg/ day) or as a saturated solution (1 g KI/ml of water) and colchicine 0.5 mg orally 3 times a day for 10–21 days. 2nd line drugs are dapsone (100–200 mg/day), indomethacin (150 mg/day for 7 days, then 100 mg/day for 14 days), clofazimine (200 mg/day for 4 weeks, then 100 mg/day for 4 weeks), cyclosporine (2–4 mg/kg/day), antibiotics like doxycycline and metronidazole in cases of secondary infections, antimetabolite agents like cyclophosphamide and anti-TNF agents such as etanercept and infliximab[4,5].

CONCLUSION:

Despite the fact that sweet syndrome has been linked to autoimmune diseases and cancers in the past, no such link was found in our case study. Nevertheless, the patient should still be monitored in case of future recurrence, inflammatory illness, or cancer. Research potential lies in defining more associations and prognostic value in other autoimmune and myeloproliferative diseases with further advancements in treatment modalities of Sweet syndrome.

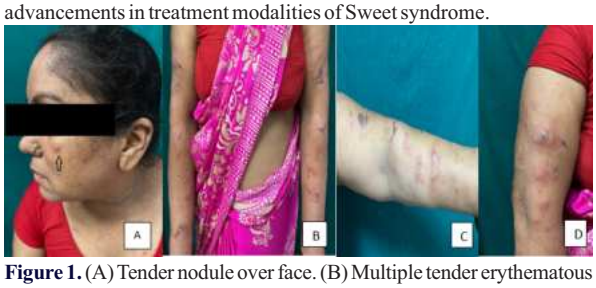

Figure 1. (A) Tender nodule over face. (B) Multiple tender erythematous nodules over bilateral upper extremities and some nodules are coalescing into plaques. (C) Multiple nodular eruptions over left arm and forearm. (D) Multiple nodular eruptions over right arm.

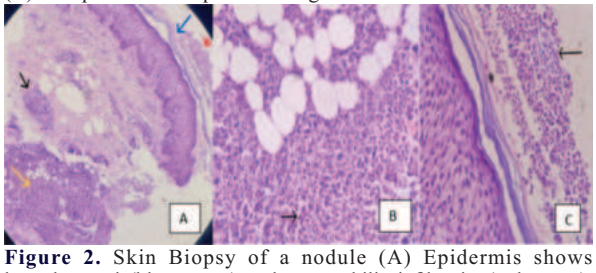

Figure 2. Skin Biopsy of a nodule (A) Epidermis shows hyperkeratosis(blue arrow) and neutrophilic infiltration(red arrow), Dermal neutrophilic micro abscess(black arrow) and neutrophilic panniculitis (yellow arrow) (low-power magnification, hematoxylin and eosin stain). (B) Neutrophilic panniculitis(black arrow) (high-power magnification, hematoxylin and eosin stain). (C) Neutrophilic exudate in epidermis (black arrow) (high-power magnification, hematoxylin and eosin stain)

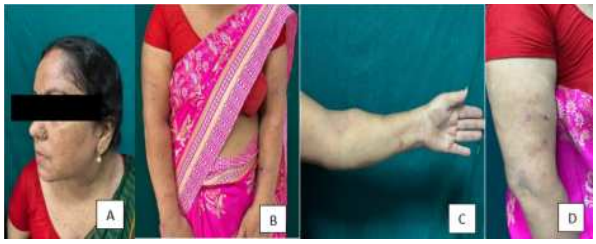

Figure 3. (A,B) Resolution of lesion over face and bilateral upper extremities respectively. (C,D) Resolution of lesion over Left arm , Forearm and Right arm respectively

REFERENCES:

1. Sweet RD. An acute febrile neutrophilic dermatosis. Br J Dermatol. 1964;76:349-356. doi:10.1111/j.1365-2133.1964. tb14541.x
2. Chaudhry A R, Iftikhar I, Choudhry S A, et al. (March 31, 2023) Fluoroquinolone-Induced Sweet Syndrome: A Case Report. Cureus 15(3): e36952. doi:10.7759/cureus.36952
3. Badiner N, Carter C, Ioffe Y, Hong L. A case of niraparib PARP-Inhibitor induced Sweet Syndrome in gynecologic cancer. Gynecol Oncol Rep. 2023 Mar 15;46:101162. doi: 10.1016/j.gore.2023.101162. PMID: 36992982; PMCID: PMC10040500.
4. Watson IT, Haugh I, Gardner AR, Menter MA. Histiocytoid Sweet syndrome successfully treated with etanercept. Proc (Bayl Univ Med Cent). 2018 May 9;31(3):347-349. doi: 10.1080/08998280.2018.1460132. PMID: 29904308; PMCID: PMC5997035.
5. Sotiropoulos C, Sakka E, Theocharis GJ, Thomopoulos KC. Sweet Syndrome Associated With Crohn's Disease Developed After a Combo Therapy With Infliximab and Azathioprine: A Case Report. Cureus. 2021 Dec 27;13(12):e20749. doi: 10.7759/cureus.20749. PMID: 35111438; PMCID: PMC8791332.