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Original Research Paper

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BULLOUS VARIANT OF SWEET'S SYNDROME- A RARE CASE REPORT

Dr. B Sruthi*	Junior Resident, Department Of Dermatology Venereology And Leprosy, Chettinad Hospital And Research Institute. *Corresponding Author
Dr. Abinaya Kuberan	Junior Resident, Department Of Dermatology Venereology And Leprosy, Chettinad Hospital And Research Institute.
Dr. Soundarya S	Associate Professor, Department Of Dermatology Venereology And Leprosy, Chettinad Hospital And Research Institute.
Dr. Jayakar Thomas	Emeritus Professor, Department Of Dermatology Venereology And Leprosy, Chettinad Hospital And Research Institute.

ABSTRACT Sweet's syndrome, or acute febrile neutrophilic dermatosis, is a rare inflammatory skin condition typically characterized by abrupt onset of tender, erythematous papules and plaques. The bullous variant is an uncommon manifestation that can closely mimic other dermatologic and infectious disorders, making diagnosis challenging. We report the case of a 66-year-old male with a two-week history of painful, erythematous bullous lesions predominantly involving the dorsolateral hands, forearms, and peri-elbow regions. The eruption was preceded by a recent upper respiratory tract infection and occurred in the absence of systemic symptoms such as fever or arthralgia. His medical history included poorly controlled type 2 diabetes mellitus and a past episode of vasculitis managed with corticosteroids and mycophenolate mofetil. Laboratory workup revealed leukocytosis with neutrophilia and elevated inflammatory markers. Histopathology from a lesional biopsy demonstrated a dense dermal neutrophilic infiltrate without vasculitis, consistent with Sweet's syndrome. Direct immunofluorescence was negative, and infectious and autoimmune markers were unremarkable. The patient responded dramatically to oral corticosteroids, though temporary discontinuation led to recurrence, which was effectively managed by reinitiating therapy with a tapering regimen. This case underscores the diagnostic complexity of bullous Sweet's syndrome, especially in the absence of classic systemic features. Early recognition and histological confirmation are crucial to prevent misdiagnosis and ensure prompt therapeutic intervention.

KEYWORDS : Neutrophilic dermatosis, Idiopathic, Bullous skin lesions, Dense Neutrophilic infiltrate, Erythema elevatum diutinum

INTRODUCTION:

Acute febrile neutrophilic dermatosis, commonly referred to as Sweet's syndrome, was initially identified by Dr. Robert Douglas Sweet in 1964.[1] It is characterized by the sudden onset of tender, erythematous skin lesions such as papules, nodules, or plaques, often accompanied by systemic symptoms like fever.[2] A bullous variant of Sweet's syndrome represents an uncommon presentation in which blister formation coexists with typical cutaneous findings.[3] It may present as classical, malignancy-associated or drug-induced forms.[4] Affected individuals may exhibit constitutional symptoms including fatigue, joint pain, muscle aches, and headache prior to or during the eruption of skin lesions.[2] While the face, neck, and upper limbs are most frequently involved, lesions can appear on any part of the skin surface.[5] Laboratory evaluation typically reveals elevated white blood cell counts, and histopathology is essential for diagnosis, showing a dense infiltrate of mature neutrophils in the dermis without evidence of vasculitis.[6] Given its clinical and microscopic resemblance to several other conditions including pyoderma gangrenosum, erythema elevatum diutinum, rheumatoid neutrophilic dermatosis, and bacterial cellulitis, accurate identification can be challenging. This report describes a rare case of the bullous subtype of Sweet's syndrome, emphasizing its diagnostic complexity and clinical significance.

Case Presentation

A 66-year-old male presented to our dermatology outpatient clinic with complaints of multiple painful, erythematous bullae over the dorsolateral aspects of both hands, forearms, and the areas near the elbows, with a duration of two weeks. The lesions were associated with pruritus and a sudden onset of pain. He reported an episode of upper respiratory tract infection approximately two weeks prior to the onset of skin lesions. He denied any history of fever, chills, muscle or joint pain. The patient had a known history of poorly controlled type 2 diabetes mellitus. He also had a previous episode of vasculitis one year ago, for which he was treated initially with oral corticosteroids, followed by a four-month course of mycophenolate mofetil at a dose of 500 mg twice daily. On cutaneous examination, multiple erythematous, tender bullae were observed symmetrically over the dorsolateral surfaces of both hands, forearms, and near the elbows. Some lesions exhibited mild induration and tenderness. The palms and soles were spared. There was no involvement of mucosal surfaces, and no recent drug intake was reported. The rest of the systemic examination was unremarkable.

Laboratory investigations revealed an elevated total leukocyte count with neutrophilic predominance. Inflammatory markers including erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were raised, while lymphocyte counts were within normal limits. A skin biopsy was performed from a lesion on the left forearm. Histopathological examination revealed epidermal hyperkeratosis, acanthosis, and spongiosis.

The dermis showed dense infiltration of neutrophils in both the papillary and reticular layers. There was also perivascular and peri adnexal neutrophilic infiltration, with focal micro abscess formation. Direct immunofluorescence studies were negative. Serological tests, including anti-streptolysin O titres, viral markers, and immunoglobulin levels, were within normal limits.

The clinicopathological findings were consistent with a diagnosis of bullous Sweet's syndrome. The patient was initiated on oral prednisolone at 30 mg/day, which resulted in marked improvement of the lesions. A relapse occurred when he discontinued therapy temporarily, but reintroduction of oral prednisolone at the same dosage followed by a gradual tapering of 5 mg/day led to clinical resolution.

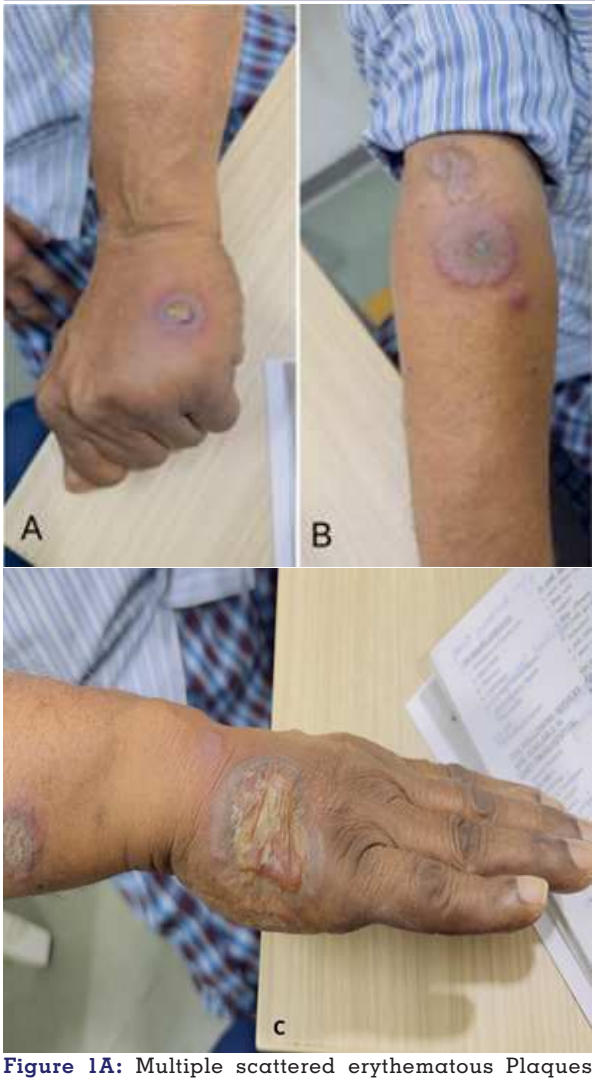

Figure 1A: Multiple scattered erythematous Plaques surmounted with bullae over Left dorsomedial aspect of hand; **Figure 1B:** Ventral aspect of the forearm; **Figure 1C:** Right dorsomedial aspect of hand

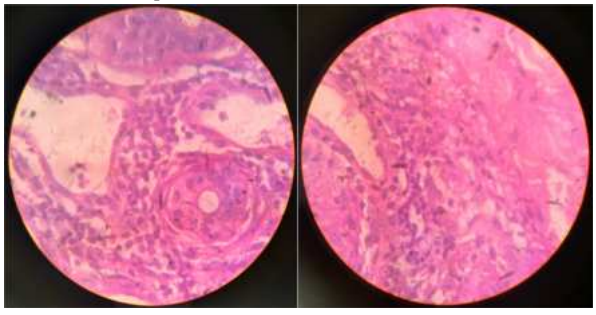

Figure 2A ; Figure 2B
2A: Under high power ,The dermis showed dense infiltration of neutrophils in both the papillary and reticular layers. **2B**: There was also perivascular and peri adnexal neutrophilic infiltration, with focal micro abscess formation.

DISCUSSION

This case illustrates the diagnostic challenge posed by bullous Sweet's syndrome, a rare variant of neutrophilic dermatosis. The patient presented with bilaterally symmetrical, painful bullous lesions over the upper limbs, following an upper respiratory infection, but without classical systemic features such as fever or arthralgia. Laboratory findings pointed to an inflammatory process, and histopathology confirmed dense dermal neutrophilic infiltrates in the absence of vasculitis or infectious agents. Direct immunofluorescence was negative, further supporting the diagnosis. A rapid clinical response to oral corticosteroids and relapse on discontinuation reinforced the diagnosis and highlighted the importance of prompt treatment and adherence to therapy.

Heath et al. [6] emphasized that Sweet's syndrome is a heterogeneous neutrophilic dermatosis with diverse triggers and unclear pathogenesis, though emerging evidence implicates inflammasome activation and genetic factors. Joshi et al. [7] detailed the expanding spectrum of Sweet's syndrome presentations and emphasized the role of corticosteroids as first-line therapy, while also highlighting steroid-sparing alternatives for refractory cases. Bhat et al. [3] described a bullous variant of Sweet's syndrome precipitated by radiocontrast, underscoring the aggressive nature of this form and its excellent response to corticosteroids. These studies collectively frame bullous Sweet syndrome as a diagnostically challenging and clinically variable entity with potential for rapid resolution upon timely immunosuppressive intervention.

Bullous Sweet's syndrome is a rare and diagnostically challenging subtype, with clinical features often overlapping with infectious, autoimmune, or drug-induced dermatoses. Comparing the present case to existing literature provides insights into variability in presentation, associated conditions, and therapeutic responses.

Sherban et al. [8] described multiple elderly patients with extensive hemorrhagic bullae involving the face, arms, and trunk, often mimicking vasculitis. In contrast to their findings of p-ANCA positivity and systemic involvement, our patient tested negative on immunofluorescence studies and exhibited no evidence of systemic vasculitis, highlighting the spectrum of serological and clinical variability in bullous Sweet's syndrome. Verma et al. [9] reported a younger patient with seasonal recurrent bullous eruptions over the forearms, bilaterally symmetrical and showed rapid response to corticosteroids which is similar to our case. However, unlike our patient, the recurrent nature of Verma's case necessitated adjunctive NSAIDs, underlining variability in disease chronicity and therapeutic needs.

Voelter-Mahlknecht et al. [10] noted rapid onset bullous lesions with systemic symptoms including fever and lymphadenopathy. Their patient responded well to moderate-dose steroids, mirroring the steroid responsiveness seen in our case. Yang et al. [11] uniquely reported hemophagocytosis on biopsy, a feature absent in our histopathology, suggesting a more aggressive immune activation in select cases.

Endo et al. [12] documented a unilateral bullous Sweet syndrome following herpes zoster infection, suggesting localized immune dysregulation. Our patient also had an antecedent infection (upper respiratory), supporting the proposed link between infections and Sweet's syndrome flare-ups, though his lesions were bilateral and widespread. Burke et al. [13] presented Sweet's syndrome in the setting of acute leukemia, with histiocytoid and subcutaneous variants-forms not observed in our patient, whose presentation remained confined to the dermis without malignancy.

Abdelnabi et al. [14] and Bhat et al. [3] described radiocontrast-induced bullous Sweet's syndrome, both showing widespread hemorrhagic bullae and systemic complications including acute kidney injury. Unlike these iatrogenic cases, our patient had no recent drug exposure, supporting an idiopathic or post-infectious etiology. The favourable response to systemic corticosteroids across these cases, including ours, affirms corticosteroids as the mainstay of therapy.

Joseph et al. [15] highlighted a localized neutrophilic dermatosis affecting the dorsal hand, initially mimicking an infectious etiology—similar to the diagnostic confusion in our case due to bullous morphology. Their patient responded to dapsone, whereas our case achieved full remission with corticosteroids alone. Finally, Almedfa et al. [16] reported Sweet's syndrome in a terminal cancer patient with multiple skin ulcers and significant systemic decline. Although histology was similar (dense neutrophilic infiltrate), their case underscores the potential for Sweet's syndrome to complicate advanced malignancies, unlike the idiopathic presentation seen in our patient.

Overall, the diversity in trigger factors like infection, malignancy, drugs, localized or widespread lesion distribution and associated systemic findings across these reports underscores the diagnostic flexibility required to recognize bullous Sweet's syndrome. Our patient's favourable response to oral corticosteroids, relapse on discontinuation, and successful re-induction with tapering align with established management protocols.

The literature suggests that early recognition and adherence to steroid therapy are critical for preventing relapses. While steroid-sparing agents such as dapsone and mycophenolate mofetil have roles in recurrent or steroid-resistant cases, they were not required in our patient's final management. This case report contributes to the limited literature on idiopathic bullous Sweet's syndrome and supports histopathological confirmation and prompt corticosteroid therapy as key pillars of management.

CONCLUSION

This case underscores the need for clinical suspicion, histopathological confirmation, and prompt corticosteroid therapy. Early recognition and adherence to treatment are key to preventing relapse and ensuring favourable outcomes.

Suggested Keywords:
Sweet Syndrome, Bullous Diseases, Neutrophilic Dermatoses, Steroid-responsive dermatosis, Inflammatory skin eruption, Relapsing dermatosis

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