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Concomitant oral and genital mucosal involvement in classical Sweet syndrome

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Dear Editor,

Sweet syndrome (SS) is a rare inflammatory dermatosis characterized by the abrupt onset of painful erythematous plaques, papules and nodules associated with a neutrophilic dermal infiltrate, fever, and peripheral neutrophilia. It is classified into three major subtypes: the classical form, frequently associated with infections, pregnancy, inflammatory bowel disease and other autoimmune disorders; the malignancy-associated form, linked to both solid-organ and hematologic malignancies, particularly acute myeloid leukemia and myelodysplastic syndromes; and the drug-induced form. SS typically affects the upper extremities, face, and neck; however, extracutaneous involvement has been reported in multiple organs, including ocular, musculoskeletal, neurologic, and visceral manifestations. Mucosal involvement is generally considered uncommon and has traditionally been regarded as a feature of malignancy-associated SS. However, robust subtype-specific data supporting this assumption remain limited.[1-3] Here, we describe a patient with classical SS presenting with simultaneous oral and genital mucosal involvement.

A 39-year-old Caucasian woman presented to the Emergency Department with a recent history of fever, malaise, odynophagia, and a painful pustular rash associated with a burning sensation. Two weeks prior to admission, she had experienced an episode of gingivitis treated with oral clarithromycin and reported a history of recurrent pharyngitis. Her past medical history was otherwise unremarkable. On admission, the patient was febrile (38°C). Physical examination revealed polymorphic, raised erythematous papulonodular lesions and vesicles with central yellowish discoloration, predominantly involving the face, neck, and lower extremities (Figure 1a). Scattered lesions were also present on the abdomen and genital area. The Nikolsky sign was negative. Painful ulcerated lesions covered by fibrin were observed on both sides of the tongue apex, the left tonsil, and the anterior palatoglossal pillar (Figure 1b). An extensive diagnostic work-up was performed, including serological and polymerase chain reaction testing for common viral and sexually transmitted pathogens (including Monkeypox, herpes simplex virus type 1 and 2, *Treponema pallidum*, and HIV), blood cultures, hepatitis panel, and autoimmune screening (antibodies, extractable nuclear antigen antibodies, anti-neutrophil cytoplasmic antibodies, tissue transglutaminase antibodies, Quantiferon-TB Gold).

All results were negative. Laboratory investigations revealed leukocytosis (12,470/µL) with neutrophilic predominance (78.6%) and an elevated C-reactive protein level (82.9 mg/L). Skin biopsies demonstrated a dense neutrophilic infiltrate within the dermis associated with marked subepithelial edema, without evidence of vasculitis or leukocytoclasia (Figure 1d). There was no evidence of an underlying malignancy. No clinical features suggestive of autoimmune or inflammatory bowel diseases were identified. On day 4 of hospitalization, a diagnosis of classical SS was established.

Treatment with intravenous methylprednisolone 60 mg/day led to rapid clinical improvement, with regression of existing lesions and no appearance of new ones. Chest X-ray and abdominal ultrasound revealed no infectious foci, and transthoracic echocardiography excluded endocarditis. The patient was discharged on a tapering course of oral prednisone 50 mg/day. Disease partially relapsed after four weeks, with the onset of pseudofolliculitis and oral aphthous lesions. Intravenous methylprednisolone (60 mg) was administered, followed by oral prednisone 50 mg/day with gradual tapering and colchicine 0.5 mg twice daily. After 2 months of treatment, near-complete resolution of both cutaneous and oral lesions was observed, along with normalization of laboratory parameters (Figure 1c).

Our patient fulfilled the diagnostic criteria proposed by Su and Liu and subsequently modified by von den Driesch, meeting both major criteria – abrupt onset of painful erythematous plaques with compatible histopathological findings – and two minor criteria, namely elevated inflammatory markers with neutrophilia and an excellent response to systemic corticosteroid therapy.[4,5] Given the traditional association between mucosal involvement and malignancy-associated SS, we performed a focused, non-systematic review of the literature to better characterize the distribution of this manifestation across SS subtypes.[2,6] We identified more than 35 cases of SS with documented mucosal involvement, most occurring in classical rather than malignancy-associated or drug-induced disease, suggesting that this manifestation may be underrecognized in the classical form. However, publication bias and the non-systematic nature of our review preclude any inference regarding the relative prevalence of mucosal involvement across SS subtypes. A recent systematic review of 539 SS patients reported oral (n=42, 7.8%) and genital (n=12, 2.2%) involvement but classified these sites as skin rather than mucosal or extracutaneous manifestations, without specifying whether they occurred concomitantly. Notably, other extracutaneous manifestations, such as ophthalmologic involvement, were compared by subtype (11% in malignancy-associated *vs.* 12.5% in non-malignant SS, p=0.645), whereas oral and genital sites were not compared by subtype. Furthermore, no significant differences in clinical skin manifestations were identified between malignancy-associated and non-malignant SS.[3] Together, these findings highlight the lack of subtype-specific data on mucosal involvement in SS, whose true distribution remains uncertain. Our case further adds to the evidence supporting mucosal involvement in classical SS. Regardless of its true frequency across disease subtypes, clinicians should be aware that mucosal involvement does not necessarily indicate malignancy and should not delay appropriate corticosteroid treatment once infectious causes have been excluded. Larger, systematic studies are needed to clarify the true prevalence, clinical relevance, and prognostic implications of mucosal involvement in SS.

References

1. Griffiths CEM, Barker JNWN, Bleiker TO, et al., eds. Rook's textbook of dermatology. 10th ed. Wiley-Blackwell; 2024.
2. Cohen PR. Sweet's syndrome--a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis 2007;2:34.
3. Orfaly VE, Shakshouk H, Heath M, et al. Sweet Syndrome: A Review of Published Cases. Dermatology 2023;239:664-9.
4. von den Driesch P, Steffan C, Zöbe A, Hornstein OP. Sweet's syndrome--therapy with cyclosporin. Clin Exp Dermatol 1994;19:274-7.
5. Su WP, Liu HN. Diagnostic criteria for Sweet's syndrome. Cutis 1986;37:167-74.
6. Raza S, Kirkland RS, Patel AA, et al. Insight into Sweet's syndrome and associated-malignancy: a review of the current literature. Int J Oncol 2013;42:1516-22.

Figure 1. a) Erythematous papulonodular lesions and vesicles involving the face and neck; **b**) ulcerated lesions covered by fibrin on the bilateral apex of the tongue before treatment; **c**) histological appearance of the biopsy specimen, papillary dermal edema with an underlying dense neutrophilic infiltrate (hematoxylin and eosin, x5); **d**) resolution of the tongue ulcers after 2 months of treatment.