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(CASE REPORT)

Entero-Behçet disease associated with sweet syndrome: A case report

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Abstract

Sweet syndrome is an acute neutrophilic dermatosis that may be associated with systemic inflammatory diseases. Behçet disease is a multisystem vasculitis whose gastrointestinal involvement, referred to as entero-Behçet disease, may be severe. The association between these two entities remains exceptionally rare.

We report the case of a 32-year-old woman diagnosed with entero-Behçet disease based on bipolar aphthosis, a positive pathergy test, and characteristic intestinal ulcerations observed on endoscopy. One month later, she developed an extensive cutaneous eruption consisting of infiltrated erythematous plaques, vesiculopustular lesions, and bullae, associated with biological inflammatory syndrome. Histopathological examination of the skin biopsy revealed a dense neutrophilic infiltrate without evidence of vasculitis, consistent with Sweet syndrome. Treatment with systemic corticosteroids combined with colchicine resulted in complete regression within 20 days.

This case highlights a rare association likely related to a shared neutrophilic hyperactivation pathway and underscores the importance of early diagnosis in order to optimize therapeutic management.

Keywords: Entero-Behçet Disease; Sweet Syndrome; Neutrophilic Dermatosis; Vasculitis

1. Introduction

Sweet syndrome is an acute neutrophilic dermatosis characterized by fever, painful cutaneous lesions, and a dense dermal infiltration of neutrophils. It may occur idiopathically or in association with malignancies, infections, or inflammatory diseases.

Behçet disease, particularly its gastrointestinal form known as entero-Behçet disease, is a rare multisystem vasculitis responsible for deep intestinal ulcerations and heterogeneous systemic manifestations.

The concomitant association of Sweet syndrome and entero-Behçet disease has rarely been reported, suggesting a potential shared inflammatory mechanism involving neutrophil activation. This overlap may pose significant diagnostic challenges and carry important therapeutic implications.

We report a case illustrating this unusual association and discuss its clinical and pathophysiological implications.

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2. Case Presentation

A 32-year-old woman presented with a 2-year history of bipolar aphthosis *(Figure 1)*. Over the preceding 3 months, she had developed non-bloody mucoid diarrhea associated with abdominal pain and a positive pathergy test. The diagnosis of entero-Behçet disease was established based on characteristic endoscopic findings and supportive histopathological examination.

Ileocolonoscopy revealed ulcerative pancolitis characterized by large geographic ulcerations separated by areas of normal mucosa, with associated involvement of the ileocecal valve *(Figure 2)*. Histopathological analysis demonstrated ulcerated mucosa with chronic lymphoplasmacytic inflammatory infiltrates and congestive vascular changes, without epithelioid granulomas or architectural distortion suggestive of Crohn's disease. These findings were consistent with entero-Behçet disease.

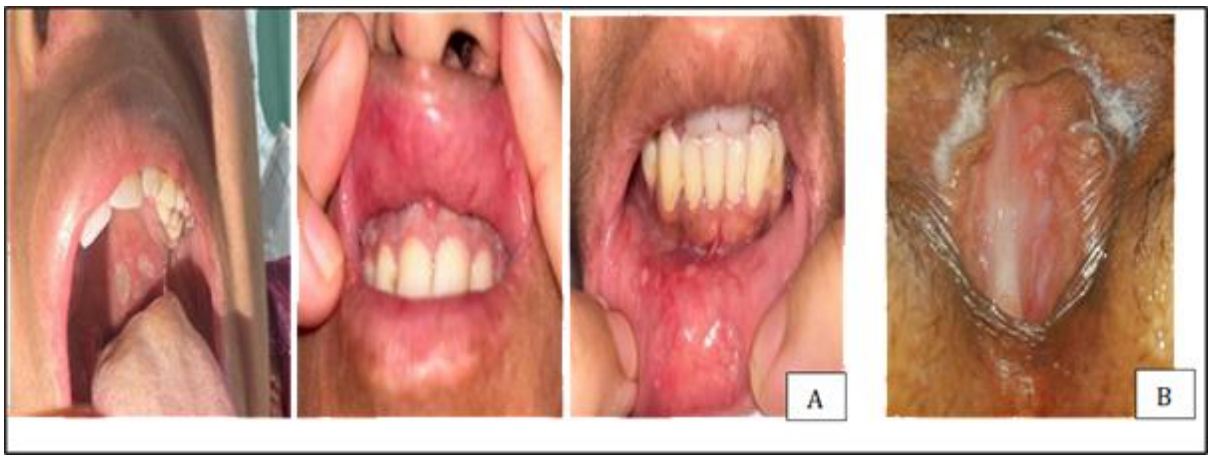


Figure 1 Bipolar aphthosis : A. Oral aphthosis B. Genital aphthosis

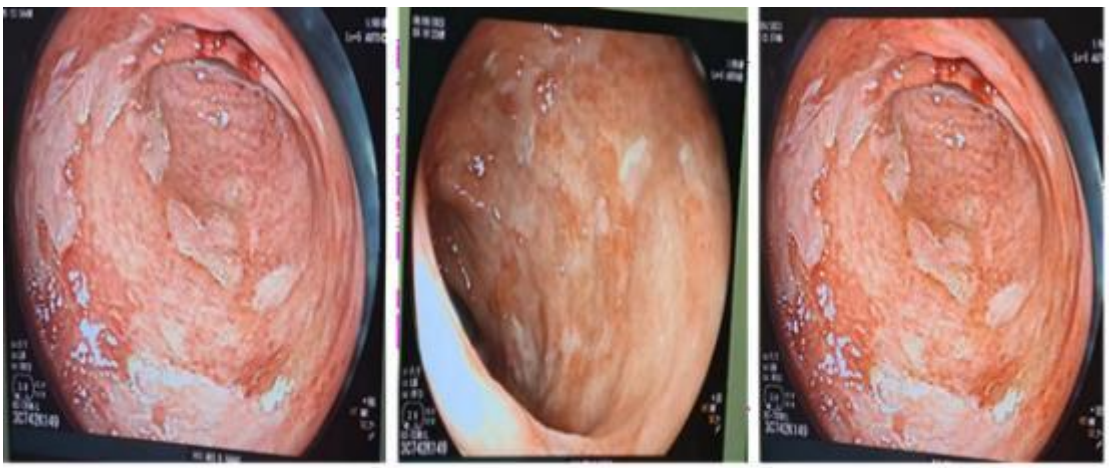


Figure 2 Endoscopic images showing geographic colonic ulcerations

One month later, the patient reported the onset of severe headaches and arthralgia, followed by the development of pruritic papules initially involving the trunk and subsequently the face, with the later appearance of several bullous lesions.

Dermatological examination *(Figure 3)* revealed bilateral eyelid edema associated with infiltrated erythematous plaques scattered with a few pustules on the face. The neck and extremities showed confluent vesiculopustular lesions forming target-like plaques. The trunk exhibited diffuse erythematous-violaceous plaques. Clear-fluid bullae, occasionally hemorrhagic, were observed on the fingers and the palm of the right hand. Cutaneous involvement affected

approximately 50% of the body surface area. The pathergy test was positive. Fluorescein angiography was normal. Examination of the skin appendages revealed several erythematous plaques on the scalp.

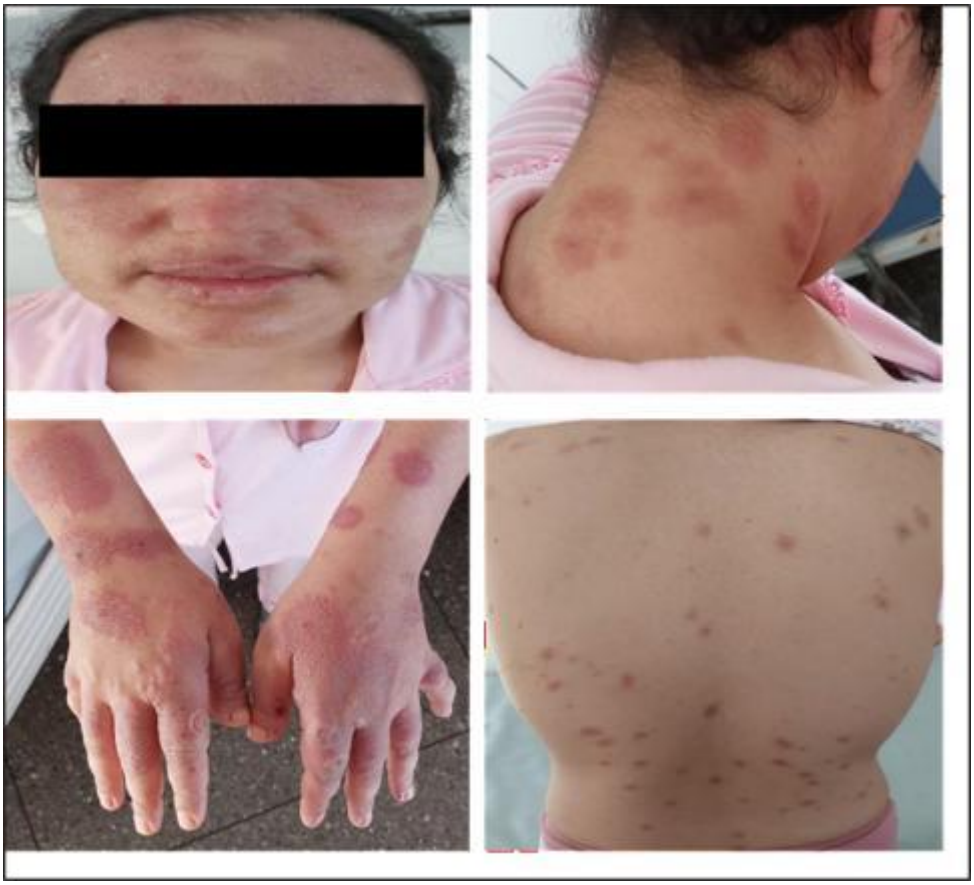


Figure 3 Dermatological manifestations of Sweet syndrome

The remainder of the physical examination, including neurological, rheumatological, abdominal, and pleuropulmonary assessments, was unremarkable.

Laboratory investigations revealed an elevated C-reactive protein level of 75 mg/L and an erythrocyte sedimentation rate of 65 mm/h. The white blood cell count and the remainder of the laboratory workup were within normal limits.

A skin biopsy performed from a forearm lesion showed an epidermis covered by an orthokeratotic keratin layer with focal areas of parakeratosis. The papillary dermis was mildly edematous, whereas the reticular dermis exhibited a dense superficial and deep perivascular and interstitial inflammatory infiltrate composed predominantly of leukocytoclastic neutrophils, with preservation of the vascular walls and no evidence of vasculitis. Overall, the histopathological findings were consistent with Sweet syndrome *(Figure 4)*.

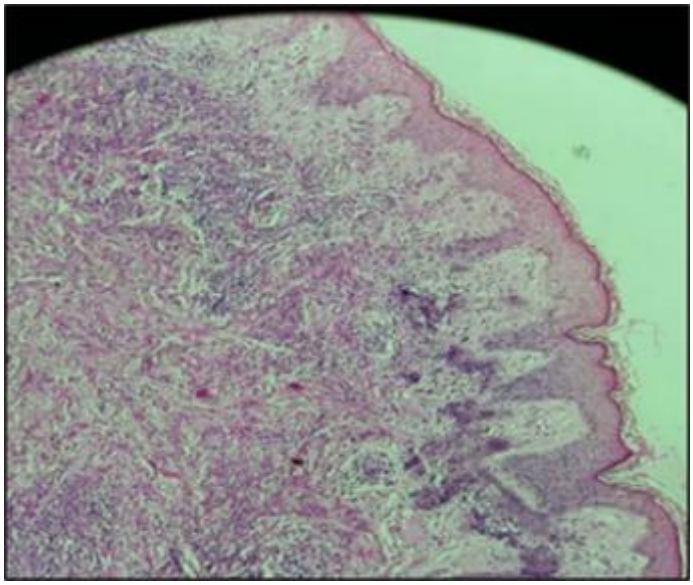
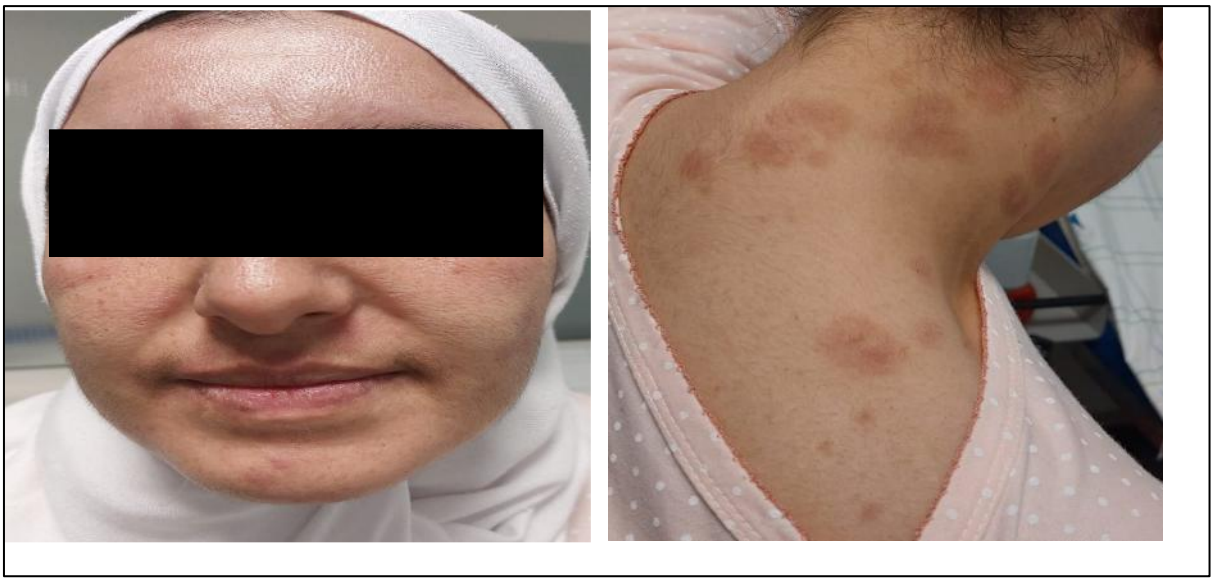


Figure 4 Dense dermal inflammatory infiltrate predominantly composed of neutrophils, without evidence of vasculitis. H&E staining, ×200

The diagnosis of Sweet syndrome associated with Behçet disease was established in our patient.

The patient was subsequently treated with systemic corticosteroid therapy at a dose of 40 mg/day with gradual tapering, combined with colchicine 1 mg/day for one month.

The clinical course was marked by progressive improvement of the cutaneous lesions *(Figure 5)*, with complete resolution achieved within 20 days.



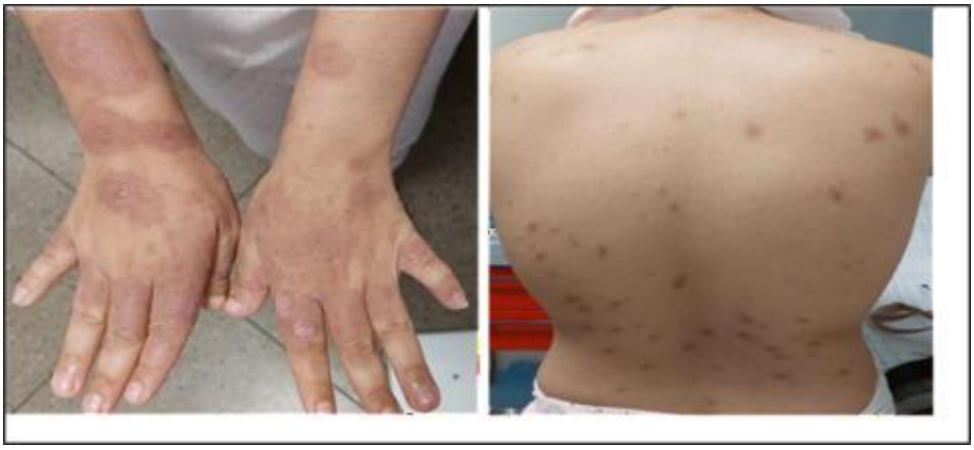


Figure 5 Improvement of cutaneous lesions following corticosteroid therapy and colchicine treatment

3. Discussion

Sweet syndrome (SS), also known as acute febrile neutrophilic dermatosis, is a rare inflammatory disorder belonging to the group of neutrophilic dermatoses, first described by Sweet in 1964. Clinically, it is characterized by the abrupt onset of painful erythematous plaques or nodules, often associated with a biological inflammatory syndrome, and histologically by a dense dermal infiltrate of neutrophils without vascular involvement [1–3]. SS is now recognized not only as a dermatologic entity but also as a cutaneous marker of systemic immune activation, potentially associated with inflammatory, autoimmune, infectious, or neoplastic diseases [3].

Behçet disease (BD) is a primary vasculitis of unknown etiology characterized by recurrent oral and/or genital aphthosis, ocular manifestations, and arthritis. Gastrointestinal involvement in BD is uncommon, with a reported prevalence ranging from 5% to 10% [4]. However, it may be particularly severe and challenging to diagnose, especially in distinguishing it from inflammatory bowel disease (IBD), notably Crohn's disease, particularly when it represents the initial manifestation. Intestinal involvement in BD, referred to as entero-Behçet disease (EBD), is defined by the presence, in a patient with BD, of gastrointestinal symptoms associated with aphthous ulcerations confirmed by endoscopic examination [5], in the absence of infectious, toxic, or other inflammatory causes. Gastrointestinal involvement in BD usually occurs 4 to 6 years after the onset of oral ulcerations [6,7].

From a differential diagnostic perspective with Crohn's disease, several features may help orient the diagnosis. Entero-Behçet disease typically presents with ileocecal involvement associated with few, large, deep, round or oval colonic ulcerations with well-defined borders, whereas Crohn's disease is characterized by more numerous lesions, often longitudinal or serpiginous, with discontinuous segmental involvement and a cobblestone mucosal appearance. In addition, perianal involvement is frequent and highly suggestive of Crohn's disease, while the coexistence of bipolar aphthosis, uveitis, or a positive pathergy test strongly supports the diagnosis of entero-Behçet disease. Histologically, Crohn's disease may exhibit non-caseating epithelioid granulomas, which, although inconsistently present, are highly suggestive of the disease. In contrast, entero-Behçet disease is more commonly associated with neutrophilic inflammation and possible small- and medium-vessel vasculitis, without specific granulomatous lesions.

The coexistence of Sweet syndrome and Behçet disease has previously been reported in the literature. However, this association remains exceedingly rare given the differences in the clinical and pathogenic characteristics of the two conditions [1].

The pathophysiological mechanisms underlying both Sweet syndrome and Behçet disease remain incompletely understood but are thought to involve immune dysregulation. Sweet syndrome is characterized by an exaggerated immune response predominantly involving innate immunity, with an inflammatory cascade mediated by several cytokines, including IL-1, IL-6, IL-8, IFN-γ, G-CSF, and GM-CSF, leading to neutrophil activation and recruitment, alongside a demonstrated contribution of adaptive immunity. In Behçet disease, an interaction between genetic predisposition and environmental factors triggers a cytokine cascade dominated by IL-1, IL-6, IL-8, IL-18, and TNF-α, resulting in neutrophil activation responsible for the clinical manifestations [1]. The involvement of helper T lymphocytes, particularly Th1 cells, has been demonstrated in both conditions, with elevated levels of Th1 cytokines reported in Behçet disease [8]. Finally, a potential role for HSV-1 and HSV-2 infection as triggering factors has also been suggested [9].

Furthermore, the cutaneous manifestations of Behçet disease may clinically mimic Sweet syndrome, particularly papulopustular or nodular lesions. In such cases, the distinction relies primarily on histopathological examination. Sweet syndrome is characterized by a dense neutrophilic infiltrate sparing the vascular walls, without evidence of necrotizing vasculitis, whereas cutaneous lesions of Behçet disease may exhibit leukocytoclastic vasculitis [3,10,11]. In our case, the absence of vascular involvement on skin biopsy allowed the definite diagnosis of associated Sweet syndrome to be established.

From a therapeutic perspective, systemic corticosteroids remain the first-line treatment for Sweet syndrome, usually inducing rapid and often dramatic clinical improvement within a few days [3,10]. Such a favorable response was observed in our patient, with regression of the lesions within less than three weeks. Colchicine, a conventional maintenance treatment for Behçet disease due to its anti-neutrophilic properties, also contributes to the control of both cutaneous and systemic inflammation [1].

Dapsone, cyclosporine, and anti-TNFα agents are recommended in cases of failure or inadequate response to first-line therapy [12].

4. Conclusion

The occurrence of Sweet syndrome in a patient with entero-Behçet disease may be considered a marker of heightened systemic inflammatory activity, warranting close monitoring and appropriate therapeutic management. This rare association deserves greater recognition, as it may influence both therapeutic strategies and long-term follow-up.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

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