

Sweet Syndrome as a Manifesting Symptom of Inflammatory Bowel Disease

Andreas Bub MD, Amr Sayed MD, Jaspreet Suri MD

Stamford Hospital Department of Medicine, Norwalk Hospital Department of Medicine, Norwalk Hospital Section of Gastroenterology

Introduction

- **Sweet's Syndrome (SS):** An uncommon skin disorder characterized by the abrupt appearance of painful erythematous skin lesions, fever, and neutrophilia. It is usually a cutaneous manifestation of an underlying disease
- **Subtypes:** Classical (Inflammatory/Infectious), Malignancy-Associated, and Drug-induced.
 - Inflammatory Bowel Disease (IBD): approx. 100 cases reported, typically with Crohn's colitis.
- **Diagnosis:** Skin biopsy showing classic findings, please refer to figures 1 through 4
- **1st Line Therapy:** Systemic and topical corticosteroids
 - **Alternative 1st Line:** Colchicine, Dapsone, and Potassium Iodide.
 - **Refractory disease:** Various oral immunosuppressants, Pulse dose corticosteroids, TNF- α inhibitors

Case Presentation Part 1

Our Patient: 18 year old male with no medical history

Presenting Symptom: 1 month of abdominal pain, oral ulcerations, hematochezia, and a left lower extremity lesion.

Physical Exam: Febrile, Diffusely tender abdomen, multiple ulcers bilaterally on his buccal mucosa, cheilitis, and a 1 cm ulcerated lesion on his left leg.

Labs: Anemia, thrombocytosis, neutrophilia, elevated inflammatory markers, and a positive C-ANCA.

Cutaneous and Histological Images

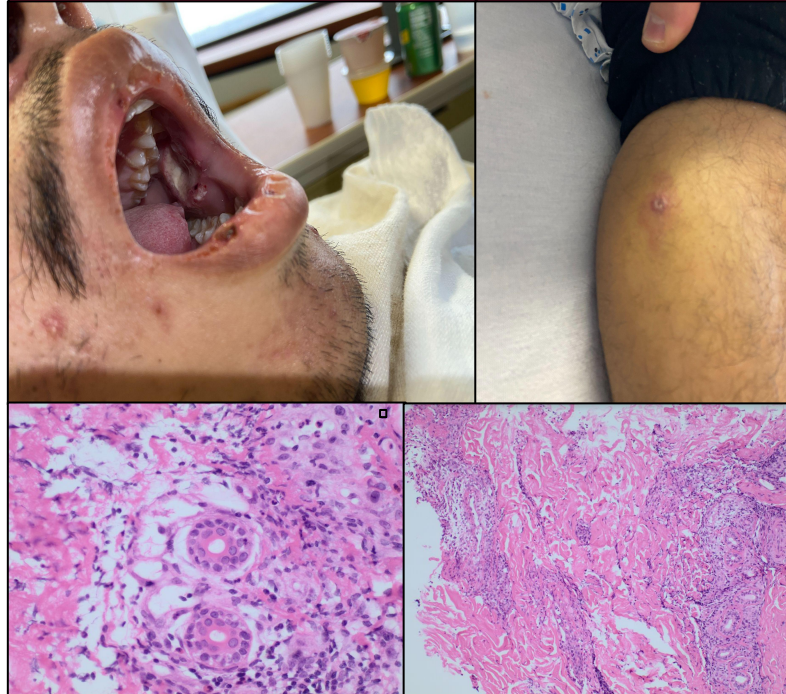


Figure 1 (Top Left): Cheilitis and large cratered ulcer on the left buccal mucosa

Figure 2 (Top Right): Painful erythematous cutaneous lesion on the right leg

Figure 3 (Bottom Left): Perivascular mixed inflammatory infiltrate without vasculitis

Figure 4 (Bottom Right): Low power view of figure 3 showing a larger section of the dermis with the mixed inflammatory infiltrate.

Case Presentation Part 2

Medical Therapy: Started on IV methylprednisolone for empiric treatment of presumed IBD.

Biopsies: The leg and oral lesions were biopsied as well as a cratered esophageal ulcer found during endoscopy. Sigmoidoscopy demonstrated a circular ulceration of the anal mucosa and severe patchy inflammation in the descending and sigmoid colon. The skin biopsy showed ND with an interstitial and perivascular inflammatory infiltrate with epidermal necrosis without vasculitis most consistent with SS.

Diagnosis and Disposition: He was diagnosed with Crohn's Disease and has seen improvement of his skin lesions, oral lesions, and abdominal symptoms after a prolonged steroid taper.

Conclusion

- SS is a rare manifestation of IBD that may occur before, concurrently, or after flares of the disease. This should be kept in mind, especially in patients with Crohn's colitis.
- Prompt evaluation of skin lesions and continuation of appropriate therapy will benefit IBD patients as the therapies used to treat SS are efficacious in IBD
- IBD associated SS has an excellent response to corticosteroids with rapid resolution of cutaneous lesions