Subcutaneous Sweet Syndrome Successfully Treated with Ustekinumab in **Patient with Ulcerative Colitis**



Kelly A. Hu, MD¹, Jeanne Shen, MD², Kerri E. Rieger, MD, PhD^{2,3}, Mike T. Wei, MD^{1,3}, John Gubatan, MD^{1,3}

1. Department of Medicine, 2. Department of Pathology, 3. Division of Dermatology, 4. Division of Gastroenterology and Hepatology Stanford University School of Medicine

Introduction

- Sweet Syndrome (SS), also known as acute febrile neutrophilic dermatosis, is a mucocutaneous manifestation of IBD that presents as erythematous papules/plaques involving the face, neck, and limbs. It is also associated with systemic symptoms of fever and arthromyalgias.¹
- SS can present before, after, or at the time of initial IBD diagnosis. It is generally associated with active IBD and reflects intestinal disease activity.²
- Here, we present the first case of subcutaneous SS with sterile osteomyelitis in a patient with ulcerative colitis (UC) successfully treated with ustekinumab.

Case Presentation

- A 50-year-old woman with a history of left-sided UC presented with abdominal pain, diarrhea, hematochezia, and progressive acuteon-chronic bilateral lower extremity pain.
- Previously failed mesalamine, azathioprine, infliximab, adalimumab, vedolizumab, fecal microbiota transplant trial.

- and negative C. diff.
- (FIGURE 1).
- \bullet abscess.



FIGURE 1: Ulcerations on Bilateral Ankles

- osteomyelitis.

References: (1) Cohen PR. Sweet Syndrome. https://rarediseases.org/rare-diseases.org/rare-diseases.org/rare-diseases.org/rare-diseases.org/rare-diseases.org/rare-diseases.org/rare-diseases/sweet-syndrome/. Published 2015. Accessed February 23, 2022. / (2) Rogler G, Singh A, Kavanaugh A, Rubin DT. Extraintestinal Manifestations of Inflammatory Bowel Disease: Current Concepts, Treatment, and Implications for Disease Management. *Gastroenterology*. 2021;161(4):1118-1132. / (3) Nasa M, Sharma Z, Lipi L, Sud R. Sweet's Syndrome in an Amplications for Disease Management. *Gastroenterology*. 2021;161(4):1118-1132. / (3) Nasa M, Sharma Z, Lipi L, Sud R. Sweet's Syndrome in an Amplications for Disease Management. *Gastroenterology*. 2021;161(4):1118-1132. / (3) Nasa M, Sharma Z, Lipi L, Sud R. Sweet's Syndrome in an Amplication State of the syndrome in a state of the syndrome in the syndro Case of Ulcerative Colitis-Case Report and Review of Literature. J Assoc Physicians India. 2019;67(4):84-85. / (4) Bento-Miranda M, Perdigoto DN, Mendes S, Portela F. Sweet's Syndrome as Manifestation of Active Ulcerative Colitis. GE Port J Gastroenterol. 2020;27(2):138-140. / (5) Bancu LA, Ureche C, Crăciun NM, Marian D. A case of Sweet's syndrome as Manifestation of Active Ulcerative colitis. Rom J Morphol Embryol. 2016;57(3):1145-1147. / (6) Rappaport A, Shaked M, Landau M, Dolev E. Sweet's syndrome in association with Crohn's disease: report of a case and review of the literature. Dis Colon Rectum. 2001;44(10):1526-1529. / (7) Rahier JF, Lion L, Dewit O, Lambert M. Regression of Sweet's syndrome associated with Crohn's disease: report of a case and review of the literature. Dis Colon Rectum. 2001;44(10):1526-1529. / (7) Rahier JF, Lion L, Dewit O, Lambert M. Regression of Sweet's syndrome associated with Crohn's disease: report of a case and review of the literature. Dis Colon Rectum. 2001;44(10):1526-1529. / (7) Rahier JF, Lion L, Dewit O, Lambert M. Regression of Sweet's syndrome associated with Crohn's disease: report of a case and review of the literature. Dis Colon Rectum. 2001;44(10):1526-1529. / (7) Rahier JF, Lion L, Dewit O, Lambert M. Regression of Sweet's syndrome associated with Crohn's disease.

On admission, she was febrile to 38.2°C and tachycardic. Labs were notable for borderline leukocytosis (neutrophilic predominance), ESR 120, CRP 32, fecal calprotectin 1655, Exam was notable for tender

subcutaneous nodules on bilateral ankles with violaceous erythema, ulceration, hemorrhagic bullae

MRI of bilateral ankles with multifocal osteomyelitis and adjacent

Bone gram stain and cultures were negative. Pathology of bone biopsies were consistent with sterile

Left ankle skin biopsies revealed spongiotic epidermis with dense granulomatous and neutrophilic inflammatory infiltrate in the deep dermis/subcutis, consistent with subcutaneous SS (FIGURE 2).

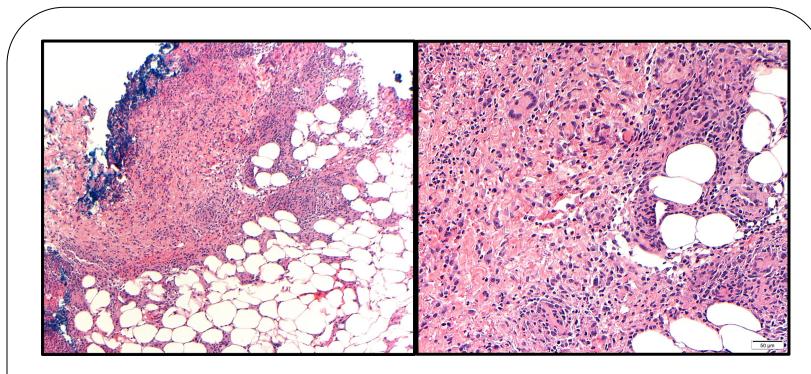


FIGURE 2: Histologic Sections from Left Ankle Skin Biopsy

Restaging colonoscopy demonstrated severe proctosigmoid UC (Mayo Endoscopy Score 3) with severe chronic active colitis on biopsy (FIGURE 3).

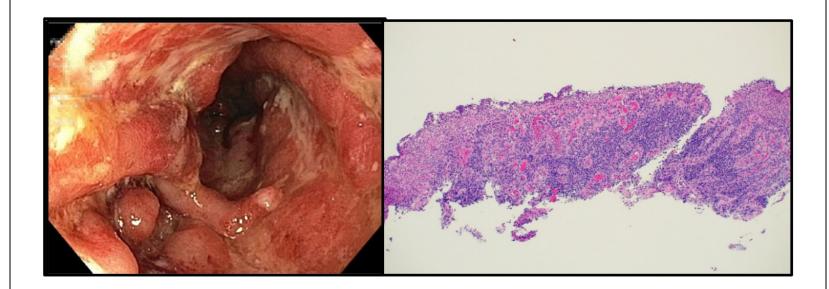
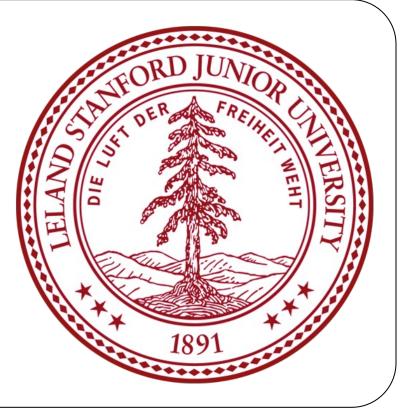


FIGURE 3: Proctosigmoid UC (*left*) with Chronic Active Colitis on Sigmoid Biopsy (right)

- Given failure of several therapies, ongoing colitis, and cutaneous SS symptoms, the patient was started on ustekinumab.
- One month later, repeat MRI demonstrated complete resolution of sterile osteomyelitis and abscesses.





- She had resolution of cutaneous SS lesions (FIGURE 4) and UC symptoms.
- Repeat colonoscopy eight months after starting ustekinumab with remission of colitis (Mayo Endoscopy Score 0), fecal calprotectin 150.



FIGURE 4: Resolution of Skin Erosions and Ulcerations After Treatment

Discussion

- The data on ustekinumab for treatment of EIMs in IBD are limited and mixed.
- There is currently no consensus on treatment of SS in IBD. Other therapies that have been used in the past include topical/IV corticosteroids, infliximab, and golimumab.³⁻⁷
- Our case highlights the first use of ustekinumab in successfully treating subcutaneous SS with sterile osteomyelitis in a patient with flaring UC.