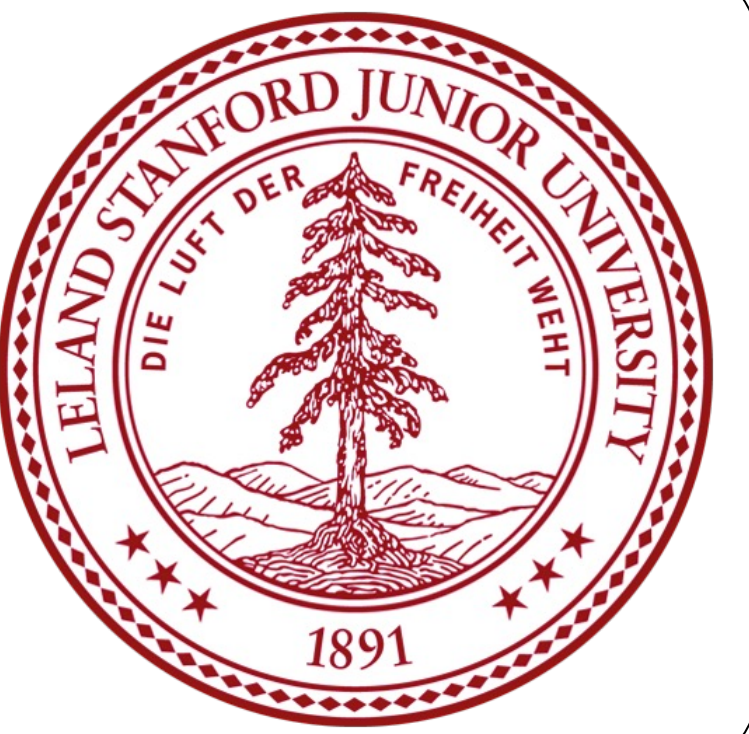




Subcutaneous Sweet Syndrome Successfully Treated with Ustekinumab in Patient with Ulcerative Colitis

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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 arthralgias.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 acute-on-chronic bilateral lower extremity pain.
- Previously failed mesalamine, azathioprine, infliximab, adalimumab, vedolizumab, fecal microbiota transplant trial.

- 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, and negative *C. diff*.
- Exam was notable for tender subcutaneous nodules on bilateral ankles with violaceous erythema, ulceration, hemorrhagic bullae (FIGURE 1).
- MRI of bilateral ankles with multifocal osteomyelitis and adjacent abscess.

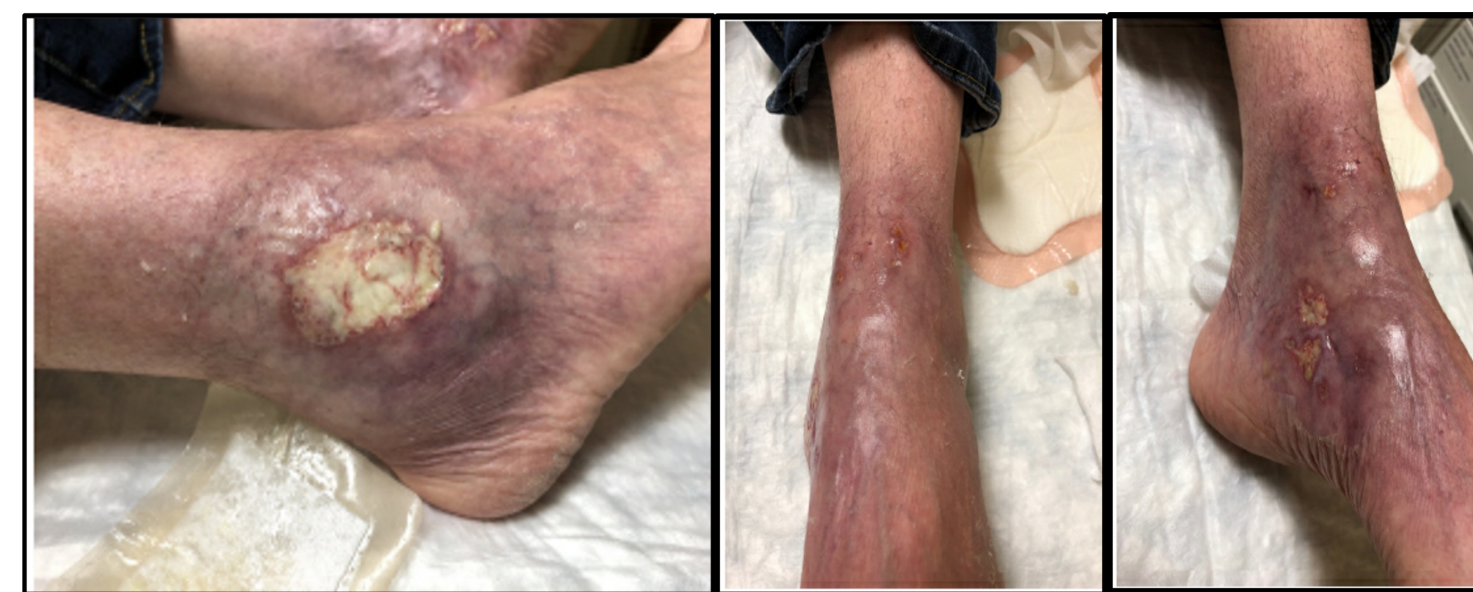


FIGURE 1: Ulcerations on Bilateral Ankles

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

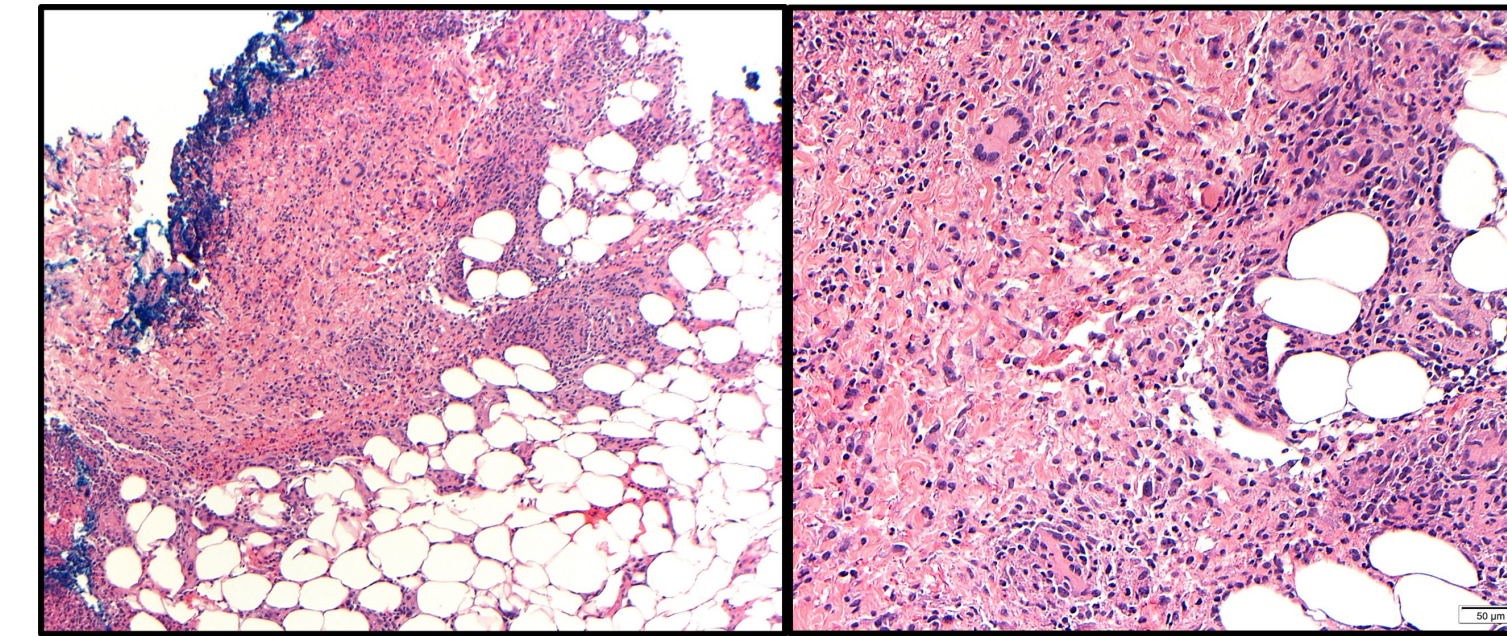


FIGURE 2: Histologic Sections from Left Ankle Skin Biopsy

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

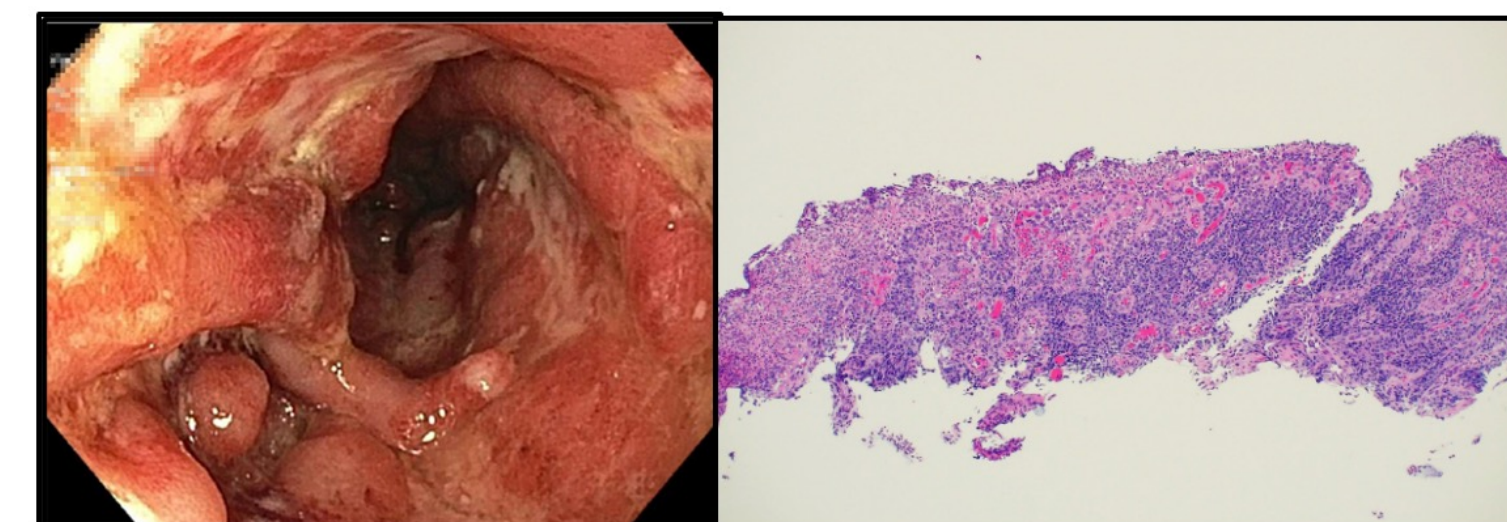


FIGURE 3: Proctosigmoid UC (left) with Chronic Active Colitis on Sigmoid Biopsy (right)

- 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.