A SWEET RESPONSE: CROHN'S FLARE LEADING TO SWEET SYNDROME

Sanskriti Sharma MD[1], Hima Veeramachaneni MD[2,4], Louis Kerkhoff MD[3,4] ,Thara Vidyasagaran MD[3,4], Karen Clarke MD[3,4], Gayle Wong MD[3,4], Francis Edward LeVert MD[2,4]

[1]Wellstar Atlanta Medical Center Internal Medicine Residency, [2]Digestive Diseases Division, [3]Department of Medicine

[4]Emory School of Medicine

Introduction

- Sweet's syndrome = acute febrile neutrophilic dermatosis (AFND)
- It is associated with underlying disorders such as IBD, infection, cancer, autoimmune disorders, & medications
- We present an uncommon case of Crohn's flare associated with bullous Sweet's syndrome, a rare entity

Case Presentation

- A 79-year-old male with history of CAD (s/p CABG), hypertension, hyperlipidemia, colonic Crohn's disease (CD) (on Infliximab), peptic ulcer disease and recurrent C. difficile infection presented with fever (102ºF), malaise and rash
- Rash is described as sudden in onset, painful, erythematous, bullous, & purulent (see physical exam images)
- Patient noted compliance with meds & noted recent decrease in Infliximab dosing

Physical Exam

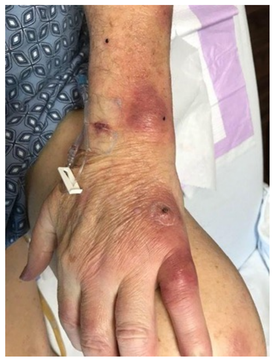


Image 1: Bullous lesions on dorsum of hand (spread to arm, bilateral knees & feet)

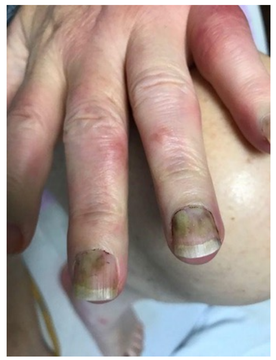
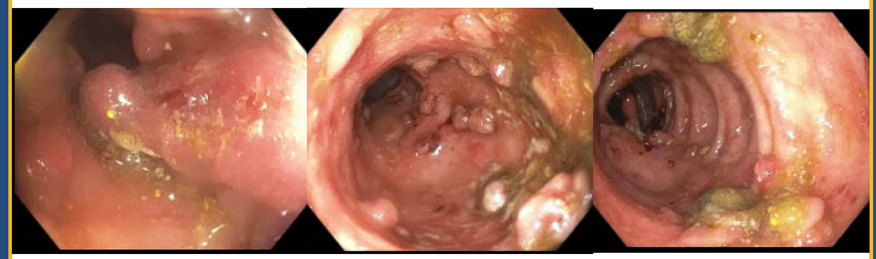


Image 2: Fingernail changes (dusky discoloration & splinter hemorrhages)

Hospital Course

- **Lab results**: lactic acid 1.96, WBC 18.5 K/uL, ESR 130 mm/hr, CRP 185.5 mg/L, positive ANA titer 1:320.
- Given elevated WBC & immunocompromised status, concern was for infectious etiology of his bullous lesions
- Blood cultures and infectious disease was consulted
- He was started on empiric antibiotics without improvement in his lesions
- Transthoracic and transesophageal echocardiogram were negative for any valvular lesions, ruling out infective endocarditis
- Gastroenterology was consulted due to Crohn's disease association with extraintestinal dermatologic manifestations (i.e., erythema nodosum [EN], pyoderma gangrenosum [PG[).
- Given his rash's atypical appearance and distribution, GI recommended Dermatology consult for skin biopsy
- Colonoscopy showed significant inflammation with large ulcers throughout the colon. Simple Endoscopic Score for CD was 25; biopsies were consistent with moderate to severe chronic active colitis.



Images from patient's colonoscopy showing active inflammation in with rectum (a), left colon (b), and right colon (c)

- Skin biopsy revealed marked, nodular dermal neutrophilic infiltrate, edema of the papillary dermis, and focal leukocytoclastic vasculitis. These findings were compatible with Sweet's syndrome.
- He was started on IV steroids and subsequently had marked symptom improvement with eventual resolution of bullous lesions
- His Infliximab dosing was increased to prior dosing to induce remission.

Discussion

- Sweet's syndrome is characterized by sudden-onset fever with tender, edematous, erythematous nodules on the extremities (bullous lesions are rare)
- 3 etiological subtypes of Sweet's syndrome:

1. **Classical** secondary to infection, IBD, or pregnancy
   Diagnosis requires 2 major & 2 minor criteria to be met:
   Major criteria: [1]abrupt onset of painful erythematous plaques/nodules & [2]histopathologic evidence of a dense neutrophilic infiltrate
   Minor criteria: [1]pyrexia >38ºC,[2]association with underlying malignancy, inflammatory disease or pregnancy, or preceded by upper respiratory or gastrointestinal infection, or vaccination, [3]excellent response to treatment with systemic glucocorticoids or potassium iodide, [4]abnormal lab values at presentation [≥3: ESR >20 mm/hour, positive CRP, WBC >8000, >70% neutrophils]
2. **Malignancy-associated**
3. **Drug-induced**

- Our patient had classical Sweet's syndrome with atypical bullous lesions and was unique given it is more common in female patients

Conclusion

- EN & PG are commonly associated with IBD; however, Sweet's syndrome is rarely associated with IBD itself
- It is important to consider immunosuppressant medication or opportunistic infections when Sweet's syndrome presents in IBD
- Consider a multidisciplinary approach with dermatology consultation for atypical skin lesions to diagnose and treat promptly
- Understanding the vast skin manifestations of IBD is key to providing comprehensive care to IBD patients