

Not So Sweet: A Rare Extraintestinal Manifestation in Crohn's

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Background

- Acute febrile neutrophilic dermatosis = Sweet's syndrome
- Rare extraintestinal manifestation (EIM) of inflammatory bowel disease
- 40 cases associated with CD¹; CD > UC
- Abrupt onset of painful erythematous nodules

Associated symptoms



↑ WBC



Phenotypes



67-80%²



87%³

Case

- 22-year-old male presents with fulminant colitis complicated by perforation requiring emergent ileostomy
- T 100.6F, HR 154, BP 97/75, WBC 45, lactate 4.45
- While diverted, diagnosed with UC on flexible sigmoidoscopy
- Started on High-dose Infliximab 10mg/kg induction
- One month after restoration of continuity as part of 3 stage TPC-IPAA, he developed fevers and wound dehiscence
- CT revealed enterocutaneous fistula raising concern for Crohn's colitis
- During hospitalization, developed red papules/pustules with peripheral scaling on his face spreading across his body with evidence of pathergy at IV sites
- Diagnosed with acute febrile neutrophilic dermatosis
- After shared-decision making discussion and consideration of risk for poor wound healing post-operatively, he was treated with IV Methylprednisolone 1mg/kg for 5 days with resolution of his lesions



Photo 1: rash that abruptly developed during hospitalization



Photo 2: pathergy at LUE IV site

Common Dermatological Manifestations of IBD

Dermatological Extraintestinal Manifestation	Anatomy Affected	Description of Lesions	IBD Activity	Management
Erythema Nodosum	Lower Extremities	Tender, raised subcutaneous lesions	parallels	Treatment of underlying IBD resolves lesions
Pyoderma Gangrenosum	Sites of trauma to skin- often lower extremities or abdominal wall adjacent to stoma after colectomy or surgical scar	erythematous papules or pustules with subsequent necrosis of the dermis and development of deep ulcerations	does not parallel	Systemic Glucocorticoids
Bowel Associated Dermatitis-arthritis	upper extremities and trunk	eruption of erythematous macules (up to 1cm) that develop a central papulovesicular or pustule	parallels	Systemic Glucocorticoids
Sweet's Syndrome	Arms, trunk, face, neck	abrupt onset of painful erythematous nodules	parallels	Systemic glucocorticoids

Discussion

- This case demonstrate a rare finding of Sweet's syndrome in a male patient, a disease associated more commonly with women
- Early recognition is important in order to start corticosteroids quickly, however corticosteroids impact wound healing mechanisms via downregulating endothelial cell expression of intercellular adhesion molecule 1 (ICAM-1) leading to reduced granulocyte adhesion/migration - a consideration for our patient with recent major abdominal surgery^{4,5}
- Animal studies have shown steroids can impair collagen turnover and decrease the strength of cutaneous wounds⁵
- A Randomized double-blind study showed no wound infections and only 1 patient with wound dehiscence⁵
- This case highlights a favorable outcome despite use of high dose steroids in this critically ill patient with recent restoration of intestinal continuity after perforation

References

1. Joseph Sleiman, Asif A Hitawala, Benjamin Cohen, Katie Falloon, Marian Simonson, Benjamin Click, Urmi Khanna, Anthony P Fernandez, Florian Rieder, Systematic Review: Sweet Syndrome Associated with Inflammatory Bowel Disease, *Journal of Crohn's and Colitis*, Volume 15, Issue 11, November 2021, Pages 1864–1876
2. Lima CDS, Pinto RDB, Góes HFO, Salles SAN, Vilar EAG, Lima CDS. Sweet's syndrome associated with Crohn's disease. *An Bras Dermatol*. 2017;92(2):263-265. doi:10.1590/abd1806-4841.20175298
3. Ali M, Duerksen DR. Ulcerative colitis and Sweet's syndrome: a case report and review of the literature. *Can J Gastroenterol*. 2008 Mar;22(3):296-8. doi: 10.1155/2008/960585. PMID: 18354759; PMCID: PMC2662205
4. Lopes Caçola R, Soares M, Cardoso C, Furtado A. Sweet's syndrome complicating ulcerative colitis: a rare association. *BMJ Case Rep*. 2016 Jan 20;2016:bcr2015212990. doi: 10.1136/bcr-2015-212990. PMID: 26791120; PMCID: PMC4735358
5. Wang A, Armstrong E, Armstrong A. Corticosteroids and wound healing: clinical considerations in the perioperative period, *The American Journal of Surgery*, Volume 206, Issue 3, 2013, Pages 410-417, ISSN 0002-9610