A Not So Sweet Complication of Crohn's Disease

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Introduction

Sweet syndrome (SS), also known as acute febrile neutrophilic dermatosis, is an inflammatory skin condition characterized by neutrophilic infiltration of the skin. SS can be idiopathic, and has also been associated with various infections, malignancy, inflammatory or autoimmune disorders, drugs, or pregnancy. We present a case of SS likely from IBD and/or azathioprine.

Case Presentation

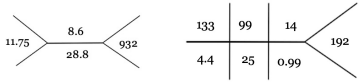
HPI:
A 34 year old male with poorly controlled Crohn's disease complicated by inflammatory arthritis and a history of IVDU and urticarial reaction to infliximab presents with a painful, pruritic rash that initially started on his face then spread down bilateral upper extremities and trunk. Additional ROS includes bilateral eye discomfort (scleritis), anorexia, weight loss, and a penile lesion.

Physical Exam Notable For:
- VSS except tachycardia, bilateral injected conjunctivae, diffuse crusted and eroded papules and plaques that are tender to palpation

Past Medical History:
- Asthma, Crohn's Disease, Inflammatory arthritis

Medications:
- Azathioprine, Montelukast, Oxycodone, Ferrous sulfate, Thiamine

Lab Findings:



- *Infectious*: RPR and HIV non-reactive
- *Inflammatory*: CRP 101
- *Immunologic*: ANA, ANCA, CCP Ab all within normal range

Diagnostic Criteria

Constant Features
(Present in all cases)
1. Abrupt onset of painful or tender erythematous papules, plaques, or nodules
2. Dense dermal neutrophilic infiltrate

Variable Features
(Present in certain cases)
1. Fever > 38°C
2. Atypical skin lesions
3. Histopathologic findings: presence or absence of leukocytoclastic vasculitis; variants including subcutaneous, histiocytoid, xanthomatoid, or cryptococcoid
4. Lab findings: elevated ESR, elevated CRP, leukocytosis, neutrophilia, anemia

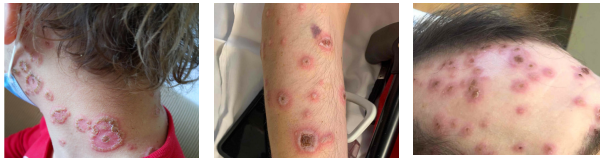
Source: Reference 3

This patient demonstrated both constant and several variable features of SS. He was ultimately diagnosed with bullous-type SS in the setting of his inflammatory bowel disease (IBD).

Skin Manifestations

Punch Biopsy (skin lesion on posterior neck): Ulcer with seropurulent crust, and a dense, superficial and deep infiltrate of numerous neutrophils, lymphocytes, and histiocytes.

Pre-treatment



This patient was treated with three days of pulse-dose steroids followed by a prednisone taper.

Post-treatment

Discussion and Learning Points

Diagnosis
- The gold-standard method to diagnose SS is a skin biopsy.
- Although idiopathic SS has been reported, it is important to evaluate for possible underlying causes or associations of SS when the diagnosis is made.
- SS is a reported complication of IBD.

Extracutaneous Features
- Fever, arthralgia, ophthalmologic inflammation, oral or genital lesions.

Inciting Factors
- SS has been associated with both IBD and azathioprine.
- Azathioprine was discontinued and the patient was re-started on infliximab to help control his Crohn's disease.

Treatment
- Systemic corticosteroid is the first line therapy for SS.
- Other oral agents include colchicine, dapsone, cyclosporine, and NSAIDs.
- Refractory cases may benefit from TNF inhibitors or IL-1 receptor antagonists.
- Withdrawal of offending agents may also lead to remission in suspected drug-induced SS.

References

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