Sweet Syndrome: Benign or Insidious?

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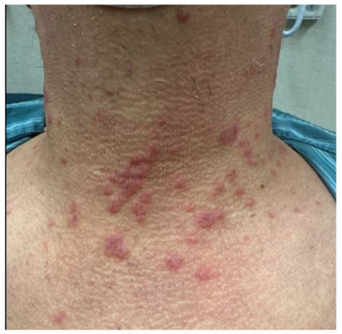
Background

Sweet Syndrome (acute neutrophilic febrile dermatosis) is an uncommon inflammatory disorder resulting in an abrupt painful, erythematous rash usually accompanied by fever and leukocytosis.

It is divided into classic/idiopathic, malignancy-associated, and drug-induced with the classic/idiopathic presentation being most common.

Figure 1



Objectives

- Describe the presentation of Sweet Syndrome
- Identify the most common disease states associated with Sweet Syndrome

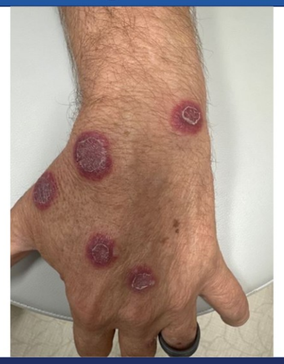
Case Description

A 66-year-old male, who worked as a radiation safety technician, with a history of a large benign bladder tumor s/p TURBT, pulmonary nodules, bicytopenia ongoing for several months, and a multiple pack-year history had previously been seen by Hematology for a workup of bicytopenia. This eventually was attributed to the aforementioned bladder tumor and no additional follow-up was pursued. He later presented with two weeks of flu-like symptoms, low-grade fevers, and fatigue along with a new spreading rash that began one week prior. Because he had recent sick contacts, he did not think much of his symptoms until the following week when he removed his work clothing to find a handful of pink, raised, non-pruritic lesions on his left forearm and one on his right. Over the next week, the lesions on the arms grew in size to become large, violaceous, and tender, while new lesions appeared on the upper back, chest, and bilateral lower extremities.

Physical exam was notable for violaceous, well-demarcated, circular, raised patches on the left forearm and hand that were mildly tender to palpation (Figure 2). Additionally, deep red/pink annular plaques with raised, irregular borders were noted on the dorsal hand, face, neck, chest, upper back, and bilateral legs (Figure 1).

Initial laboratory results were significant for pancytopenia and neutropenia without electrolyte, renal, or hepatic abnormalities. CT imaging revealed splenomegaly. A broad workup was performed to consider hematologic, rheumatologic, and infectious etiologies.

Figure 2



Discussion

Although the pathophysiology of Sweet Syndrome is not well understood, it is thought to be cytokine-induced. Classical Sweet Syndrome is usually seen in females between the ages of 30-50, often presenting 1-3 weeks after an upper respiratory-like illness, while malignancy-associated Sweet Syndrome can occur in those with a known or unknown malignancy and is frequently related to acute myelogenous leukemia (AML).

In this case, the patient was found to have hairy cell leukemia (HCL), a B-cell lymphoid malignancy seen in less than 2% of all leukemias. HCL usually presents with vague symptoms such as abdominal fullness (splenomegaly), recurrent infections (neutropenia), or easy bruising/bleeding (thrombocytopenia) and is less likely to include typical B symptoms.

Results

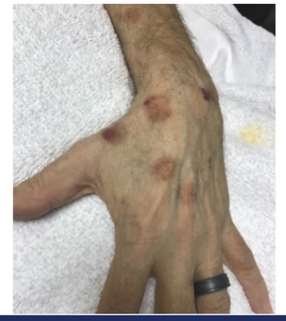
Diagnostics

Skin Biopsy: dermatopathology revealed neutrophilic and histiocytic dermal inflammation with leukocytoclasis consistent with Sweet Syndrome.

Treatment

Both oral and topical corticosteroids were started for treatment of Sweet Syndrome. Unfortunately, after completion of the initial corticosteroid regimen there was little improvement in the rash and the patient developed a worsening fever. This prompted concern for resistant Sweet Syndrome. An additional course of high-dose steroids was given. This was followed by Methotrexate and a steroid taper with subsequent clinical and symptomatic improvement (Figure 3).

Figure 3



Conclusions

It is important to keep a broad differential when new skin lesions are identified. Prompt diagnosis of Sweet Syndrome can identify an underlying malignancy resulting in fewer treatment delays.

References

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