A "Sweet" Look into a Rare Disease: Presentation and Management of Acute Febrile Neutrophilic Dermatosis

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A "Sweet" Look into a Rare Disease: Presentation and Management of Acute Febrile Neutrophilic Dermatosis

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Introduction

- Acute febrile neutrophilic dermatosis, also known by its eponym, Sweet Syndrome, is a rare disease that manifests as pyrexia with elevated inflammatory markers, neutrophilia and painful red or violaceous lesions that may be bullous, plaque like or nodular.
- Sweet Syndrome may be medication induced, associated with underlying malignancy or associated with abdominal or respiratory infections.
- This case describes an uncommon presentation of Sweet Syndrome in a previously healthy young male.

Case Presentation

- A 26-year-old male with no significant past medical history, unknown vaccination status, and recent travel to India presented with fevers, abdominal pain, vomiting and new onset, non-painful skin lesions of the left cheek, neck and shoulder (Figure 1).
- He presented in distributive shock with a temperature of 105° F and labs notable for leukopenia, bandemia, thrombocytopenia and acute kidney injury.
- Airborne precautions were instituted and he was started on doxycycline and acyclovir to cover herpes-zoster and tick-borne illnesses empirically.
- The patient quickly developed more lesions of variable morphology, now on his lower extremities (Figures 2-3), worsening leukopenia, thrombocytopenia and kidney injury.
- Extensive Infectious, Rheumatologic and Hematologic work up was unrevealing.
- Multiple punch biopsies of the skin returned a presumptive diagnosis of Sweet Syndrome with visualization of a neutrophilic invasion of the dermal and epidermal layers.
- Systemic steroid therapy was initiated on hospital day 3 with resolution of thrombocytopenia and leukopenia. The skin lesions began to heal and the patient remained afebrile. He was discharged 2 days later with outpatient follow up and showing continued clinical improvement.

Images

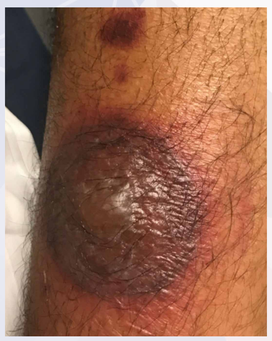

Figure 1: A pustular, raised lesion with mild surrounding erythema noted on the left side of the patient's neck.

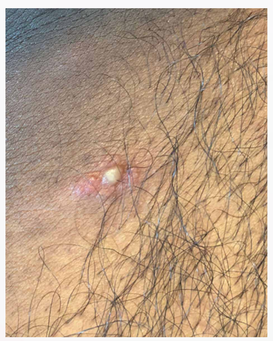

Figure 2: A well circumscribed, bullous lesion with mild surrounding erythema was noted on the patient's right medial lower extremity.

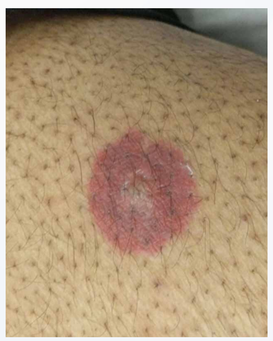

Figure 3: A well circumscribed macular, erythematous lesion found on the patient's left anterolateral thigh.

Discussion

- Although a preceding abdominal illness is a common precursor for Classical Sweet Syndrome, the patient's non-tender skin lesions, recent travel, unknown vaccination status and septic presentation resulted in a large differential diagnosis
- The leukopenia had a neutrophil trend and bandemia which coincides with the suspected pathophysiology of Sweet Syndrome as a cytokine mediated hypersensitivity reaction involving the dermal layers.

Conclusions

- Key points to this unique presentation of an uncommon disease were to quickly rule out infectious etiologies relevant to the patient and begin considering malignancies, bone marrow pathology and vasculitis. Especially beneficial to this case was obtaining an early dermatologic biopsy.
- The immediate steroid responsiveness to disease progression and severity aided in confirming the suspected cytopathological diagnosis as did keeping a broad differential that was narrowed rapidly with appropriate testing.

References

Cohen PR. Sweet's Syndrome – A Comphrehensive Review of an Acute Febrile Neutrophilic Dermatosis. *Orphanet Journal of Rare Disease*. 2007. 2:34. 1-28.

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