Masquerade of marauders: Sweet Syndrome, zoster, and hepatitis C

Madeline J. Hooper, BA[1]; Martin C. DelaTorre, DO[2]; Monica B. Mistry, DO[2]

[1]Morsani College of Medicine, University of South Florida, Tampa, FL [2]Department of Internal Medicine, Lehigh Valley Health Network, Allentown, PA

Background

- Sweet syndrome (SS) is a febrile neutrophilic dermatosis often associated with infection, malignancy, and medications.[1-7]
- SS is defined by the acute eruption of painful cutaneous lesions with histopathological evidence of dense neutrophilic infiltration. It can affect mucous membranes, the central nervous system and viscera.
- A minor diagnostic criterion of SS is excellent response to IV high-dose corticosteroids.

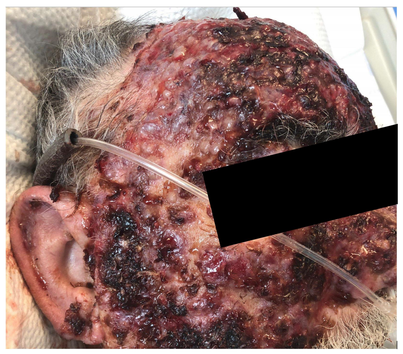
Objective

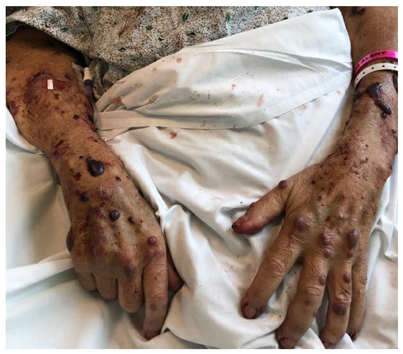
To describe a corticosteroid-refractory case of bullous Sweet Syndrome complicated by varicella zoster (VZV) and untreated hepatitis C (HCV).

Case Presentation

HPI

- 75-year-old man with end stage renal disease and untreated HCV presents with a hemorrhagic, necrotic rash on his face with oral involvement and tense bullae on his bilateral extremities (Figures 1 and 2).





Figures 1 & 2: Facial rash (1) and bullae on bilateral arms (2), Day 1.

- Five days prior, he had received IV cefepime, levofloxacin and vancomycin, and undergone contrast CT for febrile encephalopathy at an outside hospital.

Allergies

- Piperacillin-tazobactam (anaphylaxis), ciprofloxacin (rash), metronidazole (rash)

Hospital Course

Day 1

- Afebrile, hemodynamically stable
- Rash covers 10% body surface area (BSA)
- Negative Nikolsky's sign
- ***Start*** IV methylprednisolone 50mg BID

Day 2

- Burn and Dermatology favor diagnosis of bullous SS
- Punch biopsy of the scalp: diffuse dermal neutrophilic infiltrate consistent with SS
- ***Add*** hydrocortisone 2.5% and triamcinolone 0.1% ointments

Day 3

- Rash progresses to 30% BSA; larger, more numerous bullae in mouth (Figure 3) and on extremities
- Leukocyte count 10,800, 94% neutrophils
- Serology: HbsAb+, HcAb+, HCV viral load 38,400 IU/L
- Direct fluorescent antibody on leg lesion sample VZV+
- Blood cultures, HIV 1/2 Ag/Ab, HSV probe negative
- ***Start*** IV acyclovir 5mg/kg QD

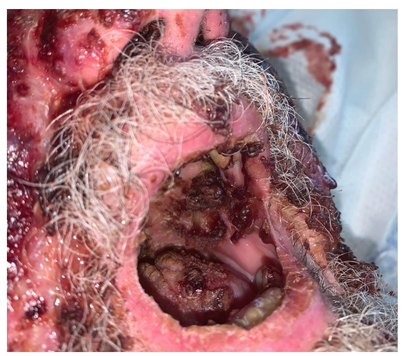


Figure 3. Oral bullae, Day 3.

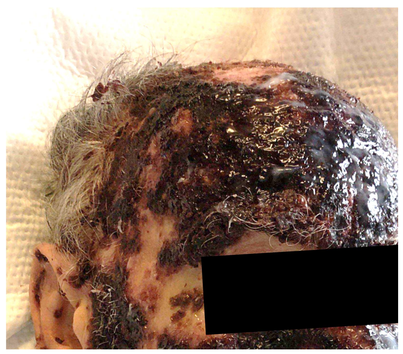


Figure 4: Facial rash, Day 8.

Day 8

- Widespread skin necrosis in areas of rash (Figure 4)
- Autoimmune panel: high levels of ANA, anti-dsDNA and anti-Ro/SSA antibodies

Day 10

- Patient died from medical complications

Discussion

Suspected trigger(s) of this patient's bullous SS

- Drug-induced
  - Contrast dye
  - Hydralazine
  - Antibiotics
- Occult hematologic malignancy[4,8]
  - Age > 65 years
  - Anemia (hemoglobin 9.4, Day 1)
  - Thrombocytopenia (platelets 90, Day 1)
  - Absence of arthralgias
- Respiratory infection, sacral wound infection

Literature review

- Only one case of dermatomal bullous SS triggered by herpes simplex virus and three cases of SS in the setting of HCV have been published.[9-12]
- No data clarify if the relationship between this patient's HCV, autoimmunity, and SS was causal or benign.

Conclusions

- Unique to our case is its myriad possible etiologies.
- Because SS may not regress without treatment of the underlying cause, elucidating the precipitating event remains important on a case-by-case basis. This element of SS management requires further study.

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