UNIDENTIFIED DIFFUSE BLISTERING DISORDER WITH TONGUE ULCERATION AND MUCOSAL SPARING

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Background

Acute neutrophilic dermatoses, are a rare group of disorders characterized by erythematous, tender skin lesions, +/- fever, and leukocytosis often-times with internal organ involvement. Malignancy, infection, drug exposure and pregnancy have all been associated with these conditions.

Since the disorder is rare in-itself; atypical presentations can pose a difficult challenge in establishing a diagnosis leading to a delay in care. Below we present a case of a patient who had multiple hospitalizations over the past year with unidentified diffuse blistering disorder, ulcerations and mucosal sparing believed to be Sweet syndrome.

Introduction

A 64 yo female with PMHx of aortic stenosis, a-fib, CVA with left sided hemiplegia, glaucoma, HTN, and T2DM presents from a nursing home with chief complaint of persistent, scattered skin “blisters”. Patient reports the lesions began three weeks prior to presentation and have progressively worsened in that period. They are mostly located on her upper body, but also include the tongue. They are described as extremely painful and itchy to the point of affecting her oral intake given their location. She denies any sick contact or persons in her facility with these symptoms or skin findings.

She was treated by her PCP with a short course of Doxycycline 100 mg BID without symptomatic relief. Prior records also show that she had a similar presentation in 2018 that was attributed to cellulitis secondary to a MRSA infection.

ED Vitals: BP 108/63 | Pulse 89 | Temp 36.7 °C (98 °F) | Resp 20 | SpO2 100 % |

ED Course: Upon presentation to the ED, routine labs and an ECG were obtained. Patient was also given 1g Ancef, 12.5mg Benadryl, 2% viscous lidocaine, and one dose of 10mg dexamethasone.

Objective Findings

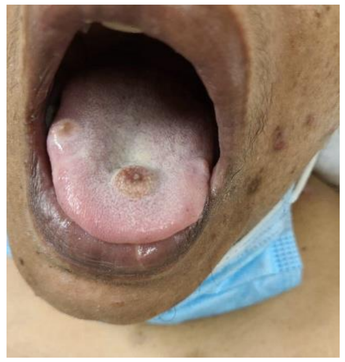
Pertinent Physical exam findings:

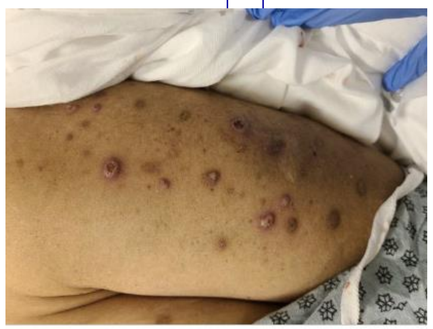
BP 106/81 | Pulse 89 | Temp 37 °C (98.6 °F) | Resp 19 | SpO2 98 % |

General: obese, ill appearing, no acute distress

HEENT: atraumatic, PERRLA, moist mucosa, normal pharynx, normal tonsils and adenoids. Papules and plaques noted on the tongue. Ulcerations from ruptured blisters present on face.

Respiratory: CTA bilaterally, decreased breath sounds.





Cardiovascular: 2/6 holosystolic murmur. RRR , No LE edema appreciated.
Abdomen: Soft, non-tender, non-distended, normal bowel sounds in all quadrants
Musculoskeletal: 3/5 LUE muscle strength and 2/5 LLE muscle strength.
Integumentary: Multiple 0.5 to 1 cm lesions with scabbed/umbilicated centers. Hyperpigmented spots noted. Multiple vesicular lesions with ulceration present.

Labs

NA 137	AST 135	HGB 11.2
K 4.3	ALT 85	PLT 200
CL 101	ALP 235	WBC 10.3
BUN 27	T. bili 1.1	HCT 36.1
CRT 1.23	D. bili 0.6	
GLU 191		

Diagnostics

Chest Xray: Unremarkable frontal view of the chest.

EKG: NSR with normal axis, Prolonged QTc with some flattened T waves.

Clinical Course

Infectious Disease: Infectious etiologies were considered including: Coccidiomycosis, Cryptococcus, molluscum. HSV less likely. Size and distribution did not fit routine bacterial process. Patient did not have any risk factors for fungal etiology.

Plan: Blood cultures, biopsy of both ulcerated and intact lesions sent to pathology for culture, stain, fungal, AFB. Continue steroids and discontinue systemic antibiotics. Punch biopsies of periumbilical and left shoulder skin area were performed by surgery and sent to in-house pathology. Specimens were also sent for a second opinion to dermatopathologist.

During the course of her admission, the patient experienced cardiac dysrhythmia prompting transfer to a tertiary care center for pacemaker implantation. She was evaluated by a dermatologist at the facility who did not believe the lesions were infectious in nature.

She was discharged home with topical steroids, one month of doxycycline, and instructed to follow up as an outpatient.

Results

- HSV 1 and HSV 2 IgG positive
- Hepatitis panel, Syphilis, HIV negative
- Coxsackie negative
- L arm bpx -Neutrophilic dermal infiltrate with leukocytoclasia. Specimen sent to outside institution
- Bpx gram stain and PAS negative for bacterial or fungal organisms.

Follow Up

One month later, the patient was admitted to ICU with altered mental status, anemia, and septic shock secondary to suspected UTI. She was treated with a 14-day course of empiric antibiotics, was placed on pressor support, and subsequently intubated for airway protection. She was downgraded from the ICU after seven days. No infectious etiology was ever isolated via blood cultures, CSF studies nor were any toxic metabolites identified on toxin screen. Three days after ICU downgrade, the patient experienced hemorrhagic shock secondary to an acute lower GI bleed prompting return-back to the unit. The following morning the she went into PEA arrest, with ROSC achieved after resuscitative measures. Her prognosis was poor and was ultimately palliatively extubated per her family’s wishes. Unfortunately, a formal diagnosis was never made.

Summary

- Neutrophilic dermatoses are conditions of inflammatory infiltrate made up of polymorphic leukocytes
- Neutrophils are usually located in the dermis in Sweet syndrome and pyoderma gangrenosum and in the epidermis in subcorneal pustular dermatosis
- Cutaneous findings include: vesiculopustules, nodules, plaques and ulcerations that are painful and non pruritic
- These syndromes have been associated with infection, malignancy, pregnancy and drug exposure
- Early diagnosis and systemic glucocorticoids are the mainstay treatments.

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