

CLINICAL VIGNETTE

Staphylococcal Scalded Skin Syndrome in an Immunocompromised 78-Year-Old-Man

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Introduction

Staphylococcal scalded skin syndrome (SSSS) is a systemic disease caused by epidermolytic exotoxins released by toxigenic strains of *Staphylococcus aureus*. The condition is characterized by bullae formation followed by desquamation, resulting in tender, erythematous areas of denuded skin. SSSS is typically seen in children and only rarely occurs in adults, usually in elderly or immunocompromised individuals. The annual incidence in adults is 0.98 cases per million compared to 7.67 cases per million in the pediatric population.¹

Although SSSS is rare in adults, it is associated with significant morbidity and mortality. The mortality rate is less than 10% in children, but up to 63% in adults, possibly due to serious comorbidities in patients affected by the disease.² Early recognition of this condition and prompt initiation of treatment is crucial. We present an interesting case of SSSS in an immunocompromised adult that required a multidisciplinary approach to establish the diagnosis. This case highlights the rare presentation of a potentially life threatening dermatologic condition in an uncommon age group and the need for clinician awareness and collaboration in such instances.

Case Presentation

A 78-year-old male with recently diagnosed squamous cell carcinoma of the neck (2018) undergoing radiation, presented to dermatology with a rash on his right upper arm. The rash began about a week prior to presentation with what he described as skin peeling and it was mildly painful. He denied any itching, burning, fevers, chills, or other associated symptoms. He saw his primary care provider who prescribed triamcinolone 0.5% cream to apply to the affected area as well as a rash diagnosed as eczema on his left cheek and right forearm. A few days later, he was seen by hematology oncology to discuss his scheduled carboplatin/paclitaxel chemotherapy later in the week. The physician noted that the rash had worsened and prescribed a 10 day course of amoxicillin 500mg by mouth 3 times daily and cotrimoxazole DS 800-160 mg by mouth two times daily. He was referred to dermatology to evaluate whether the initiation of his chemotherapy should be postponed.

Past medical history was also significant for esophageal cancer s/p radiotherapy and cefuximab, coronary artery disease s/p 4

vessel CABG, stage 2 chronic kidney disease, moderate aortic stenosis, type II diabetes mellitus, hypercholesterolemia, and hypertension. He denied any new medications, lotions, detergents, and soaps.

Physical examination revealed erythematous tender patches with desquamation on the bilateral antecubital fossae and right dorsal hand (Figure 1). Ill-defined erythematous, lichenified, scaly plaques distributed over the left cheek and right forearm were also noted. The differential diagnosis included bullous impetigo, staphylococcal scalded skin syndrome, and contact dermatitis. A punch biopsy and bacterial wound culture were obtained from an area of desquamation. The amoxicillin was discontinued, cephalexin 500 mg by mouth three times daily was initiated, and cotrimoxazole DS was continued as prescribed by his oncologist.

The wound culture grew many methacillin susceptible staphylococcus aureus, sensitive to cephalexin already prescribed. Histopathologic analysis showed a subcorneal cleft/bulla with mildly to moderately dense superficial perivascular and interstitial acute inflammatory infiltrate in the dermis, supporting a diagnosis of Staphylococcal scalded skin syndrome. The combination of the history, histologic analysis, and wound culture was most consistent with a diagnosis of SSSS.

He continued the antibiotic course and recommended wound care regimen of Aquaphor ointment twice daily. He was seen in hematology oncology clinic 5 days later and was noted to have significantly improved skin findings. Unfortunately, the patient passed away due to cancer complications several months later.

Discussion

Exfoliative toxins A and B (ETA, ETB) are responsible for the pathogenesis of staphylococcal scalded skin syndrome (SSSS) in humans.² About 30% of the population are nasal carriers of the bacteria and only 5% of all *Staphylococcus aureus* (*S. aureus*) are toxigenic.^{3,4} ETA and ETB are serine proteases that cleave desmoglein 1 in the epidermis, disrupting intercellular adhesions and causing blister formation at the subcorneal level.^{2,5} Notably, the mucous membranes are spared.

The major risk factors for SSSS in adults are kidney disease and immunosuppression. ETA and ETB are cleared renally, making patients with impaired kidney function more susceptible.⁴ Immunosuppression due to malignancy, chemotherapy, HIV, and immunosuppressive drugs reduce the formation of antitoxin antibodies, increasing the likelihood of hematogenous spread to produce widespread exfoliation.⁴ Other risk factors for SSSS in adults include IV drug use, chronic alcohol use, diabetes mellitus, cachexia, and rheumatic fever.^{1,6} SSSS should be considered on the differential diagnosis for an immunocompromised patient with a new desquamating rash. Stevens-Johnson syndrome or toxic epidermal necrolysis may also be considered, but SSSS can be readily differentiated from these conditions given the lack of mucosal involvement.²

Some of the clinical characteristics of SSSS in adults differ compared to children. For example, adults are more likely to have *S. aureus* positive blood cultures.⁴ Adults with SSSS are more likely to have several comorbidities, whereas children may be completely healthy before developing SSSS.² Since the disease is more common and thus more easily recognized in the pediatric population, skin biopsy is usually deferred. In adults where the diagnosis is uncertain, a skin biopsy is a valuable part of the work up.

The biopsy technique and transport media influence the likelihood of an accurate result. When SSSS is suspected, a full thickness biopsy of the epidermis such as a punch biopsy should be obtained.⁷ Ideally, two samples should be sent, one in a formalin bottle for hematoxylin eosin staining, and one in Michel's media for immunofluorescence; the latter was not done and not needed for diagnosis in this case.⁷ Intact bullae are typically sterile and a superficial bacterial wound culture swab should be taken from a site of suspected active infection.^{4,7} Blood cultures may be helpful if positive, but a negative result does not preclude the diagnosis of SSSS.⁴ If no source is found on initial work up, primary infections including pneumonia, osteomyelitis, and septic arthritis may also be considered.²

Most toxigenic strains of *S. aureus* can be treated with appropriate narrow spectrum antibiotics.² Especially with local disease, most patients will improve with a course of antibiotics. In a serious case of widespread exfoliation that was unresponsive to antibiotics, IVIG was used with the thought that it may neutralize the exotoxin and successfully treated the patient.⁸ Areas of desquamation should be covered with petroleum impregnated gauze and intact blisters should not be manipulated.⁹

In this case, the patient's skin condition greatly improved after the initiation of antibiotics and wound care. Unfortunately, he passed away several months later due to complications related to his cancer. His malignancy and chronic kidney disease likely made him more susceptible to developing SSSS. Given that this is a rare entity in this age group, involving dermatology in this patient's care early on was important. Clinician awareness of

this condition and interdisciplinary collaboration are vital in cases like this one and may improve patient outcomes.

Conclusion

Staphylococcal scalded skin syndrome is a rare, but potentially life threatening condition in adult patients. Early diagnosis and initiation of treatment has significant clinical and prognostic implications. Clinicians should include SSSS in their differential diagnosis for desquamating lesions in at risk populations and consider consulting dermatology to aid in the work-up.



Figure 1.

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