Unveiling an unusual case of adult-onset Staphylococcal Scalded Skin Syndrome

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Introduction

Staphylococcal Scalded Skin Syndrome (SSSS) is commonly encountered in children and neonates. Its occurrence in adults is exceedingly rare. Most adults have developed antibodies against the exotoxins, which confer immunity against the disease. When SSSS does affect adults, the outcomes can be profoundly severe, with a mortality rate exceeding 60 percent, notably higher than the mortality rate of around four percent in children[1,2].

Adult cases of SSSS typically involve patients with severe renal impairment, causing ineffective clearance of the toxin, or with chronic debilitating diseases and compromised immunity[1,3].

We describe here a case of staphylococcal scalded skin syndrome in an adult male with co-morbidities such as Diabetes Mellitus (DM), hypertension, and Chronic Kidney Disease (CKD).

Case report

A 47-year-old male was referred to us with fever, chills, and an erythematous, non-blanchable maculopapular rash over the flexor aspects of his upper extremities and abdomen, which was tender to touch. The patient had an elevated serum creatinine level of 8.2mg/dl. He reported recent hospital admission during an episode of fever, chills, and respiratory distress for which systemic Vancomycin, Piperacillin, and Tazobactam were administered. The patient did not give any history of addictions.

The patient's medical history initially presented a diagnostic dilemma, with the possibility of the current symptoms being those of either a viral exanthem, drug rash, or red man syndrome. However, upon further probing, it was learned that a blood culture obtained from an intravenous catheter sample at the time of previous admission had yielded a positive result for methicillin-sensitive *Staphylococcus aureus*. This raised clinical suspicion of staphylococcal scalded skin syndrome. Furthermore, there was a notable absence of mucosal involvement, rendering drug rash a less likely diagnosis. Nonetheless, a skin biopsy sample from the affected area was sent with a differential diagnosis of viral exanthem, drug rash, and SSSS.

Being a known case of CKD, the patient was already receiving hemodialysis but was non-compliant with the treatment.

Following the onset of skin lesions, two sessions of hemodialysis were conducted three days apart, with the first session being conducted on the second day of the presentation. He also received systemic cloxacillin. After the first session of hemodialysis, a slight reduction in the erythema of the skin was observed. Following the second session, there was a noticeable and significant improvement, and the progression of erythema was halted. The patient had simultaneously started developing periorificial scaling over the face, which eventually progressed to involve the neck and retro-auricular region, chest, abdomen, back, and upper and lower limbs (Figure 1a and 1b). For the entire course of admission, the patient was prescribed topical white liquid paraffin for symptomatic relief.

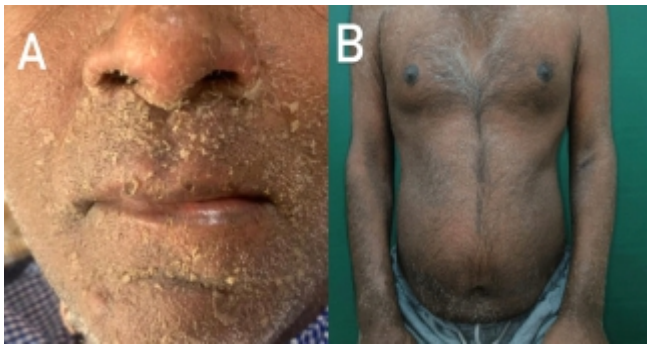

Figure 1:
1a- Clinical photograph demonstrating periorificial scaling. 1b- Subsequent progression of scaling over the chest, abdomen and upper limbs.

Histopathological examination of this patient revealed a sub-corneal split without inflammatory infiltrate, confirming the diagnosis of SSSS.

It is essential to distinguish SSSS from Toxic Epidermal Necrolysis (TEN). In SSSS, the Tzanck smear shows many acantholytic keratinocytes without inflammatory cells, whereas the smear in TEN shows few necrotic keratinocytes along with fibroblasts and inflammatory cells[4]. However, in this case, Tzank smear was not done.

After a three-week follow-up, the patient showed marked improvement with subsidence of lesions and no residual post-inflammatory hyperpigmentation.

Discussion

SSSS is a condition caused by exfoliative toxins (exfoliative toxin A, C and D) produced by certain strains of the staphylococcus species[1,5]. Five Percentage of all phage groups of *Staphylococcus aureus* produce exfoliative toxin. Exfoliative toxin B-producing strains are more commonly isolated from patients with SSSS. Exfoliative toxins are serine proteases and selectively cleave cellular adhesion molecules Desmoglein — I on epidermal Keratinocytes. Exfoliative toxin B is encoded on a large plasmid. In SSSS, exfoliative toxins are speculated to be superantigens. In some patients of SSSS antibodies to Desmoglein may be developed.

It originates following a localized infection from the upper respiratory tract, ear, conjunctiva, or infections related to the umbilical stump in infants. SSSS can be triggered in adults by conditions like abscesses or arteriovenous fistula infections[3].

Clinically, it manifests as typical periorificial scaling and crusting over the face, de-epithelization of friction zones such as flexor folds, and desquamation following initial erythroderma[5].

The resulting loss of skin poses a serious risk to patients, as it can lead to hypothermia due to reduced insulation and fluid and electrolyte imbalance. The compromised skin barrier also increases the susceptibility to infections[3].

The above case represents an unusual occurrence of SSSS in an adult, which manifested following the patient's recovery from a previous episode of Staphylococcus aureus-induced septicemia. It occurred within the context of compromised renal function. Furthermore, the patient's susceptibility to infections was heightened due to his underlying diabetes.

These unconventional cases highlight the considerable challenge clinicians face when recognizing SSSS in adult patients. Its rarity in adults can indeed lead to instances of overlooked diagnoses, emphasizing the need for heightened awareness among healthcare providers.

In this case, the patient demonstrated improvement following hemodialysis, which helped eliminate toxins from the body.

For management, antibiotics targeting Staphylococcus aureus need to be administered. Treatment options for methicillin-sensitive *Staphylococcus aureus* (MSSA) include Cefazolin, Nafcillin, or Oxacillin, while Vancomycin is suitable for suspected Methicillin-Resistant *Staphylococcus aureus* (MRSA). Systemic antibiotics are essential even for localized disease. If a secondary bacterial skin infection is suspected, antibiotics with pseudomonas coverage should be given. Intravenous fluids are necessary for the management of dehydration and/or sepsis. Emollients and non-adherent dressings promote healing and minimize heat loss. Supportive care, including dehydration management, temperature control, and nutrition, is important[3].

Furthermore, it is important to recognize that individuals with chronic kidney disease often have indwelling catheters, which increase the risk of septicemia. To prevent future episodes, the patients should receive comprehensive counseling about catheter care, adherence to hygiene protocols, and the importance of seeking prompt medical attention at the earliest signs of any potential infection, such as persistent fever, pharyngitis, or tonsillitis.

Our patient was further counseled about the nature of the disease and the need for regular check-ups for his CKD along with dialysis.

Although not a common occurrence, the presentation of fever and exanthema, especially in a patient with CKD, should point to a possible differential diagnosis of staphylococcal scalded skin syndrome, irrespective of the age of the patient[6].

Conflict of Interest: Nil

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Ethical consideration

Patient's consent was taken for publication.

Authors' Contribution

SD: Conceptualization, Design, Defining intellectual content, Literature search, Clinical study, Manuscript preparation, editing and review; ST: Design, Defining intellectual content, Literature search, Clinical study, Data acquisition, Manuscript preparation; AH: Design, Defining intellectual content, Literature search, Clinical study, Manuscript preparation, VS: Design, Defining intellectual content, Literature search, Clinical study, Data acquisition, Manuscript preparation and editing

Data availability statement

Data will be available with corresponding author on request.

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