CASE REPORT

A Report of Staphylococcus Scalded Skin Syndrome in Adult

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Summary
Staphylococcal scalded skin syndrome (SSSS) is typically a clinical diagnosis,[1] affecting primarily neonates and children. It is characterised by a diffuse skin disorder with tenderness, erythema, large wrinkled superficial blistering, and desquamation caused by the hematogenous dissemination of exotoxin-producing strains of staphylococcus aureus to the skin.[4,10] Hospital admission is required for intravenous anti-staphylococcal antibiotic therapy and supportive care.

The rarity of SSSS in adults is best explained by the presence of exotoxins neutralizing antibodies and renal elimination of the toxins.[2] Two major risk factors are kidney failure and immunosuppression. Therefore, SSSS in adults warrants thorough evaluation.[3] Mortality is also greater than 60% in adults, attributed to predisposing comorbid conditions.[1,4]

One of the mimickers of SSSS is toxic epidermal necrolysis (TEN). Here, we report a successful treatment of SSSS in an adult with recreational drug abuse and incidental liver cirrhosis possibly secondary to hepatitis C viral infection, after careful exclusion of TEN.

Key Words: *Staphylococcal scalded skin syndrome, Staphylococcus aureus, Toxic epidermal necrolysis, Immunodeficiency, Penicillin*

Introduction
SSSS commonly spread from a nidus of Staphylococcus aureus infection, with exfoliative toxins A and B cleaving desmoglein-1, leading to epidermal granular layer separation. These toxins are secreted by some 5% of Staphylococcus aureus infection and species.[3,4,8,9] Currently, the overall prevalence of SSSS in adults is approximately 1 case per 1 million adults and increasing incidence has been reported worldwide.[5] The true incidence is also possibly underestimated in Asia, where Staphylococcus aureus is an important nosocomial pathogen for developing countries[14].

Adult SSSS is frequently identified in

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individuals who have renal insufficiency and primary inherited defects in the immune system or secondary immunodeficiencies; heroin addiction, HIV, malnutrition with chronic alcohol abuse, glucocorticoids, immunomodulator drugs, malignancy, and 'immuno-paralysis' during severe sepsis.[7,12-13]

Adult SSSS is commonly misdiagnosed as toxic epidermal necrolysis (TEN).[11] Clinical presentation is as in children with striking skin tenderness, pathognomic Nikolsky sign, and typical mucous membranes sparing mark their distinction, usually. Majority of adult cases are complicated with bacteremia, a result of increased severity and the underlying comorbid conditions.[3] The poor prognostic factors are sepsis, electrolyte imbalance, and dehydration.[1,4]

Case Report

A 50-year-old gentleman an electrical labourer, presented with abrupt onset of fever accompanied by the eruption of tiny vesicles over the face that rapidly devolved into bullae with frank pus and peri-oral 'honey-crusted' erosions over the span of 2 days that subsequently became generalised. The flaccid bullae had accentuated epidermal detachment, exposing extremely tender, erythematous raw areas over flexural and decubitus areas. No enanthema was observed.

He reported dysuria and lower back pain 5 days prior and had taken diclofenac sodium (Olfen™) bought over the counter. There were no history of respiratory symptoms, no burn or trauma, and no topical application to the skin.

His past medical history was significant for chronic mechanical lower back pain, receiving regular short courses of piroxicam (Feixicam), dexamethasone (Dexasone), diclofenac (Voren) almost weekly for the past 3 years. His social history recorded active recreational drug use (heroin, morphine), last taken 2 days prior. There was no family history of any blistering skin diseases.

Vital signs on admission recorded blood pressure 100/66 mmHg, heart rate 100 bpm and temperature 36°C. Physical examination revealed skin lesions covering approximately 30% of the body surface area and Nikolsky sign was positive.

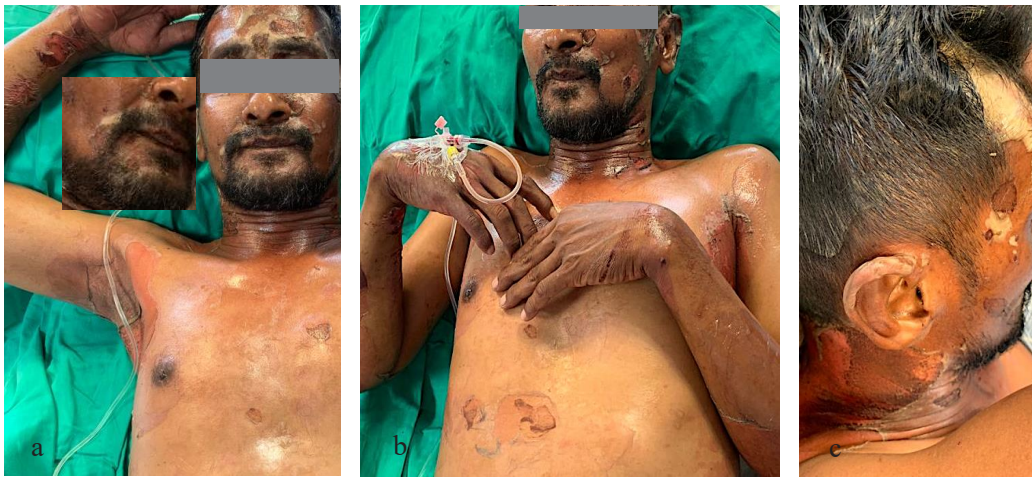
Urinalysis revealed trace leukocyte, negative nitrite, protein 1+ and erythrocyte 3+. Laboratory evaluations showed leukocytosis with left-shifted neutrophils, serum blood urea nitrogen 21.6 mmol/L, serum creatinine 190 micromol/L, serum aspartate aminotransferase 323, serum alanine aminotransferase 88, serum creatinine kinase 417 U/L, C-reactive protein 318 mg/L. Electrolytes were within normal limits. Chest X-ray had heterogeneous opacities over the left lung fields.

At that juncture, the dilemma was on whether this patient had SSSS or drug-induced TEN. However, given the overall clinical picture of urinary tract infection (UTI), pneumonia and sepsis with the lack of mucosal involvement, drug-induced TEN was less favoured. He was diagnosed with SSSS, admitted to the high dependency unit, and intravenous cloxacillin was administered with supportive care.

Microbiologic culture from forehead bullae (pus aspirate) and peripheral blood sent grew methicillin-susceptible *Staphylococcus aureus* (MSSA), resistant to penicillin G. No vegetation was found on echocardiogram. Sputum culture was not available. We had ordered infectious disease screening which was reactive for Hepatitis C Antigen. Ultrasound abdomen revealed multiple hyperechoic liver lesions in the background of liver cirrhosis and features of early bilateral renal parenchymal disease.

These were consistent with the diagnosis of disseminated *Staphylococcus aureus* bacteremia with SSSS. We did not perform staphylococcus aureus phage-typing, as this is not routinely available in our hospital laboratory and a skin biopsy was also unnecessary.

Figure 1 a & b. Superficial pus-filled blisters over face with shallow erosions over the axilla and inner forearm. Inset showed perioral honey crusted lesion. Mucosal surfaces spared. No purpuric macules or targetoid lesions. **c.** Wrinkled superficial blistering extended to nape.



He received intravenous cloxacillin 2 gram 6 hourly and his renal function improved with hydration. He was also given morphine infusion for pain control, emollients and non-adhesive dressings for thermoregulation and to enhance wound healing.

The lesions were superficial and healed without scaring barring areas with previous superimposed bacterial infection after a period of 12 days. No pathogens were isolated on repeated cultures.

Figure 2. Right back lesions healing, re-epithelialization with no significant scarring

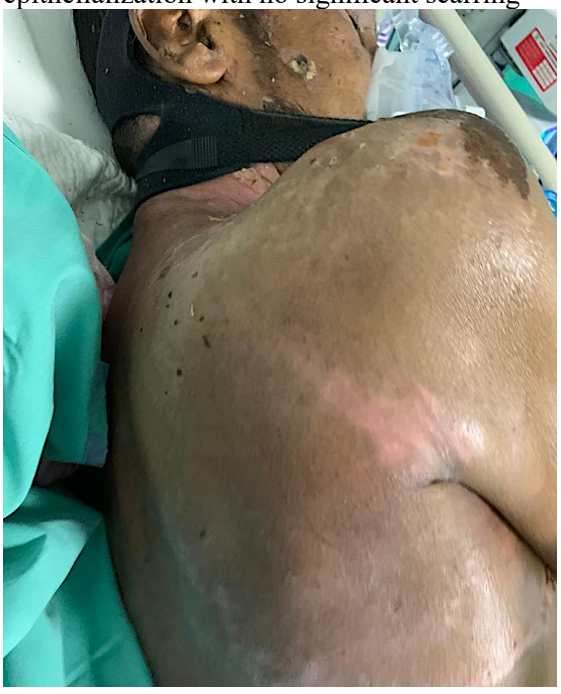


Figure 3. Sheet-like desquamation of the anterior chest wall skin and re-epithelization at axilla

Discussion

Although SSSS is also reported in healthy adults, there clearly appear to be undisputed factors that influence the susceptibility of adult individuals to SSSS. Our patient had recreational drug use, chronic glucocorticoid use, incidental liver cirrhosis and lesions suggestive of hepatocellular carcinoma with possible underlying hepatitis C, which suggest an immunocompromised state. Steroid therapy also significantly increases bacterial burden by facilitating its growth.

He presented with typical features of SSSS in which diffuse tender, erythematous shallow erosions form ruptured flaccid bullae with flexural accentuation and most importantly, without enanthema, thus ruling out the possibility of TEN. Either pneumonia or UTI could be the primary source of infection. Toxins produced could be overwhelming and exceeded what his kidneys could cope with, given the baseline renal parenchymal disease and the severity of illness.

Management of SSSS differs from TEN as systemic glucocorticoid used for TEN is contraindicated irrespective oral or topical administration.[16] Prompt adequate parenteral semisynthetic penicillins are the choice of antimicrobial as *Staphylococcus aureus* strains isolated in SSSS are found to be penicillin-resistant and penicillinase-resistant.[15] Consideration for methicillin-resistant *Staphylococcus aureus* (MRSA) coverage should also be made in endemic areas in which Panton Valentine Leukocidin (PVL) toxin associated *Staphylococcus aureus* is also a particular concern due to its association with more severe infection.

Because the extent of body surface area involved was great, corresponding to severe burns (more than 20%), the supportive management towards the loss of the skin barrier function should be intensified. Patients should remain adequately hydrated and managed like major burn patients; emollients should be applied to eroded skin areas, and antiseptics deployed to impede secondary colonization. We used an antiseptic with broad-spectrum efficacy and most of all, well-tolerated without risk of systemic absorption. Warming blankets were also used to compensate for the change in core body temperature as we had to adhere to strict cool temperature standards in the high dependency unit. Alternating pressure air mattresses are preferred to the seemingly less efficient two-hourly repositioning. This also minimises the risk of mechanical trauma and sleep deprivation.[19]

It is also imperative that pain control in this extremely painful condition is not overlooked as this could potentially cause significant distress to the patients. The World Health Organization pain ladder is used as a guide for the prescription of analgesia and the preferred choice is opiates. The role of non-steroidal anti-inflammatories (NSAIDs) in the development of SSSS remains elusive. However, it should be omitted due to its potential detrimental effect on kidney function, which further complicates the disorder.

Skin lesions heal without significant scarring after prompt and adequate management confirming the diagnosis of SSSS. As desmoglein-1 is mainly expressed in the superficial upper layers of the epidermis, skin lesions usually heal within 14 days without scarring. Mucosal membranes which lack desmoglein-1 are thus spared.[15]

Conclusion

Clinicians should be aware of the clinical spectrum of adult SSSS which is a potentially endemic, life-threatening disorder. Early recognition requires a great deal of clinical finesse for prompt management of this high mortality disease entity. It appears that prudent effort is still required for the prevention and management of SSSS since its first description in 1972 and screening for immediate family members of the diagnosed patients as potential carriers of the ET- A and B producing strains of *Staphylococcus aureus* should be done to prevent outbreaks.

Conflict of Interest Declaration

The authors declare that there is no conflict of interest in this work.

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