

Staphylococcal scalded skin syndrome: A pediatric dermatology case report

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Abstract

Staphylococcal scalded skin syndrome is a condition which predominantly affects children and causes a spectrum of skin lesions. We present a case of a 2-month-old infant with complaints of fever and fragile blisters over the body. The mucosal areas were spared. The diagnosis of staphylococcal scalded skin syndrome was reached on clinical grounds and culture report. The patient responded well to the treatment, which included an antibiotic (cloxacillin), an analgesic (paracetamol), and hydration with intravenous fluids. He was discharged after 8 days, with almost complete resolution of his skin lesions. Having a high clinical suspicion for staphylococcal scalded skin syndrome, early diagnosis/treatment, and following robust hygiene measures are imperative for the effective management of staphylococcal scalded skin syndrome. More efforts are needed to develop novel therapies for staphylococcal scalded skin syndrome.

Keywords

Staphylococcal scalded skin syndrome, desmoglein-1, superficial blisters

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Introduction

Staphylococcal scalded skin syndrome (SSSS) is a rare disorder with clinical features varying from superficial localized blisters to generalized exfoliation.¹ The epidermolytic toxins (ETs) released by *Staphylococcus aureus*, particularly ETA and ETB, are thought to lyse desmoglein-1, present on desmosomes located in the strata granulosum of the epidermis, causing a loss of cell-to-cell adhesion between the keratinocytes, finally leading to intraepidermal splitting.² SSSS predominantly affects neonates of 3–15 days of age, children less than 5 years of life, and adults with various comorbidities.³ We present a case of a 2-month-old infant with SSSS, emphasizing the role of early diagnosis/treatment and discussing the latest developments in the field.

Case report

A 2-month-old infant was presented to the pediatric outpatient department with erythematous blisters near the nose and mouth. The child was irritable and afebrile at that point of time. Systemic examination of the infant appeared unremarkable. The baby was born via normal vaginal home delivery, conducted by a traditional birth attendant of the village. Hence, Apgar scores at birth were not known. The mother was a primigravida and had no history of consumption of any drug

during the past few months, except for iron and folic acid tablets. There was no history in the family of any blistering diseases. The HIV test was negative. Immunizations were complete for his age.

Within 24 h of admission, the infant developed a mild-grade fever; diffuse erythroderma; blisters on the chest, axilla, and gluteal region, which ruptured with minimal pressure; and desquamation of the skin. The lesions encompassed around 20% of his body area. Nikolsky's sign was positive. The mucosa of the mouth and pharynx was not affected. The conjunctiva of both eyes did not show any signs of inflammation. At this point, the infant grew more irritable, refused

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Figure 1. 2-month-old infant with diffuse erythema and fragile blisters on the face, neck, and chest. Povidone-iodine ointment is applied on the blisters.

to feed, and developed tachycardia (Figure 1). A diagnosis of SSSS was reached based on the history and clinical features. A skin biopsy was not requested as the clinical features were consistent with the diagnosis.

An intravenous (IV) catheter was introduced for the infusion of IV fluids. Parental cloxacillin 25 mg/kg/day in divided doses for every 6 h was initiated. Paracetamol was used for countering the fever and pain. The skin lesions were covered with sterile gauze dressings. Povidone-iodine ointment was applied on the blisters on the face (Figure 1). The blood counts were within the normal range. Prior initiating antibiotic therapy, cultures were obtained from the blood, nasopharynx, conjunctivae, umbilical stump, and skin lesions. The nasopharyngeal culture report, as obtained after 2 days, showed the growth of methicillin-sensitive *Staphylococcus aureus* (MSSA). Cultures from other sites were sterile. Meanwhile, the patient seemed to be responding well to the antibiotic as the desquamation had halted. After 8 days of therapy, the erythroderma completely resolved, and the skin lesions were healing with incrustations. The patient was subsequently discharged.

In order to identify the source of the MSSA strain, nasal swabs for microbiological cultures were collected from the parents of the child. However, their culture reports turned out to be negative for MSSA. The traditional birth attendant, who had conducted the delivery, could not be contacted for collecting culture samples. Immunofluorescence and genetic studies, in order to detect ET toxins and ET genes, were not performed due to the lack of material in our hospital setting.

Discussion

SSSS is a rare disease with an incidence between 0.09 and 0.56 cases/ million.⁴ An infection with *Staphylococcus aureus* usually precedes SSSS. *Staphylococcus aureus* releases numerous toxins, which spread hematogenously from the locus of infection. Two ETs, particularly ETA and ETB, have been found to have an affinity toward the glycoprotein, desmoglein-1, present on desmosomes located in the

zona granulosa layer of the skin. ETA and ETB lyse desmoglein-1, thereby destroying the cell-to-cell adhesion between the keratinocytes, leading to epidermolysis.^{1,2}

Neonates and children are at a higher risk for SSSS due to their undeveloped immune system to produce antibodies against the ETs and their inadequate renal capacity to excrete the pathogenic toxins.^{1,3} Similarly, immunocompromised adults or adults with renal diseases show a higher incidence of SSSS.⁴

The clinical features of SSSS comprise a prodromal phase in which there maybe fever and the child may become irritable. This is followed by the appearance of erythematous patches over the body, on which large superficial fragile blisters develop. When these blisters rupture, the skin appears reddish and scalded.^{1,3-5} All of these clinical features were observed in our patient.

Usually, the diagnosis of SSSS is reached clinically with the help of culture reports, as we did in our case. However, if in doubt, diagnosis can be confirmed via skin biopsy, which shows intraepidermal cleavage without necrosis.⁴ Also, phage typing the *Staphylococcus aureus* is found to be useful, as almost 80% of the strains of *Staphylococcus aureus* causing SSSS belong to phage group II.³ Other sparingly used diagnostic tools are techniques measuring the titers of the ETs and isolating their gene sequences.¹

The conditions which we considered in our differential diagnosis were toxic epidermal necrolysis (TEN) and bullous impetigo. A history of drug intake usually precedes TEN. The blistering skin lesions of TEN encompass more than 30% of the body surface area. Also, mucous membranes of the conjunctiva, mouth, trachea, esophagus, anus, and genitalia are involved.^{4,5} These features were absent in our patient. In bullous impetigo, a localized form of SSSS, the blistering lesions are restricted to the area of the skin infection. Also, cultures from the skin lesions produce growth of *Staphylococcus aureus* in bullous impetigo, which was not found in our case.^{4,5} All these points favored the diagnosis of SSSS.

As most strains of *Staphylococcus aureus* causing SSSS are methicillin-sensitive, penicillinase-resistant beta-lactam agents such as cloxacillin, dicloxacillin, oxacillin, flucloxacillin, and nafcillin are the first-line antibiotics. If the patient is not responding to these agents, then methicillin-resistant strains of *Staphylococcus aureus* (MRSA) should be suspected, for which vancomycin is the drug of choice. Topical therapy should constitute either fusidic acid and/or mupirocin as adjunct therapy at the site of blisters in an attempt to eradicate colonization. Exposed, damaged areas can be treated with emollients which sooth and moisturize the skin. Other important aspects to be addressed in the management of SSSS are temperature regulation, fluid resuscitation, analgesia, sterile dressing of the lesions, and prevention of secondary infections. Paracetamol is the analgesic of choice in cases of SSSS. Corticosteroids are contraindicated as they worsen the disease. With early diagnosis and management, mortality rate of SSSS is lower

than 4% in children, and most skin lesions resolve by 2 weeks, as found in our patient.^{1,3–6}

Healthcare attendants and mothers, serving as asymptomatic carriers of *Staphylococcus aureus*, have been reported to be the source of several outbreaks of SSSS in pediatric units. Also, healthcare professionals, handling patients with SSSS, have been found to cross-contaminate other patients admitted in the unit.⁷ Hence, following robust hygiene measures, while treating SSSS patients and identifying/isolating suspected carriers at the earliest, is recommended to avert such outbreaks.

With the rise of MRSA strains and increase in the mortality rate of SSSS, some newer therapies have been investigated by researchers. Infusion of fresh frozen plasma (FFP) obtained from adults into children with SSSS has been found to be successful in a pediatric report. The use of FFP in SSSS is based on the buttress that adults more than 40 years develop antibodies against the causative ETs of SSSS.⁸ Also, infusion of IV immunoglobulins has been suggested to antagonize the ETs causing SSSS.⁹ Use of laxatives in order to excrete the pathogenic toxins of *Staphylococcus aureus* has been suggested in one report.¹⁰ There are reports suggesting better patient outcomes following the use of artificial skin substitutes over conventional sterile gauze dressings.¹¹ However, there have been no clinical trials conducted to authenticate the abovementioned therapies. Although, theoretically developing desmoglein-1 antitoxins/analogues to antagonize the ETs causing SSSS should show superior results, no clinical or experimental studies exploring this therapy are available in the literature.^{1,12} Vaccines targeting *Staphylococcus aureus* have failed in the Phase III of clinical trials and face considerable hurdles in their development.¹³

Conclusion

In conclusion, although most cases of SSSS respond well to conventional therapy, it is a potentially fatal condition. Hence, early diagnosis, prompt treatment, and following robust hygiene measures are imperative for its successful management. More efforts are required to develop novel effective therapies for SSSS.

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Ethical approval

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Informed consent

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