



A case report on Staphylococcal Scalded Skin Syndrome in a 3 year old: Clinical diagnosis and management

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ABSTRACT

Staphylococcal-scalded skin syndrome (SSSS) is a blistering skin disorder which is considered as a pediatric emergency. This life-threatening syndrome is caused by Staphylococcal exfoliative toxins produced by *Staphylococcus aureus*. Although it can affect any age group, children below 5 years of age are at utmost risk. The specific antibodies to exotoxins and increased exotoxin clearance, minimize the frequency of SSSS in adults. Mortality in infancy is low (4%) and is less than 10 % in children but is between 40% and 63% in adults. Clinical features of SSSS may range from a few localized blisters to generalized desquamation covering the entire body. The histopathological evaluation of the skin biopsy helps in the definitive diagnosis of SSSS. Here we present a case of a 3 year old female child with low grade fever associated with erythematous rashes all over the body and severe itching. A clinical diagnosis of Staphylococcal Scalded Skin Syndrome (SSSS) was made. With appropriate treatment using intravenous Flucloxacillin and supportive care, the child's symptoms were completely resolved within a week.



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but has also been described in adults (Handler and Schwartz, 2014). Mortality is low in infancy (4%), less than 10% in children, but higher in adults (between 40% and 63%), despite antibacterial therapy. Inadequate renal clearance and diminished immunity against bacterial exotoxins increases the chance of SSSS in children below 5 years of age. It's severity could range from localized blisters to generalized desquamation affecting the entire body (Amagai et al., 2000). Here we discuss a case of a 3 year child with SSSS, highlighting the importance of early diagnosis and it's treatment.

INTRODUCTION

Staphylococcal-scalded skin syndrome (SSSS), also known by the name of Ritter disease, is a superficially occurring blistering skin disorder. It is mainly caused by the exfoliative toxins of certain strains of *Staphylococcus aureus* (Leung et al., 2018). The disease especially affects infants and small children,

CASE REPORT

A 3 year old female child previously asymptomatic was brought to the pediatric department with a low grade fever associated with erythematous rashes since 1 week and severe itching that had lasted for 4 days. The baby was born through normal vaginal delivery. There was no history of joint swelling or

altered sensorium in the child. The mother had a history of atopic dry skin. Immunizations were complete for the child's age. The child was initially managed symptomatically at home. As the erythematous rashes persisted, the child was taken to the local hospital and was started on Ceftriaxone for 2 days.

At admission the child was febrile with temperature of 101.1°F. Examination showed erythematous rashes all over the body, scaling over the forehead, around the nose, mouth, retro auricular region, neck and upper back. Rest of the systemic examinations were within the normal limits. Laboratory investigation showed eosinophilia (18.1%) with negative inflammatory markers. Hepatic and renal functions were within normal limits. CVS examination also showed normal S1, S2 and no murmur. Blood cultures were stable. The HIV test was negative.

SSSS was diagnosed based on clinical findings. Biopsy of the skin was not done as the clinical features were found to be consistent with the diagnosis.

TREATMENT

A prompt therapy was then initiated with Parental Flucloxacillin 50 mg/kg/day in divided doses for every 6 h. Paracetamol was given for encountering the pain and fever associated with SSSS. Syrup Hydroxyzine was also provided for the management of the skin lesions. Supportive care including management of dehydration, temperature regulation and nutrition were given. The child responded to the treatment with parenteral Flucloxacillin which was then switched to oral Dicloxacillin 250mg qid after 3 days.

DISCUSSION

Staphylococcal scalded skin syndrome is a rare staphylococcal toxin-mediated dermatitis caused by exfoliative toxins A and B, which are capable of cleaving desmoglein 1, a glycoprotein responsible for keratinocyte-to-keratinocyte adhesion in the superficial epidermis of the skin. Cleavage of this glycoprotein leads to skin fragility and formation of superficial blisters and erosions (Amagai *et al.*, 2000).

SSSS with an estimated incidence rate of 0.09 to 0.56 case/million is considered as one of the major dermatological condition. Early and proper clinical diagnosis is the mainstay for prevention of progression of this disease. It's symptoms starts with fever, irritability, known as the prodromal phase along with widespread redness of the skin, and malaise. Red, tender and painful areas around the infection

site, dehydration and weakness are some of the other symptoms. Within 24 to 48 hours, fluid-filled blisters form with rapid progression to superficial blister formation (positive Nikolsky's sign), which is a classic feature for diagnosis.

Blood cultures, as presented in our case, typically do not help in the definitive diagnosis of SSSS, as they are often negative in children contrary to it being generally positive in adults. A secondary infection (sepsis, pneumonia and cellulitis) along with dehydration and consequential electrolyte imbalances can complicate the patient's condition (Handler and Schwartz, 2014; Farroha *et al.*, 2012). SSSS begin abruptly after an incubation period of 1 to 10 days. With prompt treatment, healing may occur within 10-14 days and recover with no problems or skin scarring (Aydin and Alsbjörn, 2016).

Although SSSS is relatively uncommon, it is easily diagnosed on clinical grounds, such as signs of bullae, tender erythroderma, and desquamation with a scalded appearance. Differentiation of SSSS from other exfoliating diseases such as Stevens- Johnson syndrome (SJS) and Toxic epidermal necrolysis (TEN) is mainly done by assessing mucosal involvement, which is absent in former. Furthermore, SSSS demonstrates a subcorneal blister usually with scant inflammation where TEN shows a dermal-epidermal blister with focal dyskeratosis and areas of full thickness with epidermal necrosis (Harr and French, 2010).

The management of SSSS involves eradication of the causative staphylococcal infection through appropriate antibiotics and supportive care to promote healing, reduce discomfort, and minimize complications. Prompt empirical therapy with intravenous anti-staphylococcal antibiotic such as Penicillinase-resistant synthetic penicillins like nafcillin, flucloxacillin or oxacillin, is necessary until cultures are available to guide therapy for methicillin-sensitive *Staphylococcus aureus* (MSSA). Antibiotics with broad MRSA coverage, such as Vancomycin or Linezolid, should be considered as the drug of choice to help fight infection in areas with significant MRSA prevalence (or if SSSS suspected of MRSA infection) (Braunstein *et al.*, 2014).

Some clinicians routinely add clindamycin as an adjunct to a penicillinase-resistant penicillin or cephalosporin based upon the theory that clindamycin may decrease ribosomal production of the pathogenic staphylococcal exotoxins. However it is not recommended as a primary treatment because of high rates of clindamycin resistance in SSSS (Braunstein *et al.*, 2014; Schlievert and Kelly, 1984; Ladhani, 2001).

Moreover, certain case reports have described rapid improvement following the addition of intravenous immune globulin or plasma exchange in patients with severe disease; however, data are insufficient to confirm efficacy of these therapies (Meshram *et al.*, 2018). Although theoretically, antagonizing activity of desmoglein-1 antitoxins against ETs causing SSSS show superior results, no experimental or clinical studies regarding this therapy are available (Gaddam and Thirunagari, 2019). Also, vaccines targeting *S.aureus* could not succeed in the Phase III of clinical trials and are still in the developmental phase.

Here early diagnosis and prompt treatment following aseptic measures were the mainstay for its effective management. Moreover, the prevalence of SSSS tends to increase over time, which is related to a number of socio-demographic factors and other infections. Therefore further studies are needed to confirm these findings and to minimize rising rates of SSSS (Staiman *et al.*, 2018).

CONCLUSIONS

The case of exfoliative toxin mediated SSSS had a good resolution and no sequelae because of appropriate case management. Therefore, early detection of this clinical disorder, adequate hygiene measures and prompt treatment spearheaded by a multidisciplinary medical team are imperative for a good prognosis in children. More efforts are required to develop novel effective SSSS therapies for SSSS to help enhance prognosis, decrease length of hospital stay, and improve quality of life.

Conflict of Interest

The authors declare that there is no conflict of interests for this study.

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