Recurrent Staphylococcal Scalded Skin Syndrome in a 20 month old – A case report

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Abstract: We present a case of a 20 month old child with a history of atopic dermatitis that exhibited recurrent erythematous-bullous lesions consistent with Staphylococcal Scalded Skin syndrome (SSSS). SSSS is an exfoliative toxin mediated skin disorder most commonly found in children. In this paper, we discuss the importance of recognizing the clinical symptomatology and progressive nature of SSSS, particularly in patients with a history of atopic dermatitis, to assure prompt treatment and resolution of the syndrome.

Main Text:

Background: Staphylococcal Scalded Skin Syndrome (SSSS) is an exfoliative skin disease that usually occurs due to a toxin produced by a *Staphylococcal aureus* infection [1]. Toxin a or b from *S. aureus* spread hematogenously and have been shown to lyse the cell-cell adhesion molecule desmoglein-1 in the superficial epidermis, causing intraepidermal splitting and thus a loss of structural integrity [2]. SSSS primarily occurs in children and neonates and is rare in the adult population [3, 4, 5] We present a case of a 20 month old patient who presented with progressive SSSS and recurrence of SSSS to emphasize the role of early treatment and diagnosis in reducing morbidity of the disease.

Case Presentation

Initial Admission:

Day 1: A 19 month old African American male presented to the ED with erythematous patches and blistering on face and neck as well as erythematous patches on buttocks (Figure 1). Patient had a history of atopic

dermatitis mostly affecting bilateral antecubital fossa. The patient had no new medications prior to this event and was up to date on relevant immunizations.

Per caregivers who were present at ED, the patient was also irritable, had reduced appetite, and appeared fatigued 3-5 days before presentation. The patient also experienced two days of afebrile dry cough, rhinorrhea, and sinus congestion, increased scratching of the antecubital fossa due to his atopic dermatitis. Upon rubbing of the perinasal and perioral area in the ED, the uppermost layer of skin sloughed off with minimal pressure (positive Nikolsky's sign). On physical exam, erythema and edema were present. The mucous membranes of the mouth and pharynx were not affected. The patient was apyretic. The patient's initial WBC was 8.3 x 10ˆ9 /L. Further investigation with blood and ear wound cultures were obtained. A diagnosis of SSSS was made based on history and clinical features. The diagnoses of bullous impetigo and TEN were also considered, but the lack of mucous membrane involvement in our patient made SSSS more likely. A skin biopsy was not performed as the clinical presentation was consistent with SSSS.

The patient received IV nafcillin 40mg/mL every 6 hours after admission as well as intravenous fluids. For pain control, the patient was given ibuprofen and Tylenol as needed. The patient's wounds were covered with sterile bandages.

Days 2-3: After discussion with the caregivers, on day 2 the patient was started on IV clindamycin 30 mg/kg/day to target the exfoliative toxin of *S. aureus.* After 48 hours, the results of the blood and ear wound cultures were negative and the urine analysis was clear. The patient's pain was controlled. However, the patient's caregivers noticed increased periorbital edema without drainage on day 2. This was thought to be due to third spacing in the setting of inflammation and fluids were decreased to KVO.

Of note, the patient's WBC count of 5.0 X 10ˆ9 /L (ANC 0.9) on hospital day 3 was thought to be caused by IV nafcillin. Repeat CBC within 24 hours showed WBC count 5.1 (ANC 1.2). On day 3, periorbital edema was decreased and skin lesions were in the process of healing with crusting (Figure 2). The patient was taking adequate oral intake, and the IV antibiotics were transitioned to oral cephalexin and clindamycin was discontinued. The patient was discharged on appropriate 7 day course of oral cephalexin, topical mupirocin and close outpatient pediatric follow-up.

Readmission, 3 weeks later: Subsequently, the now 20 month old patient presented to the ED with erythematous patches on face, neck, hyperpigmentation on buttocks consistent with SSSS. Patient's caregivers stated that patient's skin was clear for 2 weeks after initial admission and treatment, and the patient continued to scratch antecubital fossa at night due to atopic dermatitis. The patient's mother noticed an erythematous patch behind the ear which had worsened for the past two days, so she brought the patient to the ED. At readmission, there was also no involvement of mucosal membranes and thus no concern for SJS. CBC and CMP were obtained in the ED and were in the normal range except for a low Na, 133 mmol/L. Blood cultures were obtained showing no growth after 48 hours; wound cultures identified no organisms. Initially the patient was given IV nafcillin for MSSA coverage because during the prior admission this treatment healed the rash.

After discussion with the Pediatric Infectious Disease team, on day 2 the patient was switched from IV nafcillin to a ten day course of PO Linezolid. The Pediatrics ID team was concerned that, due to the rapid recurrence of SSSS, this patient may be experiencing a rare case of MRSA caused SSSS. Thus, the coverage needed to be broadened.

On day 3, the patient had fewer new lesions and previous lesions appeared to be healing with scabbing and hyperpigmentation. He was afebrile during his entire hospital stay. The patient was also well appearing with adequate PO intake during the hospital stay and was discharged on day 3 with close outpatient pediatric follow up. The patient's mother was appropriately concerned for immune deficiency given the recurrence of SSSS. Pediatrics Infectious Disease commented that his readmission was more likely due to MRSA rather than MSSA and that is the reason that SSSS recurred. The patient was also otherwise a normally developing child with excellent growth. The patient's mother was aware that she needed to return should symptoms worsen or reoccur.

Discussion:

In this case report, the prompt diagnosis and treatment of SSSS during the first ED admission allowed the patient to heal, and the subsequent readmission with broadened antibiotic coverage allowed for timely discharge and effective treatment.

SSSS, or Ritter's disease, is a rare disease process that typically occurs in children. The preceding *S. aureus* infection spreads in the bloodstream and lyses the desmoglein-1 proteins in the stratum granulosum layer of the epidermis [1, 2, 6]. This separation of the anchoring desmosomes leads to the exfoliative presentation of SSSS. SSSS typically arises from an area of infection, such as impetigo, bacterial conjunctivitis, or iatrogenic wounds [7]. The susceptibility of children to acquisition of SSSS has been postulated to be a result of the lack of protective antibodies *S. aureus* toxins and/or the insufficient excretion of exotoxins from children's kidneys [1].

The diagnosis of SSSS is mainly clinical. Prodromal symptoms of irritability, skin pain, fever, and poor feeding are common. Cutaneous erythema may initially present in the skin folds of the neck, axillae, gluteal folds before becoming generalized within the first 48 hours of presentation [7]. On exam, the skin may have flaccid bullae that erode upon minimal pressure. Additionally, superficial desquamation occurs due to the exotoxin's lytic mechanism [6]. There is typically quick progression of the disease process.

The standard treatment of SSSS includes IV antibiotic against staphlycoccal species (eg. Nafcillin, oxacillin) and supportive care [6, 7, 8, 9]. Empiric treatment with penicillinase-resistant penicillins is done initially, with broadening for MRSA coverage considered when clinical improvement is not observed or in communities with high MRSA incidence [10]. In our case, MRSA coverage was added (PO linezolid) after initial clearing and improvement was followed by a recurrence of SSSS. Clarithromycin can be given to patients with a penicillin allergy [8]. Other therapies such as IV immunoglobulin have been suggested to antagonize the exfoliative toxins of SSSS [11]. Patients may continue to have skin pain following treatment as well as post-inflammatory hypopigmentation or hyperpigmentation during the healing process [6]. SSSS carries a risk of progression to sepsis if not recognized and treated early. Other complications include secondary infection, dehydration, electrolyte imbalance, and death [5, 12]. Recurrence is rare but can occur in young infants and neonates [13].

The differential diagnosis for SSSS includes TEN (Toxic Epidermal Necrolysis) and SJS (Steven's Johnson Syndrome) [5]. Both TEN and SJS have mucous membrane as well cutaneous involvement, which was not present here. Bullous impetigo is also on the differential diagnosis, but bullous impetigo has localized skin infection rather than hematogenous spread. The former has more limited involvement and a less severe clinical presentation [6].

Conclusion:

This case report highlights the importance of prompt evaluation and antibiotic treatment as well as the consideration of MRSA in SSSS. More research is necessary to elucidate the prevalence and resistant staphylococcal strains and therapies for SSSS caused by MRSA. This case report aims to further support research in exfoliative skin conditions.

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Figure Legends:

Figure 1: Day 1 of first admission showing facial and bilateral periorbital edema with diffuse erythematous patches with overlying hemorrhagic crust.

Figure 2: Day 3 of first admission showing resolving facial and bilateral periorbital edema and improving erythematous patches.