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Case Report: Recurrent Staphylococcal Scalded Skin Syndrome in Healthy Term Neonate Despite Full Course of Antibiotic Therapy

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Abstract

Background: Staphylococcal scalded skin syndrome (SSSS) describes a blistering skin infection caused by the exfoliative toxin in the bacterium *Staphylococcus aureus*. It more commonly affects the infant population and is characterized by large blistering bullae that rupture upon application of pressure.

Case description: We describe a case of recurrent SSSS in a healthy term neonate who initially presented with a perioral rash on day of life (DOL) 11 that quickly became vesicular with new lesions on the sternum and extremities. The patient's rash began to resolve upon administration of culture-specific IV antibiotics. She was appropriately treated with a 14-day course and was discharged home.

The patient returned on DOL 35 with a perioral rash and generalized reddening of the skin. She was admitted and placed empirically on nafcillin, clindamycin, and vancomycin for concerning recurrence of SSSS. At this time, consults were placed to dermatology, as well as allergy & immunology for possible epidermolysis bullosa and immune deficiency. Skin biopsy revealed development of recurrent SSSS. Patient finished another 14-day course of IV antibiotics and was discharged home with resolution of the rash.

Conclusion: This report discusses a case of recurrent SSSS in a term neonate who received two full courses of antibiotics and has since fully recovered.

Key words: Staphylococcal scalded skin syndrome, epidermolysis bullosa, disease recurrence

Introduction

Staphylococcal Scalded Skin Syndrome (SSSS) is a clinical term that describes a host of blistering infections caused by the exfoliative toxin (ET) of the bacterium *Staphylococcus aureus* (SA). The prevalence of SSSS is greater in the pediatric population. It commonly presents between the first 2 weeks of life as large bullae that rupture upon application of pressure (Nikolsky sign). (1) This case report outlines the presentation and treatment course of a patient with recurrent SSSS within the first 5 weeks of life. We discuss the unusualness of a recurrence and our concerns regarding a possible inherited connective tissue disease. This case of recurrent SSSS is unique in that similar cases have not been commonly documented in the literature. Our case will provide physicians with a blueprint for analysis of recurrent SSSS including an exhaustive differential and possible treatment options.

Case Report

The patient was a baby girl born via normal vaginal delivery at 37-4/7 weeks to a 31-year old mother who consented to the writing of this case report at a routine clinic visit after discharge. Mother was Group B *Streptococcus* (GBS) unknown at the time of delivery, GBS positive in a previous pregnancy, and was appropriately treated with penicillin. APGARs were 9 and 9 at 1 and 5 minutes. The patient was taken to the newborn nursery and was discharged home on day of life (DOL) 2. There were no clinical signs of SSSS at this time.

On DOL 11, the patient was brought to the Emergency Department (ED) with a perioral rash that had begun as chapped lips the day before. The patient's lips appeared red and raw, and a vesicular lesion was noted at the philtrum. There were no skin lesions in the periumbilical and perineal regions, and the extremities were clear. Her parents reported that she was fussy and was taking less than usual during feedings. Family history was not significant for any skin diseases. A CBC was ordered, which showed an elevated WBC of 26,000 (normal 5,000-20,000). A BMP was also ordered and showed no signs of dehydration. Of note, the patient remained afebrile. At this time, the pediatrics team was consulted to assess the patient and arrangements for admission were made after denuded skin was noticed at the IV site.

The patient was admitted to rule-out sepsis and started empirically on vancomycin, acyclovir, and cefotaxime. The patient underwent a lumbar puncture for diagnostic aspiration of fluid. Upon completion of the procedure, the patient developed a blister-like lesion at a point of pressure and skin sloughing upon removal of the sterile drape. She developed similar lesions in the genital region after placement of a urinary catheter and on her right lower extremity after removal of tape. The vesicular culture grew methicillin-sensitive SA (MSSA). Blood cultures and CSF cultures were negative, and antibiotics were appropriately de-escalated from both antibacterial and antiviral coverage to solely antibacterial coverage based on culture results and the hospital's antibiogram. The patient was placed on nafcillin, clindamycin, and ceftriaxone. Nafcillin was continued for seven days and subsequently discontinued. Culture data showed the organism was resistant to clindamycin. However, this was felt to be a macrolide-induced resistance and clindamycin was continued as it likely retained some activity because it works at the ribosomal level to diminish toxin production. The rash resolved, the patient completed a full 14-day course of IV antibiotics as described above, and the patient was discharged home after the remainder of the hospital stay was uneventful.

On DOL 35, the patient returned to the ED with a 1-day history of facial blistering that had worsened overnight. In the interim, she had been seen by the outpatient pediatric clinic for a well child visit and was found to be doing well. She also had desquamation of the left upper extremity. She was afebrile with a WBC within normal limits. She was again admitted to the pediatrics service and was empirically started on vancomycin, clindamycin, and nafcillin for possible recurrent SSSS. Dermatology and immunology services were consulted for concerns regarding recurrent SSSS, possible congenital connective tissue disorder such as epidermolysis bullosa, and possible immune deficiency. Patient underwent a skin biopsy and immunological tests were ordered. Blood cultures were negative after 48 hours and vancomycin was stopped. Flow cytometry was performed after recommendation of Allergy and Immunology. Results were unremarkable and showed normal CD4 and CD8 counts. The biopsy revealed recurrent SSSS. The patient remained in the hospital to receive a full 14-day course of IV antibiotics and was discharged home with resolution of rash. She has been seen in clinic consistently for routine well-child checks since discharge with no noticed relapse of SSSS.

Discussion

SSSS can present with varying severity depending on the route of transmission of the ET to the skin. For patients whose total body surface area involvement is extensive, secondary infections, excessive fluid losses, and poor temperature regulation become concerns. Outbreaks of SSSS

involving neonatal and pediatric hospital wards are not uncommon, and therefore necessitate swift intervention and treatment of the disease. SSSS has a 4-10% mortality rate in the pediatric population, 50% in the adult population 100% in patients with underlying disease, despite the standard of care antibiotic treatment. (2,7)

The diagnosis of SSSS is made on clinical grounds and is verified by culture of ET producing *S. aureus* and skin biopsy. There are four types of ET that have been identified: ETA, ETB, ETC, and ETD. ETA, which was shown to be the cause of SSSS in our patient, is a serine protease. It cleaves cell-cell adhesion at the desoglein-1 molecule causing blister formation. (3) Because SSSS is readily susceptible to appropriate antibiotic treatment, we were inclined to broaden our differential for this patient upon her second admission. Of note, our literature review did not reveal any cases of recurrent SSSS in a baby born at term highlighting the importance of this case for provider reference in similar situations. (4) Recurrence of SSSS has been documented in ward outbreaks in which proper cleaning and maintenance of equipment was not performed. (1,2) It has also been documented in patients who have an underlying immunodeficiency and premature infants, but not in any term neonate without an underlying disease. (1,5,9) Interestingly, a recent 8-year retrospective study published in *Pediatric Dermatology* regarding SSSS epidemiology did not reveal a single case of recurrent disease. (8) Because recurrence is rare, it would be imperative to include bullous impetigo, epidermolysis bullosa, toxic epidermal necrolysis, neonatal pemphigus, and bullous mastocytosis as possible diagnoses. (5)

The importance of broadening the differential is evidenced by the differences in treatment protocols and the long-term prognoses of the various disease states. For example, clinical presentations of epidermolysis bullosa can range from mild to extremely severe, with patients experiencing blistering of skin at even the slightest points of contact. This condition can be lethal to the patient. A patient with bullous mastocytosis can be treated with corticosteroids and recover satisfactorily. In the case of SSSS, a 14-day course of antibiotics will usually cure the patient and prevent a recurrence. Proper hygiene and decolonization should also be discussed with both the patient and family. This should include use of mupirocin ointment and hibiclens baths. (6)

The broad range of potential diagnoses reminds us that a multidisciplinary approach to patient care can be beneficial to both the patient and primary team. In our case, consult services allowed us to exhaust our differential, arrive at the correct diagnosis, and treat the patient properly. We believe that our multidisciplinary approach is a great strength of this case report as it allowed us to broaden our differential in order to provide more comprehensive care to our patient. While a recurrent case of SSSS was indeed unusual, it was found to be true in the case of our patient.

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