Newborn with desquamating rash

The patient's age and the appearance of the lesions pointed us to the proper diagnosis in this case.

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A 9-DAY-OLD BOY was brought to the emergency department by his mother. The infant had been doing well until his most recent diaper change when his mother noticed a rash around the umbilicus (**FIGURE**), genitalia, and anus.

The infant was born at term via spontaneous vaginal delivery. The pregnancy was uncomplicated; the infant's mother was group B strep negative. Following a routine postpartum course, the infant underwent an elective circumcision before hospital discharge on his second day of life. There were no interval reports of irritability, poor feeding, fevers, vomiting, or changes in urine or stool output.

The mother denied any recent unusual exposures, sick contacts, or travel. However, upon further questioning, the mother noted that she herself had several small open wounds on the torso that she attributed to untreated methicillin-resistant *Staphylococcus aureus* (MRSA).

On physical examination, the infant was overall well-appearing and was breastfeeding vigorously without respiratory distress or cyanosis. He was afebrile with normal vital signs. The majority of the physical examination was normal; however, there was erythematous desquamation around the umbilical stump and genitalia with no vesicles noted. The umbilical stump had a small amount of purulent drainage and necrosis centrally. The infant had a 1-cm round, peeling lesion on the left temple (**FIGURE**) with a small amount of dried serosanguinous drainage and similar superficial peeling lesions at the left preauricular area and anterior chest. There was no underlying fluctuance and only minimal surrounding erythema.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE
Peeling skin on temple and chest with erythematous rash at umbilical stump and genitalia

IMAGES COURTESY OF: NATALIYA HOLMES, MD

Staphylococcal scalded skin syndrome should be considered a pediatric emergency due to potential complications such as sepsis.

Diagnosis: Staphylococcal scalded skin syndrome

Based on the age of the patient, clinical presentation, and suspected maternal MRSA infection (with possible transmission to the infant), we diagnosed staphylococcal scalded skin syndrome (SSSS) in this patient. SSSS is rare, with annual incidence of 45 cases per million US infants under the age of 2.[1] Newborns with a generalized form of SSSS commonly present with fever, poor feeding, irritability, and lethargy. This is followed by a generalized erythematous rash that initially may appear on the head and neck and spread to the rest of the body. Large, fragile blisters subsequently appear. These blisters rupture on gentle pressure, which is known as a positive Nikolsky sign. Ultimately, large sheets of skin easily slough off, leaving raw, denuded skin.[2]

S aureus is not part of normal skin flora, yet it is found on the skin and mucous membranes of 19% to 55% of healthy adults and children.[3] *S aureus* can cause a wide range of infections ranging from abscesses to cellulitis; SSSS is caused by hematogenous spread of *S aureus* exfoliative toxin. Newborns and immunocompromised patients are particularly susceptible.

Neonatal patients with SSSS most commonly present at 3 to 16 days of age.[2] The lack of antitoxin antibody in neonates allows the toxin to reach the epidermis where it acts locally to produce the characteristic fragile skin lesions that often rupture prior to clinical presentation.[2,4] During progression of the disease, flaky skin desquamation will occur as the lesions heal.

A retrospective review of 39 cases of SSSS identified pneumonia as the most frequent complication, occurring in 74.4% of the cases.[5] The mortality rate of SSSS is up to 5%, and is associated with sepsis, superinfection, electrolyte imbalances, and extensive skin involvement.[2,6]

If SSSS is suspected, obtain cultures from the blood, urine, eyes, nose, throat, and skin lesions to identify the primary focus of infection.[7] However, the retrospective review of 39 cases (noted above) found a positive rate of *S aureus* isolation of only 23.5%.[5] Physicians will often have to make a diagnosis based on clinical presentation and empirically initiate broad-spectrum antibiotics while considering alternative diagnoses.

A clinical diagnosis with a large differential

While biopsy rarely is required, it may be helpful to distinguish SSSS from other entities in the differential diagnosis (**TABLE**[2,3,7-13]).

Toxic epidermal necrolysis (TEN) is a rare and life-threatening desquamating disease nearly always caused by a reaction to medications, including antibiotics. TEN can occur at any age. Fever, diffuse erythema, and extensive epidermal involvement (>30% of skin) differentiate TEN from Stevens-Johnson syndrome (SJS), which affects less than 10% of the epidermis. It is worth mentioning that TEN and SJS are now considered to be a spectrum of one disease, and an overlap syndrome has been described with 10% to 30% of skin affected.[8] Diagnosis is made clinically, although skin biopsy routinely is performed.[7,9]

Congenital syphilis features a red or pink maculopapular rash followed by desquamation. Lesions are more common on the soles.[10] Desquamation or ulcerative skin lesions should be examined for spirochetes.[11] A quantitative, nontreponemal test such as the rapid plasma reagin (RPR) or the Venereal Disease Research Laboratory (VDRL) will be positive in most infants if exposed through the placenta, but antibodies will disappear in uninfected infants by 6 months of age.[8]

Congenital cutaneous candidiasis presents with a generalized eruption of erythematous macules, papules, and/or pustules with widespread desquamating and/or erosive dermatitis. Premature neonates with extremely low birth weight are at higher risk.[13] Diagnosis is confirmed on microscopy by the presence of *Candida albicans* spores in skin scrapings.[13]

Neonatal herpes simplex virus (HSV) symptoms typically appear between 1 and 3 weeks of life, with 60% to 70% of cases presenting with classic clustering vesicles on an erythematous base.[14] Diagnosis is made with HSV viral culture or polymerase chain reaction (PCR).

SSSS should be considered a pediatric emergency

SSSS should be considered a pediatric emergency due to potential complications. Core

TABLE

Differential Dx of desquamating lesions in neonates[2,3,7-13]

Condition	Characteristics
Congenital cutaneous candidiasis	Generalized eruption of erythematous macules, papules, and/or pustules with widespread desquamating and/or erosive dermatitis
Congenital syphilis	Red or pink maculopapular rash (more commonly on the soles), followed by desquamation
Neonatal HSV infection	Clustering vesicles on an erythematous base that subsequently ulcerate
SSSS	Macular erythema followed by diffuse epidermal exfoliation
Toxic epidermal necrolysis	Fever, diffuse erythema, extensive epidermal involvement of any part of the skin

HSV, herpes simplex virus; SSSS, staphylococcal scalded skin syndrome.

measures of SSSS treatment include immediate administration of intravenous (IV) antibiotics. US population studies suggest clindamycin and penicillinase-resistant penicillin as empiric therapy.[15] However, local strains and resistance patterns, including the prevalence of MRSA, as well as age, comorbidities, and severity of illness should influence antibiotic selection.

IV nafcillin or oxacillin may be used with pediatric dosing of 150 mg/kg daily divided every 6 hours for methicillin-sensitive *Staphylococcus aureus* (MSSA). For suspected MRSA, IV vancomycin should be considered, with an infant dose of 40 to 60 mg/kg daily divided every 6 hours.[16] Fluid, electrolyte, and nutritional management should be addressed immediately. Ongoing fluid losses due to exfoliated skin must be replaced, and skin care to desquamated areas also should be addressed urgently.

Our patient. Phone consultation with an infectious disease specialist at a local children's hospital resulted in a recommendation to treat for sepsis empirically with IV vancomycin, cefotaxime, and acyclovir. Acyclovir was discontinued once the HSV PCR came back negative. The antibiotic coverage was narrowed to IV ampicillin 50 mg/kg every 8 hours when cerebrospinal fluid and blood cultures returned negative at 48 hours, wound culture sensitivity grew MSSA, and the patient's clinical condition stabilized. Our patient received 10 days of IV antibiotics and was discharged on oral amoxicillin 50 mg/kg divided twice daily for a total of 14 days of treatment per recommendations by the infectious disease specialist. Our patient fully recovered without any residual skin findings after completion of the antibiotic course. JFP

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