Visual Diagnosis: 2-week-old Has a Red, Peeling Rash
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2-week-old Has a Red, Peeling Rash

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Presentation
A 2-week-old term female presents to the emergency department with a history of an abrupt, generalized red rash with peeling of the skin. Her mother reports that she first noticed the skin peeling around her daughter’s umbilicus 3 days ago, about the same time the umbilical cord stump fell off. She also noted some brownish discharge from the umbilicus at that time. Twenty-four hours earlier, the infant had developed irritability and a generalized red rash over her body, with areas of peeling skin. There is no history of fever, cold, hypothermia, cough, or eye discharge. Up to this point, the infant has been feeding and stooling well.

The infant was born term via a normal spontaneous vaginal delivery without complications. Birthweight was 2,700 g, and Apgar scores were 9 and 9 at 1 and 5 minutes, respectively. However, the maternal group B Streptococcus status was unknown; the mother received two doses of intravenous penicillin 4 hours before delivery.

On physical examination, the patient’s vital signs are appropriate for age, and there is no fever. The infant reacts to touch as if it hurts. Examination of the skin reveals erythroderma (scaling erythematous dermatitis) and large, thin sheets of peeling skin with underlying shallow, bright red erosions around the face (Fig 1), lower abdomen, axillae and antecubital fossae (Fig 2), and both ankles and feet (Fig 3). There are no vesicles or bullae. There is no increased erythema around, or oozing or crusting from, the periumbilical area. The mucous membranes of the mouth and anus are unaffected. The rest of the physical examination is normal for age. A clinical diagnosis is made.

Figure 1. Bright red face with thin sheets of peeling skin and underlying shallow erosions.

Author Disclosure
Drs Gupta and Jacobs have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

The editors and staff have been gratified by the large number of submissions (by readers from all over the world) to the Visual Diagnosis column. At the present time, the journal has so many cases in production that we cannot accept new manuscripts. We will indicate in the journal when readers can resume sending offers to write cases and discussions.

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Diagnosis: Staphylococcal Scalded Skin Syndrome

The diagnosis of staphylococcal scalded skin syndrome (SSSS) was based on the classic clinical presentation of a generalized, faint, orange-red, macular erythema with cutaneous tenderness and a positive Nikolsky sign (the separation of superficial skin from the deeper skin layers after application of light pressure).

Discussion

Other names for forms of SSSS are Ritter disease and impetigo neonatorum in neonates. SSSS is a toxin-mediated exfoliative dermatitis caused by toxigenic strains of Staphylococcus aureus that usually belong to phage group 2 (types 3A, 3B, 3C, 55, or 71). The exfoliative toxins (ET) usually involved are A (ETA) and B (ETB), with ETA occurring more commonly (89% of cases) than ETB (4%). Exfoliative toxins cause intraepidermal splitting by destroying the protein desmoglein 1, an important cell-to-cell attachment protein found only in the superficial epidermis. This breakdown leads to bullae formation and diffuse, sheet-like desquamation. The spectrum of SSSS from localized bullous impetigo to full-blown SSSS depends on the distribution of the toxin.

SSSS is primarily a disease of children; 62% of affected children are younger than age 2 years, and 98% are younger than age 6 years. Predisposing factors for SSSS are immunocompromise and lower titers of enterotoxin-specific antibodies, which are lowest in infants and toddlers up to age 2 years. Most maternal antibodies transferred to infants via human milk are thought to be partially protective, but neonatal disease still can occur, possibly as a result of inadequate immunity or immature renal clearance of exotoxin.

In neonates, Staphylococcus commonly colonizes the skin, eyes, perineum, wound sites, circumcision wounds, and umbilical stumps. Approximately 30% of neonates are colonized by S. aureus strains within 1 week of birth, although some studies have shown carriage rates of 60% to 90% in newborn infants discharged from hospitals, particularly when there are staphylococcal outbreaks and where the use of antiseptic umbilical cord care is discouraged.

Usually, SSSS begins with a local skin infection by an S. aureus strain that produces ET, which affects the
immediate area. By contrast, the generalized form of SSSS affects newborns (Ritter disease), infants, and children but rarely adults. In this form, fluid from intact bullae generally is sterile, and the infecting strain usually is recovered from distant sites, such as the throat or nose. Generalized SSSS is thought to arise from systemic ET absorption from local sites, followed by bloodstream diffusion to cutaneous targets. The incubation period from skin infection to the appearance of SSSS usually is 1 to 10 days.

SSSS typically presents clinically with the sudden appearance of diffuse tender erythroderma. Usually, there is a preceding upper respiratory tract infection, conjunctivitis, and, rarely, infection with staphylococcal superinfection as a complication of varicella or measles. Early SSSS commonly includes fever, irritability, and generalized, faint, orange-red, macular erythema with cutaneous tenderness. Within 1 to 2 days, a positive Nikolsky sign develops, and flaccid, thin-walled bullae appear. Within hours of bullae formation, the bullae spontaneously rupture, and the superficial epidermis separates in large sheets to reveal widespread areas of moist red undersurface. Within the next 1 to 3 days, these denuded areas of dry skin undergo a secondary flaky desquamation. Unless infection of the exposed dermis supervenes, the affected skin heals without scarring within 14 days of the onset of skin separation.

The definitive diagnosis of SSSS depends on culture and biopsy results. It is important to culture the skin sites in all patients who have suspected SSSS to identify the causative organism and to determine antibiotic sensitivities. Results of blood cultures are almost always negative in children who have SSSS. Skin biopsy results of affected skin in children who have SSSS reveal intraepidermal cleavage, with splitting beneath and within the stratum granulosum. The cleavage space may contain free-floating or partially attached acantholytic cells. Unaffected surrounding epidermis appears unremarkable, and the dermis contains no inflammatory cells.

**Differential Diagnosis**

The differential diagnosis for SSSS includes streptococcal scarlet fever, Stevens-Johnson syndrome, toxic epidermal necrolysis (TEN), toxic shock syndrome, and Kawasaki disease. Distinguishing features are severity of the disease, percentage of total body surface area affected, involvement of mucous membranes, skin biopsy findings, and systemic signs and symptoms. For example, SSSS differs from the more severe TEN by histology and absence of mucosal pathologic evidence. In SSSS, there is intradermal cleavage of the skin; in TEN, there is necrosis of the full epidermal layer. The Table presents ways to differentiate SSSS from TEN.

**Management**

The mainstay of therapy for SSSS is eradication of the staphylococcal focus of infection by using antibiotic medications, as well as supportive care with careful attention to hydration and monitoring of electrolyte levels due to

| Table. Differentiating Staphylococcal Scalded Skin Syndrome and Toxic Epidermal Necrolysis |
|---------------------------------|------------------------------------------------------------------------------------------|
| **Features**                    | **Staphylococcal Scalded Skin Syndrome**                                                | **Toxic Epidermal Necrolysis** |
| Patients affected               | Infants, young children, immunocompromised adults                                         | Older patients                 |
| Patient history                 | Recent staphylococcal infection                                                          | Drug use, infections, and renal failure |
| Level of epidermal cleavage     | Within the granular cell (outermost) layer of the epidermis as shown below               | Between the epidermis and dermis or at the level of the basal cell as shown below |
| Mucous membrane involvement     | Mucous membranes usually are not involved                                               | Mucous membranes are involved |

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fluid loss through the denuded skin. Fluid rehydration plays an important role if a large percentage of body surface area is affected. Management of the skin in SSSS is similar to treating severe burns. Topical wound care of denuded areas is important and includes applying a bland emollient to decrease pruritus and tenderness. In addition to the use of bland emollients on the skin, nonadherent dressings such as petrolatum-impregnated gauze can be used to cover erosions.

SSSS in infants should be treated with a parenteral β-lactam antimicrobial agent such as nafcillin or oxacillin or, if methicillin-resistant *S aureus* is a consideration, with intravenous vancomycin or oral clindamycin to decrease the staphylococcal bacterial load. Corticosteroids are not indicated, and topical antimicrobials are of no benefit.

Of note, ongoing research has demonstrated that infusing anti-ET antibodies into mice can halt progression of exfoliation. However, this technique has not yet been tested in humans.

**Patient Course**

The patient was admitted to the PICU for observation, intravenous fluid and vancomycin therapy, and burn care. Both the infectious disease and dermatology services were consulted. Skin care included applying petroleum jelly to keep the skin moist and reduce water permeability. A complete blood count on the peripheral smear revealed a normal white blood cell count of 5,200 per cubic mm, with 25% neutrophils, 3% bands, 52% lymphocytes, and 20% monocytes. Wound cultures from the mouth, nose, umbilicus, and skin lesions grew *Staphylococcus epidemidis*, which was most likely commensal. Pain was relieved with rectal acetaminophen. The infant was transferred to the hospital floor in 2 days. She remained afebrile and her skin condition improved without further peeling after 7 days. The patient was discharged from the hospital on oral clindamycin to complete 14 days of therapy.

**Summary**

- Although staphylococcal scalded skin syndrome (SSSS) is relatively uncommon, usually diagnosed on clinical grounds, and readily treated with conventional antibiotics, it is important to emphasize that mortality rates are still unacceptably high and outbreaks are difficult to control.
- SSSS can occasionally lead to serious complications, such as pneumonia, septic arthritis, hypothermia, dehydration, and secondary infections, particularly in newborns, in whom complications may be lethal.
- Early diagnosis and appropriate treatment can prevent the morbidity and mortality associated with these complications. With appropriate management, mortality due to SSSS in children remains below 5% compared with approximately 60% in adults.

**Suggested Reading**


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