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photo quiz

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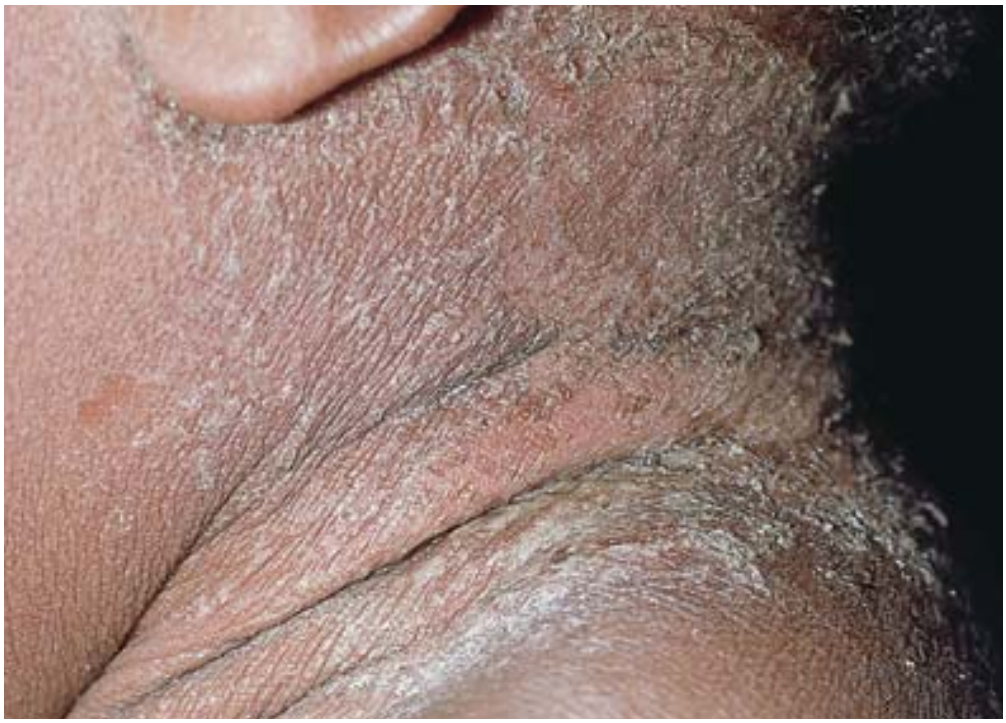

A 9-month-old child developed fever followed by generalized erythema, skin tenderness, and rapidly progressive desquamation beginning on the face and neck. Crusting is noted below the nares.

What is your diagnosis?

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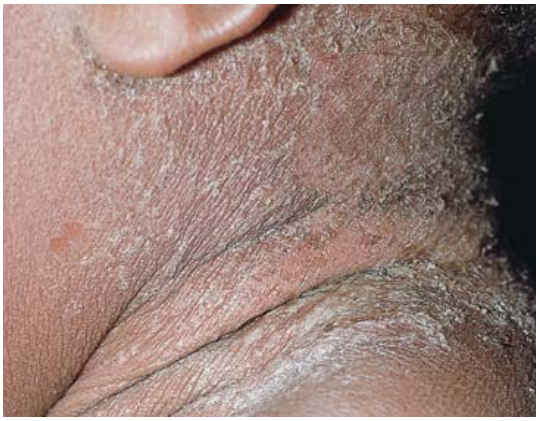
The Diagnosis: Staphylococcal Scalded Skin Syndrome



Staphylococcal scalded skin syndrome (SSSS), or Ritter's disease, is seen in neonates and young children as a manifestation of infection with a toxin-producing strain of *Staphylococcus aureus*. Initial features include fever, generalized erythema, and skin tenderness. Within 24 hours, desquamation begins. With gentle traction, the skin easily shears away, leaving a moist base. This phenomenon, called the Nickolsky sign, is easily elicited during routine movement and care of the child. The site of the staphylococcal infection can vary, but involvement about the nares is common and characteristic. Other common sites of infection include the umbilicus, the conjunctiva, or a surgical wound. Nurses caring for children in a nursery can spread the infection, resulting in outbreaks of SSSS in the neonatal unit. Because the desquamation is toxin mediated, skin cultures are generally negative unless secondary colonization has occurred later in the course of the illness.

Differentiation from toxic epidermal necrolysis (TEN) is important. TEN commonly follows drug therapy with an anticonvulsant agent or a nonsteroidal anti-inflammatory drug. Because TEN is characterized by full-thickness necrosis and sloughing of the epithelium at the level of the basal layer, the level of cleavage is deeper. Conjunctival and circumoral erythema are more characteristic of SSSS than of TEN.

A diagnosis can be established by a frozen-section examination of the sloughed skin. Tzanck smears from the base of the split have also been used.[1] Histologically, this split occurs within the granular layer, in a pattern resembling pemphigus foliaceus. Although serum and neutrophilic crusting may be seen, immunofluorescent studies are negative or nonspecific.

The toxin that produces SSSS is exfoliative and is produced by staphylococci of phage group II, usually types 3A, 3B, 3C, 55, or 71. Occasionally, groups I and II have been implicated.[2] Exfoliative toxin A and B genes can be detected by a polymerase chain reaction.[3] In some cases, the toxins responsible for toxic shock syndrome have been implicated in cases of SSSS.[4]

SSSS is rare in adults because the adult kidney efficiently eliminates the toxin. Cases have appeared in adults with renal failure, overwhelming sepsis, or immunosuppression by disease or corticosteroid administration.[5] Staphylococci produce superantigens that bypass conventional antibody production and bind directly to class II major histocompatibility molecules. Cytokine production may be involved in some of the manifestations of the disease.[6]

Antibiotic therapy should be initiated based on known susceptibility patterns in the hospital or community, although antibiotic-resistant strains have been implicated in some cases of SSSS.[7] Cultures should be performed and treatments modified as appropriate.

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