Case Report

Neonatal *Staphylococcal* scalded skin syndrome complicating ileal atresia

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ABSTRACT

Staphylococcal scalded skin syndrome is a toxin mediated Staphylococcal infection, the toxin produced by *staphylococcus aureus* type 2 phage types (55,71,3A,3B,3C). There is a generalized tender erythema which commences on the head and neck, accompanied by fever, irritability, continuous cry and miserable look. The erythema is followed by cleavage of the upper epidermis in a large sheets mainly in the head, neck and the flexures, with formation of bullae (Nikolsky sign). It is most common in infants and children under 5 years. Most cases respond to antibiotics with other supportive measures. The prognosis is good, and the skin lesions disappear without a residual scar.

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Staphylococcal scalded skin syndrome (SSSS), was first described by Gottfried Ritter Von Rittershain in 1870, who observed the disease in Prague. He eventually called the disease "dermatitis exfoliative neonatorum".[1] Approximately 100 years later, the pathogenesis and the scope of the disease order were clearly and beautifully demonstrated by Melish and Glasgow.[2,3] Staphylococcal scalded skin syndrome is a toxin-mediated *Staphylococcal aureus* (*Staph. aureus*) infection, in which a circulating toxin (epidermolysin) is elaborated by group 2 *Staph. aureus* phage type (55,71,3A,3B,3C).[4,5] The clinical disease occurs when there is sufficient toxin load produced for an infection with these organisms.[6] There are 2 types of the toxin A and B.[7-9] Type A which is a heat stable toxin and encoded by bacterial chromosomal genes, and type B which is a heat labile toxin and encoded on a 37.5 Kilobase plasmid.[10] The toxin will lead to cleavage of the epidermis through the malpegian or the granular layer[2-4] by selective destruction of the intracellular cutaneous.[9,11] The mechanism of action is still not well understood, although some evidence suggests that they act as serine proteases.[8,12] Although the toxin is responsible for the skin loosening seen in this syndrome, it does not account for all the symptoms of the disease, there may be other factors which are involved in the pathogenesis of the disease, for example delta hemolysin.[13]

Case Report. A female neonate, was admitted to the Surgical Department, Al-Fateh Teaching Hospital, Benghazi, Libya, with intestinal obstruction. At laparotomy, we found ileal atresia, and at the age of 7 days, the patient developed sudden onset bright red skin rash which started in the head and neck, and within 2 days it became generalized, and associated with mild fever (37.8^{0}C). Due to irritability, the baby was always crying, after another 3 days, there were areas of skin pealing leaving superficial wet glistening areas, specially in the head and neck, flexures (axillae, cubital and popliteal fossae) (**Figure 1**). During the next 6 days, the exfoliative skin became dry, and crusted, with scales and fissuring appeared around the mouth (**Figure 2**). The conjunctivae were congested, the oral cavity was normal, and a small-sized bullous appeared on the right

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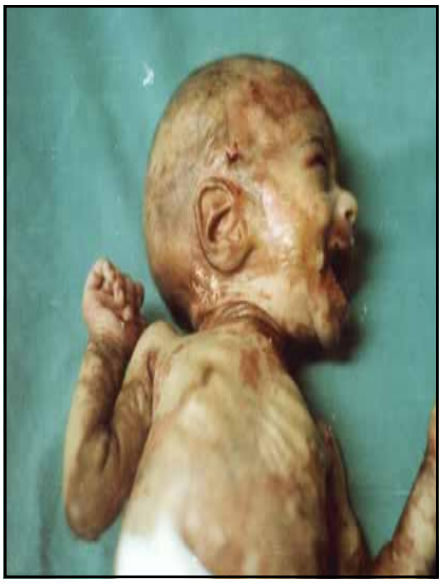

Figure 1 - Shows the area of skin pealing and superficial wet glistering areas specialty in the head, neck and axillae.

Figure 2 - Shows the dry, crusted skin with scales and fissuring around the mouth.

shoulder (Nikolsky sign), which ruptured after few hours. The patient remained in the hospital under incubator care, and put on intravenous fluids, nothing per-oral, complete blood picture showed only leukocytosis with relative neutrophelia (18,000/cmm), blood sugar, blood urea and serum electrolytes were all normal. Culture were taken from the denuded skin, and urine were negative, swab cultures were taken from the throat and nares were negative, blood culture was positive for coagulase positive *Staph. aureus* type-2 phage type 3a, 3b. Histopathologicaly skin biopsy showed superficial epidermal separation, with no cellular infiltration. The patient was treated by systemic antibiotic (Cloxacillin), maintenance of her hydration and local application of emollient. After 12 days from the onset of the rash, the patient became well, the skin lesions disappeared without any residual scar.

Discussion. Staphylococcal scalded skin syndrome is a dramatic generalized blistering and exfoliative skin condition, predominantly affecting infants and children under the age of 5 years.[5,14-17] Maternal fetal transmission with intrauterine SSSS is manifasted a few hours after the birth was reported.[18,19] It is very rare in adults, but it can affect immuno-compromised adult patients, where the symptoms are similar to those of pediatric disease but blood culture are often positive for diagnosis.[7,14,20] It can occur in outbreak, specially in the neoborn nurseries where epidemic or outbreak have occurred.[21,22] The primary site of infection may not be apparent, non-cutaneous sources of infection must be sought (sinuses, nares, throat, lungs, deep wounds), it appears in most cases to be introduced through the respiratory system, in neonates the common site is the umbilicus.[6,23] A short prodromal phase of conjunctivitis, rhinitis, or respiratory infection, with an associated malaise, fever, and irritability may proceed the phase of cutaneous manifestation. The skin lesions usually start as macular sun-burn like erythema,[16,24] which appear first in the head and neck, as was described in our patient, this faint erythematous rash spreads rapidly and may extend within a few hours to become generalized eruption.[25] The skin becomes quite tender, the baby will cry and draw back when picked up or touched, with a miserable look, (**Figure 2**). The mucous membranes are spared except for conjunctivae.[4,23,26] Within 2 days, the skin may become wrinkled and the exfoliative stage starts, where the superficial layers of the epidermis start to peel off in a large superficial wet sheets or flaccid bullae (Nikolsky sign) with little oozing.[4,10,27] The exfoliation is commonly emphasized in the flexures especially the neck, axillae and around the body orifices,[10,11,25] over 2-3 days the glistening areas become dry and crusted scaling flakes appear, with a distinctive fissuring developing around the mouth and eyes, the peeling rash around the mouth often occurs in a radial pattern resembling rhagades. (**Figure 2**).[10] If the erythematous but unpeeled skin is rubbed side ways, the superficial epidermal layer will separate from the deeper areas and blister may appear (Nikolsky sign), although Nikolsky sign and shedding of the skin are typical of SSSS, still in the abortive form, the skin erythema does not progress to blister formation and so Nikolsky sign is negative.[10] Widespread bullae formation may occur in cases with decreased antibody titer and decreased renal excretion of the toxin.[24] Secondary infection of the glistening denuded areas is rare but when it happens severe sepsis and electrolytes disturbance will supervene. Identification of the

micro-organisms are not easy, culture from the skin, blisters, and blood are usually negative,[6,10,28] wright or giemsa stained section of the peeled epidermal fragment with hematoxyline and eosin staining can detect the level of splitting,[6] culture from the nose, throat, nasopharynx and conjunctivae may be carried out if there is no apparent area of obvious infection, as it may reveal the coagulase positive *staphylococcal aureus*. Histologically, there is high epidermal separation with minimal acantholysis, the rest of the epidermis and dermis are intact, the intercellular septums widen without cell lysis or loss of the mucopolysaccharide cell coat, there is minimal or no inflammatory cell infiltration of the superficial epidermis that underlies the forming blister.[4,29] Frozen section of the detached epidermis permit even emergency diagnosis.[30] Early differential diagnosis during the erythrodermic phase is difficult,[23] specially from bullous impetigo and toxic epidermal necrolysis (TEN), this differentiation is important due to the different treatment the former entities required. In TEN; there is full-thickness epidermal damage which may have sub-epidermal separation, as well as the blisters and the skin erosions are hemorrahagic and the mucous membranes are commonly involved.[2,3,31] In cases of bullous impetigo there is an obvious inflammatory infiltration of the superficial epidermis that underlies the blisters and the organisms are readily obtained directly from skin lesions.[24] Other less common conditions which may simulate SSSS are scarlet fever, toxic shock syndrome, and drug induced TEN, but all these show involvement of the mucous membrane, strawberry tongue and rarely demonstrate frank blistering.[6] The treatment of SSSS is primarily supportive, it includes adequate fluid and nutritional intake to maintain hydration and electrolyte balance. If oral fluids are refused or not adequately ingested intravenous fluids are indicated, and that is why in seriously ill patient, admission to the hospital with parenteral therapy is mandatory. Sterile sheets and burn cardles are beneficial, saline compresses to decrease fluid loss, antibiotic cover (penicillinase-resistant) to reduce the chance of septic complications or spread to other children, and topical application to decrease itching are useful once the skin desquamation commences. Others advice topical applications only in neonates where silver sulfadiazine or other burn therapy may be needed. The patient is better to be nursed naked with as little handling as possible. Complete healing is the rule without any remnant scar, the disease will last 10-14 days from the onset of the symptoms.[4,6,24] The prognosis is good since sepsis is not common. The mortality rate is 7-8% in most series, death due to sepsis or bacterial endocarditis is usually attributed to spread of the micro-organisms from its nasal, nasopharyngeal or cutaneous sources.[23,29]

In conclusion, SSSS is mainly a disease of infancy and early childhood below 5 years. Proper differentiation of this disease from other similar entities is critical for the appropriate management. It is highly responding to medical treatment without residual damage.

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