Unusual presentation of more common disease/injury

Staphylococcal scalded skin syndrome in a preterm newborn presenting within first 24 h of life

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Summary

A preterm neonate was noted to have diffuse blanching erythema around the mouth followed by appearance of bullous lesions on the upper back, lower neck and right scapular areas at 23 h of life. The bullae subsequently ruptured leaving an extremely tender, erythematous, denuded area of the skin, which extended over next few hours to involve most of the upper back and right shoulder regions. Nikolsky sign was positive. Clinical diagnosis of staphylococcal scalded skin syndrome was made. The throat, blood, urine and cerebrospinal fluid cultures did not yield any growth, but wound culture was positive for *Staphylococcus aureus*. Treatment included administration of intravenous fluids and vancomycin for 10 days. The wound area was covered with vaseline and sterile gauze dressings. On day 5 of life, epithelialisation began and was complete on the seventh day of life. She was discharged home with intact skin, without scars, on day 12 of life.

BACKGROUND

Staphylococcal scalded skin syndrome (SSSS) is an extensive desquamative erythematous condition of the skin characterised by blistering and epidermal peeling. It is induced by two epidermolytic toxins, A and B (ETA and ETB) produced by *Staphylococcus aureus*. This disease usually occurs in children under 5 years old, especially in neonates with an onset between the first 3 to 16 days of life. SSSS is rare in premature infants and to the best of our knowledge, SSSS presenting within the first 24 h of life has only been reported in two term and one preterm infants. We present a case of preterm neonate with onset of SSSS within the first 24 h of life, who survived and was eventually discharged home.

CASE PRESENTATION

This preterm neonate was born at 34 weeks gestation to a 28 year old gravida 11, para 4 mother by C-section. Indication for C-section was history of previous C-sections and presence of placenta previa, placenta accreta and percreta. Mother had received regular prenatal care during this pregnancy. Her obstetric history was positive for one full term vaginal delivery, two preterm C-sections, one full term C-section, one ectopic pregnancy, two first trimester spontaneous abortions and three voluntary terminations of pregnancy. She denied any history of tobacco, alcohol or illicit drug use during this pregnancy. Her labs were; blood type B positive, hepatitis B surface antigen negative, HIV negative, gonorrhea and chlamydia negative by DNA amplification, rubella immune, urine drug screen negative and urine culture negative. She received two doses of betamethasone prior to delivery. Rupture of membranes was at delivery and amniotic fluid was clear. Baby received bag and mask ventilation after birth and was eventually intubated due to poor respiratory efforts and poor tone. Apgar scores were 6, 7 and 7 at 1, 5 and 10 min respectively.

Baby was started on mechanical ventilation with moderate support and admitted to neonatal intensive care unit (NICU). Growth parameters were appropriate for gestational age with weight at 25th percentile (2080 g), length at 25th percentile and head circumference at 50th percentile. Vital signs at admission to NICU were stable except for low blood pressure. Temperature was 36.8 degree celsius, heart rate was 162 beats/min, respiratory rate was 46 breaths/min, blood pressure was 52/16 with mean arterial pressure of 30 mm Hg and oxygen saturation was 96%. Physical examination was normal except for presence of mild intercostal retractions. Peripheral perfusion was good with a capillary refill time of <2 s. Chest x-ray showed mild granularity in bilateral lower lung fields with slightly decreased lung volumes suggestive of mild hyaline membrane disease. First capillary blood gas showed a pH of 7.27 with carbon dioxide of 50 and bicarbonate of 23. Baby received one dose of surfactant (survanta) via endotracheal tube. After that, ventilator support was weaned and she was extubated to a nasal canula on day 2 of life and then weaned to room air by day 3 of life. She received one normal saline bolus of 10 ml/kg initially for low blood pressure and afterwards, her blood pressures remained stable with good peripheral perfusion. Her complete blood count showed a white blood cell count of 20 500/µl with neutrophilic predominance (79%), haemoglobin of 15.8 g/dl and platelet count of 306 000/µl. She was started on ampicillin and gentamicin on day 1 of life after sending a blood culture.

At approximately 23 h of life, she was noted to have diffuse blanching erythema around the mouth followed by appearance of bullous lesions on the upper back, lower neck and right scapular areas. The bullae subsequently ruptured leaving an extremely tender, erythematous, denuded area of the skin (figure 1). This denuded area extended over next few hours to involve most of the upper back and right shoulder regions (figure 2). Nikolsky sign (gentle pressure

Figure 1 Ruptured bullous lesions leaving a large denuded area of the skin with diffuse, tender, erythematous rash on upper back, lower neck and right scapular regions at 23 h of life. Nikolsky sign was positive.

applied to the skin resulting in separation of the upper epidermis) was positive. Clinical diagnosis of SSSS was made. The throat, blood, urine and cerebrospinal fluid cultures did not yield any growth, but wound culture was positive for *S aureus*. Herpes simplex virus (HSV) 1 and 2 PCR on blood, cerebrospinal fluid HSV 1 and 2 direct fluorescent assay and HSV culture; all were negative.

TREATMENT

Treatment included administration of intravenous fluids (adjusted to compensate for evaporation through the wound surface) and vancomycin for 10 days. Ampicillin and gentamicin were discontinued. Clindamycin was given for initial 5 days of treatment to decrease the toxin production. The wound area was covered with vaseline and sterile gauze dressings. Acyclovir was started initially, but discontinued later on as HSV 1 and 2 PCR on blood, cerebrospinal fluid HSV 1 and 2 DFA and HSV culture; all were negative.

OUTCOME AND FOLLOW-UP

No new lesions appeared after starting vancomycin. On day 5 of life, epithelialisation began (figure 3) and was complete on the seventh day of life. She was discharged home with intact skin, without scars, on day 12 of life. At 4 months follow-up visit, she was doing fine with no residual scars and had normal development.

DISCUSSION

We describe a preterm infant with SSSS presenting within 24 h of birth. The clinical symptoms of diffuse blanching erythema around the mouth, fragile bullous lesions, tender, erythematous rash and positive Nikolsky sign; all were compatible with the diagnosis of SSSS. The patient

suffered from mild respiratory distress syndrome as a result of prematurity. SSSS is rare in premature infants. To the best of our knowledge, SSSS presenting within the first 24 h of life has been reported in two term infants^{1 2} and only one preterm infant.³ Haveman LM *et al* reported a case of congenital SSSS in a premature infant with fatal outcome. Despite adequate antibiotic treatment, the infant died within 24 h after birth because of respiratory failure. Although, our patient had also presented within 24 h after birth, she responded well to the treatment and was eventually discharged home.

SSSS is a rare dermatologic disorder caused by *S aureus* exotoxins, ETA and ETB.⁴ These toxins cause cleavage of desmoglein 1 complex, a protein in the desmosomes, which hold keratinocytes together in the zona granulosa of the epidermis.⁵ This leads to the formation of flaccid bullae, that rupture easily.⁶ In a study by Patel and Finlay,⁵ 91% of adults older than 40 years had antibodies to ETA, whereas only 41% of children aged 2 to 5 years had antibodies to ETA. Neonates are more susceptible to dissemination of these exfoliative toxins, because of the decreased renal clearance of these toxins in them.

In neonates, presentation usually occurs at 3 to 16 days of age.⁶ Affected infants may present with fever, irritability and diffuse blanching erythema beginning around the mouth. Fragile bullous lesions appear within 1–2 days. Nikolsky sign (gentle pressure applied to the skin resulting in separation of the upper epidermis) is positive. In severe cases, the whole upper epidermis may be shed. Mucous membranes are not involved, but may look hyperaemic. During the healing phase, flaky desquamation may occur. Lesions heal without any residual scarring due to their intraepidermal location.⁷

Figure 2 Same patient a few hours later. The rash has extended to involve most of the upper back and right scapular areas.

Figure 3 Beginning of epithelialisation on day 5 of life.

In infants with suspected SSSS, cultures should be obtained from blood, urine, nasopharynx, abnormal skin, or any suspected focus of infection. Diagnosis of SSSS is usually clinical, but may be confirmed with skin biopsy that shows intraepidermal and subcorneal division with minimal necrosis.⁸ However, biopsy is usually reserved for the doubtful cases that fail to respond to initial management.

The differential diagnosis of SSSS in neonates includes bullous impetigo, toxic epidermal necrolysis (TEN), epidermolysis bullosa, bullous mastocytosis, herpetic lesions and neonatal pemphigus.⁹ The most important dermatologic condition to be considered is TEN, as it has an overall mortality of 30%,¹⁰ whereas SSSS has a mortality of about 4% in children.⁵ TEN, more commonly seen in adults or older children, is usually due to a drug reaction.¹¹ The

distinguishing clinical features between TEN and SSSS are the presence of mucosal involvement and the presence of the Nikolsky sign in only affected areas in TEN, whereas in SSSS there is no mucosal involvement and the Nikolsky sign is also present in areas that are not visibly affected.¹² On histopathological examination, TEN has epidermal necrosis with dermal sparing, whereas SSSS will show intraepidermal, subcorneal division.¹³

Treatment of SSSS consists of:

- ▶ Prompt administration of intravenous penicillinase-resistant penicillin, or vancomycin in areas with a high prevalence of methicillin-resistant *S aureus*.¹⁴
- ▶ Supportive skin care with the use of emollients to improve barrier function and
- ▶ Monitoring of fluid and electrolyte status with replacement of fluid losses as needed.

Clindamycin can be used in infections with susceptible strains, and there is also evidence that it may inhibit toxin formation.^{15 16} In addition to eradicate the focus of infection, antibiotics can help in preventing superinfections, which may occur secondary to the breakdown of the epidermal barrier and increase the risk of mortality. Early diagnosis and prompt supportive therapy along with antimicrobial therapy improve outcomes in patients with SSSS.

Learning points

- ▶ SSSS is an extensive desquamative erythematous condition caused by exfoliative toxins of *S aureus*.
- ▶ This disease usually affects neonates at 3–16 days of age. Although rare, SSSS may present within the first 24 h of life.
- ▶ Early diagnosis and prompt supportive therapy along with appropriate antimicrobial therapy improve outcomes in neonates with SSSS.

Competing interests None.

Patient consent Obtained.

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