Intrauterine staphylococcal scalded skin syndrome in a preterm infant

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Abstract

Staphylococcal scalded skin syndrome (SSSS) in premature infants is a rare condition. We present SSSS in a preterm infant, who developed all signs and symptoms of SSSS at birth, with a fatal outcome due to fungemia caused by Candida parapsilosis. The clinical presentation was challenging to differential diagnosis. SSSS diagnosis was

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Intrauterine staphylococcal scalded skin syndrome in a preterm infant**Abstract**Staphylococcal scalded skin syndrome (SSSS) is a rare condition in premature infants. We present SSSS in a preterm infant, who developed all signs and symptoms of SSSS at birth, with a fatal outcome due to fungemia caused by Candida parapsilosis. The clinical presentation was challenging to differential diagnosis. SSSS diagnosis was confirmed by skin biopsy. This was presented to emphasize the importance to recognize the clinical manifestations and early diagnosis of SSSS for clinicians. Intrauterine SSSS in preterm infants is a potentially fatal condition, and early recognition and prompt supportive therapy along with antimicrobial therapy improve outcomes could improve outcomes and be life-saving.**1 INTRODUCTION**SSSS is a blistering, toxin-mediated skin disorder caused by*Staphylococcus aureus* producing exfoliative toxins, A and

B (ETA and ETB). [1] It usually affects neonates between the first 3 and 16 days of life. It is rare in premature and low-weight newborns. We present a case of SSSS in a premature newborn, who developed all symptoms of SSSS at birth, with a fatal outcome due to fungemia caused by *Candida parapsilosis*. 2 **CASE PRESENTATION**After 33 weeks 5 days of gestation, a 1,680 g male neonate was born via vaginal delivery. His mother was a 37-year-old, gravida 4 para 4. Prenatal sonography revealed small size for gestational age. Amniocentesis and prenatal laboratory tests were within normal limits. No medical procedure during pregnancy was performed. His mother received one dose of betamethasone prior to delivery. Membranes ruptured 3 hours prior to delivery, and amniotic fluid was clear. After birth, bradycardia, poor muscle tone, and respiratory failure were noted. The infant was intubated, chest compression was performed, and endotracheal epinephrine was administered. Apgar scores were 2 and 7 at 1 and 5 min after birth, respectively. Physical examination highlighted diffuse erythematous patches and blisters with wide-spreading exfoliation on the face, trunk, and limbs, without ectropion. Nikolsky sign was positive. Mucous membranes were intact (Figure 1). With these clinical manifestations, four differential diagnoses were suggested: (1) epidermolysis bullosa, (2) SSSS, (3) ichthyosis, and (4) immune deficiency syndromes. To confirm the diagnosis, skin biopsy was performed. Skin biopsy revealed infant skin with superficial acantholysis in the subcorneal and intragranular cell layers and mild dermal inflammation (Figure 2). These findings can be seen in either pemphigus foliaceus or SSSS. Indirect immunofluorescence (IIF) was negative. All these findings were found to be consistent with SSSS. Oxacillin and gentamicin were initiated. The wound area was covered with petroleum jelly and sterile gauge dressings. Diffused and marked erythema with progressing to flaccid blisters and desquamation developed. The blisters ruptured, leaving changes resembling a burn (Figure 3). Meanwhile, shock with acute renal failure developed on the day of life (DOL) 5. Antibiotics were adjusted to oxacillin, clindamycin, meropenem, and fluconazole for prophylaxis. Cultures on blood samples collected on the DOL 1, 3, and 5 yielded negative results. However, *Candida parapsilosis* grew in blood cultures performed on the DOL 8 and 10, and yeast was identified in cultures on the neck, extremities, and inguinal pus. Caspofungin was added. Due to septic shock with persistent fungemia, the infant's clinical condition deteriorated and he expired on DOL 13.**3 DISCUSSION**We describe a premature infant with SSSS presenting at birth. Congenital SSSS presenting within the first day of life has only been reported in 2 term infants [2,3] and 2 preterm infants,[4,5] and only 1 term infant [3]developed symptoms before delivery (Table 1). Distinguishing skin lesions in infants is important. Diffuse blanching erythema, generalized erythema, fragile bullous lesions and positive Nikolsky sign were all compatible with SSSS. [6] Despite the failure for pathogen isolation, histopathology was used to diagnose SSSS. SSSS is a dermatologic disorder caused by ETA and ETB produced by*Staphylococcus aureus*. [1] Immature renal systems with decreased renal clearance of epidermiolytic toxins, increased amount of desmoglein-1 in the skin at an early age, and lack of protective antibodies to these toxins may account for the increased incidence in neonates and children. [6]SSSS has diffused tender erythroderma and wrinkling of the skin accentuated in periorificial and flexural area. Mucosal membranes are characteristically spared. [6] Fragile bullae and erosions develop, and the overlying epidermis loosens and peel like a scald, which can be extended after 24–48 h. [1]Affected skin may have desquamation due to the loss of the roof of the blister, leaving an erythematous area, resulting to the scalded appearance. Nikolsky sign was shown to be positive. [7]SSSS diagnosis is mainly based on the clinical symptoms of bullae, tender erythroderma, and desquamation with a scalded appearance especially in friction regions, absence of mucosal involvement, periorificial scabs, and positive Nikolsky sign. The diagnosis can be confirmed by *Staphylococcus aureus* culture from suspected sites of infection, including the conjunctiva, nasopharynx, umbilicus, and diaper area. Blood culture is usually negative and typically unhelpful in the diagnosis of SSSS. [8] In neonates with a blistering rash and diffuse desquamation, epidermolysis bullosa, and ichthyosis also remain high on the differential diagnosis. The differential diagnoses by category included cutaneous infections (common), inherited genodermatoses (rare), and autoimmune blistering diseases (very rare). Reaching an etiological diagnosis on the clinical information alone is usually difficult. [9] Skin biopsy for histopathological examination is extremely helpful in these situations. [10] Biopsies of SSSS reveal superficial intraepidermal separation along the granular cell layer.[11]Prompt empiric treatment with intravenous anti-staphylococcal antibiotics/beta-lactams and clindamycin, which have a capability to inhibit production of exfoliative toxins. [6]Complications of SSSS include dehydration, hypothermia, electrolyte imbalance, and secondary infection.

Most patients resolve without sequelae within 2–3 weeks of acquiring proper treatment.[12] Skin lesions heal without scarring is superficial. The mortality rate is less than 5% in pediatric age group.[11]Despite the ease of treatment, SSSS occurrence calls for an emergency and is potentially fatal in neonates. Therefore, early diagnosis and treatment is essential.

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