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CASE REPORT

Congenital staphylococcal scalded skin syndrome in a preterm infant

Ting-Yu Lee | **Tzu-Yu Liu**

Department of Pediatrics, Hsinchu Municipal MacKay Children's Hospital, Hsinchu City, Taiwan

Correspondence
Tzu-Yu Liu, Department of Pediatrics, Hsinchu Municipal MacKay Children's Hospital, No. 28, Jiangong 2nd Road, East District, Hsinchu City, Taiwan (R.O.C.).
Email: a0922603203@gmail.com

Key Clinical Message

Staphylococcal scalded skin syndrome (SSSS) is a rare condition in premature infants. We report a case of SSSS in a preterm neonate who displayed all clinical manifestations at birth, leading to a fatal outcome from *Candida parapsilosis* fungemia. The clinical presentation was challenging to differential diagnosis. SSSS diagnosis was confirmed by skin biopsy. This case emphasizes the significance of early recognition and diagnosis of SSSS promptly for clinicians. Congenital SSSS in premature infants can be fatal, but with early recognition and appropriate supportive and antimicrobial therapy, outcomes can be improved and lives can be saved.

KEYWORDS

preterm, staphylococcal scalded skin syndrome (SSSS)

1 | INTRODUCTION

SSSS is a skin disease characterized by blister formation and is mediated by toxins produced by *Staphylococcus aureus*, namely exfoliative toxins A and B (ETA and ETB).[1] It usually affects neonates within the first 3–16 days of life and is rare in premature and low-weight newborns. We report a case of SSSS in a premature neonate, who developed all symptoms of SSSS at birth, with a fatal outcome from *Candida parapsilosis* fungemia.

2 | CASE PRESENTATION

After 33 weeks 5 days of gestation, a 1680 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 examinations were within normal limits. No

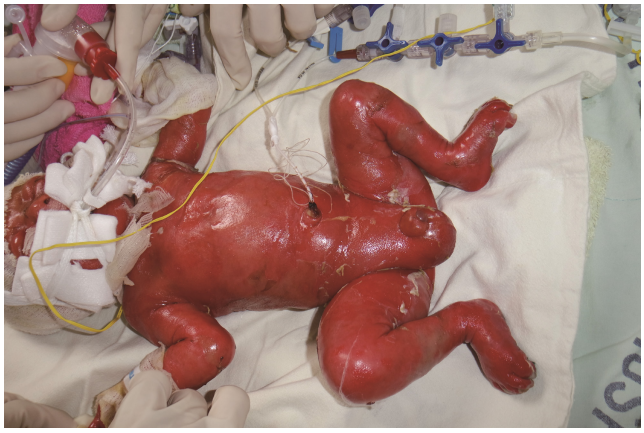

FIGURE 1 A preterm neonate presented with diffuse erythematous patches and blisters with wide-spreading exfoliation on the face, trunk, and limbs, without ectropion after birth. Nikolsky sign was positive. Mucous membranes were intact.

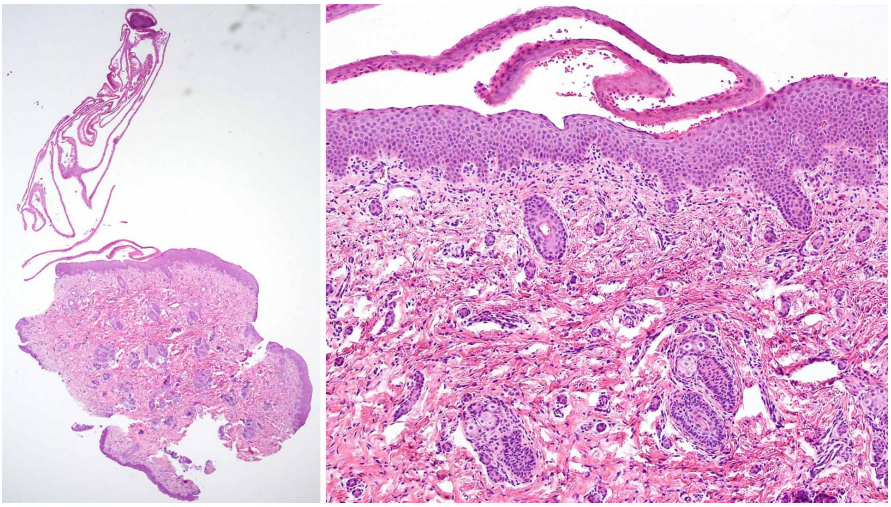

FIGURE 2 The skin biopsy showed infant skin with superficial acantholysis in the subcorneal and intragranular cell layer and mild dermal inflammation.

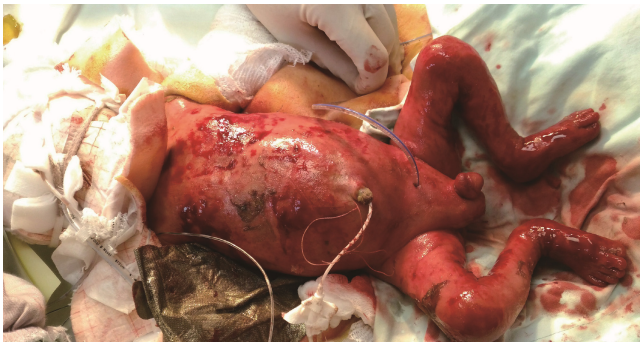

FIGURE 3 Diffuse, markedly erythematous rash with progression transitions to flaccid blisters and desquamation developed. The blisters ruptured, and the resulting area appeared as a burn on DOL 5.

medical procedure during pregnancy was performed. His mother received one dose of betamethasone prior to delivery. Membranes ruptured 3 h 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 present the case of a premature infant with SSSS presenting at birth. Such cases of congenital SSSS presenting within the first day of life have been previously reported in only two term infants[2,3] and two preterm infants,[4,5] and only one 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 disease resulting from ETA and ETB produced by *Staphylococcus aureus*.[1] The increased incidence of SSSS in neonates and children may be attributed to their underdeveloped renal systems, leading to reduced renal clearance of epidermolytic toxins, an

TABLE 1 Clinical characteristics of neonates with SSSS within the first 24 h of life.

Author	Gestational age	Time of illness	Maternal risk	Outcome
Loughead, J.L., et al.	Term	8 h after delivery	Mother developed fever and abdominal tenderness at the day of delivery	Remission
Lo, W.T., et al.	Term	At birth	Previous amniocentesis procedure	Remission
Haveman, L.M., et al.	Preterm (GA 33 + 5 weeks)	12 h after delivery	Mother developed fever 1 week before delivery	Died due to pulmonary hemorrhage
Arora, P., et al.	Preterm (GA 34 weeks)	23 h after delivery	Not mentioned	Remission

elevated level of desmoglein-1 in the skin at a young age, and a lack of protective antibodies against these toxins.[6]

SSSS has diffused tender erythroderma and skin wrinkling accentuated in periorificial and flexural area. Mucosal membranes are typically spared.[6] Fragile bullae and erosions develop, with the epidermis peeling like a scald, extending within 24–48 h.[1] Affected skin may undergo desquamation as the blister roof is lost, revealing an erythematous area, resulting in a scalded appearance. Nikolsky sign was shown to be positive.[7]

The diagnosis of SSSS primarily relies on the clinical manifestations, including the presence of bullae, tender erythroderma, and desquamation resembling a scalded appearance, particularly in intertriginous areas., absence of mucosal involvement, periorificial scabs, and positive Nikolsky sign. To confirm the diagnosis, *Staphylococcus aureus* culture can be performed on suspected infection sites, such as the nasopharynx, conjunctiva, umbilicus, and perianal areas. 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 are also 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 a separation of superficial intraepidermis along the granular cell layer.[11]

Furthermore, the patient indicated the presence of *Candida parapsilosis* in blood cultures and yeast in the skin pus cultures. Neonatal sepsis is frequently caused by organisms colonizing the skin and mucosal surfaces. *C. parapsilosis* is a third of all neonatal Candida infections, with the risk factors of prematurity, parenteral nutrition, intravascular catheters, and use of antibiotics.[12] However, *C. parapsilosis* is comparatively rare in cutaneous infections. Dermatologic manifestations include onychomycosis and ulcer, which were mentioned in a case report.[13] In our case, the clinical manifestations were diffuse erythematous patches and blisters with wide-spreading exfoliation, which is feasible SSSS initially, and was confirmed diagnosis by skin biopsy.

Intravenous anti-staphylococcal beta-lactams and clindamycin, have the ability to inhibit the production of exfoliative toxins.[6] Complications of SSSS include dehydration, electrolyte imbalance, hypothermia, and secondary infection. Most patients received appropriate treatment and recovered without sequelae within 2–3 weeks.[14] Superficial skin lesions typically heal without scarring. 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.

AUTHOR CONTRIBUTIONS

Ting-Yu Lee: Project administration; writing – original draft. **Tzu-Yu Liu:** Conceptualization; supervision; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

Ting-Yu Lee and Tzu-Yu Liu have no conflicts of interest to disclose.

DATA AVAILABILITY STATEMENT

The authors confirm that the data supporting the findings of this study are available within the article.

STATEMENT ON CONSENT FOR PUBLICATION

Informed consent for patient information and images to be published was obtained from the patient's legal guardian.

STATEMENT ON ETHICAL APPROVAL AND INFORMED CONSENT

This case report received IRB and Ethical approval. Informed consent was obtained from the patient's legal guardian for this study.

ORCID

Tzu-Yu Liu https://orcid.org/0000-0002-1801-7105

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