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STAPHYLOCOCCAL SCALDED SKIN SYNDROME WITH ATYPICAL PRESENTATION IN AN INFANT: A CASE REPORT

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ABSTRACT

Staphylococcal Scalded Skin Syndrome (SSSS) is a rare, toxin-mediated condition caused by *Staphylococcus aureus*, typically presenting with acute fever, blistering, and skin peeling in children. While most cases resolve rapidly, atypical chronic presentations remain underreported, posing diagnostic dilemmas. This study aims to report an unusual case of SSSS in an infant with a 3-month history of persistent exfoliative dermatitis, highlighting deviations from classic clinical features and the role of histopathology in diagnosis. A 7-month-old infant presenting with generalized erythema, desquamation, and dehydration underwent detailed clinical, laboratory, and histopathological evaluations. Skin biopsy and cultures were performed to confirm SSSS, and the patient received intravenous cefotaxime, supportive care, and topical treatments. The infant exhibited chronic skin peeling without fever or bullae, a positive Nikolsky sign, and histopathology revealing subcorneal blistering consistent with SSSS. The patient improved after 8 days of hospitalization with antibiotics and hydration, underscoring the efficacy of cefotaxime in this context. This case underscores the need to consider SSSS in chronic exfoliative dermatitis, even without classic symptoms. Histopathology is critical for differentiating SSSS from mimics like SJS/TEN or Leiner's disease. The findings advocate for heightened clinical suspicion in atypical presentations and validate cefotaxime as a therapeutic option, contributing to broader management strategies for SSSS.

Keyword: *Staphylococcal Scalded Skin Syndrome, SSSS, Atypical Presentation, Infant*

Introduction

Staphylococcal scalded skin syndrome (SSS) is caused by epidermolytic exotoxins produced by some strains of Staphylococcus aureus, a possibly fatal illness. The incidence of SSSS is estimated 9,09 until 0,56 cases per one million people. Children under six are most often found to have SSS. Lack of protective antibodies against exfoliative toxins and lower renal clearance of the toxins resulting from immature renal function explain the higher prevalence in young children. This disease usually occurs after a local infection in the upper respiratory tract, ear, conjunctiva, or pus. Desmoglein-1, which is found in the skin's outer layer, breaks down as part of the intricate pathophysiology of SSSS (Brazel et al., 2021; Pratiwi et al., 2024).

Clinically, it is characterized by denudation of the skin and presents as large superficial blisters. The sickness often starts with a fever, followed by a red rash that spreads quickly throughout the body. The rash is typically accompanied by discomfort, tenderness, and blistering on the affected area. SSSS is characterized by skin peeling within hours of rash start. Skin peeling usually starts around the mouth and eyes and progresses to the rest of the body, including the trunk and extremities. Skin peeling typically begins with tiny blisters that erupt, leaving behind rough, denuded skin. Affected skin may appear glossy, red, wet, and painful to touch. Nikolsky's sign refers to the skin's tendency to quickly detach with rubbing. The presence of fever, erythematous rash, and skin peeling helps distinguish SSSS from other disorders. (Brazel et al., 2021; Medugu et al., 2023)

Severe cases of SSSS can result in widespread skin peeling, leaving huge portions of the body with raw skin. This significantly increases the risk of subsequent bacterial infections, dehydration, and electrolyte abnormalities. Possible complications of SSSS include sepsis, pneumonia, and kidney failure.(Medugu et al., 2023)

This research aims to reporting a rare and atypical presentation of SSS in an infant with a chronic 3-months history of persistent exfoliative dermatitis, highlighting the diagnostic challenges posed by prolonged course and unusual cutaneous distribution, which deviates from the typical acute and rapidly progressive nature of SSSS. The current research presents a rare and atypical chronic presentation of Staphylococcal Scalded Skin Syndrome (SSSS) in a 7-month-old infant, contrasting with the typical acute and rapidly progressive cases described in the literature (Brazel et al., 2021; Mishra et al., 2016). Unlike the classic SSSS presentations featuring fever, blistering, and rapid skin denudation within days (Medugu et al., 2023; Grama et al., 2016), this case exhibited a 3-month history of persistent exfoliative dermatitis without fever or preceding bullae, resembling the scarlatiniform variety noted by Nusman et al. (2023). The chronicity of symptoms and absence of mucosal involvement further distinguish it from Stevens-Johnson Syndrome/Toxic Epidermal Necrolysis (SJS/TEN) and other differentials (Tóth et al., 2023). While Shelley et al. (2002) reported a chronic SSSS case in an immunocompromised adult, this study highlights a similar prolonged course in an otherwise healthy infant, emphasizing diagnostic challenges and the necessity of histopathology for confirmation. Additionally, the successful use of cefotaxime, supported by Aldridge (1995), offers insights into alternative antibiotic regimens for SSSS management.

Case Presentation

A 7-month-old infant with 7 kg weight and 64 cm height, brought by his mother to the dermatology outpatient clinic at Abdul Manap Regional Hospital Jambi City, with chief complaint of reddish skin peeling on the face and almost the entire body for the past 3 months. The mother reported that the skin peeling started from the face and then increased to the head, hands, body, groin, and feet. During this complaint, the baby was reported to frequently scratch the lesions

and often cry and be more fussy than usual. The mother denies the presence of blisters appearing before the skin peeling occurred. The consumption of drugs before the lesion appeared had also been denied. The baby has also been experiencing watery and mushy diarrhea for the past 3 days with a frequency of 3-4 times. The mother also reported that her baby has no appetite but is still able to drink well. Fever was denied by the mother. Therefore, the mother took the initiative to take her baby to the dermatology clinic. For the past 3 months, the mother has reported that if her baby's skin is rubbed, lesions will appear. At first it will go reddish then the skin will peeling. The patient's mother said that the skin lesions were previously treated by a dermatologist through an online consultation and received cefadroxil as an antibiotic and a compounded cream. The condition improved at the beginning of the treatment but then returned. The patient has a history of urinary tract infection at the age of 2 months. At the age of 1 week, there were pustules filled with pus in the abdominal area, but after being treated, these complaints healed and never appeared again. The patient is the second child of two siblings and born full term. there is no history of the same disease in the family. History of drug allergy, history of allergy, and history of cancer in family were denied by patient's mother.

Vital signs within normal limits, but upon general examination, sunken eyes, ectropion of the eyelid, decreased skin turgor, and increased bowel sounds were found. Dermatological examination revealed generalized erythema accompanied by tender, circumscribed erythematous patches and plaques, distributed over regio facialis, scalp, neck, antecubital fossae, wrists, interphalangeal flexural creases of manus and pedis, distal phalanges of manus and pedis, dorso manus, dorsum pedis, inguinal folds, gluteus, and femur distal. The facialis regio exhibited marked erythema with areas of crusting, fissuring, superficial erosions, and desquamation (lamellar type of squama), particularly involving the perioral, perinasal, and infraorbital regions. Honey colored crusts are seen on the perioral and perinasal area. The scalp demonstrated scaly plaques with patchy alopecia. A positive Nikolsky sign was noted over the affected areas. There were no mucosal lesions involved.

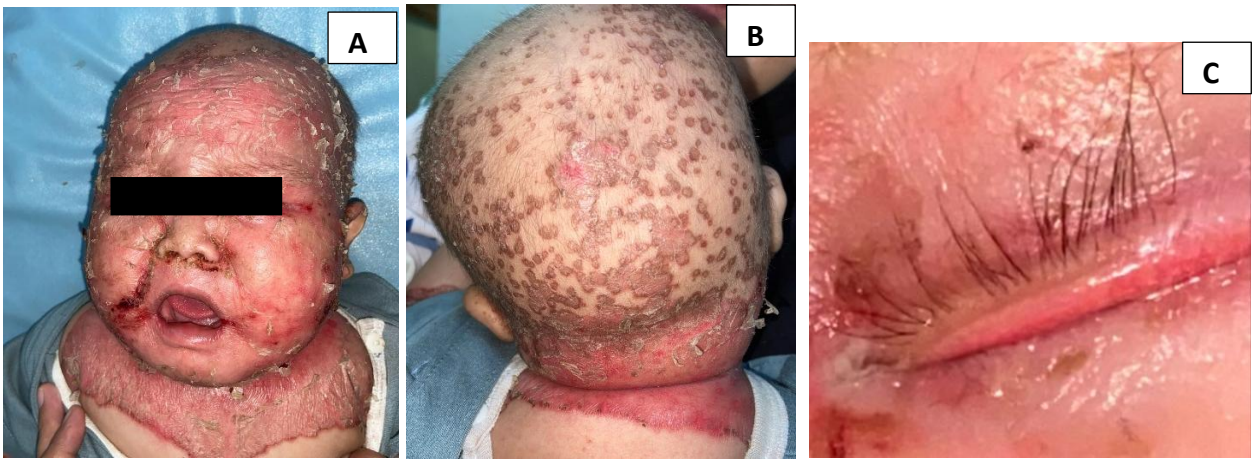

Figure 1. A. Skin manifestation on face and neck, Figure B. Skin manifestation on scalp and neck, Figure C. Ectropion on the eyelid

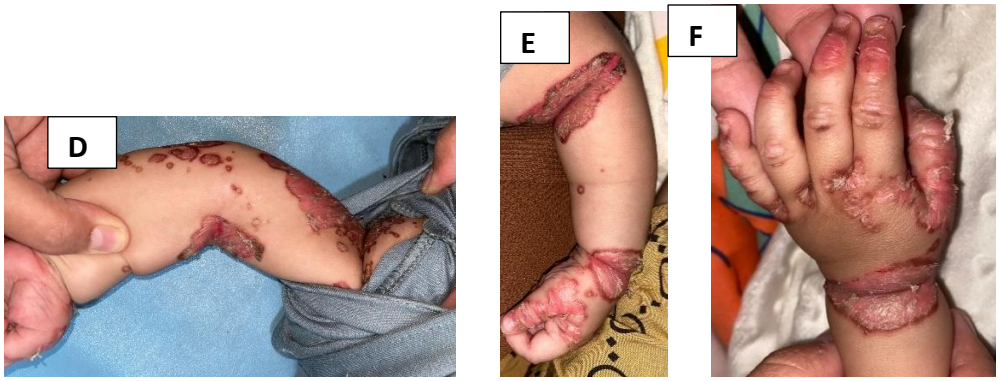

Figure 2. D, E, and F. Erythema and desquamation on hand and finger

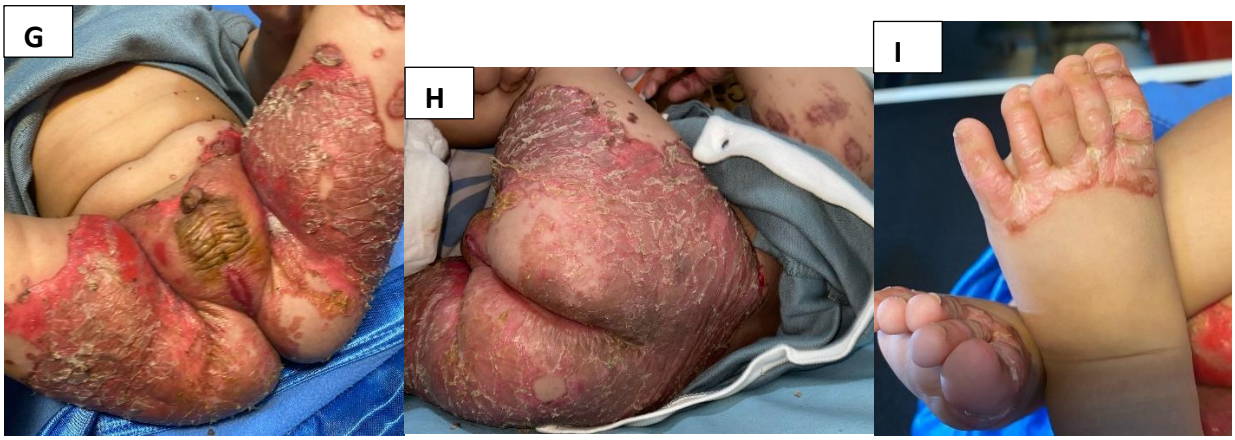

Figure 3. G, H, and I. Skin presentation on diaper area and foot

In the laboratory examination, an increase in leukocytes to 11,800 µL was found, a decrease in urea (2 mg/dL) and creatinine (0.2 mg/dL), a decrease in total protein (5.2 g/dL), and a decrease in albumin (2.6 g/dL), while routine urine examination and electrolyte levels remained within normal limits. A biopsy was also performed on the lesion on the left thigh using an elliptical incision method, taking part of the lesion that borders normal skin. The lesion sample was then sent to the laboratory for histopathological examination. The histopathological examination results show that the epidermis is lined with keratinized complex squamous epithelium with parakeratosis, and blisters are observed in the superficial intradermal layer in the subcorneal layer containing erythrocytes without inflammatory cells. the dermis consists of dense connective tissue with a slight network of inflammatory cells, lymphocytes, and plasma cells, among which skin appendages are visible. based on histopathology, the lesion's appearance suggests to Staphylococcal Scalded Skin Syndrome (SSSS).

The patient is treated with the involvement of a pediatrician and an ophthalmologist. During the hospitalization, the patient received fluid therapy, namely Ringer's lactate 20 tpm, cefotaxime injection 3x250 cc, albumin infusion 100 cc, gentamicin eye ointment 4x1 ods, gentamicin eye drops 3x1 ods, paracetamol syrup 3x70 mg, cetirizine syrup 1x2.5 mg, and moisturizer lotion. The patient was hospitalized for 8 days and showed improvement during the stay. The discharge medication prescribed is cefadroxil syrup 2x125 mg and cetirizine syrup 1x2.5 mg

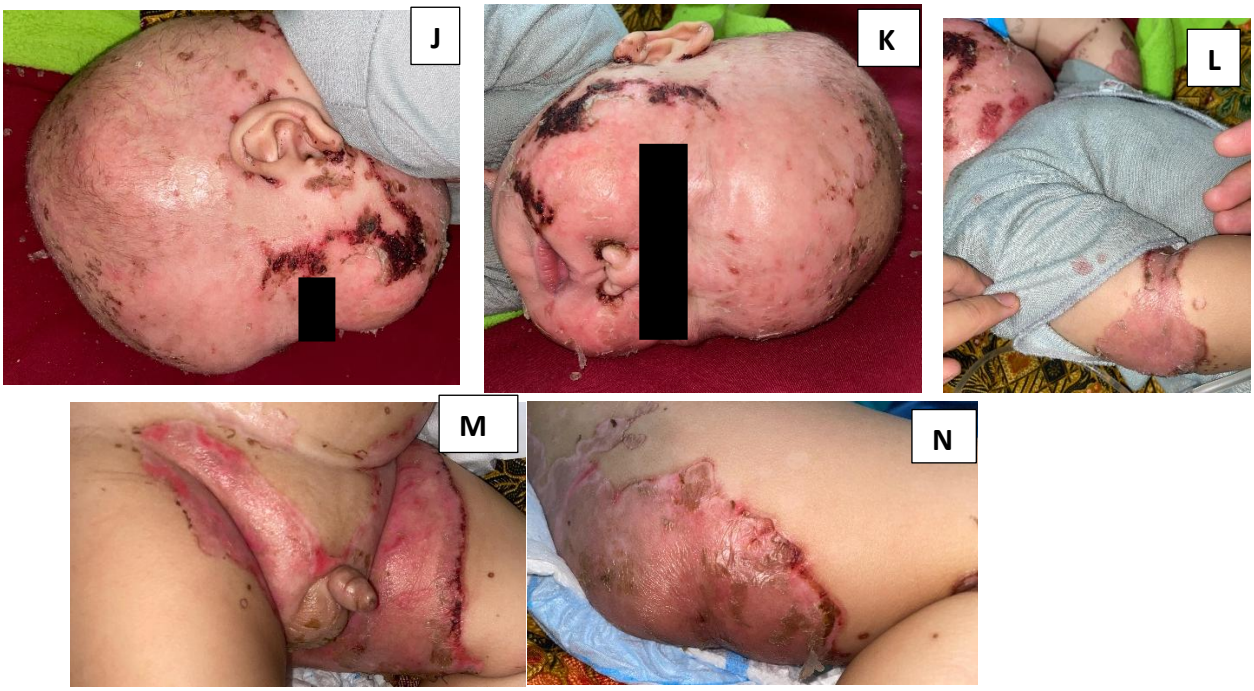

Figure 4. J, K, L, M, and N. Inward Day 2 showed significant improvement

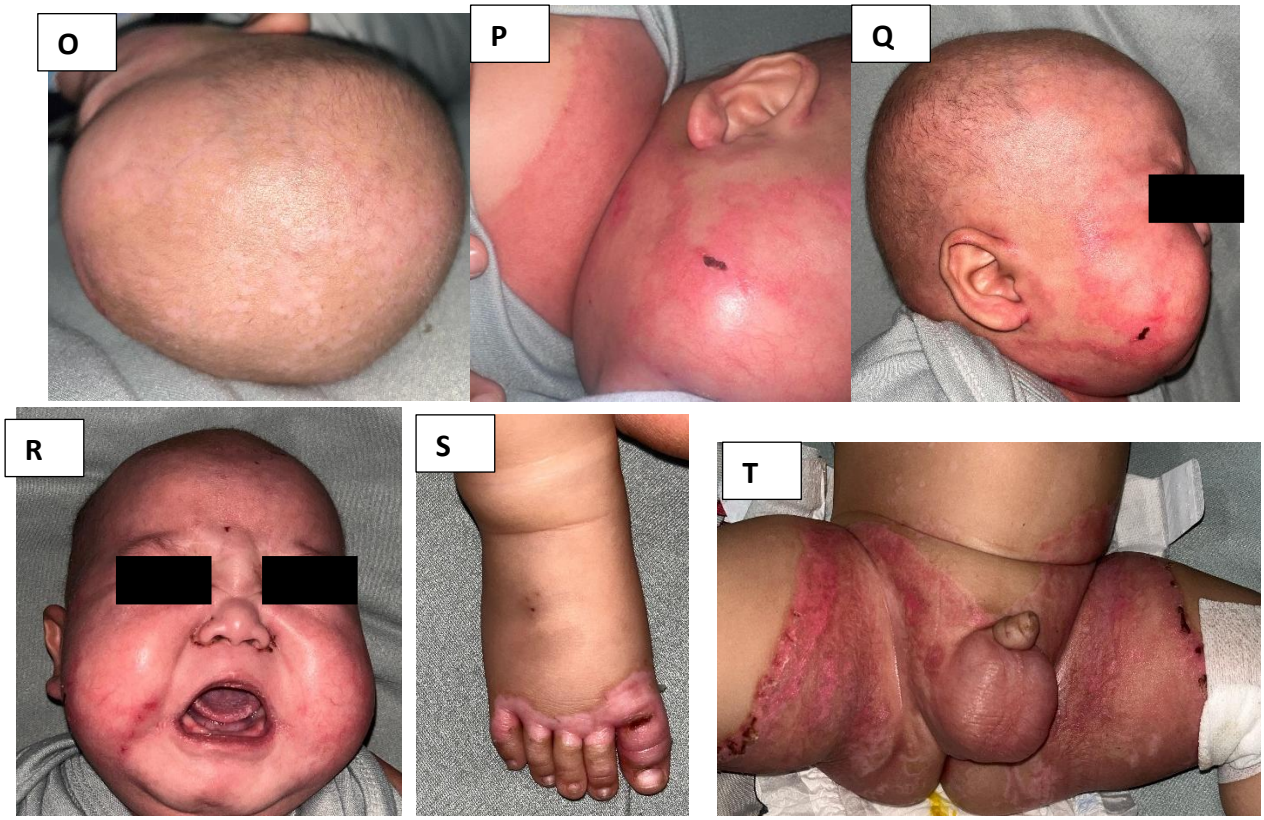

Figure 5. O, P, Q, R, S and T. Last Day Inward

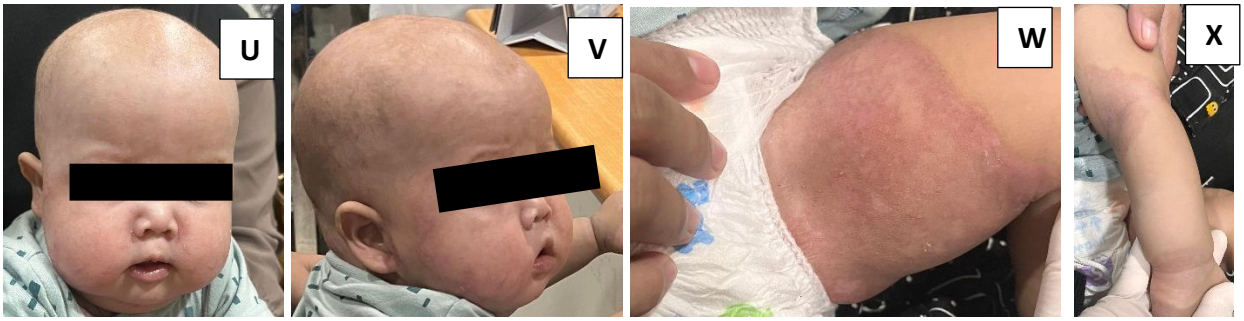

Figure 6. U, V, W, and X. Day-8 After Inward, desquamation are gone and the erythem are minimal

Discussion

S. aureus-specific ETs (serological forms of staphylococcal) mediate the progression of the disease's clinical manifestations. ET is a serine protease exotoxin that is divided into ETA and ETB. These two parts are referred to as epidermolytic toxins, epidermolysins, and exfoliatins. The ETs that cleave the protein desmoglein (Dsg)-1 in the superficial epidermis induce disturbance of keratinocyte adhesion and consequent intra-epidermal splitting at the level of the granulosa layer. These toxins are heat-resistant and resistant to proteases and acidic pH, allowing them to survive in harsh environments and cause damage to the skin. Both ETA and ETB can cause considerable harm to the skin, resulting in the development of blisters, bullae, and skin exfoliation. The typical

skin signs of SSSS include generalized erythema, blistering, and desquamation, with a preference for friction zones and sparing mucosal membranes (Brazel et al., 2021; Kusumo & -, 2022; Medugu et al., 2023).

In typical presence, the first symptom of SSSS is fever and redness across the whole surface of the skin. Within 24-48 hours, blisters with fluid collection appear all over the body. The blisters break, resulting in a burn. A large part of the skin's surface has peeled off or fallen away, indicating exfoliation or desquamation. Following a 24-hour infection, the skin's surface peels off in little sheets, leaving the topical surface wet and red. Fluid-accumulated blisters that resembled tissue paper were found in the groin, armpits, nose, and ears. The skin rashes quickly extended to the trunk and other legs. Skin rashes linked to SSSS are frequently found in the diaper area of babies (Mishra et al., 2016).

Based on anamneses, a 7^{th} year old infant is having reddish skin peeling on the face and almost the entire body for the past 3 months, started from the face and then increased to the head, hands, body, groin, and feet. The lesion is itchy and making patients feel uncomfortable. If there is a trauma on the skin, the lesion will appear. There were no fever or bullae appereance in these case. In a case report by Nusman et al (2023), desquamation without preceding blister formation is known as the scarlatiniform variety (Nusman et al., 2022).

SSSS mainly happens acutely, but in this case it happens in 3 months. The patient did not take any medication except cefadroxil and compounded cream by an online dermatologist. In the case of SSSS, a 14-day course of antibiotics will usually treat the patient and prevent a relapse. If there isn’t any complication and the disease is not severe, usually SSSS will be self-limited. Recurrence in SSSS is rare, but it has also been reported in patients who have immunodeficiency, premature infants, and underlying disease. Proper hygiene and decolonization should be discussed with both the patient and their family. This should include the application of mupirocin ointment and hibiclens baths (Bhavsar et al., 2016). The chronic occurrence of SSSS was also reported by Shelley et al (2002), a staphylococcal scalded skin syndrome (SSS) that continued for two years in a 50-year-old lady with cerebellar ataxia and epilepsy.(Shelley et al., 1998) Dehydration, weakness, exhaustion, and severe pain surrounding the infection site are further signs of SSSS. Secondary infection can also happen because of skin barrier damage. In this case, infants showed diarrhea, dehydration, and discomfort.

The differential diagnoses of SSSS in this case are SJS/TEN, Epidermolysis Bulosa (EB), pemphigus, and Leiner disease. SJS/TEN can be recognized by the history of drug consumption, inflammation and ulceration of mucocutaneous, excessive epidermal and keratinocyte necrosis, and ocular complaints such as conjunctivitis, visual impairment, ulceration, symblepharon, and ectropion. Skin examination showed a positive nikolsky sign. Histological analysis demonstrates full-thickness epidermal necrosis and a subepidermal split, in contrast to the intra-epidermal split in SSSS. In this case, it is became a differential diagnosis with SJS/TEN because of positive Nikolsky sign, inflammation and erosion of cutan, and ectropion. However, in this case, there is no mucosal

involvement and from the biopsy blisters are observed in the superficial intradermal layer in the subcorneal layer and there is no epidermal necrosis (Nusman et al., 2022; Tóth et al., 2023).

EB differentiate with SSSS by the level of blister formation: simplex (intra-epidermally at the basal membrane), junctional (at the epidermal-dermal junction), and dystrophic (intradermally) while pemphigus has a distict clinical presentation with deep epidermal skin splitting and oral mucosa involvement in pemphigus vulgaris and the presence of intra-epidermal immunoglobulins in pemphigus foliaceus (Nusman et al., 2022).

Infants who have seborrheic dermatitis may develop Leiner's illness, also known as erythroderma desquamativum, which is characterized rough and scaly erythema (erythroderma) and frequently a secondary bacterial infection. There is vomiting, diarrhea, and anemia. Erythema on the body and rough scale (from pytiraisiform to lamellar squama) were the skin manifestations, which were followed by fever, diarrhea, anemia, vomiting, failure to thrive, and weight loss. Usually occurring between weeks three and twenty-three, this illness develops after birth. In Leiner, the squama are reddish on scalp, face, or the area around the mouth and gluteal that turn into yellowish white color throughout the body with multiple varying sizes. The histological changes consist of acanthosis, parakeratosis, hyperkeratosis, parakeratosis, papillary edema, and mild lymphocytic infiltration in the epidermis, papillary and deeper vascular layers. Although this case is having lamellar squama, erythema, and secondary bacterial infection, the patients did not have history of allergy leads to seborrheic dermatitis, and the biopsy result is different from Leiner’s (Halim et al., 2023).

The diagnosis of SSSS is made clinically and confirmed by culture of ET generating S. aureus and a skin biopsy (Nusman et al., 2022). Although having atypical presentation, from skin biopsy it is confirmed that this case is SSSS. SSSS can be treated by an intravenous anti-staphylococcal antibiotic, such as nafcillin or oxacillin, which should be administered right away. If treatment is not working or if MRSA is detected, vancomycin should be tried. Clindamycin may diminish the formation of exfoliative toxins, although resistance prohibits monotherapy. Supportive care, including adequate IV fluids, temperature control, and wound care, is also required. Children's skin is expected to heal without scars in around two weeks (Grama et al., 2016).

This case using cefotaxime for intravenous antibiotic therapy, based on Cefotaxime therapy for staphylococcal infections has resulted in clinical cure rates ranging from 78%-100% and bacteriologic eradication rates ranging from 85%-100% in a wide range of illnesses (Aldridge, 1995).

Conclusion

Staphylococcal Scalded Skin Syndrome (SSSS) caused by epidermolysis exotoxins produced by some strains of Staphylococcus aureus and typically characterized by denudation of the skin and presents as large superficial blisters. Although it can be diagnosed clinically, sometimes SSSS having atypical presentation like in this case and can mimic another diagnosis. Therefore, to confirm it, the role of a skin biopsy or culture is needed.

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