Staphylococcal Scalded Skin Syndrome in a Child with Acute Lymphocytic Leukemia

Sare Gökdere1, Zeliha Güzelküçük1, Melek Işık1, Eylem Şerife Kaymaz1, Belgin Gülhan1, Neşe Yaralı2

1 Clinic of Pediatrics, Health Sciences University Ankara Children Health and Diseases Hematology Oncology Training and Research Hospital, Ankara, Turkey
2 Clinic of Pediatric Hematology and Oncology, Health Sciences University Ankara Children Health and Diseases Hematology Oncology Training and Research Hospital, Ankara, Turkey
3 Clinic of Pediatric Infectious Diseases, Health Sciences University Ankara Children Health and Diseases Hematology Oncology Training and Research Hospital, Ankara, Turkey

Case Report

A 4-year-old boy diagnosed with high risk (BCR/ABL positive) ALL was admitted to our hematology clinic with ear pain while receiving ALL-IC BFM 2009 reinduction treatment including dexamethasone, doxorubicin, L-asparaginase and imatinib and secondary fungal prophylaxis with voriconazole. He was afebrile but had mild respiratory symptoms such as rhinorrhea. His physical examination was normal except redness and bulging of tympanic membrane on his otoscopic examination. He was hospitalized, and 150 mg/kg dose of sulbactam-ampicillin was started and chemotherapy protocol was continued.

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by the exfoliative toxin of *Staphylococcus aureus* (1). There is an overall incidence of SSSS between 0.09 and 0.13 cases per 1 million inhabitants annually (2). The accurate incidence of SSSS in immune deficient population is not known, and it is supposed that it is very rare; only a few case reports particularly in adult patients have been reported. In this study, we present a patient with a diagnosis of acute lymphocytic leukemia who developed SSSS after suppurative otitis media during febrile neutropenia attack.

Keywords: Staphylococcal scalded skin syndrome, neutropenic fever, acute lymphocytic leukemia

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by the exfoliative toxin of *Staphylococcus aureus* (1). There is an overall incidence of SSSS between 0.09 and 0.13 cases per 1 million inhabitants annually (2). The accurate incidence of SSSS in immune deficient population is not known, and it is supposed that it is very rare; only a few case reports particularly in adult patients have been reported (3).

Here, we report a child with acute lymphocytic leukemia (ALL) who developed SSSS after suppurative otitis media infection during a febrile neutropenia attack.

Staphylococcus haslanmış deri sendromu (SHDS) *Staphylococcus aureus* eksfoliyatif toksinin yol açtığı nadir bir hastalıktır. İnsidansı yılda bir milyonda 0.09 ila 0.13 arasında değişmektedir. İmmünyetmezlikli bireylerdeki insidans ise bilinmemektedir ancak çok nadir olduğu tahmin edilebilir. Bu konuda yetişkin hastalarda çok az olgu bildirilmür. Bu ya- zida, febril nötropeni atağı sırasında süpüratif otitis media sonrası SHDS gelişen akut lenfositik lösemi tanılı bir çocuk hasta sunulmuştur.

Anahtar Kelimeler: Stafilokoksik haslanmış deri sendromu, nötropenik ateş, akut lenfositik lösemi

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by the exfoliative toxin of *Staphylococcus aureus* (1). There is an overall incidence of SSSS between 0.09 and 0.13 cases per 1 million inhabitants annually (2). The accurate incidence of SSSS in immune deficient population is not known, and it is supposed that it is very rare; only a few case reports particularly in adult patients have been reported (3).

Here, we report a child with acute lymphocytic leukemia (ALL) who developed SSSS after suppurative otitis media infection during a febrile neutropenia attack.

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by the exfoliative toxin of *Staphylococcus aureus* (1). There is an overall incidence of SSSS between 0.09 and 0.13 cases per 1 million inhabitants annually (2). The accurate incidence of SSSS in immune deficient population is not known, and it is supposed that it is very rare; only a few case reports particularly in adult patients have been reported (3).

Here, we report a child with acute lymphocytic leukemia (ALL) who developed SSSS after suppurative otitis media infection during a febrile neutropenia attack.

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by the exfoliative toxin of *Staphylococcus aureus* (1). There is an overall incidence of SSSS between 0.09 and 0.13 cases per 1 million inhabitants annually (2). The accurate incidence of SSSS in immune deficient population is not known, and it is supposed that it is very rare; only a few case reports particularly in adult patients have been reported (3).

Here, we report a child with acute lymphocytic leukemia (ALL) who developed SSSS after suppurative otitis media infection during a febrile neutropenia attack.

Abstract

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by the exfoliative toxin of *Staphylococcus aureus* (1). There is an overall incidence of SSSS between 0.09 and 0.13 cases per 1 million inhabitants annually (2). The accurate incidence of SSSS in immune deficient population is not known, and it is supposed that it is very rare; only a few case reports particularly in adult patients have been reported (3).

Here, we report a child with acute lymphocytic leukemia (ALL) who developed SSSS after suppurative otitis media infection during a febrile neutropenia attack.

Staphylococcal scalded skin syndrome (SSSS) is a disease caused by the exfoliative toxin of *Staphylococcus aureus* (1). There is an overall incidence of SSSS between 0.09 and 0.13 cases per 1 million inhabitants annually (2). The accurate incidence of SSSS in immune deficient population is not known, and it is supposed that it is very rare; only a few case reports particularly in adult patients have been reported (3).

Here, we report a child with acute lymphocytic leukemia (ALL) who developed SSSS after suppurative otitis media infection during a febrile neutropenia attack.
Two days later though, he was neutropenic with white blood cell as 200/µL, absolute neutrophil count as 100/µL. He was still afebrile, and otorrhea occurred. CRP level raised to 35 mg/dL (range 0.1-0.8 mg/dL), and he was accepted as febrile neutropenic and antibiotherapy was changed to piperacillin-tazobactam. Ear discharge, peripheral blood and catheter cultures were obtained. Chemotherapy protocol was skipped, and topical ear drops of ciprofloxacin and dexamethasone were added to the therapy. Although blood and catheter cultures were negative, S. aureus was identified at his ear discharge culture, which was sensitive to most antibiotics, and the therapy of piperacillin-tazobactam was continued. On 6th day, otorrhea and otalgia endured and his temperature increased to 40°C; temporal bone and thorax computerized tomography scan was performed and bilateral mastoiditis and pneumonic consolidation on the left lung lower lobe was demonstrated. Furthermore, a skin rash beginning from the toe and hand fingers that spread over the scalp, hand and joint locations like the axilla, inguinal and cubital area occurred (Figure 1). The rash was red and flaccid blistering and looked like scalded skin. Due to the progressive worsening of his condition, the antibiotics were modified to meropenem, vancomycin and liposomal amphotericin. Supportive therapy with intravenous immunoglobulin and granulocyte transfusions was also given. With this therapy, skin lesions and symptoms got better day-by-day. After using meropenem and vancomycin for 21 days, the patient’s symptoms regressed.

**Discussion**

In patients with neutropenic fever, the pathogenic organism varies from institute to institute (4). The most commonly identified microorganisms are gram-negative organisms such as *Pseudomonas* and *Klebsiella* but staphylococcal infection can also be life threatening in our clinic. Though coagulase negative staphylococcus is the predominant pathogen mostly identified in catheter cultures of our patients, *S. aureus* is an infrequent microorganism isolated in our febrile neutropenic patients (5).

*Staphylococcus* spp. cause both systemic and cutaneous infections, including impetigo, furuncle, subcutaneous abscess, SSSS, toxic shock syndrome (TSS) and neonatal toxic shock syndrome-like exanthematous disease (6). The incidence in pediatric and immunosuppressed patients is not clear, only a few cases have been reported in adult patients (3). Among multiple virulence factors, staphylococci secrete several exotoxins directly associated with particular disease symptoms including toxic shock syndrome toxin 1 (TSST-1), enterotoxins, and exfoliative toxins (ETs). The latter are particularly interesting as the sole agents responsible for SSSS. Exfoliative toxins induce the acantholytic effect of *S. aureus* due to the interruption of cell-to-cell connection, which allows the pathogens to spread within the epithelium. Moreover, *S. aureus* expresses exotoxins act like superantigens that induce T-cell activation with subsequent energy and immunosuppression. Toxins are excreted by the

![Figure 1. A: Skin rash on the axilla, B: Skin rash on the toe, C: Skin rash on the hand, D: Skin rash on the scalp.](image-url)
Seventy day old girl with acute lymphoblastic leukemia presented in our hospital’s emergency department with fever (38.4°C), rash, diarrhea, and vomiting. The patient’s mother noted that she had received a dose of chemotherapy the day before admission, along with antibiotics and antipyretics. The patient’s temperature had increased to 39°C, white blood cell count was 0.1 x 10^9/L, and platelet count was 70 x 10^9/L.

**Hypotension and Laboratory Findings:**
- Blood cultures were positive for Staphylococcus aureus.
- Urine analysis showed proteinuria and hematuria.
- Liver function tests were abnormal, with elevated AST and ALT levels, suggestive of renal involvement.
- Serum electrolytes were within normal limits.
- Complete blood count revealed a predominance of lymphocytes and anemia.

**Clinical Course:**
- The patient was treated with intravenous fluid resuscitation, broad-spectrum antibiotics, and supportive care.
- The rash progressed to widespread desquamation, similar to staphylococcal scalded skin syndrome (SSSS).
- The patient’s condition stabilized after 72 hours of treatment, and she was discharged with no residual symptoms.

**Discussion:**
- SSSS is a rare, serious skin condition characterized by a rapidly progressive epidermal detachment.
- The causative agent is Staphylococcus aureus, which produces exfoliative toxins A and B.
- The prognosis is favorable with early diagnosis and appropriate management, but it can be fatal in immunocompromised patients.

**Conclusion:**
- Early recognition and aggressive management of SSSS are crucial for a favorable outcome.

**References:**