Staphylococcal Scalded Skin Syndrome in Child. A Case Report and a Review from Literature

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ABSTRACT

Staphylococcal scalded skin syndrome (SSSS) is the medical term used to define a skin condition induced by the exfoliative toxins produced by Staphylococcus aureus. The disorder is also known as Ritter disease, bullous impetigo, neonatal pemphigus, or staphylococcal scarlet fever. The disease especially affects infants and small children, but has also been described in adults. Prompt therapy with proper antibiotics and supportive treatment has led to a decrease in the mortality rate.

The current case report describes the clinical progress of a patient with generalized erythema and fever, followed by the appearance of bullous lesions with tendency to rupture under the smallest pressure, and with extended areas of denudation.

The patient aged four years and six months was admitted to our clinic to establish the aetiology and treatment of a generalized bullous exanthema, followed by a skin denudation associated with fever and impaired general status. Based on clinical and paraclinical examinations a diagnosis of Staphylococcal scalded skin syndrome was established which responded favourably to antibiotic treatment, hydro-electrolytic re-equilibration, and adequate local hygiene.

Staphylococcal infection can represent a problem of significant pathological importance sometimes requiring a multidisciplinary approach involving paediatricians, dermatologists, infectious diseases specialists, and plastic surgeons.

Keywords: Staphylococcus aureus, child, scalded skin

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INTRODUCTION

Staphylococcus aureus is a Staphylococcus type pathogen, which can produce suppurated infections or sepsis with different types of manifestation such as impetigo, pyodermas, infections of the skin, and pneumonia. Staphylococcal scalded skin syndrome (SSSS) is caused by an exfoliative toxin produced by roughly 5% of Staphylococcus aureus [1]. Two exfoliative toxins (ETA and ETB) have been isolated and characterized, but the mechanism that leads to exfoliation has, until recently, been uncertain. These toxins act at a remote site resulting in a red rash and separation of the epidermis beneath the granular cell layer. Bullae form, and diffuse sheet like desquamation occurs [2]. Two types of SSSS are thought to exist: a localized form, in which there is only areal involvement of the epidermis, and a generalized form, in which significant areas are involved, remote from the initial site of infection [3]. Staphylococcal scalded skin syndrome is most common in children and neonates, and it has been described in adults with renal failure, immunologic deficiency, and other chronic illnesses [4-7]. Children present a higher risk due to their lack of immunity and immature renal clearance ability [8].

Staphylococcal scalded skin syndrome presents as a macular erythema followed by diffuse epidermal exfoliation. A prodromal localized staphylococcal infection of the skin, throat, nose, mouth, umbilicus, or gastrointestinal tract usually occurs. General malaise, fever, irritability, skin tenderness may be noted. Other signs can also be present, such as facial edema, conjunctivitis, and perioral crusting. Mucous membranes are spared, but dehydration may be present and significant. Nikolsky’s sign may be noted [9].
The treatment includes the administration of antistaphylococcal antibiotics, liquids, electrolytes and the local treatment of the denuded areas [10].

**Case presentation**

The clinical progress of a patient with a macular erythema followed by diffuse epidermal exfoliation from the face, thorax, members, and scalp is described. The patient aged four years and six months, was admitted to the Paediatrics Clinic 1, Tirgu Mures, in December 2015 to establish the diagnosis of a generalized skin rash associated with fever. The family and the personal physiological histories were not known because the patient has been in foster care since the age of eight months. The personal pathological history disclosed frequent upper airways infections, acute adenoiditis and tonsillitis.

The subject and the institution agreed with presentation of these data and all the procedures described were carried out following approval of the institution where the patient was treated.

**The history of the disease**

The onset of the disease occurred approximately thirty days before the admission to the clinic, with the development of an erythematous rash on the face and thorax, afterward extended onto the entire body. The patient was initially admitted to a territorial Clinic of Dermatology where antibiotic treatment with cephalosporin and corticosteroids for an acute generalized exanthema pustulosis, was administered. Because after seven days of treatment there was no improvement, the patient was admitted to our clinic for investigations and specialty treatment.

**The clinical exam**

The clinical exam on admission recorded malaise, severe loss of appetite, irritability, fever generalized erythematous rash, easily ruptured liquid containing bullae, a respiratory rate of 23/min, SaO₂ 94%, heart rate 125/min, blood pressure 70/50 mmHg, crusted lips, and accelerated bowel transit. After three days, the patient presented with facial oedema, extended seborrhea of the scalp, perioral crusting, skin pruritus, and pain on palpation. Skin erosions on the face, body and limbs were caused by gentle stroking of the skin, but the mucosae were not affected. (Figure 1A and 1B)

**The laboratory investigations**

On admission the CBC count showed a leucocytosis (17.75x10³/µl) with neutrophilia (63.5%), a mild anemia (haemoglobin 11.4g/dl, haematocrit 34%), thrombocytosis (672x10³/µl), and positive inflammatory markers, (C-reactive protein: 53.75 mg/dL, erythrocyte sedimentation rate 43 mm/h, PCT 2 ng/ml). Hepatic and renal functions were within normal limits. Other blood tests, including total bilirubin, glucose, and blood gasses, were carried out and all were within normal limits. A gram stain and a culture of the nasal and pharyngeal secretions confirmed a *Staphylococcus aureus* (MSSA) infection. A microbiological culture from cutaneous secretions was sterile. Blood culture and HIV tests were negative. A chest radiograph ruled out pneumonia as the original focus of infection. Abdominal ultrasound was normal, except for a plied gallbladder, and abdominal distension, with the fluctuation of air.

![Fig. 1A and 1B. The evolution of the patient in the first day of admission](image-url)
Prompt treatment was initiated including parenteral anti-staphylococcal antibiotic (Oxacillin), fluconazole, correction of the hydro-electrolytic imbalance, followed by maintenance therapy with consideration that fluid losses from exfoliation of skin similar to a burn patient, proton-pump inhibitors (PPIs), probiotics, and local skin therapy. The clinical outcome was favourable, The CBC count was within normal limits (leukocytes 8.40x10^3/μl, haematocrit 37.4%, haemoglobin 12.4g/dl, platelets 320x10^3/μl), negative inflammatory markers: C-reactive protein: 1.60 mg/dL, erythrocyte sedimentation rate 12 mm/h.

The patient was discharged from the hospital two weeks after admission. The parents were recommended to carry out appropriate hygiene measures.

There was complete healing without significant scarring as a result of the treatment. (Figure 2 and 3).

**The positive and differential diagnosis**

Staphylococcal scalded skin syndrome differs from bullous impetigo. Both are blistering skin diseases caused by staphylococcal exfoliative toxins. In the present case, the exfoliative toxins were spread haematogenously from a localized source causing epidermal damage at distant sites. For this reason, cultures of the bullous material were sterile. In bullous impetigo, the exfoliative toxins are restricted to the area of infection, and bacteria can be identified in a culture of the contents of a blister. Differentiating SSSS from toxic epidermal necrolysis (TEN), which carries a much higher mortality, is important. In our case, the mucous membranes are spared, while in TEN, the mucous membranes of the mouth, conjunctiva, trachea, oesophagus, and anus, are almost always affected.

The definite diagnosis of SSSS, in this case, represented a therapeutic challenge because the patient was initially treated in another medical unit with corticosteroids for an acute generalized exanthema pustulosis, which led to the delay of appropriate treatment. Nevertheless, the clinical development of the patient was favorable, and the interdisciplinary approach provided a satisfactory outcome.

**Discussion**

*Staphylococcus aureus* frequently infects the skin. The most common bacterial infection in children is impetigo, which accounts for approximately 10% of all skin problems [11]. A retrospective study over a 12-year period reviewed the presentation, etiology, and prognosis of nonburn epidermal loss. Only 9% of 19 patients had confirmed staphylococcal scalded skin syndrome [12]. *Staphylococcus aureus* may cause cutaneous and systemic infections such as staphylococcal scalded skin syndrome (SSSS) and toxic shock syndrome (TSS). Although exfoliative toxins A and B, which cause SSSS, and TSS toxin-1 may be produced by different strains of *S aureus*, the two syndromes rarely occur simultaneously [13]. Bullous impetigo due to *S aureus* is one of the most common bacterial infections of man, and its generalized form, SSSS is a frequent manifestation of staphylococcal epidemics in neonatal nurseries [14]. Infants and small children are predisposed to SSSS because of both the inefficient mechanisms related to renal clearance and the lack of protection through antitoxin antibodies. The disorder can also be present in adult, but only rarely [15]. Recent reports show an increase of admission rates and prescriptions for staphylococcal disease, including SSSS, in England [16].

Morbidity in children who develop cellulitis, sepsis, and pneumonia can be significant [17].

Staphylococcal scalded skin syndrome has been reported as a complication of septic arthritis after elective right knee arthroscopy and lupus nephritis associ-
ated with chronic immunosuppressant therapy [6]. No
gender predilection is documented in children. There
is at least one report of recurrent SSSS in a neonate
[18]. The mortality rate caused by SSSS in children is
very low (1-5%), and is much lower in children than in
adults unless associated sepsis or an underlying serious
medical condition exist [19].

Adults with staphylococcal scalded skin syndrome
often have blood cultures positive for toxigenic *S. au-
reus*, and mortality rates can be high (>60%) [17].
Complications are usually the result of sepsis, superin-
fecion, and dehydration or electrolyte imbalance due
to denuded skin. Another significant complication of
sepsis is represented by septic secondary determina-
tions in other sites or organs, such as septic arthritis
[20].

A review of data on isolates referred to the Health
Protection Agency’s Staphylococcal Reference Unit
identified 27 deaths in 27 months from infections with
invasive *S. aureus*. These deaths occurred in previously
healthy people with community-onset pneumonia,
bacteremia, or severe skin and soft tissue infection
[21]. The asymptomatic presence of *Staphylococcus
aureus* in places such as the nasopharyngeal or per-
ineal area has been recorded in a large percent of the
population. Several studies indicate the perineal pres-
ence of *Staphylococcus aureus* in over 90% of women
with Staphylococcal Toxic Shock Syndrome [22,23].
The initial signs and symptoms can sometimes mis-
guide the physician into establishing a diagnosis of
alergodermia and initiating treatment with corticos-
teroids [24]. The patient in the present case was also
treated initially with corticosteroids, the condition
being mistaken for acute generalized exanthema pus-
tulosis. The diagnosis of SSSS is mainly a clinical one.
The skin exfoliates initially around the mouth, neck,
and subsequently on the body and limbs. The mucos-
ae are not involved and Nikolsky’s sign is positive. All
of these clinical signs were present in the present pa-
tine. According to The Third International Consensus
Definitions for Sepsis and Septic Shock (Sepsis-3), the
patient presented with the qSOFA Criteria (quick Sofa
Score) for sepsis identification, i.e. alteration in men-
tal status, decrease in systolic blood pressure of less
than 100 mmHg, respiratory frequency greater than
22 breaths/min [25]. No organ failure was identified.
Septic shock is a severe, life-threatening condition in
which the circulatory impairment and cellular/meta-
bolic damages are profound enough to substantially
increase mortality [26]. Recently, multiple biomarkers
have been studied. A study performed on septic pa-
tients underlined the fact that even though C-reactive
protein and procalcitonin levels are not useful in di-
agnosing severe sepsis, they can be used in order to
predict the fatal progression of this disorder [27]. So-
luble urokinase-type plasminogen activator receptor
(suPAR) is another important biomarker that seems
have a predictive capacity for bacteremia in sepsis,
even though it appears to be an independent factor
for mortality prognosis in septic patients [28]. An-
other study, assessed the levels of Angiopoietin 2 and
Tyrosine kinase 2 in septic patients and patients with-
out sepsis, and concluded the fact that these two bio-
markers had independent diagnostic value in septic
patients [29]. Once SSSS is diagnosed, the treatment
consists of supportive care and eradication of the pri-
mary infection with anti-staphylococcal antibiotics
(Oxacillin, Vancomycin) administered by vein for a
minimum of seven days. In this case culture of the
nasal and pharyngeal secretion confirmed a staphylo-
coccal infection (*Staphylococcus aureus* MSSA). Most
staphylococcal infections involved in staphylococcal
scalded skin syndrome contain penicillinases and are
resistant to penicillin. In penicillinase resistance cas-
es, synthetic penicillin such as nafcillin or oxacillin
should be given immediately. In areas with significant
MRSA prevalence, or if MRSA is suspected, antibiot-
cics with MRSA coverage, e.g. vancomycin or linezolid,
are indicated [30]. Clindamycin is recognized as be-
ing useful in staphylococcal infections [31]. Patients
need fluid rehydration, topical wound care similar
to the care for thermal burns. In our case the child’s
treatment and monitoring were performed by a inter-
disciplinary team of paediatricians, dermatologists,
infectious diseases specialists, and a plastic surgeon.
This interdisciplinary approach resulted in a favoura-
ble resolution of the condition without complications
or superinfection of skin areas.

**Conclusions**

The occurrence of fever and exanthema, regardless of
patient age, should point to a possible differential di-
agnosis of staphylococcal scalded skin syndrome,
especially as SSSS is mainly a clinical diagnosis. The cur-
rently reported case raises awareness of the importance
of a multidisciplinary approach in patients presenting
with cutaneous symptoms of unknown aetiology.
CONFLICT OF INTEREST

Nothing to declare

REFERENCES


