

Case Report

Staphylococcal scalded skin syndrome: case report

Síndrome da pele escaldada estafilocócica: relato de caso

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ABSTRACT: Staphylococcal scalded skin syndrome is a bullous disorder induced by exfoliative toxins associated with *Staphylococcus aureus* infection. Its exact mechanism is still uncertain, but it manifests with the formation of bullous lesions spread throughout the body and it usually affects children under 5 years of age. The case reported presents a 4-month-old child with skin lesions of recent onset, hyperemia, and blisters that evolved to desquamation on the trunk, upper limbs and face, in addition to peripalpebral edema, bilateral eye discharge, and crusts in the perioral region. The prognosis is favorable and the treatment consists of systemic or oral antibiotic therapy and supportive therapy to manage hydroelectrolytic changes and thermoregulation. As the lesions improved and the patient was in good general condition, and considering that the antibiogram showed sensitivity, the regimen was changed to outpatient treatment with Sulfamethoxazole + Trimethoprim for another 7 days. The patient had a good response to the treatment and evolved with no scars.

Keywords: Staphylococcal scalded skin syndrome; *Staphylococcus*; Infant; Impetigo.

RESUMO: A síndrome da pele escaldada estafilocócica é um distúrbio bolhoso induzido por toxinas esfoliativas associado a infecção por *Staphylococcus aureus*, cujo exato mecanismo ainda é incerto manifestando-se com a formação de lesões bolhosas difusas pelo corpo que, usualmente, afeta crianças menores de 5 anos. O caso relatado apresenta uma criança de 4 meses de idade que manifestou lesões na pele de início recente, com hiperemia e bolhas no tronco, membros superiores e face, as quais evoluíram com descamação, além de edema peripalpebral, secreção ocular bilateral e crostas em região perioral. O prognóstico é favorável e o tratamento consiste em antibioticoterapia sistêmica ou oral, além de terapia de suporte na vigência de alterações hidroeletrólíticas e da termorregulação. Como o paciente apresentou melhora das lesões e encontrava-se em bom estado geral, o esquema foi trocado por Sulfametoxazol + Trimetoprima após o antibiograma evidenciar sensibilidade, por mais 7 dias a nível ambulatorial. O paciente apresentou boa resposta ao tratamento e evoluiu sem cicatrizes.

Palavras-chave: Síndrome da pele escaldada; Estafilococcia; Lactente; Impetigo.

Institution where it was conducted: Hospital São José do Avai

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INTRODUCTION

Staphylococcal Scalded Skin Syndrome (SSSS) is predominantly caused by group II staphylococcus, particularly strains 71 and 55, present in infection sites¹.

The clinical symptoms of Staphylococcal Scalded Skin Syndrome are caused by hematogenous dissemination, in the absence of specific antitoxin for staphylococcal exfoliative or epidermolytic toxins A and B, which are protease enzymes that react against a protein called desmoglein-1¹. Epidermolytic toxin A is heat-stable and encoded by bacterial chromosomal genes, and exfoliative toxin B is heat-labile and encoded by 37.5 kb plasmid. These toxins are located in the desmosomes of the granule cells in the granular layer, and cause their rupture without affecting neighboring cells^{1,2}.

SSSS occurs predominantly in infants and children under five years of age and includes a variety of diseases, going from bullous impetigo to generalized skin involvement with systemic disease³.

The diagnosis is clinical and differential diagnosis with other bullous disorder should always be performed. A skin biopsy can be performed to confirm the diagnosis when a subcorneal cleavage of the granular layer is found. The absence of inflammatory infiltrate is common, and the histology is identical to that seen in pemphigus foliaceus and subcorneal pustular dermatosis^{1,4}.

The prognosis is favorable, and the treatment consists of systemic or oral antibiotic therapy and supportive therapy³.

The objective of this study is to report a case of Staphylococcal Scalded Skin Syndrome and discuss a conservative approach, with an emphasis on diagnosis and treatment, contributing to early recognition and immediate management, since SSSS is rare in medical practice.

MATERIAL AND METHODS

The present study is a case report of a 4-month-old child admitted to Hospital São José do Avaí – Itaperuna/RJ with skin lesions of recent onset.

The study addresses a clinical case related to SSSS, based on a literature review on the subject, conducted in the databases Latin America and the Caribbean Literature on Health Sciences - LILACS, Medical Literature Analysis and Retrieval System Online – MEDLINE, and Scientific Electronic Library Online – SciELO as well as Google Scholar and books, with an emphasis on management, early diagnosis, pathophysiology, clinical picture, diagnosis, and treatment of the syndrome in children.

The database search was conducted in January

2021, using the Health Sciences Descriptor (DeCS): Staphylococcal Scalded Skin Syndrome; Pediatrics; Case Reports.

The articles were selected according to the following inclusion criteria: articles in Portuguese or English, available in full, published in the last 20 years (2001-2021), and covering the guiding question of the study by addressing SSSS, its pathophysiology, clinical picture, diagnosis, treatment, main complications, prevention, health education, and the role of pediatrics and dermatology professionals in the early diagnosis of the pathology.

The research instrument was based on medical records and laboratory and complementary exams, which supported patient diagnosis and treatment. After obtaining the written consent from the patient's guardian and from the health institution, the group members collected the variables in the information system of Hospital São José do Avaí.

CASE REPORT

A male 4-month-old child was admitted to Hospital São José do Avaí with skin lesions that had started 2 days before admission. The lesions began as hyperemia and blisters on the trunk, upper limbs, and face, and evolved to desquamation, palpebral edema, bilateral ocular discharge, and crusts in the perioral region. Blood count revealed a left shift (Leukocytes: 9,100 mm³; Band Neutrophils: 7%; Segmented Neutrophils: 33%; Eosinophils: 14%; Basophils: 0%; Monocytes: 5%; Lymphocytes: 41% and no alterations in inflammatory markers (C-reactive protein 0.4 mg/dl).

Eye discharge and skin lesion samples were collected with the aid of a swab (sterile swab used to collect microbiological exams for the purpose of clinical studies or research) and testing revealed *Enterobacter aerogenes* and *Staphylococcus epidermidis*, respectively. Scalded skin syndrome was diagnosed after evaluation by the dermatologist. Endovenous treatment with Oxacillin and Ceftriaxone was started, in addition to bathing with cleansing lotion for sensitive skin, application of sunflower oil after bathing, topical Mupirocin 2% in the nasal bridge twice a day, and Ciprofloxacin with topical Dexamethasone around the eyes. After the result of the antibiogram, which showed resistance to Oxacillin, Vancomycin was started. However, on the 4th day on the medication, the patient had an allergic reaction.

Considering the improvement of the clinical picture and the result of the antibiogram, the regimen was changed to outpatient treatment with Sulfamethoxazole + Trimethoprim for another 7 days. The patient had a good response to the treatment and evolved with no scars.



Figure 1: Child with skin lesions during the acute phase of the disease.

DISCUSSION

In SSSS, the onset of the skin rash may be preceded by prodromes such as malaise, fever, irritability, and pain on palpation^{3,5}. In the case reported, the patient was irritable before the onset of the skin lesions.

Other focuses of infection include the nasopharynx and, less commonly, the umbilicus, the urinary tract, skin excoriations, the conjunctivae, and the blood. Exotoxins can be produced following infections such as purulent conjunctivitis, otitis media, nasopharyngeal infection or localized bullous impetigo⁴. In the present case, we observed the presence of bilateral conjunctivitis with significant ocular discharge. A culture of the discharge was performed and showed growth of *Enterobacter aerogenes*, which was treated with antibiotic eye drops.

Scarlatiniform rash spreads diffusely and is accentuated in flexural and periorificial areas. Inflamed conjunctiva may become purulent. The skin with severe erythema can quickly become wrinkled, and in severe cases, flaccid blisters and lesions may appear diffusely. Eventually, perioral erythema can be found, as well as crusting and radial fissure around the eyes, mouth, and nose, as seen in Figure 3².

Blisters are commonly found on friction areas such as the trunk and extremities, and very extensive lesions can serve as a gateway for other invading agents. These blisters are generally intact and sterile, unlike those in bullous impetigo. However, culture samples from all suspected localized infection sites and from the blood must still be collected to identify the source of epidermolytic toxins^{3,4}.

At this stage, the Nikolsky sign can be found (Nikolsky's sign: partial or total displacement of the epidermis after application of perilesional pressure with a finger or a blunt object.), which is present when delicate tangential pressure on the epidermis results in skin peeling¹. As large sheets of skin peel away, denuded, moist, shiny areas are revealed, initially on the joints and subsequently on most of the body surface. This can lead to secondary skin infection, sepsis, and hydroelectrolytic disorders³.

The shedding phase begins 2-5 days after the onset of erythema; healing occurs without scarring in 10-14 days. Patients may have pharyngitis, conjunctivitis, and superficial lesions on the lips, but intraoral mucosal surfaces are not affected. Pain is accentuated on palpation of the skin^{2,5}.

Figure 2 shows a significant area of shedding on the trunk, which is an area with a lot of friction in this age group.



Figure 2: Shedding of an extensive area of the epidermis in the head and trunk.



Figure 3: Skin shedding in the thorax/abdomen region and involvement of the face with perioral and periocular lesions

The classic form of SSSS is known as Ritter's disease (Ritter's disease: formation of superficial skin blisters caused by infection with *Staphylococcus aureus*) and usually presents with the prodromes of malaise, irritation, fever, and skin erythema, which evolves within 24 to 48 hours to flaccid blisters all over the skin. Its mildest form is localized and is known as bullous impetigo. It usually has a good prognosis and, due to the superficial cleavage of the epidermis, the lesions tend to heal with little or no scarring^{1,4}.

Regarding the differential diagnosis, SSSS can be confused with other bullous and exfoliative disorders, including bullous impetigo, epidermolysis bullosa, epidermolytic hyperkeratosis, pemphigus, drug eruption, erythema multiforme, and drug-induced toxic epidermal necrolysis⁵.

Toxic epidermal necrolysis can often be differentiated by a history of drug ingestion, presence of Nikolsky's sign only on erythematous areas, absence of perioral crusting, full-thickness epidermal necrosis, and a cleavage plane of the blister in the lowest layer of the epidermis¹.

The biopsy can also contribute to this differentiation. It will reveal the cleavage site in the skin, which is

superficial in SSSS and much deeper in toxic epidermal necrolysis.

In the case described, a biopsy was not performed, as the patient did not have a suggestive clinical history, such as drug ingestion before the appearance of the lesions, and because *Staphylococcus epidermidis* was identified on the swab culture of skin lesions. However, in cases of doubt, skin biopsy allows differentiating SSSS from other bullous disorders.

Treatment should be with microbial agents, applied topically or systemically. In the case presented, systemic treatment was chosen, as the patient had an extensive area of involvement.

In topical treatment, emollients can be used to provide lubrication and decrease discomfort. Baths and compresses are also recommended for the debridement of necrotic tissue³.

Some authors claim that the use of topical antibiotics is unnecessary. However, reports in the literature indicate that agents such as Silver Sulfadiazine, Bacitracin, and Mupirocin are recommended, especially for localized infections, with attention not to use topical sensitizing agents⁵.

Systemic treatment should be oral in cases of localized involvement, or parenteral, with penicillinase-resistant semisynthetic penicillin, as staphylococci are usually resistant to penicillin. Clindamycin may be added to inhibit bacterial protein (toxin) synthesis. Oxacillin can be used for a minimum of seven days. Vancomycin can be used in case of superinfection or in patients allergic to penicillin. Finally, the use of aminoglycoside should be considered. It is important to avoid the use of steroids and non-steroidal anti-inflammatory drugs, as they impair immune and renal function, respectively^{1,3,5}.

Although recovery is usually quick, complications such as excessive fluid loss, electrolyte imbalance, pneumonia, septicemia, and cellulitis can increase the morbidity of SSSS².

CONCLUSION

The report presented alerts to the importance of an early recognition and diagnosis of SSSS, so that early treatment can be provided, avoiding the risks of inadequate treatment and the natural complications of the disease. In this way, it is possible to achieve a positive outcome in a disease with low prevalence and high complication rates in children. It is extremely important to differentiate the disease from other bullous disorders through histopathological analysis of skin lesions, due to their different forms of treatment and severity.

Participation of the authors: *Carmo RSV* - Responsible for data search and writing of the case report. *Pillo MD* – Elaborated the text. *Bartels R* - Conducted the literature review. *Cascabulho AME* – Responsible for proofreading the case report. *Verdan TVQ* - Responsible for the patient's hospital care. *Frizzo APM* - Responsible for reviewing the article. *Bastos MIR* - Responsible for the patient's hospital care and guidance. *Pimentel TM* – Responsible for the patient's hospital care. *Lessa LRK* - Responsible for the patient's hospital care and guiding of the report.

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