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CASE REPORT

A case report: Staphylococcal scalded skin syndrome in a minor infant

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***Abstract*:** Staphylococcal scald skin syndrome is an entity first described by Ritter Von Rittershain in 1878, which was called neonatal exfoliative dermatitis. This situation belongs to a group of pathologies caused by toxins produced by Staphylococcus aureus, called exfoliate A and B, which are characterized by a wide range of segmental lesions and degeneration. It is not a common pathology, but it most often occurs in newborns and children under the age of 5. This study describes a clinical case of a minor baby transferred to the emergency room by his mother, characterized by a systemic rash followed by follicle lesions, which is consistent with the Chikungunya fever (ChikV) outbreak reported in Maracaibo since June 2014.

Keywords: *Staphylococcus aureus*; exfoliatin; Chikungunya fever; neonatal exfoliative dermatitis

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Introduction

The staphylococcal scalded skin syndrome (SSSS) is a generalized cutaneous-ampullary pathology that belongs to a group of pathologies caused by Staphylococcus aureus toxins, a ubiquitous Gram-positive germ that is considered part of the human microbiota as it is found on the skin of healthy individuals (nostrils in 20%–40% of adults, intertriginous folds, perineum, axillae and vagina). It should be noted that 30%–50% of healthy adults are colonized, and 10%–20% remain persistently colonized; however, when the skin's defense systems are impaired, this microorganism can cause disease. In this regard, the main risk groups are individuals with type 1 diabetes, intravenous drug users, hemodialysis patients, surgical patients and individuals with acquired immunodeficiency syndrome[1–3]. This entity was first described by Ritter Von Rittershain in 1878, who called it neonatal exfoliative dermatitis, which is why it is also known as Ritter's syndrome[4].

The SSSS is not a common pathology, but it is more frequently observed in newborns and children under 5 years of age[5,6]. Its pathophysiological basis is based on the action of two exotoxins produced by *Staphylococcus aureus*[7]. Exfoliatin A and B are responsible—together with the immune function of the host—for the appearance of any of the three clinical forms identified in this syndrome: a) the localized form, known as bullous impetigo (most frequent form), which manifests as single blisters of small quantity because the toxins limit their action only to the infected area, causing an isolated bullous lesion or a

regional grouping of lesions; b) the abortive scarlatiniform form and c) the generalized form, in which staphylococcal toxins are released into the bloodstream from the primary infectious focus, acting at a distance and evolving clinically in three phases[8,9]:

· **First phase or erythrodermic stage**: in which a uniform erythematous rash is observed that respects the mucous membranes accompanied by pain on mobilization. The child cannot stand lying down or in the mother's arms, although it does not look toxic.

· **Second phase or blistering stage**: characterized by the appearance of flaccid blisters usually located in flexion areas and near the anatomical orifices. This stage has an important semiological feature: the positive Nikolsky's sign[10,11].

· **Third or desquamative stage**: it presents with detachment of the epidermis in large sheets that leave areas of moist desquamation and may resemble a scald burn. The appearance of the patient in this phase usually generates concern in family members, but the patient is usually stable and in good general condition unless complicated by infectious processes such as pneumonia or sepsis[12].

The purpose of the present study is to describe the clinical case of a minor infant transferred by his mother to the emergency department characterized by the appearance of a generalized rash that coincided with the onset of the *Chikungunya* virus (ChikV) fever outbreak in the city of Maracaibo, Venezuela.

Case report

This is an 8-month-old male infant who, according to maternal reference, began his illness on 11/11/2014, characterized by a fever that ranged between 38 °C and 40 °C (axillary), which improved with the administration of oral antipyretics (acetaminophen) and completely disappeared 24 hours later. However, the following day she reported the appearance of an erythematous and generalized skin rash that produced weeping on skin palpation and mobilization. After 24 hours this condition evolved to vesicular lesions on the upper and lower limbs, accompanied by greenish liquid evacuations in numbers of six, without mucus or blood, because it was decided to go to the doctor who indicated treatment with antipyretics, antihistamines, probiotics and antidiarrheals for four days without improvement, which is why she came to our health care center where after medical evaluation it was decided to admit her to the emergency department.

Perinatal history

Product of a 32-year-old mother in her IV gestation and III delivery, who was born vaginally without immediate complications with a birth weight of 2.8 kg and a height of 41 cm after a controlled pregnancy of 41 weeks duration according to the date of the last menstrual period. Serology for HIV, VDRL, TOXOTEST was negative. She had a urinary tract infection and leucorrhea in the II and III trimester of pregnancy, whose treatment was not specified.

Regarding feeding, the patient has been breastfed until now with NAN^{R} starter formula since 15 days of age at normal dilution without specifying the amount. From the psychomotor point of view and his psycho-biological habits, it is reported that he held his head at two months of age, having a quiet sleep only interrupted for feeding. Daily bowel habits, reporting liquid bowel movements with the current disease. Incomplete vaccination schedule and not documented at the time of history taking.

Family history

32-year-old mother is apparently healthy. 51-year-old father is with type 2 diabetes mellitus (DM2), and paternal grandparents also are with DM2.

Physical examination on admission (positive findings to the exam)

Age: 3 months. Weight: 5.8 kg. Height: 59 cm. Skin: Dermatosis characterized by small vesicles located mainly on the upper limbs extending to the trunk and lower limbs, which converge in some areas (feet) and form flaccid blisters, with positive Nikolsky's sign. And in other areas crusts are observed. Head: CC: 41 cm. Anterior fontanelle is normotensive. Thorax: CT: 40 cm, rough vesicular murmur, in both lung fields without aggregates, FR: 34x. Cardiac: HR: 125; RsCsRs, S/S. Abdomen: CA: 39 cm, soft abdomen depressible, not distended, RsHAs present. Skeletal muscle: edema is observed in both lower limbs, especially in the dorsum of the bilateral foot that leaves fovea. Neurological: Active, loud cry, normotonic and normoreflexic, with fasciitis on mobilization. Chest X-ray: (11/11/2014) Chest in normo configuration, no areas of consolidation.

Admission diagnoses

- Chikungunya fever under study.
- Acute febrile diarrhea with some degree of dehydration.
- Normal nutritional status by anthropometric assessment.

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- 5% of body surface area burned, type A-AB in both upper and lower limbs?
- Child abuse syndrome?

Laboratory studies

Table 1. Laboratory studies

Test	11/11/2014	15/11/2014	17/11/2014	Normal values
White count (× mm^3)	9,200	7,700	7,700	5,000–12,000
Segmented (%)	39.1	60	29	32
Lymphocytes (%)	45.3	35	61	61
Monocytes (%)	15.6	5	9	5
Eosinophils (%)	-	-	1	3
Hemoglobin (g/dL)	8.6	8.9	8.2	10.5–14.0
Hematocrit (%)	28.0	28.1	26.3	32–42
Platelets (× mm^3)	135.000	70.000	256.000	150.000–450.000
Glycemia (mg/dL)	-	80	-	60–99
Creatinine (mg/dL)	-	0.3	-	0.4–0.5
Sodium (mequiv/L)	-	133.5	-	135.0–145.0
Potassium (mequiv/L)	-	4.85	-	3.5–4.5
ESR mm 1 h Wintrobe	-	-	7	
Corrected ESR	-	-	0	
Hemoculture	-	Negative	-	-
Rotavirus	-	-	-	-

Antibiotic therapy received

Vancomycin (40 mg/Kg/day) and Cefepime (150 mg/Kg/day) for 10 days, due to regular general conditions, extensive areas of denudation and cutaneous involvement, and need for central nervous system (CNS) coverage in view of being a minor infant and risk of dissemination to CNS.

During her hospital stay, daily treatment with antiseptic soap is indicated, and also application of mupirocin ointment on the lesions located on the dorsum of the foot, three times a day, and moisturizing cream should be applied daily. In view of the conditions of the service, dermal exposure and serous secretion in some of the lesions, the patient was placed in isolation and the use of mosquito netting 24 hours a day was indicated. Subsequently, the lesions evolved into a dermatosis characterized by thick desquamation that resolved by day seven of in-hospital evolution, followed by the presence of hypochromic macules at the sites of vesicles and blisters. After 10 days of intravenous antibiotics, she was discharged due to clinical improvement, with subsequent follow-up 15 days after discharge, where an almost complete improvement of the pigmentary alterations was evidenced, without scars.

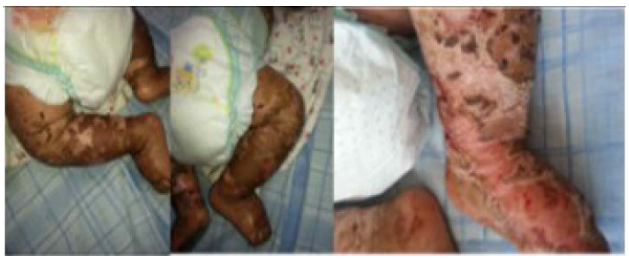

Figure 1. Extense area of moist cutaneous denudation due to blistering. Positve Nikolsky's sign.

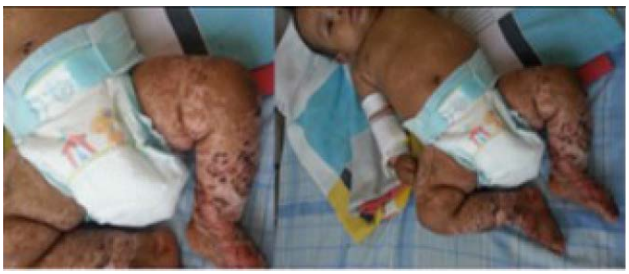

Figure 2. Desquamation in large squamous-crusted sheets.

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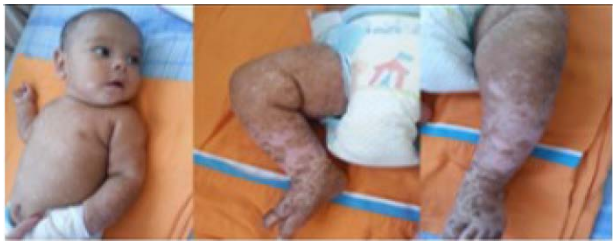

Figure 3. Flaky scales and residual hypopigmented areas.

Figure 4. Residual hypopigmented areas.

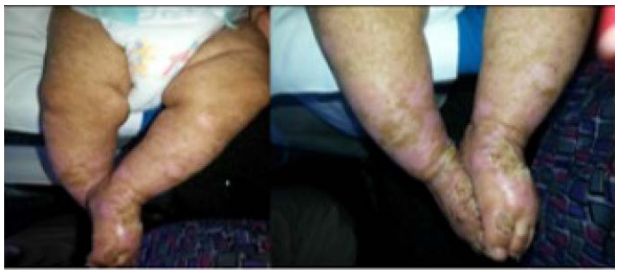

Figure 5. Clincial improvement with areas of reepithelialization and residual scaling.

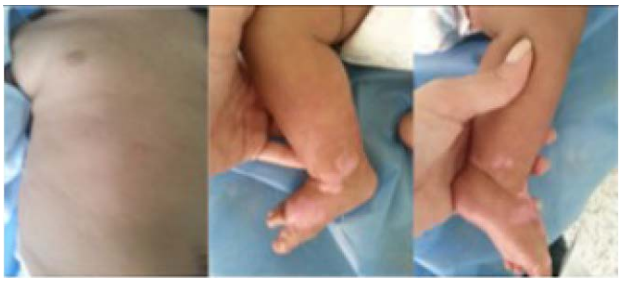

Figure 6. Evaluation 15 days after discharge. Improvement of cutaneous dyschromia and absence of scars.

In view of the clinical evolution, the discharge diagnosis is concluded: 1) staphylococcal scalded skin syndrome and, in a complementary manner; 2) normal nutritional status by anthropometric assessment, 3) Anemia.

Discussion

The staphylococcal scalded skin syndrome (SSSS) originates in an infectious focus involving Staphylococcus aureus, a bacterium that produces toxins known as exfoliatins (exotoxin type A and type B), which are serine proteases highly specific against cadherin desmoglein I, an adhesion protein present in the desmosomes of the stratum granulosum that facilitates adhesion between keratinocytes[9]. The resulting vesicles are intraepidermal clefts between the stratum corneum and stratum spinosum located above the basal cells (suprabasal). This condition is very similar to the autoimmune skin disorder pemphigus vulgaris produced by IgG antibodies against desmoglein[9].

This bacterium is usually located in the nasopharynx or conjunctiva, sites from which the toxins pass into the bloodstream and then spread to the skin where they affect the desmoglein I complex. It is important to note that the excretion of toxins is renal, therefore, immaturity or decreased renal functionalism may affect the course of the disease. It is for this reason that newborns are the most affected, and in cases where it has been observed in adults there is some type of comorbidity that directly affects the kidney[6].

The symptoms usually begin with fever accompanied by irritability and a diffuse erythematous rash on the palms, soles and mucous membranes, accompanied by pain in the affected areas that worsens with mobilization. Around 24 to 48 hours of evolution, flaccid blisters begin to appear that easily break (Nikolsky's sign)[10,11], and frequently look like a burn, a fact that should be taken into account for the possible differential diagnosis of this disease[1,7,13]. When the extension of the skin affected by the formation of phlyctenas or denuded areas is very extensive, dehydration and water and electrolyte disorders can be observed, especially in neonates and young infants.

The diagnosis of SSSS is purely clinical because the picture is quite characteristic, although skin biopsy can be performed to make a definitive diagnosis, its usefulness is more important in atypical cases. Paraclinical studies are not usually useful since neither hemogram nor acute phase reactants usually provide much information, and in children blood cultures are usually negative, as are cultures of blister secretions; toxins can be detected by ELISA or polymerase chain reaction (PCR) tests, but these are not usually done routinely[14–16].

The most important differential diagnosis is with toxic epidermal necrolysis or Lyell's syndrome but the patient looks toxic and with touch of general condition, also among the differential diagnoses we can include sunburn, drug reactions, Kawasaki disease, bullous impetigo, viral diseases, pemphigus and various pathologies where the main lesion are vesiculoampullary, but in all cases the diagnosis is usually mainly clinical1. Mortality in this condition is rare, and does not exceed 3% of cases,

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which is directly related to the number of comorbidities and complications during the disease[16].

Early evaluation and timely diagnosis to initiate early parenteral antibiotics has been shown to reduce complications and guarantee treatment success; cephalosporins and beta-lactamase inhibitors are the most frequently used, with good results; in the experience of our service, the use of glycopeptides in view of the increase in the frequency of methicillin-resistant Staphylococcus aureus infections in our population has also proven to be a valid option in the treatment of this disease. In addition to antibiotics, there are also general measures such as controlling the patient' s environment and improving general skin conditions to ensure early recovery of skin turgor, pigmentation or macroscopic appearance.

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