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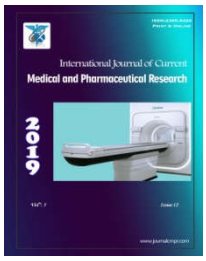

Case Report

TOXIC EPIDERMAL NECROLYSIS: A CASE REPORT IN PEDIATRICS

Mario Jorge dos Santos Noel Filho[1], Laís Viana Lopes Sato[1], Juliana Vieira de Oliveira[1], Marcella Lopes Abtibol[1], Rafaela Monique Mendonça Barros[1], Renata da Silva Almeida[2] and Felipe Alves Barbosa[3]

[1]Pediatric Resident Doctor at the Tropical Medicine Foundation Doutor Heitor Vieira Dourado, Brazil
[2]Pediatric Resident Doctor at Amazonas State University, Brazil
[3]Doctor at West Side Child Hospital, Brazil

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ABSTRACT

Toxic Epidermal Necrolysis (Lyell's syndrome) is a rare but very serious dermatological lesion. Treatment includes mainly the immediate suspension of the inducing drug and the precocious admission of the patient in a hospitalar facility with the capacity to provide intensive support care and to minimize the infectious risk, having also the conditions for the execution of surgical debridement and covering of the affected areas, that is to say in Burn Units. These illnesses are characterized as dermatological emergencies and its adequate management and cares must be part of the routine knowledge of the intensive care doctors.

INTRODUCTION

Ease of access to medicines and their uncontrolled use by population are public health issues, considering their risks, adverse events and toxicity. Negative consequences of drug use and occurrence of adverse events lead to an increase in hospital admissions and deaths. Adverse drug reaction (ADR) can be defined as the harmful and unintended response to a drug, which occurs at doses normally used for prophylaxis, diagnosis or treatment of a disease, or for the modification of a physiological function.1

ADR may occur in the form of mild skin rashes, which can sometimes be severe and lethal, such as in Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN).2 In SJS, body surface area affected does not exceed 10%.3 Cutaneous detachment between 10% and 30% characterizes a transition zone between SJS and TEN. Epidermis detachment superior to 30% characterizes TEN, also known as Lyell's syndrome.4

They are characterized by cutaneous erythema with formation of blisters and hemorrhagic erosions of mucous membranes, such as stomatitis, balanitis, colpitis, severe conjunctivitis and blepharitis. Despite severity of these conditions, especially TEN, there is still no consensus on the most appropriate management of patients, and there is great variability in the management of patients with SJS and TEN.

In order to evaluate prognosis of patients affected by SJS and TEN, there is a score that measures severity of cases (SCORTEN) and that must be performed within the first 48 hours of the disease.5

CASE REPORT

C.S.N 11 years old, male, born in COARI (Amazonas), diagnosed with epilepsy using carbamazepine for 06 months. Started unverified fever associated with bullous lesions on the skin and oral cavity.Sought medical attention complaining of otalgia, intense odynophagia, ulcerated lesions in the oral mucosa and polymorphic lesions (vesicles, rash and papules) with progression of 03 days. There was a worsening of the lesions and general condition during this period. Due to worsening he was transferred to an intensive care unit with more than 90% of the body surface compromised by the injury. Physical examination revealed vesicles with blackened areas (central necrosis) in the face, trunk and extremities, as well as periorbital edema associated with conjunctivitis. During hospitalization he was diagnosed with toxic epidermal necrolysis (TEN) triggered by carbamazepine and was classified with the severity score "SCORTEN: 4", performed biopsy of skin in the anterior region of the left thigh that confirmed the clinical diagnosis with the following findings:

Corresponding author:* **Mario Jorge dos Santos Noel Filho
Pediatric Resident Doctor at the Tropical Medicine Foundation Doutor Heitor Vieira Dourado, Brazil

epiderme with keratinocyte necrosis expense with presence of epithelium cells, circumscribed detachment of epiderme and derme.In derme there is a perivascular mononuclear infiltrate. Patient remained in the intensive care unit for 45 days, progressing satisfactorily and complete remission of the lesionsdespite the high risk of SCORTEN obtained a good response to clinical treatment.

DISCUSSIONS

Stevens-Johnson Syndrome (SSJ) and Toxic Epidermal Necrolysis (NET) are spectra of the same disease: severe drug-reactive eruptions such as sulfonamides, phenobarbital, carbazepine, and lamotridine.6In the pediatric population, cases of SSJ and NET are believed to be related to upper respiratory tract infections.7In cases considered as triggered by drugs when exposure to the drug occurs within 8 weeks of the onset of clinical manifestations.

The initial clinical picture of SSJ is nonspecific, characterized by fever and asthenia that progresses to macular rash affecting less than 10% of the body surface. Mucosal involvement in the oral, ocular and genital region may be painful and hemorrhagic in 90% of the cases.8

The diagnosis is clinical, considering that there is no laboratory test that establishes which drug caused the erythema, thus being the empirical diagnosis. Provocation tests are not indicated as exposure to the agent may trigger a new severe episode of SSJ / NET.9

Differential diagnosis should not forget toxic shock syndrome, graft versus host disease, generalized acute pustular disease, burns, Kawasaki disease, autoimmune dermatoses, among others.10

The most effective treatment is supportive treatment, similar to the treatment of burn patients.11 Patient survival depends on access to treatment and discontinuation of the causative drugs. 12

CONCLUSION

SSJ and NET characterize frequent dermatological emergencies and knowledge of their proper handling and early diagnosis are care that should be part of the routine of pediatric intensive care units.

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