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Toxic Epidermal Necrolysis (TEN) in a Patient with Human Immunodeficiency Virus: Case Report

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ABSTRACT

Stevens-Johnson Syndrome (SJS)/Toxic Epidermal Necrolysis (TEN) is a spectrum of mucocutaneous hypersensitivity reactions. This report details the case of an18-year-old male patient with a history of HIV who developed extensive skin lesions and mucosal involvement after using carbamazepine. The diagnosis was confirmed clinically, and it was treated with intravenous corticosteroids, crystalloid solutions, supportive care, and discontinuation of the implicated drug, resulting in favorable clinical evolution. This case highlights the importance of early recognition of SJS/TEN and immediate withdrawal of the causative agent to improve prognosis.

KEYWORDS: SSJ/TEN drug reaction systemic dermatoses HIV dermatosis in hiv

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INTRODUCTION

Stevens-Johnson Syndrome was first described in 1922 by pediatricians A.M. Stevens and F.C. Johnson, who reported two cases of children from New York City with a febrile mucocutaneous eruption of presumed infectious etiology. Initially, both were misdiagnosed as hemorrhagic measles or "black measles," until Stevens and Johnson recognized the unique clinical characteristics and associated symptoms1. Stevens-Johnson Syndrome (SJS)/Toxic Epidermal Necrolysis (TEN) are mucocutaneous hypersensitivity reactions, mostly induced by medications and associated with significant morbidity and mortality.

Stevens-Johnson Syndrome (SJS) is defined as less than 10% of the body surface area (BSA) being detached, with an overlap of SJS/TEN involving 10-30% BSA detachment, and TEN with more than 30% BSA detachment4.SJS has a global distribution, affects all races, and is more common in women with an approximate mortality rate of about 10%. In contrast, TEN is rare, with an annual incidence of approximately one case per million people. It affects all races and is more common in populations with higher drug consumption (elderly and women). The mortality rate for TEN has declined from nearly 80% to approximately 30%, though it remains high in the elderly (~70%)5. Among the risk factors that increase the likelihood of developing SJS/TEN are HIV infection, connective tissue disease, cancer patients, older adults, individuals of Black or Asian descent, increased drug dosages, and impaired renal function.

Over 80% of cases are associated with exposure to medications. The medications most frequently involved include sulfonamides, anticonvulsants (phenytoin, carbamazepine, lamotrigine), oxicams, allopurinol, nonsteroidal anti-inflammatory drugs (NSAIDs), pyrazolones and their derivatives, aminopenicillins, cephalosporins, macrolides, imidazoles, quinolones, oral hypoglycemics, and diuretics. Most cases occur between 4-28 days after initial exposure to the drug.

The pathophysiology of the disease involves type IV hypersensitivity reactions (delayed), mediated by T cells. The primary pathogenic event involves the interaction between the drug, human leukocyte antigen (HLA), and T-cell receptors, leading to CD8+ T-cell activation and subsequent release of cytotoxic proteins. This leads to epidermal necrolysis due to keratinocyte death.

The prodromal symptoms begin suddenly. Fever, typically ranging from 39°C to 40°C, is often the first symptom. General malaise, myalgia, sore throat, and conjunctivitis are

also included and may precede or occur simultaneously with the mucocutaneous presentation.

In the following days, mucocutaneous symptoms begin. Dermatosis appears on the skin, initially affecting the face and thorax, and predominantly involving the face, trunk, hands, and feet. It is characterized by atypical target-like macular lesions that often become confluent. These progress to vesicles and blisters that may be hemorrhagic, resulting in erosions and meliceric crusts. Papules and purpuric petechial lesions also occur. Finally, they develop into full-thickness necrosis with skin detachment and desquamation. The Nikolsky sign is positive. The affected skin is usually very painful.

Stomatitis manifests as vesicles on the lips, tongue, cheeks, soft palate, and pharynx. These vesicles lead to hemorrhagic ulcers and pseudomembranes, resulting in difficulty swallowing and excessive salivation.

Conjunctivitis is purulent and bilateral, presenting as conjunctival hyperemia, formation of pseudomembranes, or complete corneal epithelial defects. Rhinitis with crust formation and epistaxis are observed. The vagina and penis can blister, and urethritis may occur.

There are no standard diagnostic criteria, although the presence of target-like macular lesions, involvement of two mucous membranes, recent exposure to drugs, and corresponding histopathology are suggestive.

The histological characteristics of SSJ/TEN include full-thickness epidermal necrosis, apoptosis of keratinocytes, basal vacuolar change, subepidermal blisters, subepidermal clefts, and a mild infiltrate of T cells. Biopsies of drug-induced SSJ/TEN may show dermal infiltration with a large number of eosinophils or neutrophils.

The management of SJS/TEN remains controversial, with immunosuppressive agents frequently utilized. First-line treatments include systemic corticosteroids, cyclosporine A (which can be nephrotoxic), and etanercept. Second-line treatments involve intravenous immunoglobulin and plasmapheresis. However, in some countries, supportive therapy is considered more appropriate because immunosuppressive agents increase the susceptibility to infections and contribute to mortality.

In mild cases, the disease may be managed conservatively with supportive care. Supportive management focuses on meticulous wound care, draining blisters, removing crusts, applying warm compresses or baths with mild antiseptics followed by sterile drying powders, managing fluids and electrolytes, providing nutritional support, controlling temperature and pain, preventing and managing infections, providing Ophthalmologic care and supporting organs as needed.

The withdrawal of the causative drug and all non-essential medications is crucial, followed by hospitalization and supportive care. A useful prognostic tool for SSJ/TEN is the SCORTEN severity scale (Table 1), which is based on seven clinical and laboratory variables. It has been shown that SCORTEN is a reliable parameter for predicting mortality (Table 2).

Table 1. SCORTEN scale. Adapted from: Dobry AS, Himed S, Waters M, Kaffenberger BH. Scoring Assessments in Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis. Front Med. 2022; 9. doi: 10.3389/fmed.2022.883121.

SCORTEN	
RISK FACTOR	**SCORE**
Age >40 years old	1 point
Malignancy	1 point
Heart Rate >120 beats per minute	1 point
Initial epidermal detachment body surface area >10%	1 point
Serum urea >10 mmol/L	1 point
Serum glucose >14 mmol/L	1 point
Bicarbonate <20 mmol/L	1 point

Table 2. Mortality rate according to score on the SCORTEN scale. Adapted from: Dobry AS, Himed S, Waters M, Kaffenberger BH. Scoring Assessments in Stevens - Johnson syndrome and Toxic Epidermal Necrolysis. Front Med. 2022; 9. doi: 10.3389/fmed.2022.883121.

SCORTEN SCORE	***MORTALITY RATE***
0-1	3.2%
2	12.1%
3	35.3%
4	54.3%
≥5	90%
Bicarbonate <20 mmol/L	1 point

CLINICAL CASE

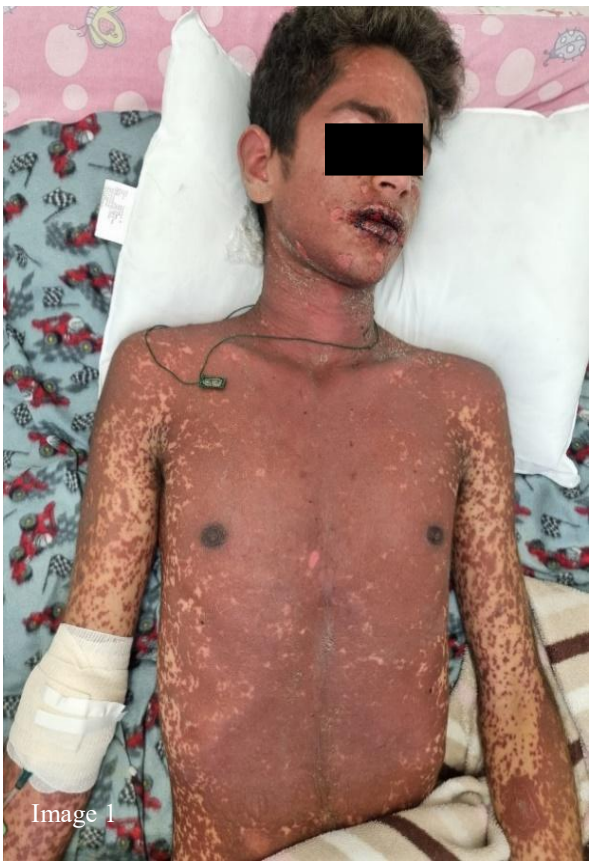
An 18-year-old male patient with a significant medical history of HIV, who discontinued treatment three years ago, with an unknown disease duration. He has a history of crack cocaine and methamphetamine use, starting at age 14 and consumed alternately, along with alcohol use since age 12, leading to weekend intoxication. His condition began with a fever of 39°C, fatigue, adynamia (loss of strength), complete loss of force, and ocular prodrome characterized by pain, tearing (lacrimation), and yellowish discharge. He reported consuming carbamazepine, paracetamol (acetaminophen), and hydroxyzine. He attended the emergency service at Hospital General de Chetumal due to generalized dermatosis characterized by erythematous macules and ulcerative lesions on mucous membranes and genitals.

PHYSICAL EXAMINATION

The patient is conscious, alert, and oriented to person, place, and time, with a Glasgow Coma Scale score of 15/15. The pupils are isocoric and reactive to light. There is conjunctival redness with tearing and purulent discharge. The oral mucosa shows ulcerative lesions with hemorrhagic crusts. The neck is symmetrical and of normal contour, with no lymphadenopathy or palpable masses. The trachea is midline, mobile, and without signs of jugular venous distension. At the Integumentary level, there is a generalized erythematoviolaceous eruption with blisters and vesicles and areas with serous exudate having a foul odor predominantly on the trunk involving 50% of the total body surface area; Nikolsky's sign is positive up to the abdominal level. The thorax appears short, with no structural abnormalities; chest expansion is symmetrical, with normal breath sounds and adequate air movement; no pleuropulmonary syndrome was identified. Heart sounds have good tone and intensity without added murmurs. The abdomen is soft and depressible, with tenderness on deep palpation in the right hypochondrium. Genitals examination is consistent with age and sex but show ulcerative lesions on the glans penis. Upper and lower extremities are intact without signs of edema; capillary refill time is 2 seconds; peripheral pulses are present; muscle strength (Daniels scale) 5/5.

Laboratory studies:

On admission: Creatine phosphokinase-MB 63 u/L, Creatine phosphokinase 78.15 u/L, Lactate dehydrogenase 411 u/L, Total bilirubin 0.47 mg/dl, direc bilirubin 0.12 mg/dl, indirect bilirubin 0.35 mg/dl, glutamic oxaloacetic transaminase 42 U/L, glutamic pyruvic transaminase 21 U/L, total serum protein 7.0 g/dl, alkaline phosphatase 67.56 U/L, albumin 4.6 g/dl, globulins 2.4 g/dl, gamma-glutamyl transferase 27 U/L, sodium 131 mmol/l, potassium 4.17 mmol/l, serum chlorine



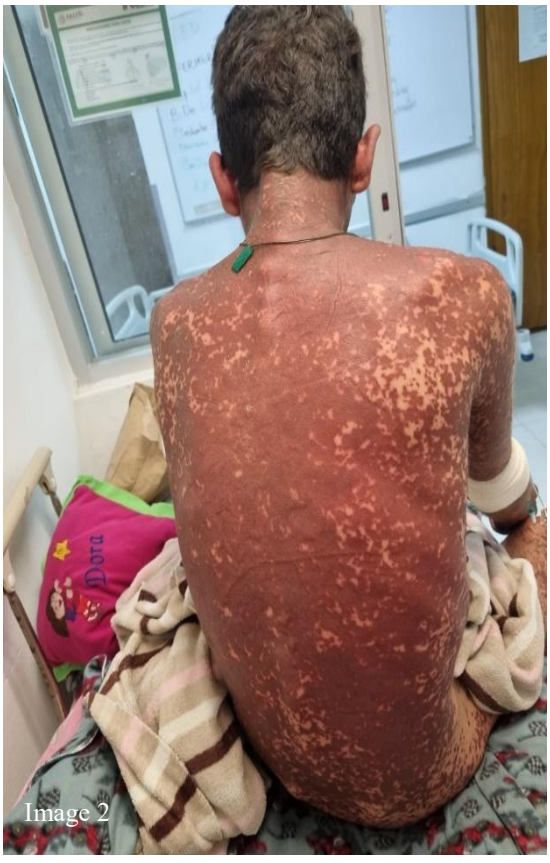
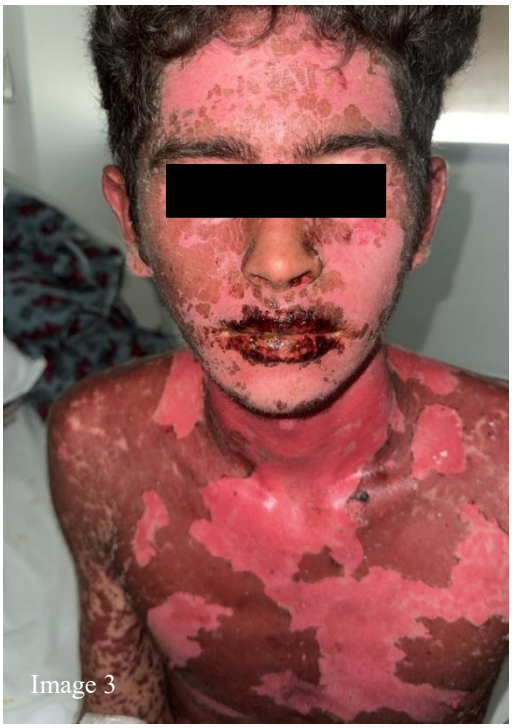


Image 1 and 2. Patient's dermatosis on admission

97mmol/L, phosphorus 2.6 mg/dl, magnesium 1.7 mg/dl, calcium 9 mg/dl, hemoglobin 16.7 g/dl, hematocrit 48.6%, leukocytes 6.9 10^3 uL, platelets 112 10^3uL.



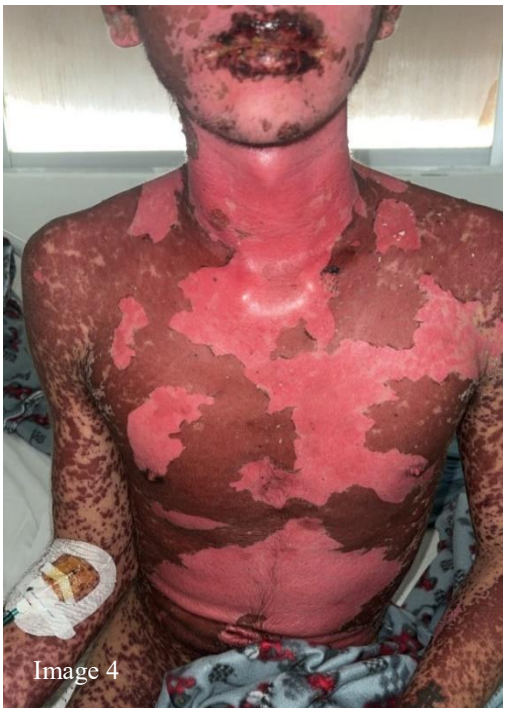


Image 3 and 4. Patient's dermatosis 72 hours after admission

On disscharge: Glucose 79 mg/dl, serum urea 36mg/dl, blood urea nitrogen16mg/dl, creatinine 0.6 mg/dl, uric acid 4.4 mg/dl, cholesterol 124mg/dl, triglycerides 161 mg/dl, sodium 140 mmol/l, potassium 3.9 mmol/l, serum chlorine 103mmol/l phosphorus 4 mg/dl, magnesium 1.9 mg/dl, calcium 8.8 mg/dl, hemoglobin 14.9 g/dl, hematocrit 43.5%, leukocytes 11.1 10^3 uL, platelets 416 10^3uL. Anti HIV antibodies 1 y 2 reactive, VDRL test negative.

The patient is evaluated by the dermatology service, which concludes a diagnosis of Toxic Epidermal Necrolysis and suggests treatment with crystalloid solutions, empirical antibiotics (clindamycin 300 mg IV every 8 hours and metronidazole 500 mg IV every 12 hours), antifungal medication (fluconazole 100 mg IV every 8 hours), and steroid pulses for systemic inflammatory response (methylprednisolone 500 mg IV every 12 hours in 250 cc of saline solution at a concentration of 0.9% for five days). As a complementary measure, silver sulfadiazine is prescribed for application on lesions on the thorax and back. Daily bathing with emollients, covering lesions with petrolatum-impregnated gauze, and performing colloidal baths every 24 hours are advised. Initiation of an antiretroviral therapy protocol as soon as possible is strongly recommended.

DISCUSSION

We present a clinical case of a patient from Hospital General de Chetumal with Toxic Epidermal Necrolysis and a significant history of being HIV seropositive with poor adherence to treatment. According to reviewed literature, this is a key risk factor for developing the disease, as patients with HIV infection have been shown in European cohort studies to have approximately 12 times greater risk of SSJ/TEN compared to the general population.

Additionally, the patient has a significant history of carbamazepine use, which is among the common pharmacological triggers for SSJ/TEN. Although in 15-30% of cases, no triggering agent can be identified. There is evidence linking the HLA-B*15:02 genotype to an increased risk of SSJ/TEN induced by carbamazepine and oxcarbazepine.

The clinical picture of the patient represents a typical evolution of SSJ/TEN, as it began with previously described prodromal symptoms followed by the presence of dermatosis, adding mucosal involvement (conjunctivitis and penile lesions). Upon admission, the patient had a SCORTEN score of 1 due to more than 10% body surface area involvement, with other prognostic factors being normal. This classified him with a mortality rate of approximately 3.2%.

During his stay at Hospital General de Chetumal, supportive management was implemented with daily wound cleaning, silver sulfadiazine, petrolatum-impregnated gauzes, and colloidal baths. Additionally, fluid management was performed using crystalloid solutions. One of the actions taken due to the patient's poor adherence to treatment because of comorbidities was empirical prophylaxis with antibiotics using clindamycin and metronidazole, as well as fluconazole due to the high risk of hospital-acquired infections. Although prophylactic antibiotics are not typically recommended as part of routine supportive care, immediate use of antibiotics is crucial in the context of a clinical infection.

Due to the involvement of more than 50% of the body surface area, it was managed with pulses of corticosteroids (methylprednisolone) since no other systemic medication alternatives were available at the hospital. Japanese treatment guidelines recommend pulse therapy with corticosteroids as one of the first-line treatments for SSJ/TEN , particularly in settings with limited resources, due to its effectiveness and low cost.. The patient presented a complete remission from TEN without complications.

CONCLUSION

This case highlights a typical presentation of SSJ/TEN, emphasizing the importance of a high index of suspicion for early diagnosis and appropriate management to prevent complications and reduce morbidity and mortality in patients with this condition. Additionally, proper management of human immunodeficiency virus (HIV) is crucial as it can significantly influence patient outcomes and potential complications.

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