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A Case Report of Steven Johnson Syndrome in the Dominican Republic

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ABSTRACT

This case report details the presentation, treatment, and clinical course of an 8-year-old boy with epilepsy who developed Stevens-Johnson syndrome (SJS) following an adjustment in his anticonvulsant regimen. The patient, from Los Guaricanos, Dominican Republic, was admitted to Hospital Dr. Robert Reid Cabral after reducing the dose of levetiracetam to 500 mg and increasing the dose of lamotrigine to 25 mg. Upon admission, the patient presented maculo-papular erythema, accompanied by facial edema, lip swelling, and lesions that later formed pustules and scabs, on the face, neck and chest with mucocutaneous manifestations, conjunctival injection. Given the suspicion of SJS, multidisciplinary interventions were initiated. During the subsequent days management led to notable clinical improvement. However, on the fifth day, he had three tonic-clonic seizures, necessitating a neurological reevaluation and anticonvulsant adjustment. This case highlights the importance of

recognizing and managing rare adverse drug reactions such as SJS in pediatric patients with epilepsy.

Keywords: Steven-Johnson syndrome; Lamotrigine; Levetiracetam; Epilepsy; Drug reaction

INTRODUCTION

Stevens-Johnson syndrome often but not exclusively seen in pediatric patients is characterized by extensive involvement of various mucosal and cutaneous sites including the lips oral mucosa conjunctiva urethra genital regions and perianal as described in Robbin Pathology. The distributions of lesions vary widely. This case report detailed an 8-year-old boy presenting with facial edema and multiple erythematous macules, purulent ocular discharges, and generalized pleomorphic, erythematous and purpuric macular lesions throughout the body, characterized by progressive worsening of the lesions. At the same time, the patient had an elevated touch temperature, as reported by the guardian, with no documented fever or seizures. It should be noted that the patient has a medical history of epilepsy treated with lamotrigine and levetiracetam, with recent dose adjustments that precede this clinical condition, specifically an increase in lamotrigine to 25 mg and a decrease in levetiracetam to 500 mg.

CASE REPORT

Day 1 - April 11th 2024.

On April 11th, an 8-year-old male with a known medical history of epilepsy was admitted to the emergency department of Dr. Robert Reid Cabral Hospital in the Dominican Republic. Several days preceding admission, adjustments were made to the patient's anticonvulsant regimen where the Levetiracetam dosage was reduced from 750 mg to 500 mg, while Lamotrigine dosage was escalated from 20 mg to 25 mg. Notably, the patient had been under treatment for epilepsy with Atremperator (Valproic acid) since January 2024, transitioning recently to the Lamotrigine and Levetiracetam combination.

Upon presentation, the patient, accompanied by a report of fever from the mother, noted facial edema, primarily localized to the frontal and perilabial regions. Concurrently, the lips manifested signs of swelling, erythema, fissuring, and scaliness, accompanied by oropharyngeal hyperemia. Extensively distributed maculopapular lesions of variably defined purpuric hue, interspersed with erythematous patches and mucocutaneous involvement were evident across the face, neck, anterior and posterior thoracic regions, abdomen, and extremities, exhibiting compromised mucosal integrity. Furthermore, bilateral eyelid edema was observed, compounded by purulent ocular discharge, impairing the patient's ability to fully open their eyes.

The patient's mother reported the abrupt onset of lesions the previous day, initially appearing small in size and progressively enlarging. Preliminary medical attention was sought at a local health community center, where permethrin cream and acetaminophen were administered.

However, due to worsening symptoms, the patient was subsequently referred to the emergency department on the following day. Suspecting Stevens-Johnson syndrome, the infectious disease specialists at Dr. Robert Reid Cabral Hospital initiated a comprehensive diagnostic and therapeutic intervention regimen, as detailed in (Table 1).

Figure 1: Day 1 - April 11th 2024

Day 2 - April 12th 2024

On the second day of hospitalization, the patient's physical examination revealed no discernible alterations. At 5pm, the patient registered a temperature of 38 C. Throughout the day, the patient's clinical status remained stable, with no modifications warranted to the existing medication regimen. Following comprehensive

evaluation by the pediatric neurology team, the therapeutic approach was augmented to include oral Clonazepam, administered at a dosage of 5 drops every 12 hours.

Day 3 - April 13th 2024

The patient's fever abated with sustained hemodynamic stability thereafter. Demonstrating over 30 hours of afebrile status and tolerating oral intake satisfactorily, the patient continued under the care of a multidisciplinary team comprising dermatology, ophthalmology, neurology, and infectious disease specialists. Adherence to the established treatment protocol and prior recommendations was diligently upheld.

Day 4- April 14th 2024

The patient exhibited no notable changes in clinical status, and the treatment regimen remained unaltered. The mother reported the child is feeling better and has not presented fever. On physical exam there was still mild pallor of skin and mucous membranes, multiple generalized maculo-papular lesions of ill-defined purplish color, on erythematous skin, on face, neck, anterior and posterior thorax, abdomen, back, upper and lower extremities, in addition to presenting loss of integrity at the oral mucosa level (Figure 2).

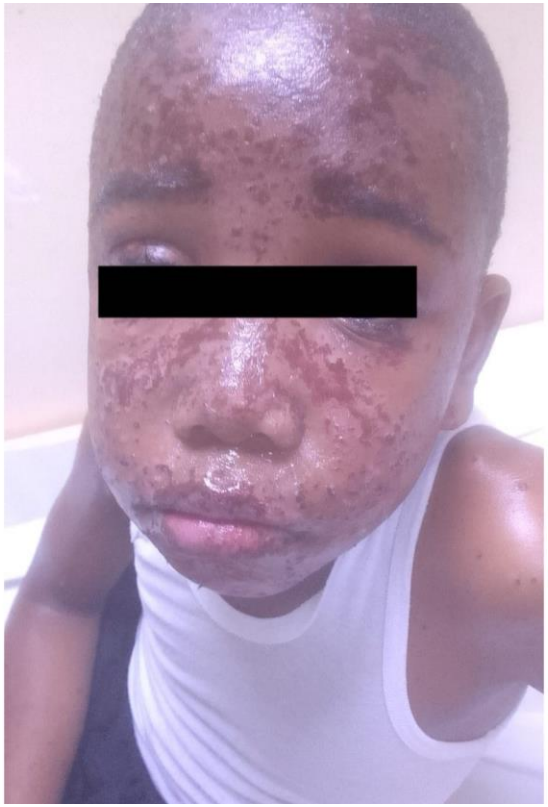

Figure 2: Day 4 - April 14th 2024

Day 5 - April 15th 2024

On day five the patient remained in the same condition. The mother reported a seizure that lasted less than 5 seconds. The treatment regimen remained the same with Diazepam and Clonazepam since said seizure took place.

Day 6 - April 16th 2024

Day 6 of hospital stay the patient maintained hemodynamic stability, alertness, and afebrile status, while still exhibiting tolerance to oral intake. Notably, during the early morning hours, the patient experienced three tonic-clonic seizures, each lasting approximately 1 minute. These seizures were characterized by involuntary

extremity movements, muscle rigidity, loss of consciousness without sphincter relaxation, and rapid recovery. To combat the seizures, the pediatric neurology team gave a loading dose of Diphenylhydantoin 15mg/kg/dose and a maintenance dose of 5mg/kg/day. Subsequent evaluation by the pediatric neurology team will require extending the patient's stay for an additional 48 hours in the center (Figure 3).

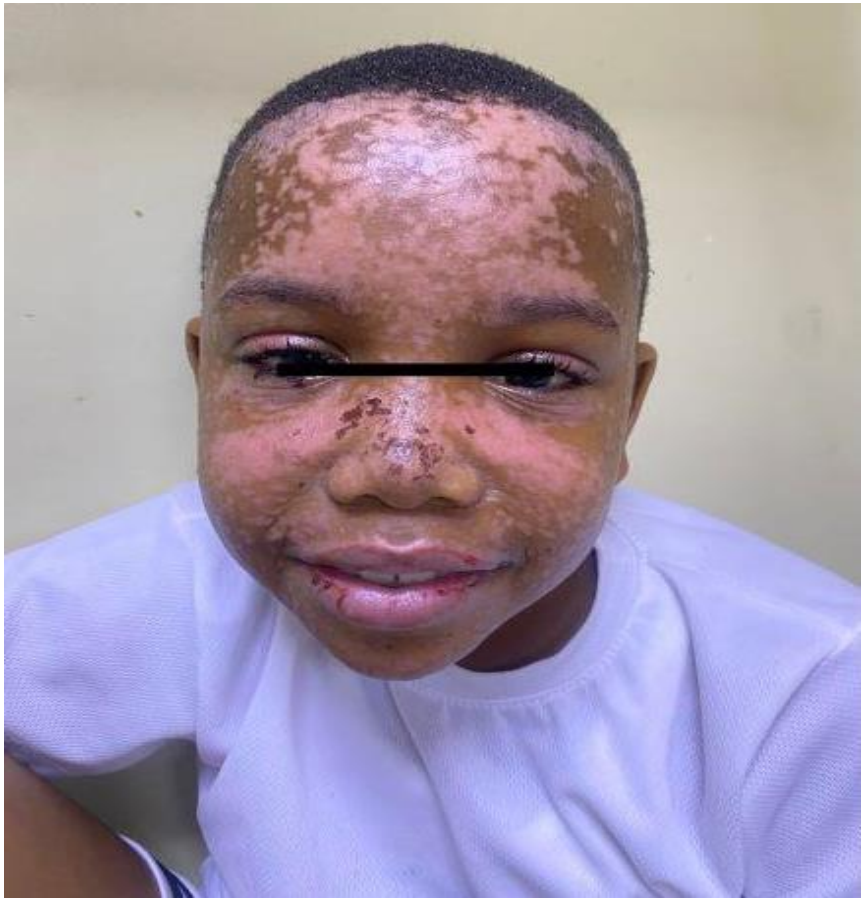

Figure 3: Day 6 - April 16th 2024

Day April 17th 2024

On Day 7, the patient remained seizure-free and was discharged with his treatment regimen to be continued at home.

Table 1. PRN = Pro re nata, IV = Intravenous therapy

Day 1 MEDICAL ORDERS:	
HYDRATION:	**USE:**
Sodium chloride .33% with alpha dextrose 5% (H-S 100%)	1,700 mL-150 mL of dilution: 1,550 mL/24 hrs go to 64.5 mL/hr per infusion pump + baxter pump downpipe (IV)
MEDICATIONS:	**USE:**
Fosfomycin (200 mg/kg/day)	500 mg every/8 hours, diluted in 50 mL of Saline Solution .9% (IV)
Methylpredinsoline (1 mg/kg/dose) Maintenance	15 mg every/6 hours (IV)
Parecetamol (10 mg/kg/dose)	300 mg PRN (IV)
Diclofenac (3 mg/kg/dose)	30 mg every/8 hours (IV)
Diphenhydramine (1 mg/kg/dose)	30 mg every/8 hours (IV)
Diazepam (0.3 mg/kg/dose)	9 mg = 1.8mL PRN (IV)
A-Derma Epitheliale A.H Ultra Soothing Repairing Cream	Over the affected areas every 12 hours
Lagricel (Sodium Hyaluronate 0.4%) ophthalmic drops	2 drops per eye every 12 hours
Tobramycin ophthalmic drops	2 drops per eye every 8 hours
Eucerin aquaphor	Over all mucosal surfaces, multiple times per day
Day 2 MEDICAL ORDERS:	
There were no changes to the medical order on this day.	
Day 3 MEDICAL ORDERS:	
MEDICATIONS:	**USE:**

Clonazepam drops	5 drops every/12 hours (PO)
Day 4 MEDICAL ORDERS:	
There were no changes to the medical order on this day.	
Day 5 MEDICAL ORDERS:	
MEDICATIONS:	**USE:**
Fosfomycin (200 mg/kg/day)	2 gr every 8 hours, diluted in 50 mL of Saline Solution 0.9% (IV)
Day 6 MEDICAL ORDERS:	
HYDRATION:	**USE:**
Sodium chloride .33% with alpha dextrose 5% (H-S 100%)	850 mL/24 hours and 35 mL/hr per infusion
MEDICATIONS:	**USE:**
Fosfomycin (200 mg/kg/day)	10 gr every 8 hours (PO)
Methylprednisolone (1 mg/kg/dose) Maintenance dose	5 mg every 8 hours (IV)
Paracetamol (10 mg/kg/dose)	12 mg PRN (IV)
Diclofenac (3 mg/kg/dose)	Suspended
Diphenhydramine (1 mg/kg/dose)	Suspended
Diphenylhydantoin (15 mg/kg/dose) Loading dose	450 mg diluted in 100 mL of Saline Solution 0.9% passed in 2 hours (IV)
Diphenylhydantoin (5 mg/kg/dose) Maintenance dose	75 mg every 12 hours (IV)

DISCUSSION

Stevens-Johnson syndrome (SJS) represents a life-threatening, delayed-type hypersensitivity disorder characterized by mucocutaneous epidermal necrolysis and detachment of the epidermis. The disease typically initiates with a prodromal phase, presenting as flu-like symptoms including malaise, fever, and rhinorrhea. Subsequently, patients develop a mucocutaneous rash accompanied by a positive Nikolsky sign, which is pathognomonic for SJS. Despite its rarity.

SJS bears a considerable mortality rate of approximately 10%, necessitating prompt initiation of supportive therapies. The pathogenesis of SJS is attributed to the body's failure to detoxify drugs and their peptide metabolites, which are perceived as foreign entities. This leads to the activation of cytotoxic T cells, prompting an attack on keratinocytes and epithelial cells within the mucosa and epidermis. Consequently, cell death ensues, culminating in the sloughing of the skin due to the release of cytolytic proteins such as perforin and granzymes by T-lymphocytes and natural killer cells[1].

A myriad of medications and infectious agents have been implicated as causative agents of SJS. Common offending drugs include allopurinol, non-steroidal anti-inflammatory drugs (NSAIDs) like piroxicam, antibiotics, immune modulators such as sulfasalazine, and anticonvulsants[2]. Antibiotics, particularly sulfonamides, are recognized for eliciting the highest rates of cutaneous drug reactions. Among anticonvulsants, lamotrigine, phenytoin, and carbamazepine are frequently associated with SJS. Immunizations and infections, although less common, can also precipitate SJS, with pathogens such as mycoplasma pneumonia, cytomegalovirus, herpes simplex virus (HSV), coxsackievirus, and echovirus being implicated.

In the context of this case report, Lamotrigine, an anticonvulsant and mood stabilizer, was identified as the causative agent. It is noteworthy that Lamotrigine carries a black box warning for Stevens-Johnson syndrome[3], underscoring the importance of vigilance and prompt recognition of potential adverse drug reactions associated with its use.

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