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Case report

Stevens-Johnson Syndrome with marked mucous presentation

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Abstract

Introduction: Stevens-Johnson Syndrome (SJS) is a rare mucocutaneous disease whose incidence increases with age and about 20% of cases occur in children and adolescents. Mortality reaches 5%.

Case description: We present a male adolescent, 11 years old, admitted due to oral mucosa erosions, lips edema and erythema, enanthema, cutaneous target lesions and edemaciate, erythematous and exudative glans, treated with non-steroidal anti-inflammatory drugs one day before the onset of cutaneous symptoms. The diagnostic tests didn't show alterations and microbiological tests and serologies were negative. The patient was admitted to the Pediatrics Department with diagnosis of SJS. He was hospitalized for 10 days with intravenous analgesia, mucositis solution and topical emollient, hydric and nutritional reinforcement. He exhibited successive improvement of lesions with complete resolution after 2 weeks.

Conclusions: The diagnosis of SJS is clinical, with special emphasis to the use of drugs or the presence of previous infection. Its recognition is important so that clinical intervention can occur as early as possible, decreasing the likelihood of complications and death.

Keywords

Adolescent, drugs, mucous membrane, epidermal detachment, early intervention, Stevens-Johnson Syndrome.

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Introduction

Stevens-Johnson Syndrome (SJS) is a severe dermatologic reaction with mucocutaneous involvement and an incidence of 1.2 to 6 cases per million/year. It is estimated that 20% of the total cases correspond to pediatric patients [1, 2].

The etiology is not fully understood, but skin damage is believed to result from cell-mediated and often drug-specific cytotoxic reactions against keratinocytes, leading to widespread dermal-epidermal detachment [3]. Most cases are drug-related (50-80%), although certain infections may also be involved in its etiopathogenesis [1]. The most common and frequently associated drugs are allopurinol, antibiotics, in particular ß-lactams and sulfamides, non-steroidal anti-inflammatory drugs (NSAIDs) and anticonvulsants [4]. Other causes described are viral infections [1, 4, 5].

The initial manifestations can be nonspecific with prodromal influenza-like symptoms. The cutaneous lesions typically first appear on the face, before spreading to other areas, in a symmetrical distribution. Mucosal involvement, mainly oral, ocular and genital, occurs in nearly all cases. [3, 6].

SJS and toxic epidermal necrolysis (TEN) are considered variants on a severity spectrum of the same disease and are defined based upon the amount of epidermal detachment [7]. The first involves less than 10% body surface area detachment while the second is characterized by extensive involvement of greater than 30% body surface area [7].

SJS is associated with considerable morbidity and presents a mortality rate that reaches 5% [4, 8].

The objective of this presentation is to raise awareness of SJS, since timely recognition and early intervention are associated with a better prognosis.

Case report

We present an 11-year-old boy without relevant personal or family history, with adequate weight and height and complete vaccination schedule for age.

He was admitted to the paediatric emergency with a history of bullous lesions, edema, hyperemia and exudate of the oral and genital mucosae. The day prior to the appearance of the cutaneous symptoms, he reported a sore throat and took two doses of ibuprofen. On the following day, bullous cutaneous lesions of serous content appeared on the lower limbs and wrists.

The skin lesions spread from the 3rd to the 5th day, affecting the oral and genital mucosa. Due to progressive worsening, he went to the Emergency Department on the 6th day of illness.

The patient denied ingestion of other drugs, fever, ocular symptoms, weight loss, asthenia or other constitutional symptoms. There had been no sexual contact or family context of acute illness.

On examination, he had swollen, friable, bleeding and ulcerated lips, with yellowish exudate (**Fig. 1**) and had difficulty opening his mouth, with gingival hypertrophy and halitosis. On the genitals, the glans had edema, erythema and an exudate (**Fig. 2**),

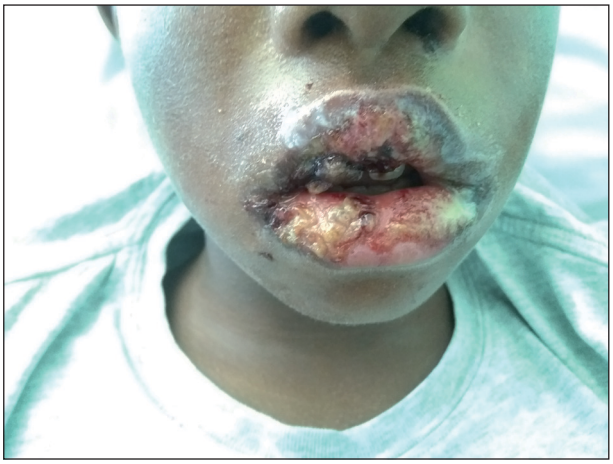

Figure 1. Swollen, friable, bleeding and ulcerated lips, with yellowish exudate.

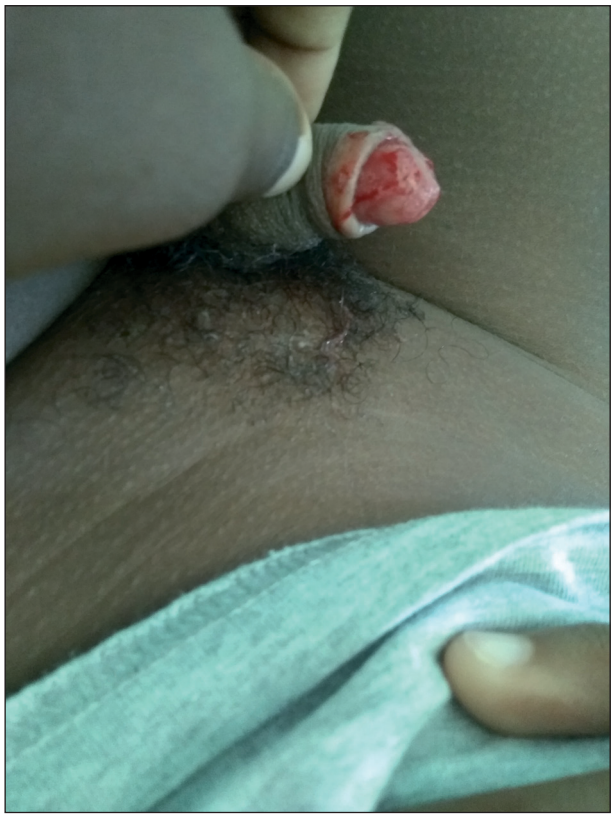

Figure 2. Swollen, erythematous and exudative glans.

and he had several cutaneous target lesions on the limbs reaching the palmar surface (**Fig. 3**).

Laboratory tests including full blood count, acute infection parameters, kidney and liver function were normal. Serologies for *Mycoplasma pneumoniae*, EBV, herpes simplex virus (HSV), and cytomegalovirus (CMV) were negative. Microbiological tests were negative.

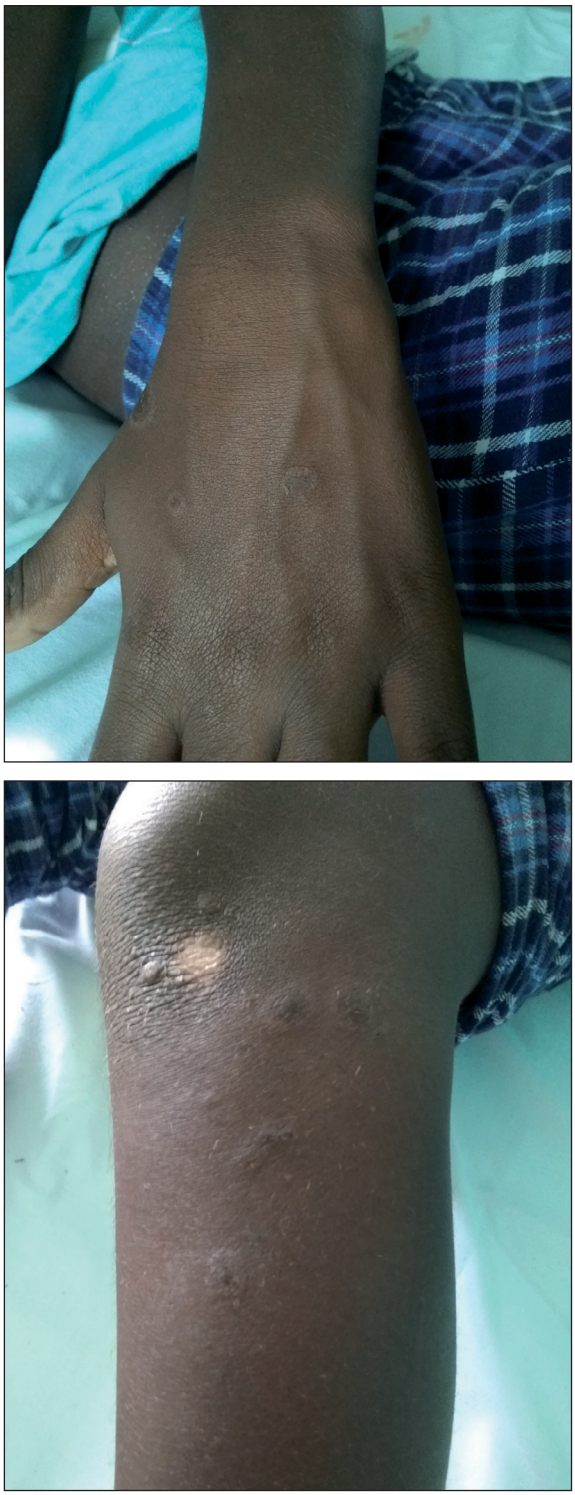

Figure 3. Cutaneous target lesions on hand (**A**) and leg (**B**).

The patient was admitted to the Pediatrics Department with a diagnosis of SJS. The diagnosis was clinical, based on the recent exposure to a NSAID, along with the targetoid skin lesions with central necrosis and mucosal involvement.

Treatment included the suspension of all prior medication and non-pharmacological and pharmacological measures. Non-pharmacological measures included: cardiorespiratory monitoring, temperature control, moisturizing the lips with vaseline and eyes with artificial tears, intravenous fluid therapy and enteral nutrition according to tolerance. Pharmacological measures included: oral sucralfate, systemic analgesia with intravenous paracetamol, mouthwash of saline and lidocaine and topical ocular antibiotic (gentamicin).

During hospitalization, he was evaluated by an ophthalmologist that maintained topical eye treatment.

There was a clinical improvement with complete resolution after 2 weeks of supportive treatment.

Discussion

SJS is a rare disease, with rapid progression and high mortality rates. In the pediatric age group, it is often associated with an adverse reaction to medication or infectious agents such as *Mycoplasma pneumoniae* and HVS [8, 9].

When the etiology is pharmacological, clinical manifestations appear on average 7 to 21 days after the start of the drug involved. Studies in children suggest that upper respiratory infection coexists in about 80% of cases [1, 10].

The diagnosis is clinical and supported by histology, with keratinocyte necrosis and the cleavage of the dermo-epidermal junction as the main finding [3].

Differential diagnosis includes erythema multiforme, widespread acute exanthematic pustulosis, staphylococcal scalded skin syndrome, pemphigus paraneoplastic and graft versus host disease [3, 11].

The key to the diagnosis for the case presented was the temporal relationship between taking the drug and appearance of the mucocutaneous features of SJS – skin rash with some atypical target lesions and the exuberant involvement of the oral and genital mucosae. The mucosal involvement is usually more intense and more frequent than the cutaneous involvement [11], as

was seen in this patient. In about 25-50% of cases, it only affects the oral mucosa, with a predilection for the lips, rarely affecting the gums [1].

The recent start of NSAIDs, potentially implicated in this syndrome, made us suspect that the etiopathogenesis was an adverse reaction to the ibuprofen.

Currently, there is no clear consensus on the best treatment for SJS. Management includes identification of the underlying cause and its immediate withdrawal, which decreases mortality and improves prognosis. Aggressive supportive care is the mainstay of treatment: proper wound care, fluid and electrolyte management, nutritional support, ocular care and pain control [3, 6].

In some cases, systemic immunomodulatory therapies (corticosteroids, human intravenous immunoglobulin [IVIG], cyclosporine and tumor necrosis factor-alpha inhibitors) may be considered; however, the treatment with these therapies is still controversial [12, 13].

A meta-analysis of 11 studies evaluating the effectiveness of glucocorticoids versus supportive care only, showed comparable results between the two groups with no difference in mortality. In the same meta-analysis, the effect of IVIG versus supportive care was compared in terms of mortality and IVIG was not found to be superior to supportive care [13, 14].

In the presented case, we chose to maintain supportive care only with adequate pain control, using intravenous paracetamol and topical lidocaine.

The main problem during management is the transdermal fluid loss, which leads to hypovolemia and changes in electrolyte levels, which in this case did not occur.

The combination of septicemia and hypovolemia increases the risk of shock and multiorgan failure, making sepsis the main contributor to mortality [3]. SJS may lead to long-lasting sequelae depending on the mucosal involvement in the acute stage [6].

One in 5 patients has a recurrence, and a third have multiple recurrences, mostly between 2 months and 7 years after the first episode [1].

Conclusion

The Authors present a case of SJS, a rare but potentially serious disease, with the objective of promoting early recognition and clinical intervention, thus decreasing the likelihood of complications and death.

Declaration of interest

None of the Authors have conflicts of interest to state. No sources of funding have been attributed to this paper. The corresponding Author is not a recipient of a research scholarship.

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