Early Diagnosis of Stevens-Johnson Syndrome in Dental Clinic

Wisam Alhathlol 1, Asma AlMeslet 2

1, General practitioner in Security of forces Hospital, Hail, Saudi Arabia

2, Lecturer in Oral Maxillofacial Surgery and Diagnostic Sciences Department, Riyadh Elm University, Riyadh, Saudi Arabia

Abstract:
Stevens-Johnson syndrome (SJS) is a mucocutaneous illness in which the patients are usually presented with a spectrum of mucocutaneous rash involving the oral, ocular, or genital mucosa. It is associated with several factors including the commonly used medications in clinical practice. In this report, we describe a case of SJS that developed after exposure to antibiotics in a patient who presented with acute oral lesions to dental emergency clinic.

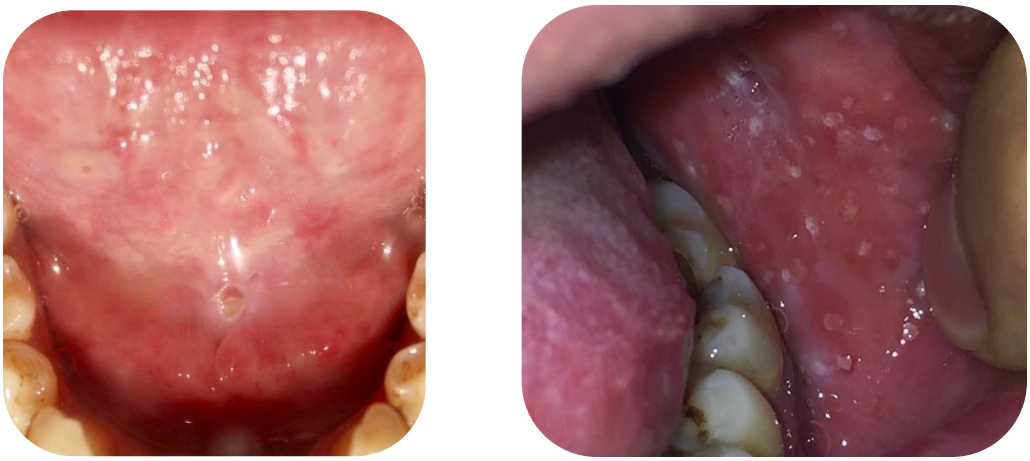
Introduction:
Stevens–Johnson Syndrome (SJS) is a rare mucocutaneous condition. It has been related to several risk factors including medications. It is commonly presented with skin rash that usually involve oral mucosa or conjunctivae but it could be life-threatening in rare cases (1). Stevens–Johnson syndrome is part of a spectrum of diseases ranging from SJS when presented with less than 10% skin detachment to toxic epidermal necrolysis when presented with greater than 30% skin detachment (2). Cases of SJS have been increasingly reported as unfavorable drug effects, yet the exact pathology is not entirely understood (3). With use of commonly prescribed medications, such as antibiotics, SJS as an adverse drug reaction represents a continuing challenge for the emergency health providers.

Case Report:
A 37-year-old Saudi male presented to dental emergency clinic of Riyadh Elm University complaining of burning sensation in the mouth and difficulty in swallowing with diagnosed oral candidiasis. Five days prior to this visit he was prescribed antibiotics by private clinic that resulted in another attack of burning sensation followed by more difficulty in swallowing and generalized skin rash. **Medical History:** Fever since 2 days, joint pain and blurred vision since 2 days with no history of any kind of allergy. The family history was unremarkable. **At clinical presentation:** He was sick and febrile with a temperature of 40 C. **General examination** showed enlarged cervical lymph nodes, skin rash over the upper limbs (**Figure 1**). There was no joints swelling. **Local examination** showed blisters on the floor of the mouth and buccal mucosa with scattered oral ulcers with erythematous base (**Figure 2**), and coated tongue (**Figure 3**). The purpose of this report was to highlight the early signs and symptoms of SJS, which may have been overlooked and misattributed to other causes. **Differential Diagnosis:** Herpes simplex virus (HSV), bullous pemphigoid, HIV and autoimmune vasculitis. **Management at our center:**

Initially he was treated with topical betamethasone for 14 days, topical chlorhexidine10 days and nystatin suspension: 100,000 units PO. The patient was reviewed 3 weeks later in our clinic and his clinical features showed afebrile, active with improved general and local clinical manifestations. (**Figure 4**). **Management at referral center:** He was referred to an infectious disease consultant for second opinion in another hospital. Laboratory investigations showed negative results in pan cultures, HIV and hepatitis screening. The diagnosis was mild Steven-Johnson syndrome versus immuno-deficiency disorders.



Figure1: showed skin rash over the upper limb.



(A) (B)

Figure 2: showed blisters on the floor of the mouth (A) and buccal mucosa with scattered oral ulcers with erythematous base (B).



Figure 3: showed coated tongue



Figure 4: Three weeks after treatment, the patient showed improved the mucocutaneous lesions.

Discussion:

This case report highlights the early clinical features of SJS and also stresses the importance of early recognition. Stevens–Johnson syndrome results from extensive keratinocyte apoptosis, or programmed cell death, which is provoked by a cell-mediated cytotoxic reaction involving T lymphocytes and natural killer cells (5).

Research has shown that the concentration of granulysin in patient blisters correlates with the severity of the disease. Furthermore, granulysin from the blisters of patients has been shown to be sufficient to induce keratinocyte cell death and blistering in mouse skin in a dose-dependent manner (6). Another theory is that the drug react with the major histocompatibility complex that is responsible for activation of cytotoxic T lymphocytes. Consequently, this process induces apoptosis in keratinocytes throughout the entire depth of the dermis resulting in painful blistering of the skin and mucous membranes (7).

The management of SJS is challenging. Several clinicians are usually involved in the care of these patients including a dermatologist, intensivist, pulmonologist, nephrologist, ophthalmologist, plastic surgeon and gastroenterologist. The essential treatment of these patients is the wound care. In addition, the pharmacist should closely monitor the medications that the patient is using to prevent exacerbation of the condition (8,9).

The management outcome of patients with SJS depends on the extent and severity of skin involvement. For example, those with a mild eruption, the lesions usually improve in 12 to16 weeks. Mild scarring may appear, but there is usually no systemic functional loss unless the eyes and other mucous membranes are involved. It has been reported that the mortality rates of 1-27% when the involved skin area is more than 20%. Furthermore, the mortality rate in SJS can be worse with the concomitant bacterial infection. Additional risk factors that adversely affect the outcome include advanced age, leucopenia, presence of a malignancy, renal dysfunction and hyperglycemia (10,11).

Stevens Johnson syndrome is still an ongoing challenge for emergency department providers, and misdiagnosis of this condition may lead to severe consequences. Thus, recently it has been suggested that clinicians need to be familiar with the clinical features of SJS to facilitate prompt recognition and appropriate care (12). Obviously, this rare condition would be much more challenging for dental practitioners. However, obtaining a thorough medical history and performing a proper examination including oral health assessment are essential for early diagnosis and management of this condition.

Our case is unique due to early recognition of this critical syndrome by a dental clinician and transferring the patient to an appropriate level of care. Thus, this case is emphasizing on the need for dental practitioners to be aware of early recognition of SJS, and to enable them to promptly manage this critical clinical condition, and thus to avoid any adverse outcome.

Conclusion:

Stevens-Johnson syndrome is a rare mucocutaneous illness that commonly related to several risk factors including medications such as antibiotics. The general dental practitioner should be aware of this rare illness and should perform a comprehensive evaluation for any patient attending the dental clinic with oral lesions to facilitate early recognition and appropriate care.

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