Steven Johnson Syndrome Two Cases Report

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Abstract

Stevens-Johnson syndrome (SJS) is a severe mucocutaneous disease that can progress to toxic epidermal necrolysis (TEN), known as Lyell’s syndrome, that might be sometimes lethal. Systemic corticosteroid use in the treatment of SJS has been debated in the medical literature for many decades. The current reported cases of SJS showed improvement after uses of systemic corticosteroids along with maintenance of the oral hygiene health.

Keywords: Steven Johnson Syndrome; Systemic Steroids; Target Lesions

Introduction

Steven Johnson syndrome (SJS) is severe and potentially lethal disease that involves the skin and mucous membranes due to immune complex mediated hypersensitivity reaction [1]. SJS mostly occurs due to a drug reaction whereas Erythema multiforme is mainly linked with infections. SJS clinical presentation is based upon total body surface area (TBSA) of affected skin, with the most severe form referred to as toxic epidermal necrolysis (TEN), or Lyell’s syndrome, in SJS less than 10% TBSA is affected. Whereas SJS/TEN overlap affect 10% - 30% TBSA, and TEN affect more than 30% TBSA [2]. SJS may occur at different age group but more common in children and at the elderly [3].

Initial presentation can be followed by nonspecific prodromal symptoms: fever, malaise, cough, sore throat or eye discomfort, all appearing before the cutaneous manifestations. Upon the onset of mucosal ulcerations and then progression to cutaneous, dusky red vesiculobullous-appearing atypical target-like lesions on the trunk and face appear in 2 to 15 days [3,4], where cutaneous manifestations have a positive Nikolsky sign due to the dermal-epidermal junction involvement [5].

Initial care should include discontinuation of the cause of the hypersensitivity mediated by immune complex reaction and when further management is supportive to avoid complications. Disease prognosis is variable, with the risk of mortality increasing with the severity of the syndrome. SJS has a mortality rate of approximately 5% [literature citation: Sekula P., et al. Comprehensive survival analysis of a cohort of patients with Stevens-Johnson syndrome and toxic epidermal necrolysis. Journal of Investigative Dermatology May (2013); 133:1197. (http://dx.doi.org/10.1038/jid.2012.510)] and TEN has the worst mortality 16% to 55% [5,6].

Case Scenario I

- A 67 years old Sudanese female came to the maxillofacial clinic complaining from burning sensation, difficulty in eating and swallowing started suddenly 3 weeks ago.
- No significant medical or social history.

A clinical examination was demonstrated a tender diffuse irregular shallow non-indurate bloody crusty ulcer affected the lower lip, palatal mucosa and gingiva without evidence of cervical lymphadenopathy (Figures 1-2). Besides, there were many targets like lesions affected skin of the hands.

**Figure 1:** Diffuse shallow bloody crusty ulcers affected the lower lip.

Incisional biopsy was taken and the histopathological examination revealed features of Erythema multiforme.

Patient was treated by high dose of oral systemic steroids, chlorhexidine mouth wash and broad-spectrum antibiotic (which antibiotic?) to prevent the secondary infection. The lesions showed remarkable improvement within weeks (Figure 3, 4 and 5).

**Figure 2:** Diffuse irregular shallow ulcers without apparent discharge affected the palatal mucosa and gingiva.

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Case Scenario II

- A 38 years old Sudanese female came to a private clinic complaining from burning sensation, difficulty in swallowing and urticaria started suddenly 18 days ago after application of the new lip stick.
- No significant medical or social history.
- Clinical examinations show a tender diffuse irregular shallow non-indurated bloody crusty ulcer affected the lower lip, tongue and palatal mucosa without evidence of cervical lymphadenopathy (Figures 6,7).

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Figure 6: Diffuse shallow bloody crusty ulcers affected the lower lip and tongue.

Figure 7: The image of diffuse irregular shallow crusty ulcers without apparent discharge affected the palatal mucosa.

- Lesions affected skin of the hands (Figures 8).

Incisional biopsy was taken and the histopathological examination revealed features of Erythema multiforme.

Patient was treated by high dose of oral systemic steroids, chlorhexidine mouth wash and broad-spectrum antibiotic to prevent the secondary infection. The lesions showed good improvement after 12 days (Figure 9).

Discussion

The use of corticosteroids in SJS treatment is controversial. The general negative idea regarding corticosteroids exists probably due to late administration, low dose healing phase; it may indeed impair wound healing and promote sepsis. However, short courses of high-dose corticosteroids in early SJS/TEN have a good rationale as immune mechanisms are directly responsible for the cascade of events leading to apoptosis [7]. In confirmation, we present two cases of SJS treated by systemic corticosteroids.

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SJS may be triggered by numerous agents, particularly bacteria and viruses, immune conditions, non-infectious agents such as food additives or chemicals (benzoates and nitrobenzene) and drugs [8]. In the second case triggered by chemicals found in lip stick, while the first case had no history of any triggering factor.

The initial symptoms of SJS are always non-specific and the clinical presentation may mimic different oral inflammatory diseases. (Hence the advice that general practitioner should pay more attention to the oral examination) SJS can be difficult to differentiate from vesiculobullous disorders, and it appears as acute rapid Blistering and ulceration of the lips, appearing as crusted lips associated with skin lesions with characteristic clinical presentation called target lesions [9]. Let say that the typical presentations were reported in the current two cases.

No specific diagnostic tests for SJS, but the diagnosis is supported by tissue biopsy and exclusion of other causes; biopsy is mandatory, the H histopathology shows intraepithelial edema and spongiosis early on, with (please more clear!) satellite cell necrosis and vacuolar degeneration of the basement membrane zone and monocyte infiltrate with full-thickness epidermal necrosis [3].

Study carried by Christa, et al. [10] revealed that SJS is more frequent in females than in males (7: 5), with an average age of 44 ± 28 years, which in line with the current two cases: both were females and the age was within the reported age range.

In previous study it is reported that oral steroids in treatment of SJS decrease the duration of the symptoms (15 days compare with reported typical course of 1 month) enhance the healing without any complications [11]. That is in agreement with the present result: the first case healed within 15, while the second for 12 days. That’s all approving that steroids was the drug of choice for SJS treatment.

Conclusion

The SJS is a life-threatening condition very similar to burns, where supportive care and rehydration is an essential part of therapy because the patient can experience rapid dehydration. Systemic corticosteroids are the standard drugs for the treatment of SJS as very effective in limiting the inflammatory response hence limiting the disease progression and enhance the healing.

Bibliography


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