Oral Manifestations in Stevens Johnson Syndrome

Authors: Bhavesh A. Mehta*, Alka Raka**, Vikas Sinha***  
*Professor and Head Dermatology, **Resident Dermatology, ***Dean, Professor E.N.T.

Institution: Department of Dermatology and Department of Otolaryngology and Head and Neck Surgery  
M.P.Shah Medical College, Jamnagar (Gujarat), India

Corresponding Author:  
Dr. Bhavesh A. Mehta  
Prof & Head Dermatology  
M.P.Shah Medical College  
Jamnagar -361 008 (Gujarat), India  
E mail drbhavesh_skin@rediffmail.com

Abstract

Stevens Johnson Syndrome is an acute life threatening mucocutaneous reaction characterized by extensive necrosis and detachment of epidermis with mucous membrane involvement. The aim of study is to determine the commonest etiology, the incidence of oral involvement, the most likely presentation and the survival rate of Stevens Johnson Syndrome. A total of 20 cases were included in this study. All casese were diagnosed in the department of Dermatology in association with the E.N.T. Department at M.P. Shah Medical College, Jamnagar, India. The most common skin signs were erythematous dusky red macules having a positive pseudo-Nikolsky Sign. Oral involvement was present in all patients. The oral cavity had painful hemorrhagic ulcerative lesions over the palate, tongue and lateral mucosa; with crusting and white membrane over the lips. Stevens Johnson Syndrome was associated with other mucous membrane involvement, including ocular in 90% of cases, genital in 60% of cases, nasal mucosa in of 20% cases and anal mucosa in 5% of case. All patients were given symptomatic treatment. 90% of patients survived after successful treatment.

Introduction

Stevens Johnson Syndrome is an acute life threatening mucocutaneous reaction characterized by extensive necrosis and detachment of the epidermis and involves the mucous membranes. Stevens Johnson Syndrome can be differentiated from toxic epidermal necrolysis by the extent of area involvement. When 10% or less body area is involved the patient is defined as having Stevens Johnson Syndrome. The overall incidence of Stevens Johnson Syndrome is 1 to 6 per million per year. Stevens and Johnson first reported...
two cases of disseminated cutaneous eruption associated with erosive stomatitis and severe ocular involvement. The incubation period is typically a few days to 3 weeks. Most cases of Stevens Johnson Syndrome are drug induced but the reaction is independent of the dosage and it is idiosyncratic. Other rare causes of Stevens Johnson Syndrome include food additives, contact with chemicals and graft versus host reactions. Stevens Johnson Syndrome is a severe illness that usually has a sudden onset, associated with marked constitutional symptoms of high fever, malaise, myalgia, arthralgia and extensive erythema multiforme of the trunk with occasional skin blisters and erosions. There is significant involvement of mucous membranes. Ashby and Lazar, in their review of 81 cases, found involvement of oral mucosa in 100%, eyes in 91%, male genitalia in 57% and anal mucous membrane in 5%, while bronchitis and pneumonitis occurred in 6% and 23% of cases, respectively. Many times the diagnosis at the initial stage pays a very crucial role in management. Early diagnosis increases the likelihood of a good outcome and also decreases complication.

**Aim and Objective:** The Aim and objective of this study is to determine the incidence of Stevens Johnson Syndrome, its oral involvement, presentation, complication and prognosis.

**Methods**

Case studies on 20 patients with Stevens Johnson Syndrome were carried out in the Department of Dermatology in consultation with the Department of ENT at M. P. Shah Medical College Jamnagar, India. The cases presented to our department between June 2010 to June 2011. Each patient was analyzed in detail with respect to history, presenting symptoms and clinical examination. Special emphasis was given on the history of drug use, prescription or otherwise.

**Results**

The maximum number of cases were in the age group 20 to 30 years. The youngest patient in this study was a 12 years old female, and the oldest was an 80 year old male patient. The male to female ratio was 1 to 2.3. Most of the patients gave a history of preceding drug usage of 6 to 8 weeks duration with non specific symptoms such as fever, headache, rhinitis, and myalgia. 50% of patients gave a history of anticonvulsant such as phenytoin, 30% of patients gave a history of NSAIDS such as Paracetamol or Salicylates, 10% gave history of antibiotic usage such as Cotrimoxazole and pneumonitis in 5%, while bronchitis and pneumonitis occurred in 6% and 23% of cases, respectively. Many times the diagnosis at the initial stage pays a very crucial role in management. Early diagnosis increases the likelihood of a good outcome and also decreases complication.

Mucosal involvement preceded skin involvement in 60% of patients (n=12). Patients complained of pain on swallowing and burning in the oral cavity, along with stinging and redness in their eyes. The remaining 40% of patients (n=12) gave a history of oral lesions accompanying the skin lesions. Most of the patients presented with mucosal symptoms which began with erythema followed by painful erosions of the buccal cavity. The buccal erosions were associated with lip swelling that had painful hemorrhagic erosions coated with a grayish white membrane and crusting over the lips (Figures 1A, 2A, and 3A). There was also involvement of other mucous membranes including the eyes which presented with conjunctivitis, redness, photophobia, and discharge with burning.

**Enlarged Pictures at End of Manuscript**
Skin lesions were characterized by erythematous, dusky red purpuric macules. (Figures 1A, 2A, 3A). On examination lesions were bilaterally symmetrical with a pseudo-Nikolsky sign. The dislodgement of epidermis by lateral pressure was present in the erythematous zones. The sign is elicited by applying lateral pressure with thumb or finger pad on the skin over a bony prominence. This results in shearing force that dislodges the epidermis hence this sign is also known as epidermal peeling sign. The underlying mechanism is the necrosis of epidermal cells and not acantholysis as a true Nikolsky sign. The pseudo-Nikolsky sign (epidermal peeling sign) is positive in Stevens Johnson Syndrome, toxic epidermal necrolysis and in some cases of burns. It can be elicited only on involved or erythematous areas.

Oral cavity and tongue involvement was present in all of our patients. Other associated mucous membrane involvement was present in most cases. This involvement included the eyes in 90% of cases, nasal mucosa involvement in 20% of cases and anal mucosa involvement in 5% of cases. Oral cavity lesions ranged from lip edema, hemorrhagic crusting and erosions. The oral cavity showed painful hemorrhagic ulcerative lesions with a perilesional pale halo over the palate, tongue (Figures 1A, 2A, 3A), and lateral oral mucosa.

**Treatment:** Systemic treatment was given, which included fluid replacement, aggressive nutritional support and a course of steroids. Intra venous fluids included normal saline, dextrose with normal saline, Ringer lactate with intra venous multivitamin supplements. Prednisolone was given in the dose of 2 mg/kg body weight per day, which was then tapered by 10 mg/week. The oral antibiotic (Azithromycin) were given to prevent secondary infection. Topically antibiotic creams were applied to the skin lesions. Erosion of the skin were also treated topically with soframycin cream (framycetin 1%) or Fusidic acid (2%). If erythema was severe, a mild steroid such as betamethasone valerate 0.05% or mometasone furoate (0.1%) was added to the cream.

Oral cavity lesions were treated with systemic and topical antifungal agents along with a mild steroidal preparation.
Figure 1B: After treatment for hemorrhagic erosion and crusting over the lips.

Figure 2B: After treatment for ulcerative lesions over the tongue.

Figure 3B: After treatment for hemorrhagic erosion and crusting over lips and ulcerative lesions over the tongue.

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Discussion

In our study of 20 patients with Stevens Johnson most patients were in the group of 20-30 years of age.

<table>
<thead>
<tr>
<th>Age group In years</th>
<th>Number of patients</th>
<th>% of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>10-20</td>
<td>1</td>
<td>5%</td>
</tr>
<tr>
<td>21-30</td>
<td>11</td>
<td>55%</td>
</tr>
<tr>
<td>31-40</td>
<td>4</td>
<td>20%</td>
</tr>
<tr>
<td>41-50</td>
<td>3</td>
<td>15%</td>
</tr>
<tr>
<td>51 &amp; above</td>
<td>1</td>
<td>5%</td>
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</tbody>
</table>

In present study, oral involvement was seen in 100% of patients, eye involvement in 90% of patients, genital involvement in 60% of patients, nasal mucosal involvement in 20% of patients, and 5% of patients showed anal involvement. This distribution of presentation is in accordance with other publish reports.\(^5\)
The commonest etiology of Stevens-Johnson syndrome is drug induced, which is similar to other study. Our study of 20 cases show the most common drugs causing Stevens Johnson Syndrome were anticonvulsants such as phenytoin which was taken by 50% of our patients. Twenty-five percent of patients gave a history of nonsteroidal anti-inflammatory drug use. 15% of patients gave a history of using an antibiotic similar to cotrimazole. 10% of patients did not give any history of drug usage. Our study found that the most common cause of Stevens Johnson Syndrome were anticonvulsants. The next most common cause were non-steroidal anti-inflammatory drugs. Stern and Bigby have also reported a wide range of cutaneous reactions to nonsteroidal anti-inflammatory medications. The overall mortality is 10% in our study which is similar to other authors.

### Conclusion

Stevens-Johnson syndrome is an acute life threatening mucocutaneous reaction whose most common etiology is related to drug usage. It was further observed that the majority of cases presented with oral
involvement, which was a significant cause of morbidity. Early diagnosis and treatment helps in the reducing morbidity and mortality and gives a favorable outcome (Figures 1B, 2B, 3B).

References


