Late Sequeleae in a Survivor of Stevens Johnson Syndrome in Childhood

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Received date: Apr 08, 2016, Accepted date: Jun 29, 2016, Published date: Jul 01, 2016

Abstract

Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are among the most important severe skin hypersensitivity reactions affecting cutaneous and mucous membranes. The etiology of SJS/TEN is not precisely known, but it’s triggered either by drugs most common being anticonvulsants, antibiotics or nonsteroidal antiinflammatory drugs, pregnancy, vaccines or by infections such as Mycoplasma pneumonia, especially in children and adolescents. Present here, a rare case of severe SJS, diagnosed at age of 2 and followed-up till to 17 years, who has long term sequelae of bronchiolitis obliterans, lip adhesion, hematocolpos, finger contractures and ophthalmologic involvement.

Conclusion: Multidisciplinary approach is essential in the long term follow up of SJS for the sequelae.

Keywords: Stevens-Johnson syndrome; Children; Hematocolpos; Bronchiolitis obliterance

Introduction

Stevens-Johnson Syndrome (SJS) and Toxic Epidermal Necrolysis (TEN) are among the most important severe skin hypersensitivity reactions affecting cutaneous and mucous membranes [1,2]. Sharing common causes and mechanisms, SJS and ten are considered a single disease entity with different degrees of severity [2,3]. Based on the involvement of Body Surface Area (BSA), epidermal detachment of more than 30% of BSA is defined as TEN, whereas epidermal detachment of less than10% of BSA is defined as SJS. The involvement of 10%-30% of BSA is named as SJS/TEN overlap. Traditionally, SJS/TEN have been classified in the Erythema Multiforme (EM) spectrum because of similar skin lesions and histologic findings. In the current literature, they are considered as separate entities with different clinical picture and etiology [4]. EM are generally triggered by infections and in EM major typical lesions distributed acrally, whereas in SJS trunkal and facial involvement predominates. Characteristic features of SJS/TEN and EMM are given in Table 1, according to the expert’s definition [2,5]. The etiology of SJS/TEN is not precisely known, but it’s triggered either by drugs most common being anticonvulsants, antibiotics or nonsteroidal antiinflammatory drugs, pregnancy, vaccines or by infections such as mycoplasma pneumonia, especially in children and adolescents [1,2,6]. Some several genetic risk factors have been identified for TEN/SJS associated with drugs. Carbamazapine-induced SJS/TEN was strongly associated with HLA-B*1502 in Asian patients [3].

<table>
<thead>
<tr>
<th>Classification</th>
<th>EMM</th>
<th>SJS</th>
<th>SJS/TEN</th>
<th>TEN with macules (spots)</th>
<th>TEN on large erythema (without spots)</th>
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<tr>
<td>Skin detachment</td>
<td>&lt; 10%</td>
<td>&lt; 10%</td>
<td>10-30%</td>
<td>&gt; 30%</td>
<td>&gt; 10%</td>
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<tr>
<td>Typical target lesions</td>
<td>Yes</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
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<tr>
<td>Atypical target lesions</td>
<td>Raised</td>
<td>Flat</td>
<td>Flat</td>
<td>Flat</td>
<td>-</td>
</tr>
<tr>
<td>Macules (spots)</td>
<td>-</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>-</td>
</tr>
<tr>
<td>Distribution</td>
<td>Mainly (Acral)</td>
<td>Widespread (Trunkal/facial)</td>
<td>Widespread (Trunkal/facial)</td>
<td>Widespread (Trunkal/facial)</td>
<td></td>
</tr>
</tbody>
</table>

Table 1: Characteristic features of SJS/TEN and EMM [2,5].

Although SJS/TEN is a rare phenomenon with an incidence of 1.5-1.8/1,000,000 per year. It is a public health concern. Because of the high mortality rate and associated incapacitating sequelae [7,8]. We hereby report a severe case of SJS survivor who experienced serious long-term sequelae with respiratory, ophthalmologic, genitourinary and cutaneous involvement.

Case Report

An 18 year-old female patient was first admitted to the pediatric emergency unit at the age of 2 with complaints of difficulty in breathing, fever and rash. According to the patient’s hospital records, her history was significant for use of ampicillin due to an upper respiratory tract infection within a week. At the initial examination,
target shaped erythematous and bullous lesions all over the body, hyperemic conjunctivae and edematous eyelids, and erosive mucosal lesions in the mouth were observed. She was transferred to The Pediatric Intensive Care Unit (PICU) with a preliminary diagnosis of SJS/TEN, where systemic steroid and supportive therapy including ophthalmologic care were given. Her skin biopsy was found to be compatible with SJS/TEN. On the 18th day of hospital admission she could be transferred to the pediatric ward where her cough complaint was persisted. She was prescribed inhaled corticosteroids. On ophthalmic examination, eyelid margin keratinization, symblepharon causing fornical shortening and severe corneal staining was detected. Symblepharon release was performed and aggressive medical treatment including artificial teardrops and topical steroids was continued. During follow-up, entropion surgery was performed additionally due to the cicatricial entropion of the lower eyelids, amniotic membrane transplantation for the treatment of ocular surface problems was also done. Unfortunately, the visual acuities of both her eyes decreased due to ocular surface complications despite frequent screening.

Figure 1: Thorax CT showing bilateral bronchiectasis at age 17.

During her stay at the hospital, contractures had developed on the left 5th finger, which prompted a plastic surgery consult, resulting in an operation. Long-term follow up was performed at the pediatric outpatient clinic by irregular appointments because of poor adherence. Inhaled corticosteroid and chest physiotherapy was continued for lower respiratory symptoms and signs. Spirometric tests could not be performed due to lack of cooperation because of lip contracture. She had frequent upper respiratory tract symptoms due to chronic sinusitis. A thorax computed tomography at 10 years of age revealed slight interstitial thickening at the upper and middle lobes of the right lung and bilateral bronchiectasis at the lower lobes. At the age of 14, she complained of dysuria and abdominal pain. On physical examination, a fine labial stenosis around her clitoris and urethral meatus, which had been noticed by her mother at infancy, with no vaginal opening was observed. Ultrasonographic examination revealed a fluid-filled uterus and vagina, hematometra and hematocolpos. Rectal examination under general anesthesia confirmed vaginal distension. She started to menstruate after surgical intervention and she was instructed to use vaginal dilators to maintain patency. At the age of 16, pale and tender lips was noticed on dental examination. Oral biopsy specimen demonstrated mild inflammatory cell infiltration and congestion of capillary vessels which might be compatible with SJS/TEN sequela. Periodontal assessment revealed grade 3 mobility. At the age of 17, varicose bronchiectasis and mucous impaction findings were detected on thorax CT (Figure 1). She is still on respiratory physiotherapy and inhaled bronchodilators combined with inhaled steroids. On her last ophthalmologic examination, visual acuities were light perception in the left eye and 20/60 in the right eye. Biomicroscopic examination revealed total corneal vascularization and glaucoma in the left eye. Severe fornical shortening and corneal staining were noted in the right eye (Figure 2). Currently, she has scarring of the skin and hypopigmented areas in various parts of her body (Figure 3).

Figure 2: A heavy pannus and corneal vascularization of the right eye and bilateral symblepharon.

Discussion

SJS/TEN are life-threatening conditions and long-term sequelae affect life quality of survivors with significant morbidity. Our patient experienced most of the complications of the disease and still has many related ongoing problems.

Clinically, in the acute stage, patients show generalized macules with purpuric centers progressing to large conflating blisters with subsequent epidermal detachment. Involvement of mucous membranes resulting hemorrhagic mucositis can be seen in mouth, nose, eyes, genitalia, vulva, gastrointestinal tract and bronchi [6]. Treatment is symptomatic, apart from treating the triggering infections and elimination of causative drugs. The use of systemic corticosteroids and intravenous immunoglobulins is controversial [9].

In the chronic stage healing of the cutaneous and mucosal lesions can lead to long-term sequelae with scarring, stenosis or strictures. The most common reported long-term sequelae are dermatological and
ocular. Hyper- or hypopigmentation of skin was frequently observed, whereas scarring and hypertrophic changes of skin, loss of nails are reported infrequently. Ocular complications including myopia, photophobia, trichiasis, corneal scarring, symblepharon, dryness, visual acuity problems are among the reported complications [6,8].

Figure 3: Scarring of the skin at age 17.

Oropharyngeal involvement of the mucous membranes including mouth and lips are almost always found in the acute stage. Oral symptoms such as blisters and ulcerations in lips are the most characteristic findings in SJS/TEN patients. However, ulcerative lesions are rarely seen on gingiva. Oral hygiene and symptomatic care of the gingivitis and an inappropriate care such as management of extensive dermatological involvement, infections, nutrition, pulmonary, ophtalmological, and genitourinary complications in the acute phase; and screen for delayed, debilitating complications in the long term follow-up. Last but not least, multidisciplinary approach in the treatment and follow-up of patients with this disease may be the most important factor for the elimination of complications.

References