

Administration of Intravenous Immunoglobulin in the Management of Two Different Cases of Stevens-Johnson Syndrome

Abdel Hamid El- Gamal, MD
Radhir Kadian, MD
Sahar Nazmi, MS
Nabeel Najem, MD

Department of Dermatology, Al-Adan hospital, Kuwait

Corresponding author:
Abdel Hamid El- Gamal, MD
Department of Dermatology, Al-Adan hospital, Kuwait
Email:abd17mid@yahoo.com

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Abstract

Two different cases of Stevens - Johnson syndrome are reported. We used intravenous immunoglobulin in their management and compared the results of treatment with the conventional lines of therapy in a historical control. (Iran J Dermatol 2009;12: 20-21)

Keywords: immunoglobulin, intravenous, Stevens-Johnson syndrome

Introduction

Stevens - Johnson syndrome is an immune complex mediated hypersensitivity reaction. It typically involves the skin and mucous membranes. It is considered as a serious systemic disorder with the potential for severe morbidity and even death. No specific etiology has been identified as a direct cause but drug reactions, viral infections and malignancies may be implicated ¹.

Case No.1

In September 2006, a female child aged 12 years was admitted to the pediatric unit of Al-Adan Hospital. She complained of difficulty in swallowing with multiple erosions affecting eyes and lips with a preceding history of fever and cough. She had received amoxicillin for 2 days prior to admission. Dermatology consultation was sought on the 2nd day of admission. On examination, she was irritable and febrile (40°C). Raised atypical target lesions were observed over the palms and tips of fingers. Hemorrhagic crustations were seen over the lips. Also, there was puffiness and erythema of the eye lids associated with congestion of the eyes. External genitalia were free. Complete blood count was normal with increased ESR (92 mm/hour) and elevated CRP (70 mg/L). Chest radiography

showed left sided interstitial pulmonary infiltration. Mycoplasma cold agglutination was positive.

The diagnosis of Stevens - Johnson syndrome was made. She received erythromycin in a dose of 40 mg/kg/day and intravenous fluids. A full ophthalmological assessment was made. Intravenous immunoglobulin in a dose of 1 gm/kg was given for 2 days. On the 3rd day, patient's irritability lessened, fever decreased and muco-cutaneous lesions showed mild improvement. IV immunoglobulin was continued for another 2 days. On the 5th day, the patient reported improvement in swallowing and the pain over the lesions markedly decreased. On 7th day, target lesions over the palms and the crustations over the lips showed marked improvement. Also, the puffiness and congestion of the eyes subsided. Complete recovery occurred at the end of the 3rd week.

Case No. 2

In May 2007, a 29-year-old female patient was admitted into ICU at Al-Adan Hospital after an alleged road traffic accident. She was put on ventilator with diagnoses of right pneumothorax, Acute Respiratory Distress syndrome(ARDS) and associated pelvic fracture due to the car accident. She received multiple drug therapy in the form of first generation cephalosporin, ceftriaxone,

metronidazole, gentamycin, ranitidine, lasix, etc. Ten days later, she began to develop skin rashes. Dermatology consultation was taken. On examination, she was unconscious, afebrile and her blood pressure was 125/80 mmHg with a regular pulse rate of 80/min. Flat atypical target lesions were observed over the shoulder areas and chest. Multiple denuded lesions with some crustations were seen over both upper limbs. Hemorrhagic crustations over the lips, nostrils and eye lids were seen. External genitalia were free of lesions. The diagnosis of Stevens - Johnson syndrome was made. A full eye assessment was done. Complete blood count, liver and renal function tests, And ECG were normal. Chest radiography showed right pneumothorax. All suspected offending drugs were stopped and IV immunoglobulin in a dose of 1 gm/kg was administered for 3 consecutive days.

On the 4th day, stoppage of the appearance of new lesions was observed and old lesions began to improve. On the 6th day, there was marked eye improvement with significant disappearance of crustations over the lips. On the 9th day, the crustations over the denuded areas of upper limbs showed marked improvement and some skin lesions subsided. By the end of the 3rd week, all crustations had fallen down with complete re-epithalization.

Discussion

The pathogenesis of Stevens - Johnson syndrome is unknown but is consistent with cell mediated cytotoxic reactions against epidermal cells. The hapten binds to keratinocytes making them immunogenic ². The time from exposure to the hapten and the onset of symptoms is usually a few days. The patient may experience fever and influenza-like symptoms prior to the onset of mucocutaneous lesions³.

Controversy exists concerning routine use of systemic corticosteroids in the management of Stevens - Johnson syndrome. Some claim beneficial effects particularly if corticosteroids are given early in high doses while others discourage its use ^{4, 5}. The use of intravenous immunoglobulin by continuous

drip, early in the eruption, has been anecdotally reported to be beneficial. The eruption usually starts improving on the 4th to 5th day. Epithelization begins within a few days and is completed in a few weeks ^{6, 7, 8}.

In our patients, beginning of improvement was observed on the 5th day in the 1st case and on 4th day in the 2nd case. Recovery was complete by the end of the 3rd week in both cases.

The optimal management of Stevens - Johnson syndrome should include immediate removal of the suspected offending agent(s) and early attention to fluid and electrolyte status. Use of systemic corticosteroids remains controversial. Intravenous immunoglobulin therapy was successful in reducing the pain and promoting wound healing in these two different cases of Stevens - Johnson syndrome.

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