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Stevens-Johnson Syndrome

Stevens-Johnson Sendromu

Murat KARAMAN, MD, Adem Emre İLHAN, MD

Ümraniye Training and Research Hospital, Department of Otorhinolaryngology, İstanbul

ABSTRACT

We present a case of Stevens-Johnson Syndrome (SJS) and aim to stress its importance in otolaryngologyic practice, as it may cause severe respiratory distress. The patient applied to the otolaryngology outpatient department with complaints of painful skin rashes, redness of eyes, sores in the mouth and dysphagia. He was on penicillin treatment for upper respiratory tract infection for the last five days. We stopped the antibiotic treatment (penicillin) and started high dose steroid (prednisolone) for its anti-edema and anti-inflammatory effects. Patient showed complete and dramatic recovery after proper treatment. SJS may effect upper respiratory tract causing severe respiratory distress and even surgical interference may be needed. Following proper treatment, patients with SJS usually recover completely and unbelievably.

Keywords

Stevens-Johnson Syndrome; diaper rash; erythema multiforme; deglutition disorders

ÖZET

Ciddi respiratuvar distrese neden olduğundan, Kulak Burun Boğaz hastalıklarına ışık tutması açısından Stevens-Johnson Sendromlu (SJS) bir olgu sunduk. Hasta ağrılı deri döküntüsü, gözlerde kızarıklık, boğaz ağrısı ve yutma güçlüğü şikayetleriyle KBB polikliniğine basvurdu. Hastanın hikayesinde, son 5 gün içinde üst solunum yolu enfeksiyonu nedeniyle penisilin kullanmı mevcuttu. Hastanın antibiyotik (penisilin) tedavisini sonlandırıp antiödem ve antienflamatuvar etkisi nedeniyle yüksek doz steroid tedavisi (prednizolon) başladık. Hasta verilen tedavi sonrasında dramatik ve tam bir iyileşme gösterdi. SJS cerrahi müdahele bile gerektirebilecek ciddi respiratuvar distres oluşturarak üst solunum yolunu etkileyebilir. SJS'lu hastalar uygun tedavi sonrasında dramatik ve tam bir iyileşme gösterirler.

Anahtar Sözcükler

Stevens-Johnson sendromu; diyaper raş; eritema multiforme; yutma bozuklukları

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Correspondence

Murat KARAMAN, MD

Ümraniye Training and Research Hospital,

Department of Otorhinolaryngology, Ümraniye, İstanbul

GSM: 05055664178

Fax: +90 2166417101

E-mail: karaman1398@yahoo.com

INTRODUCTION

Stevens-Johnson Syndrome (SJS) was first described in 1866 by Hebra as a self-restricting acute disease of skin and mucous membranes and it is characterized by skin lesions which have the tendency of recurrence and show symmetrical extension especially in extremities.[1] SJS is a member of erythema multiforme complex together with erythema multiforme major, erythema multiforme minor and toxic epidermal necrolysis (TEN).[2] SJS and TEN are usually caused by exposure to drugs or their metabolites and may result in serious conditions due to wide mucocutaneous involvement.[3] Supportive treatment is the standard therapy for SJS and TEN.[4] Systemic steroids,[5,6] and immunosuppresives and immunoglobulins[7-10] can also be used as alternative treatment options, although controversial. In this article, we present a case who referred to otolaryngology outpatient department from dermatology department. The case is discussed in the light of the literature, and we aimed to direct attention to this syndrome in otolaryngologic practice since it is rarely seen.

CASE REPORT

The patient was referred from The Department of Dermatology to The Department of Otorhinolaryngology in Bezm-i Alem Vakıf Gureba Research and Training State Hospital with the complaints of painful skin rashes, redness of eyes, sores in the mouth and dysphagia. He was on penicillin treatment for upper respiratory tract infection for the last five days. The patient had no respiratory distress. Physical examination revealed widespread maculopapular rash in his whole body (Figure 1, informed consent for publishing of this figure was taken from the patient). There were edema and membranous lesions in his lips and oral mucosa (Figure 2, informed consent for publishing of this figure was taken from the patient). His eye examination showed conjunctivitis. We stopped the antibiotic treatment (penicillin) and admisitered high dose steroid (prednisolone 16 mg tb, 1 mg/kg per day) for three weeks because of its anti-edema and anti-inflammatory effects. We also started intravenous fluid replacement and analgesics. Oral hygiene was achieved with debridement of membranous lesions and oral topical anti-inflammatory solutions. Patient showed total and unbelievable recovery after the proper treatment at the end of the 3rd week. The informed consent for publishing of this case report was taken from the patient.

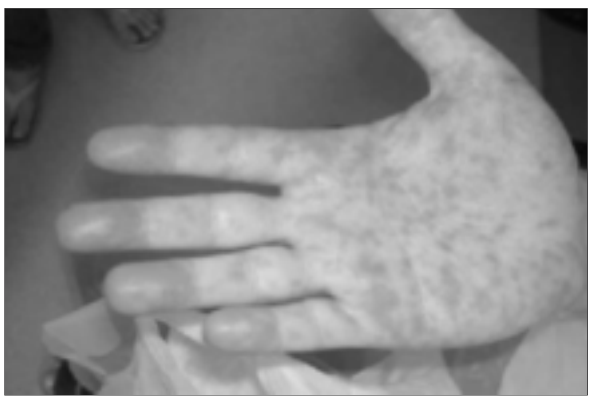

Figure 1. Widespread maculopapular rash in whole body

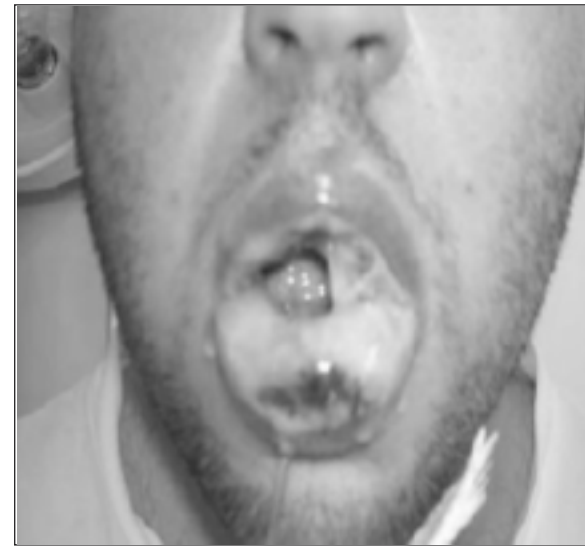

Figure 2. Edema and membranous lesions in his lips and oral mucosa

DISCUSSION

SJS is a member of erythema multiforme complex and is usually caused by exposure to drugs or their metabolites. SJS is characterized by widespread mucocutenous involvement resulting in photophobia, dysuria, respiratory distress, bacterial superinfection, synechiae, electrolyte and fluid loss. Characteristic erythema multiforme lesions are acute inflammatory skin lesions which have a central erythematous disc surrounded by an edematous ring and then again by an outermost erythematous disc.[3] We can observe flu-like symptoms one to three days prior to typical mucocutaneous lesions.[11] Symptoms manifest themselves one to three weeks following exposure to causative agent.[12]

Although many drugs may be thought to be responsible, the probable causative agent in this patient was penicilin, in accordance with a history of penicillin intake. Mortality due to SJS is higher with drugs having longer half lives.[13] SJS can be seen after vaccination with smallpox vaccine.[3,14] SJS may also occur after herpes and mycoplasma infections especially in children.[15]

Pathogenesis of SJS is thought to be associated with immune mediated keratinocyte apoptosis resulting in epidermal dissociation and mucosal involvement.[16] TEN and SJS belong to the same group and sometimes they cannot be differentiated. According to a classification, if epidermal dissociation covers 10% of the whole body surface, then it is defined as SJS; if it covers >30% of the whole body surface, then it is defined as TEN and between 10% and 30% it is defined as TEN-SJS overlapping disease.[2] However, widespread epidermal dissociation without mucosal involvement is much more characteristic to TEN.[17]

SJS is more likely triggered with drugs. High fever, sore throat and fatigue like symptoms can be seen before rashes. Symptoms with rashes become manifest 1 to 3 weeks after exposure to a drug. In some patients, mucocutaneous involvement may be limited. In our patient, physical examination showed widespread maculopapular rash in his whole body.

SJS treatment requires a multidisciplinary approach. Intravenous fluid and electrolyte replacement, analgesics, nutritional support and prophylactic antibiotics for bacterial superinfection must be administered. Debridement of necrotic lesions is necessary and ophthalmologic agents are also useful for treatment.[3] Steroid administration is still controversial and is said to increase the risk of septicemia and gastrointestinal bleeding.[5,6] There are different opinions about immunosuppressives and intravenous immunoglobuline (IVIG) treatment.[7-10] In this case, we stopped the antibiotic treatment (penicillin) and started intravenous fluid replacement, analgesics and high dose steroid (prednisolone) for its anti-edema and anti-inflammatory effects. Oral hygiene was maintained with debridement of membranous lesions and topical anti-inflammatory wash-outs.

Ocular, nasal, pharyngeal and laryngeal mucosa can be effected.[18] In a study with 28 SJS patients, it was found that 26 (93%) patients showed head and neck manifestations. In these patients the most effected areas include lips (93%), conjunctiva (82%), oral cavity (79%) and nose (36%).[19] In our case, there were also edema and membranous lesions in his lips and oral mucosa; his eye examination also revealed conjunctivitis. Upper respiratory tract involvement may cause severe respiratory distress.[19,20] Reason for respiratory distress is glossitis and involvement of supraglottic area.[20,21] Even sudden death has been seen in one case who was diagnosed as SJS by autopsy.[21] Esophageal involvement may cause hemorrhagia due to mucosal eruption and bulla formation in the early phase, whereas esophageal stricture and web formation may present in the late phase.[22,23]

CONCLUSION

In conclusion, SJS may affect upper respiratory tract causing severe respiratory distress and even surgical intervention may be needed. Patients may also need nutritional support and additional care for possible stricture and web formation if esophageal involvement ensues. Following proper treatment, patients with SJS usually show total and unbelievable recovery.

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