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Case Report

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Drug Induced - Stevens Johnson Syndrome: A Case Report

Abstract
SJS is an acute, self-limited disease, Presenting as severe mucosal erosions with widespread erythematous, cutaneous macules or atypical targets. Majority of cases are drug- induced, affecting oral & peri-oral region. We report a case of drug induced SJS in a 16 yr old female which was treated lucratively with steroids. Diagnostic criteria have changed, and more data exist on drugs with an increased risk. Although there is no standardized treatment for all patients with SJS/TEN, options that have been used include cyclosporine, corticosteroids, and intravenous immunoglobulin.

Key Words
Bleeding, Syndrome, Immunoglobulin, Ulcer

[1] Veena Naik
[2] Shivaprasad S
[3] Ashok L

[1] P.G. Student
[2] MDS, Professor
[3] MDS, Professor and Head
Dept. of Oral Medicine and Diagnostic Radiology,
Bapuji Dental College and Hospital, Karnataka

Address For Correspondence:
Dr. Veena Naik
Post Graduate Student
Dept of Oral Medicine and Diagnostic Radiology,
Bapuji Dental College and Hospital,
Davangere-577004, Karnataka, India
E Mail Id : dr_veenasnaik@yahoo.co.in
Phone No : 09886027748

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Introduction

"A new eruptive fever with stomatitis and opthalmia" was described as a severe variant of erythema multiforme & was termed by Steven and Johnson in 1922. By the 1940's it was commonly called as "Steven Johnson's syndrome (SJS)". The concept of the spectrum of erythema multiforme has been widely accepted since that time[1]. SJS is an acute, self-limited disease, Presenting as severe mucosal erosions with widespread erythematous, cutaneous macules or atypical targets[2].

Although SJS is rare with an incidence of 0.05 to 2 persons per 1 million populations per year, it has significant impact on the public health in view of its high morbidity and mortality[3]. Majority of cases are drug-induced[4].

SJS is one such disease which could manifest with extensive oral and peri-oral involvement, where oral physicians come into picture. Hence we are presenting a case of SJS, which has been successfully treated.

Case Report

16 year old female came to our Department with a complaint of painful ulcers in the mouth & bleeding from lips since a day. She presented with fever a week back for which she was prescribed antibiotics & analgesics after which she developed ulcers in the mouth & bleeding from lips, associated with pain which was sudden in onset, burning type, continuous, localized, and severe in intensity, aggravated on touching, speaking, eating food & there was no relieving factor. She also had burning micturation & watering of both eyes since 4 days. Her past medical history revealed that she was suffering with fever a week back & was on tab. Ciprofloxacin 500mg tid for 5days & tab Paracetamol 500mg + Diclofenac sodium 50mg tid for 3days for the same. Her past dental & surgical histories were non contributory. Patient had elevated temperature of 100F. Bilateral submandibular lymph nodes were palpable, tender, mobile, firm in consistency. Extra orally there was limited mouth opening & erythematous crusted areas on both upper & lower lip extending upto the vermilion border with fresh bleeding spots, surface appeared rough & scaly, on palpation the periphery of the lesion was hard & tender **(figure 1)**.

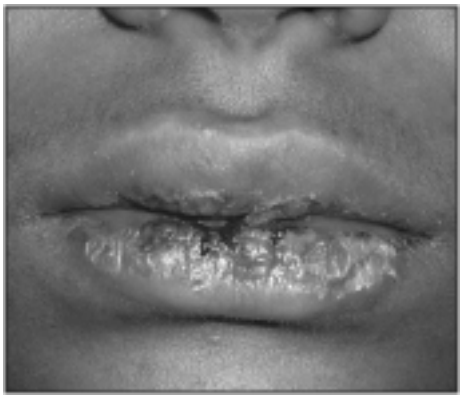

Figure 1 - Erythematous Crusted Areas On Both Upper & Lower Lip Extending Upto The Vermilion Border With Fresh Bleeding Spots

Bleeding was evident on touching with peeling up of overlying skin of lips. There was solitary ulcer seen in conjunctiva bilaterally, associated with watering of eyes & pus discharge **(figure 2)**.

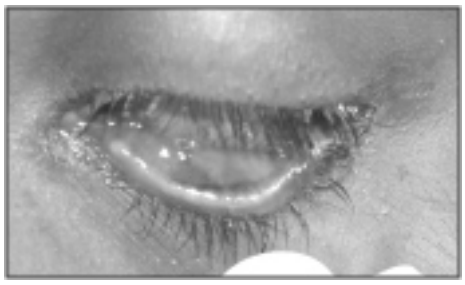

Figure 2 - Solitary Large Ulcer On The Conjunctiva Bilaterally

The vaginal lesion was confirmed with examination in department of Veenerology. Bilateral solitary submandibular lymph nodes were palpable, tender, mobile, firm & measured 1x1 cm. Intra oral examination revealed clusters of ulcers on lower labial mucosa, right & left buccal mucosa **(figure 3)** & right lateral border of tongue **(figure 4)** with fresh bleeding spots, each ulcer measured about 1×0.5 cm, surrounded with erythematous halo. Floor of ulcers revealed a whitish slough. Slight provocation initiated bleeding.

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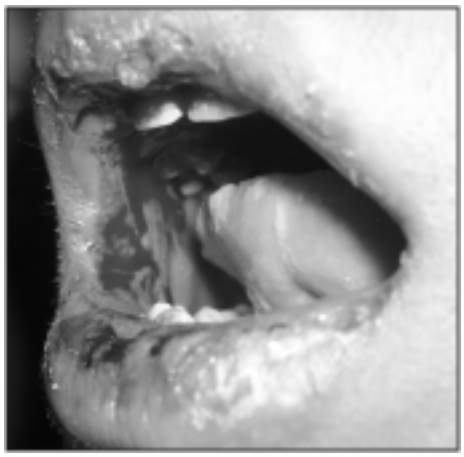

Figure 3 - Fresh Bleeding Spot With Ulcers On Right Buccal Mucosa

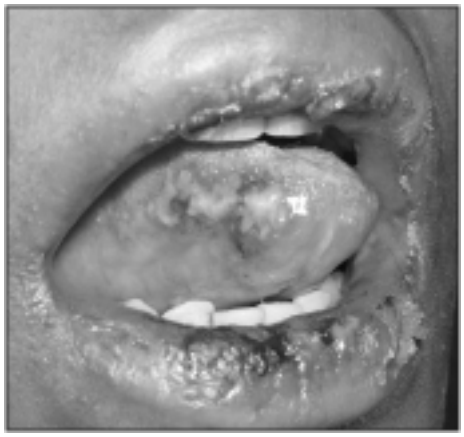

Figure 4 - Clusters Of Ulcers On Right Lateral Border Of Tongue

Based on this our clinical diagnosis was Stevens Johnson Syndrome as the lesion noticed in eyes & genitals. Differential diagnosis thought were phemphigus vulgaris & stomatitis medicamentosa. We could subject the patient to only the hematologic investigation as the lesion being acute; the patient was under severe discomfort. Her complete blood picture revealed raised ESR - 50mm/1st hr & total leucocyte count was 12000 cells/mm^3. Rest other findings were within normal range.

We treated her with systemic steroids, tab prednisolone 10mg qid for 7 days, which was gradually tapered to 10mg tid for 7days, 10mg bid for 5days & 10mg once daily for 5days respectively, benzydyamine hydrochloride 0.15% oral rinse for oral ulcers. Gention violet application for lip lesions. Clotrimazole cream 1% for vaginal lesion & ofloxacin eye drops 0.3% for eye lesion. Liquid & soft diet was advised. All the lesions healed within 15 days **(figure 5)**; there was absence of burning micturation & lacrimation **(figure 6)**.

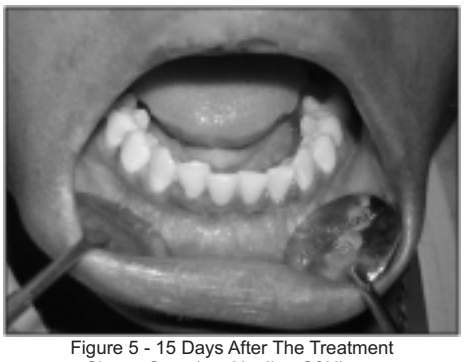

Figure 5 - 15 Days After The Treatment Shows Complete Healing Of Ulcers

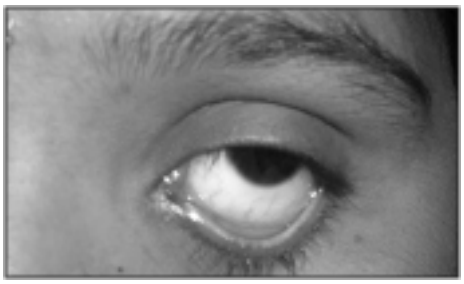

Figure 6 - Eye Lesions Healed

Discussion

Stevens-Johnson syndrome is a severe, episodic mucocutaneous intolerance reaction described by Hebra[5] in 1866 and Albert Mason Stevens and Frank Chambliss Johnson in 1922. Erythema multiforme (EM) and Stevens-Johnson syndrome are part of a clinical spectrum[6].

SJS is said to be associated with drugs. More than 100 different medications have been implicated. The syndrome usually begins within 1-14 days of ingestion of the offending agent[7]. In our case the interval was one week. SJS and TEN are considered T cell mediated disorders in which activation of CD8 T lymphocytes lead to destruction and apoptosis of keratinocytes[8]. Drugs can activate T cells by acting as a hapten, as a prohapten or by direct pharmacologic interaction among the drug, Major Histocompatibility Complex (MHC) molecule and a T cell receptor. It is postulated that drugs can bind with the MHC and T cell receptor causing activation of T cells contributing to SJS. Yap et al[3] determined that the drugs especially anticonvulsants and allopurinol were the major causes of SJS. Jean-Claude Roujeau, et al[9] in 1995 conducted a study on medication use & risk of SJS & TEN, according to whom the commonest drugs causing SJS included most classes of antibiotics, including cephalosporins, quinolones, aminopenicillins, tetracyclines, and imidazole antifungal agents. A recent study by Gokhan Okan, et al[7] in 2008 reported a case of ciprofloxacin induced SJS. This holds well in our case where the patient was on ciprofloxacin for a week. Kristina E, et al[10] stated that among the available NSAIDs, oxicam derivatives have the greatest association with SJS & the risk of SJS or TEN in patients receiving NSAIDs is extremely low; whereas older patients, women, and patients within the first month of treatment initiation have the greatest risk. Mainly drug- induced SJS-TEN Overlap and TEN have high mortality. The reported mortality rate ranges from 5% to 40% [11], [12]. The syndrome may be diagnosed on clinical criteria. Signs and symptoms consist of a non-specific prodrome of malaise, fever, headache, sore throat, cough, chest pain, vomiting, diarrhea or myalgias[6].

Systemic corticosteroids has unproven benefit in early cases of SJS and TEN and deleterious in the advanced forms[13]. Other treatment includes cyclosporine and intravenous immunoglobulin but new treatments, such as amniotic membrane support for ocular damage, may need to be considered[14]. To conclude drug induced SJS is more common & is considered to have high mortality rate, herewith we presented a case of drug induced SJS which was treated successfully with systemic steroids.

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