Case Report

STEVEN JOHNSON SYNDROME, AN UNUSUAL CLINICAL EXPERIENCE, A CASE REPORT

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ABSTRACT

Objective: *Steven Johnson syndrome is a rare, life threatening disorder characterized by skin condition with bullous formation, ocular lesions; genital and anal lesions/ulcers.It's usually a reaction to a medication or an infection. Often, Stevens-Johnson syndrome begins with flu-like symptoms, followed by a painful red or purplish rash that spreads and blisters. Then the top layer of the affected skin dies and sheds. This case report is about maxillofacial management of a 26 year old female patient came to the emergency of Abbasi Shaheed Hospital with compromised breathing, severe skin lesions with bullous formation and conjunctivitis. Initial diagnosis of Steven Johnson Syndrome was made and the patient was admitted in ICU and put on ventilator for 02 days. This case report elaborates the role of misuse of drugs mostly antibiotics/anti-virals given to common ailments like fever, sore throat, and fatigue which may result in life threatening disorder like Steven Johnson Syndrome as in this case where she received intra venous medicine (un-known) from local General Practitioner. Regular follow-up revealed marked improvement and resolution of oral, genital and skin lesions.*

Key words: *Steven Johnson Syndrome, Immune System Disorder.*

INTRODUCTION

Stevens–Johnson Syndrome (SJS) is a rare life-threatening skin condition. The syndrome is thought to be a hypersensitivity complex that affects the skin and the mucous membrane[1]. Stevens-Johnson syndrome occurs most often in children and young adults. Incidence ranges from 1.2 to 6 cases per million per year; the condition is fatal in 5% of treated cases and in 15% of untreated cases[2]. The condition is more common in adults than in children. Women are affected more often than men, with cases occurring at a two to one (2:1) ratio. The condition was first described in 1922 by Stevens and Johnson as a febrile illness with stomatitis, purulent conjunctivitis, and skin lesions[3]. Stevens–Johnson syndrome (SJS) is thought to arise from a disorder of the immune system[4]. The immune reaction can be triggered by drugs or infection[5]. Genetic factors are associated with a predisposition to SJS[6]. Although Stevens–Johnson Syndrome can be caused by viral infections and malignancies, the main cause is medications[7]. A leading cause appears to be the use of antibiotics particularly sulfa drugs[8].

Stevens–Johnson syndrome (SJS) usually begins with fever, sore throat, and fatigue, which is commonly misdiagnosed and therefore treated with antibiotics. Ulcers and other lesions begin to appear in the mucous membranes of the mouth, lips, genital and anal regions. Ulcers in the mouth are usually extremely painful and reduce the patient's ability to eat or drink. Conjunctivitis occurs in about 30% of children who develop SJS, a rash of round lesions about an inch across arises on the face, trunk, arms and legs, and soles, but usually not the scalp[4].

SJS (with less than 10% of body surface area involved) has a mortality rate of around 5%.The risk for death can be estimated using the SCORTEN scale, which takes a number of prognostic indicators into account[9]. Other outcomes include organ damage/ failure, corneal scratching, and blindness.

CASE REPORT

A 26 year old female patient reported to the emergency of Abbasi Shaheed Hospital with compromised breathing, severe skin lesions with bullous

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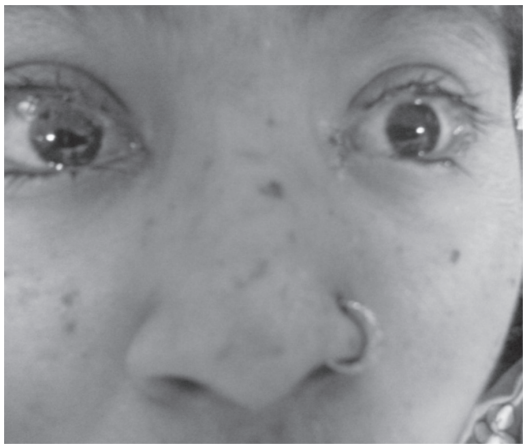

Fig - I: Patient with Conjunctivitis and Watery Eyes

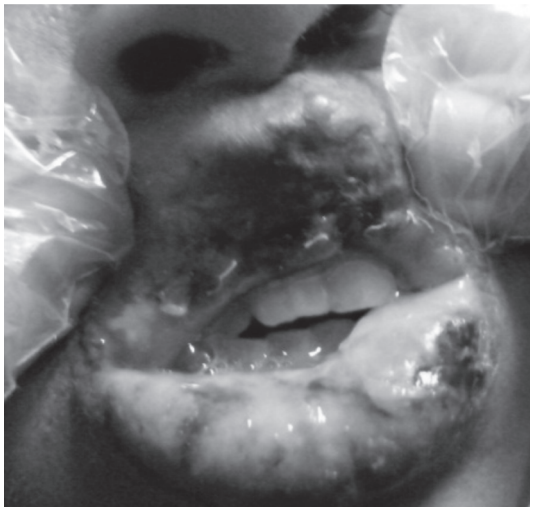

Fig - II: Lip Ulceration and Mucosal Desquamation

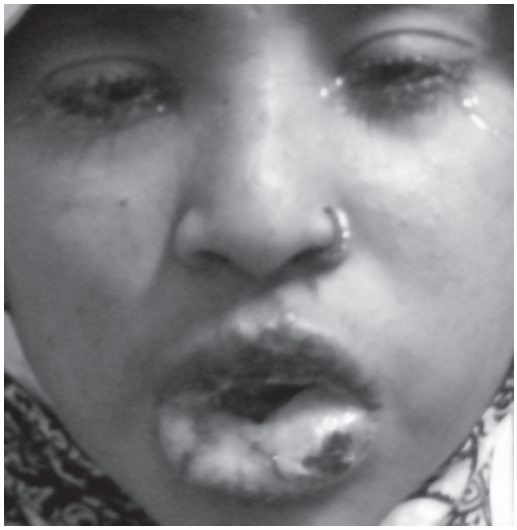

Fig - III: Watery Eyes and Mucosal Desquamation

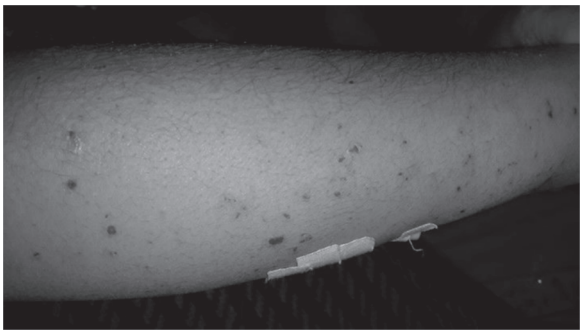

Fig - IV: Target Lesions on the Arm

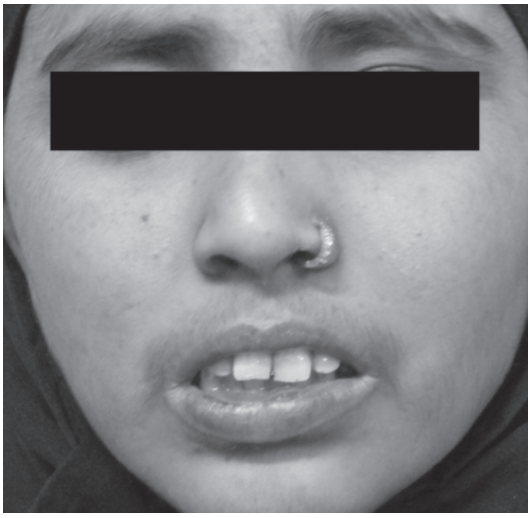

Fig - V: Follow up after 3 weeks

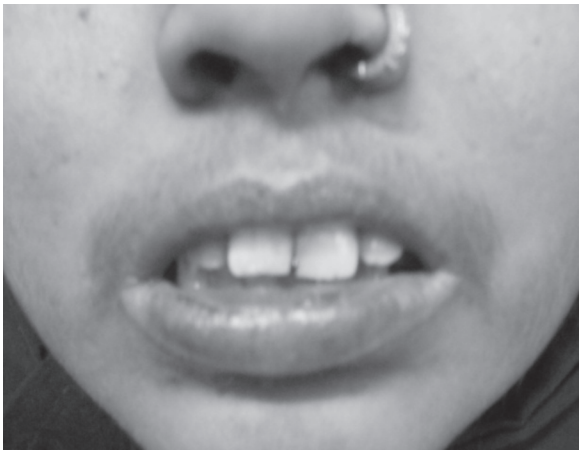

Fig - VI: Lip ulceration and mucosal desquamation recovered after 3 weeks

formation, conjunctivitis and watery discharge from eyes (Figure-I). Ulcerations were noticed on lips, oral mucosa, while genital and anal regions were also involved. The patient was a known case of epilepsy since childhood and was taking oral Phenytoin.

The patient gave a history of lips and oral ulceration for which she visited a nearby clinic where she was injected with some unknown medicine for 02 days, on 3rd day she developed skin lesions with bullous formation and difficulty in breathing. On physical examination patient was disoriented, her vitals were: temp 102°F, Pulse rate 88 beats/min, Respiratory rate 22/min, Blood pressure 80/50 mm/Hg. On oral examination lips were swollen, cracked, crusted while bleeding was evident. Intra oral examination reveals mucosal desquamation (Figure-II and Figure-III). Her general physical examination revealed target lesions on both arms and legs (Figure-IV). On systemic examination, patient had dyspnea on auscultation, bronchospasm was noted while cardiovascular and abdominal examination were unremarkable. She was immediately admitted in ICU and placed on ventilator for 02 days, later she was shifted to the ward were she remain for 15 days. Patient's laboratory Investigations showed white blood cell count 7.2 x 10/uL, Neutrophils count 82.5%. INR was also elevated 1.9, Patient's renal profile and serum electrolytes were normal. During her stay in ICU patient was managed for shock and to maintain respiration, while in ward she was given symptomatic treatment: A combination of Amoxicillin and clavulanic acid 1.2 gm, metronidazole 500 mg and Dexamethasone 8 mg were given intravenously 8 hourly, analgesic Diclofenac sodium 75 mg was given intramuscularly twice daily, Tab Carbamazepine twice daily, analgesic and antiseptic mouth rinses and gels for oral ulcers, while for skin lesions topical anaesthetics and antiseptics were given. Ophthalmologic consultation was obtained to address ocular symptoms.

Regular follow-up revealed marked improvement and resolution of oral, genital and skin lesions (Figure-V and VI)

DISCUSSION

Stevens-Johnson syndrome can be preceded by a prodrome consisting of fever, malaise, sore throat, nausea, vomiting, arthralgias, and myalgias[10]. This prodrome is followed within [14] days by conjunctivitis and by bullae on the skin and on the mucosal membranes of the mouth, nares, pharynx, esophagus, urethra, and vulvovaginal as well as anal regions. Stevens-Johnson syndrome commonly affects multiple organs, and esophageal strictures develop in some patients[11].

Ocular complications occur in about 70% of patients which are photophobia, purulent conjunctivitis, corneal ulcerations, anterior uveitis, corneal opacity, and blindness[10].

A sepsis from widespread skin infection, respiratory track involvement such as tracheobronchial ulcerations, pneumonia, renal failure and cardiac complications, can lead to death in particular complications[12].

In 70% of SJS cases, drugs are found to be causative agents and more than 100 such agents have been reported. In SJS, it's necessary to take drug history carefully and repeatedly before the causative agent can be identified[13]. Short courses of sulfonamide, aminopenicillin, quinolone, and cephalosporin drugs all increase risk of Stevens-Johnson syndrome. Longer-term therapy with anticonvulsant agents, oxicam, nonsteroidal anti-inflammatory drugs (NSAIDs), or allopurinol has also been named as a possible cause of Stevens-Johnson syndrome[14]. Stevens-Johnson syndrome also has been linked to herpes simplex virus, mycoplasma bacterial species, and measles vaccine[15]. Neoplasms and collagen diseases have also been pointed out as possible causes[4]. The cause of SJS is unknown in one quarter to one half of cases[6].

The criteria for diagnosis of SJS are epithelial detachment less than 10% of Body Surface Area (BSA) and widespread erythematous or purpuric macules of flat atypical targets. SJS has to be clinically differentiated from viral stomatitides, pemphigus, Erythema Multiforme (EM), toxic epidermal necrolysis (TEN) and the sub-epithelial immune blistering disorders like pemphigoid. Skin biopsy and tissue biopsy gives the confirmatory results.

Treatment for Stevens-Johnson syndrome is as diverse as the symptoms but should begin by withdrawing any offending agent identified. Many skin lesions can be treated with any of various topical mixtures[16]. However; extensive skin involvement requires the treatment on the guide lines of a major burn unit. Affected patients and their first-degree relatives should be instructed to avoid any identified drug or chemical that may be responsible[17].

Ocular involvement can be treated with topical

corticosteroid agents, artificial hydration, and antibiotic agents when indicated.

Treatment of oral symptoms depends upon severity. Milder forms may be treated with topical anesthetics and analgesics, proper wound care and soft diet. The condition may resolve within 2-6 weeks. For more severe cases systemic fluid replacement and topical antihistamines and corticosteroids may be needed[18].

CONCLUSION

Stevens-Johnson syndrome is a potentially fatal multi-organ disease with a strong etiologic link to certain medications. There is a need to create awareness among the medical professionals regarding medications which may cause SJS. Early diagnosis with the prompt recognition and withdrawal of all potential causative drugs is essential for a favorable outcome, affected patients and their first-degree relatives should be instructed to avoid any identified drugs or chemicals that may be responsible.

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