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CASE REPORT

STEVENS-JOHNSONS SYNDROME AND TOXIC EPIDERMAL NECROLYSIS: A CASE REPORT

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ABSTRACT

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are serious systemic disorders in which there are vesicobullous lesions involving the skin and mucous membranes, most commonly triggered by medications. It is a potentially fatal condition that damages multiple organs. A 22 years male patient admitted with the complaint of sudden appearance of blisters first over oral cavity after taking Ibuprofen. He developed generalized macular lesions over trunk, chest, face and lower limbs. He was treated with steroid, antibiotics and conservative management.

Key words: Ibuprofen, Steroid, Stevens Johnsons Syndrome.

INTRODUCTION

Stevens-Johnson syndrome (SJS) and toxic epidermal (TEN) are severe necrolysis mucocutaneous reactions, most commonly triggered by medications, characterized by extensive necrosis and detachment of the epidermis. SJS and TEN are considered a disease continuum and are distinguished chiefly by severity, based upon the percentage of body surface involved with skin detachment. They differ mainly in the extent of detachment, which is limited in SJS (<10% body surface area), more widespread in TEN (>30%), and in-between in SJS/TEN overlap (10%-30% of body surface area) 1.

This condition was first described in 1922 by Stevens and Johnson as a febrile illness with stomatitis,

purulent conjunctivitis, and skin lesions and was described as vesiculobullous erythema multiforme of the skin, mouth, eyes, and genitals². Although SJS is rare with an incidence of 0.05 to 2 persons per million populations per year, it has significant impact on the public health in view of its high morbidity and mortality³.

SJS is a type of hypersensitivity reaction due to medications (sulfonamides, penicillins, barbiturates, and phenytoin), infections (herpes simplex and mycoplasma), or illness. Approximately 90% of SJS cases are associated with herpes simplex or mycoplasma infections. In Stevens-Johnson syndrome, the systemic symptoms are severe and

the lesions are extensive, involving multiple body areas, especially the mucous membranes. Toxic epidermal necrolysis (TEN syndromes or Lyell's syndrome) involves multiple large blisters (bullae) that coalesce, followed by sloughing of all or most of the skin and mucous membranes⁴. Severe TEN is similar to extensive burns; patients are acutely ill, may be unable to eat or open eyes, and suffer massive fluid and electrolyte losses. They are at high risk of infection, multiorgan failure, and death. With early therapy, survival rates will be 90%⁵.

CASE PRESENTATION

A 22 years male patient came to emergency in Chitwan Medical College, Bharatpur on 071/06/07 at 12:24 am with chief complaint of sudden appearance of blisters first over oral cavity after taking Ibuprofen which was used by the patient for eye pain on 2071/05/29. He had generalized appearance of macular over trunk, then chest, face, lower limbs and also bullae over chest, trunk, and lower limbs for 3 days. On arrival in emergency, he was well-oriented, B/P: 130/90 mmhg, Pulse: 110/min, R: 24/min, T: 99°F, SPO2: 98% and on examination, generalized maculopapular and bullous eruptions on the neck, face, external ear.

Then the patient was admitted to Burn unit of surgical ward on 071/6/8. He was then transferred to ICU on same day at 5:15pm due to painful oral erosions with severe crusting of the lips and increased salivation

(Figure 1) lead to difficult in swallowing. While involvement of genitalia led to painful micturition. Patient had plaques with vesicles, sloughed off more than 50% of BSA (Figure 2) and Sheet like desquamation on the foot (Figure 3). Vein assess cannot be done so CVP insertion done. Patient was on NG feeding. Patient was treated with antibiotics, steroids, ciprofloxacin eye drop and ointment, Mupirocin ointment locally, dressing of eyelids with moist cotton.



Fig 1: Extensive sloughing on the face

NURSING DIAGNOSIS

- Impaired skin integrity related to inflammatory dermal and epidermal
- Activity Intolerance related to physical weakness
- Acute pain related to inflammation of the skin
- Imbalanced nutrition less than body requirements
 related to difficulty swallowing
- Lack of knowledge about the disease process

associated with less information.

Potential secondary infections associated with side effects and therapeutic steroid infusion





Fig 2: Extensive sloughing and blistering on the body



Fig 3: Sheet like desquamation on the foot

On the basis of above nursing diagnosis, nursing care was provided. Gradually his general condition improved, shifted to normal diet and was discharged on 2071/6/29 without any complications.

DISCUSSION

Steven Johnson's Syndrome is a serious systemic disorder. It can result as an immune response to an antigen or as a drug reaction. Most often it is considered as an allergic reaction. It is a self-limiting condition which responds to immediate management or may result in fluid loss, sepsis and death⁶.

In a study conducted on 225 references in India, 10 references were included as per selection criteria. The major causative drugs were antimicrobials (37.27%), anti-epileptics (35.73%) and non-steroidal anti-inflammatory drugs (15.93%), Carbamazepine (18.25%), phenytoin (13.37%), fluoroguinolones (8.48%) and paracetamol (6.17%). Total 62.96% of patients showed systemic complications. Most common complications were ocular (40.29%) and septicemia (17.65%). Higher mortality was observed for TEN as compared to SJS (odd ratio-7.19; 95% confidence interval (CI) 1.62-31.92; p = 0.0023). Duration of hospital stay was significantly higher in TEN (20.6 days; 95% CI 14.4-26.8) as compared to SJS (9.7 days; 95% CI 5.8-13.6; p = 0.020). Cost of management was significantly higher in TEN as compared to SJS. No statistical data were described for steroid use in the studies included⁷.

Fortunately SJS/TEN is a very rare complication of medication use (estimated to be 1-2/million each year for SJS, and 0.4-1.2/million each year for TEN). But anyone on medication can develop SJS/

TEN unpredictably. It can affect all age groups, both sexes and all races. It is more common in association with human immunodeficiency virus infection (HIV), which may reflect the increased use of medications by HIV patients⁸.

Drug-induced SJS presents with fever and influenzalike symptoms after the application of the suspected drug. One to 3 days later, signs begin in the mucous membranes, including eyes, mouth, nose, and genitalia in up to 90% of cases. Skin lesions manifest as generalized macules which progress to large blisters with subsequent epidermal detachment. In the following 3 to 5 days, separation of the epidermis progresses and leads to large denuded areas. The large wound area leads to extreme pain, massive loss of fluid and protein, bleeding, evaporative heat loss with subsequent hypothermia, and infection⁹.

Confirm the diagnosis by biopsy (showing necrotic epithelium) if clinical characteristics (e.g., target lesions progressing to bullae, ocular and mucous membrane involvement, desquamation in sheets) are inconclusive⁵.

CONCLUSION

Stevens-Johnson syndrome is a potentially fatal multiorgan disease with a strong etiologic link to some medications. Treatment with steroid agents may be helpful, but remains controversial. 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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