ORIGINAL RESEARCH PAPER

Plastic Surgery

EXTENSIVE TOXIC EPIDERMAL NECROLYSIS TREATED AT A BURN CENTRE – A CASE REPORT

KEY WORDS: Toxic epidermal necrolysis, steven Johnson syndrome, drug induced, burns ICU

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ABSTRACT

Toxic epidermal necrolysis (TEN) is one of the most severe drug-induced mucocutaneous reactions, characterised by extensive necrosis and detachment of the epidermis. In majority of cases, the administration of pharmaceutical drugs is thought to be the primary cause of Steven Johnson Syndrome (SJS)/TEN. Erythema, epidermal detachment that manifests as blisters, and denuded skin patches are the defining features of this pathology. In this article, we report a case of a 28-year-old male patient who presented to the emergency with a generalized maculopapular rash all over the body and was taken off the drug immediately and was referred to the burns unit with extensive blisters and skin eruptions all over the body. The patient was treated with intravenous fluids, iv antibiotics, parenteral nutrition and moisture retaining daily dressings. Patient was hospitalised for a period of 2 months and was healed completely without surgical intervention.

INTRODUCTION

Definition:

Toxic: Pertaining to, due to or the nature of a poison or toxin, manifesting the symptoms of severe infection

Epidermal: Pertaining to or resembling the epidermis

Necrolysis: Separation or exfoliation of tissue due to necrosis

Toxic epidermal necrolysis (TEN) is one of the most severe drug-induced mucocutaneous reactions, characterised by extensive necrosis and detachment of the epidermis. Stevens–Johnson syndrome (SJS) also exists along the same disease spectrum, but TEN is more severe.[1] The incidence of TEN/ SJS ranges from 2 to 7 cases per million people per year.[2] The overall mortality rate of TEN is 50%,[3] and the risk of death persists for up to 1 year after disease onset.

Patients are classified into three groups based on the body surface area involved, Stevens-Johnson syndrome if less than 10% BSA is involved.

SJS/TEN overlap between 10% and 30%.
TEN if more than 30%BSA is involved.

The Scottish dermatologist Alan Lyell first used the term TEN to describe a rare critical disorder known as Lyell's syndrome in 1956.[4] In majority of cases, the administration of pharmaceutical drugs is thought to be the primary cause of SJS/TEN.

Anticonvulsants (phenytoin, carbamazepine, lamotrigene, phenobarbitone), non-steroidal anti-inflammatory drugs, allopurinol, corticosteroids, nevirapine(anti-retroviral) and antibiotics (sulphonamides) are few of the drugs that might result in TEN[5]. Anticonvulsants are one of the primary causes of SJS/TEN and among them, carbamazepine is responsible for the majority of the instances [6,7].Erythema, epidermal detachment that manifests as blisters, and denuded skin patches are the defining features of this pathology. The underlying pathology of the skin lesions remains unclear. Some studies have suggested that apoptosis results from a cell-mediated cytotoxic reaction against keratinocytes.

CASE STUDY

In this article, we report a case of a 28-year-old male patient who gave history of fever and headache for which he received some medication from a general practitioner. Following this, the patient presented to the emergency with a generalized maculopapular rash all over the body and was taken off the drug immediately and was referred to the burns unit with extensive blisters and skin eruptions all over the body. He also had pain, redness, and discharge from both eyes.

On examination: Clinical examination revealed a non-blanching erythematous maculopapular rash with blisters all over the body including eyes and conjunctiva(fig 1). Nikolsky sign was positive over the back (fig 2). Swelling of the oral and nasal mucosa was also present. The patient was febrile, but all other vital signs were stable. His abdomen was mildly distended, and chest examination revealed no wheezing.

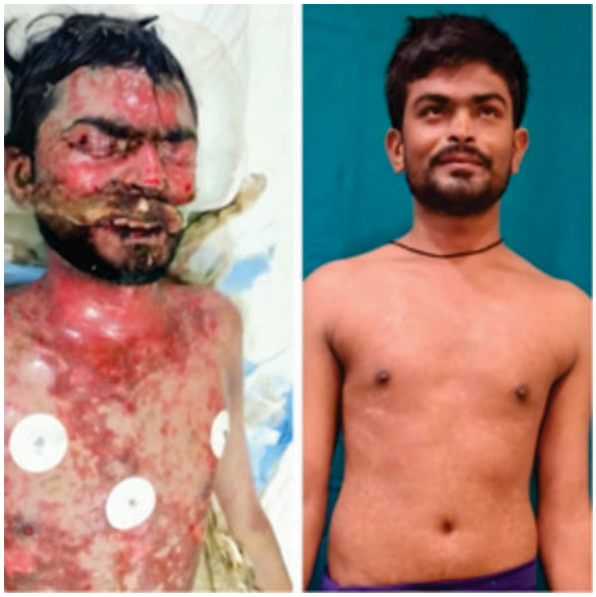

Fig1: showing the non-blanching erythematous rash with blisters over the chest and face region during admission and healed skin after treatment

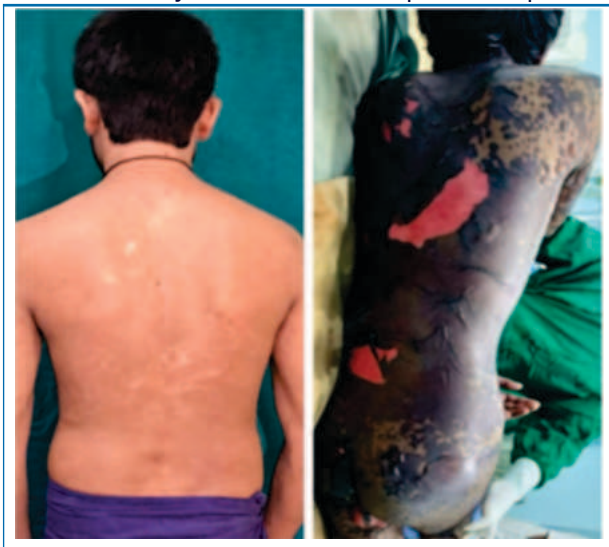

Fig2: Shows Nikolsky sign positive over the back region during admission and healed skin after treatment.

On investigating the patient during admission, the patient had leucocytosis with a white cell count of 12,800 cells/cu mm, haemoglobin of 10.2gm% and platelet count of 2.28 lakh/cu mm. His serum electrolytes, urea, creatinine, liver profile, prothrombin time, partial thromboplastin time and International Normalised Ratio were with in normal limits. His C-reactive protein concentration was 89.99mg/L. Ultrasound of the abdomen showed grade 1 fatty liver, and an echocardiogram showed mild pericardial effusion. Histopathology examination of the skin was done which revealed full thickness epidermal necrosis with sub epidermal blister formation and presence of perivesicular lymphocytes as well as dermal inflammation. The patient was provisionally diagnosed as TEN.

Treatment:- The patient was immediately transferred to the burns ICU because of the progressive worsening of the skin lesions involving the entire body, including the genitalia. In addition, the skin showed a tendency to peel off easily. Central venous catheter was inserted.

The patient was treated with intravenous fluids (normal saline and ringer lactate), intravenous antibiotics with Inj. colistimethate 2MIU for 7days followed by tab. cefixime 200mg for 7 days, intravenous dexamethasone 16mg/day in two divided dosages for 10 days then 8mg/day in two divided doses for 10 days and tapered with 4mg tablet usage for 7 days and stopped. Parenteral nutrition was started because of feeding intolerance and for calorie supplementation in the initial stages and then enteral feeding was optimized as his tolerance increased and semisolid high protein diet later before discharge. Moisture retaining daily dressing was done with ointment silver sulfadiazine and Vaseline-soaked gauze. The treatment regimen continued for three weeks and was stopped when the skin lesions were scarce and there was an improvement in the overall health of the patient.

Intravenous pantoprazole and Syrup Mucaine gel were given orally to prevent gastrointestinal bleeding. Inj albumin 20% was administered in view of hypoalbuminemia. Multivitamin injections, Vitamin C supplements, antioxidants and probiotics were administered on daily basis. DVT prophylaxis was given accordingly throughout the hospital stay.

The skin lesions improved, and the patient was transferred from ICU to general ward within forty days of admission.

The ophthalmology team was consulted regarding the conjunctival redness, corneal ulcers and pain in the eyes and chloramphenicol eye ointment, moxifloxacin eyedrops, Gramicidin Neomycin Triamcinolone ointment application, tobramycin eye drops, hydroxyl methyl cellulose eye ointment, atropine eye drops to relieve pain were administered every 2nd hourly and the patient was managed conservatively with regular follow-up plans.

The patient was treated with daily dressing with Vaseline-soaked gauze and silver sulfadiazine ointment with moisture retaining dressings. The patient's renal and liver functions were monitored throughout the treatment course. Adequate urine output was maintained throughout the hospital stay.

Aseptic precautions were maintained throughout his hospital stay. Culture and sensitivity reports were used to manage his skin infections with appropriate antibiotics. He was decannulated before discharge from the ICU. Early mobilization and regular physiotherapy also helped to achieve an overall good outcome. Patient was hospitalised for a period of 2 months and was healed completely without surgical intervention.

DISCUSSION

TEN is a medical emergency and requires rapid diagnosis and treatment in an ICU or major burn unit. Both SJS and TEN are now thought to have the same disease mechanism, with the skin surface area being the sole difference 8. A minor version of TEN is one in which less than 10% of the body surface area (BSA) is involved; 10% to 30% of the BSA overlaps with SJS/TEN, and involvement of more than 30% of the BSA is regarded as TEN 9.

The syndrome includes flu-like symptoms, extensive skin involvement, and mucosal lesions. The lesions are usually erythematous morbilliform rashes that first appear on the face and trunk10. Later, the lesions progress with sloughing of the skin and formation of vesicles and bullae. The scalp, palms, and soles are rarely involved. The lesions are fragile and easily slough off with gentle pressure (positive Nikolsky's sign). The mucosal lesions are painful erosive lesions resulting in crust formation. About 80% of patients with TEN develop ocular abnormalities such as purulent conjunctivitis, corneal ulcers, and uveitis11.

The management of TEN includes cessation of the causative cause, multidisciplinary intensive care unit (ICU) care, prevention and early detection of sepsis, fluid and electrolyte balance, adequate analgesia and temperature control, proper organ support, aggressive nutritional management, and good psychological support. The pharmacological therapy for TEN includes corticosteroids, intravenous immunoglobulin, and cyclosporine. The key elements of management are aseptic care and regular moisture retaining dressing of the skin.

CONCLUSIONS

TEN is a life-threatening drug reaction that can be caused by various pharmacological agents including NSAIDS, antibiotics, sulpha drugs. TEN is clinically diagnosed by the presence of skin lesions and a history of exposure to the causative agent. The management of TEN includes cessation of the causative cause, multidisciplinary intensive care unit (ICU) care, prevention and early detection of sepsis, fluid and electrolyte balance, adequate analgesia and temperature control, proper organ support, aggressive nutritional management, and good psychological support. The pharmacological therapy for TEN includes corticosteroids, intravenous immunoglobulin, and cyclosporine. TEN is associated with high mortality if not managed in a systemic and protocolized way. To conclude even extensive TEN can be salvaged with proper management similar on the guidelines of extensive burns.

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