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OCULAR TOXIC EPIDERMAL NECROLYSIS- A RARE CASE REPORT ON TREATMENT TRIGGERING THE DISEASE.

Ophthalmology

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ABSTRACT

We report a case of toxic epidermal necrolysis with ocular involvement in a 20 year old female who presented with confluent purpuric and erythematous dusky red macules evolving to flaccid blister and epidermal detachment on the whole body with conjunctival injection following consumption of tab itraconazole for tinea corporis.Toxic epidermal necrolysis (TEN) and Stevens-Johnson syndrome (SJS) are life-threatening diseases of the skin and mucous membranes. It is typically a side effect of some medications. Toxic epidermal necrolysis can be better managed, with higher survival rates and fewer sequelae if the management concepts are understood better.

KEYWORDS

Toxic Epidermal Necrolysis, Adverse Drug Reaction, Itraconazole, Supportive Care

INTRODUCTION

Toxic epidermal necrolysis (TEN) or Lyell's Syndrome is a rare idiosyncratic life-threatening severe cutaneous adverse reaction (SCAR) predominantly involve the skin and mucous membrane.[1] It is characterized by extensive detachment of the epidermis and erosion of mucous membranes[2]. Stevens Johnson syndrome and toxic epidermal necrolysis are on a spectrum of a severe, immune-mediated, mucocutaneous disease, a diagnosis of SJS is defined as when the skin detachment is less than 10% of the body surface area, and a diagnosis of TEN is defined as when over 30% of the body surface area is involved. A diagnosis of "overlapping SJS-TEN" is defined as when the detachment involves 10% to 30% of the body surface area.[3] They are considered two of the most devastating ocular surface diseases causing corneal damage and threatening vision.

OBJECTIVE

Early intervention during the acute phase to decrease and prevent long term ocular compromise

Case Report

A 20year female was brought to our facility presenting with fever, malaise, difficulty in swallowing and painful generalized erythematous vesiculobullous rash with skin sloughing. (FIG 1). She also complained of pain, redness and difficulty in opening eye. The patient had antecedent significant history of generalised itchy lesions 4 days ago, diagnosed as tenia corporis and treated with Tab Itraconazole 200mg. On examination widespread Skin vesicles and papules involving oral mucosa, neck, and upper extremities with widespread dissemination to the trunk and legs was seen sparing the palms and soles. Nikolsky sign was positive on erythematous zones. Ocular Examination revealed total sloughing of ocular surface and eyelid margin epithelium with conjunctival hyperaemia and injection. Fluorescein dye assessment of ocular surface showed 2x2mm epithelial defect involving pupillary region. Laboratory findings including complete blood cell count, electrolyte/BUN/Cr analysis, liver function test, urine analysis, and tests for autoimmune diseases (anti-nuclear antibody, anti-Smith antibody, anti-neutrophil cyto-plasmic antibody, and smooth muscle antibody) were within normal range. Frequent saline rinse, moxifloxacin and carboxymethylcellulose eye ointment were used. Clinical signs and symptoms rapidly progressed with increasing epithelial defect, sterile infiltrates and conjunctival pseudomembrane. (FIG 2) Saline flushes, pseudomembrane debridement with cotton tipped applicator, moxifloxacin, topical steroid drops with antibiotic-steroid combination ointment to the eyelid, Preservative-free artificial tears, soft therapeutic contact lenses to aid healing and minimize trauma were the treatment employed. Placement of amniotic membrane transplant though treatment of choice was not possible due to instability of the patient's condition and issues with patient cooperativity. Because the total area of epidermis detachment was greater than 30% of total body surface and significant drug history, diagnosis of toxic epidermal necrolysis was made. Patient was managed conservatively. Follow up showed eyelid cicatrization with healed epithelial defect (FIG 3) with visual acuity 6/9 in both eyes without any symblepharon formation and was managed with topical lubricant eye drops and eye ointments.

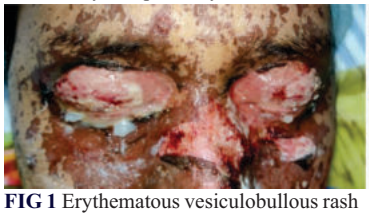

FIG 1 Erythematous vesiculobullous rash with skin sloughing

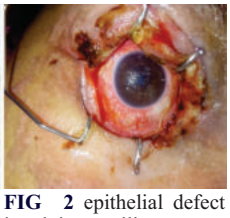

FIG 2 epithelial defect involving pupillary zone

Conjunctival Pseudomembrane

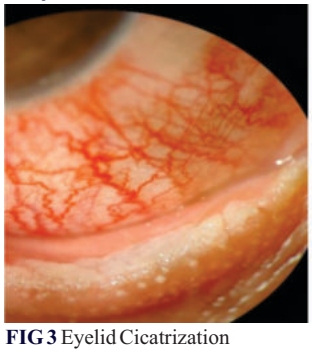

FIG 3 Eyelid Cicatrization

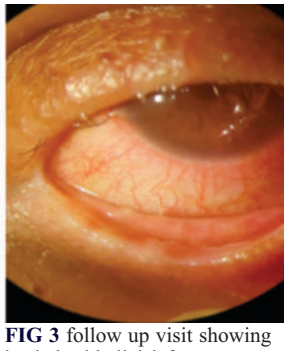

FIG 3 follow up visit showing healed epithelial defect.

DISCUSSION

Toxic epidermal necrolysis (TEN) a rare condition affecting approximately 1 or 2/1,000,000 annually. TEN is considered medical emergency as they are potentially fatal. Cases are typically linked to drug exposure. A number of medications, Genetic factors associated with drug hypersensitivity and HLA typing has been identified as etiopathogenesis behind TEN.[4]

Ocular surface is one of the most commonly involved mucosal surfaces in TEN.[5] Pathophysiology behind is Extensive skin loss is also associated with loss of the barrier function to infections and an increased risk of infection and sepsis by microorganisms that colonize the skin and local inflammatory response associated with release of cytokines into the circulation and a systemic inflammatory response.[6,7]

In 50% to 88% of instances, TEN involves the eyes, which can lead to serious consequences.

Application of lubricating ointment, use of antibiotics to prevent infection, steroid use to control inflammation, and periodic symblepharon lysis are all part of ocular care. AMT is regarded as a recent method for treating TEN's acute stage, which prevents sight-threatening cicatrical consequences by reducing inflammation, preventing the development of ulcers, and promoting healing.[8,9,10]. Ocular problems in the current case were treated with supportive care and eye drops and ointment as AMT could not be done due to issues with patient stability and cooperation and showed promising recovery.

CONCLUSION

SJS multisystem, immune-mediated mucocutaneous disease commonly involves ocular surface, has potential to result in corneal blindness. At the acute stage, ocular involvement is often easily overlooked because of high mortality rates associated with serious systemic diseases. Thus, strict attention must be paid to ocular involvement at the acute stage. The ophthalmologist is a critical caretaker in acute and long-term treatment of these patients. Early aggressive intervention using a standardized protocol is vital to reduce and/or prevent chronic ocular morbidity and in centres where AMT option is not available or patient factors for AMT are not favourable, prompt conservative management can prevent ocular morbidity in the acute stage and in long run too.

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