

Case Report

MULTIDISCIPLINARY CARE OF PATIENT WITH TOXIC EPIDERMAL NECROLYSIS AND SECONDARY SEPSIS

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ABSTRACT

Toxic epidermal necrolysis (TEN) is an acute life-threatening mucocutaneous disease that is mostly drug-related. The diagnosis of TEN can be confirmed by histopathology demonstrating necrotic keratinocytes with separation at the dermal-epidermal junction. The risk of death of TEN can be accurately predicted by the Severity of Illness Score of Toxic Epidermal Necrolysis (SCORTEN). Multidisciplinary care of management is critical for the outcome. We described a patient presented to our hospital with acute life-threatening toxic epidermal necrolysis secondary to ibuprofen; he has diabetes mellitus, hypertension, acute kidney injury, and sepsis. The case substantiates other studies that recommend admission to burn unit and multidisciplinary care in the management of toxic epidermal necrolysis, as well as early consideration of sepsis secondary to staph aureus.

Keywords: Multidisciplinary care, sepsis, Toxic epidermal necrolysis

INTRODUCTION

Toxic epidermal necrolysis (TEN) is an acute and life-threatening mucocutaneous disease occurs rarely with the incidence rate of 0.4-1.2 million persons-year (Lipozencic *et al.*, 2013; See & Mumford 2001) and the mortality rate is up to 30-40% (Lissia M *et al.*, 2010; Wolkenstein & Revuz 2000) reported mostly due to drug-related (Prins 2003) with the characteristics of erosions of mucous membranes and detachment of large epidermal sheets on more than 30% of the body surface area (Roujeau JC 2005). TEN is caused by inappropriate immune activation triggered in response to certain drugs or their metabolites (Downey *et al.*, 2012). Blisters evolve, and large sheets of epidermis slough off, leaving an expose weeping dermis. Early recognition and multidisciplinary management approach are crucial in this regard (Mustafa *et al.*, 2018).

CASE

A 50-year-old male has had hypertension and diabetes presented to the emergency room with fever and skin rash for seven days. The patient reported upper respiratory tract infection and had received amoxicillin-clavulanic acid and ibuprofen, three days later the patient developed abrupt onset fever and skin rash as well as the genital, oral, conjunctival and pharyngeal lesion. On examination, he was alert pulse rate 100 bpm, BP 153/67, T 39.7, and oxygen saturation was 96% on room air. Examination of his face: There was multiple erosion involving the whole face there was conjunctivitis and no corneal involvement; examination of his mouth showed multiple erosion involving the lips, buccal and tongue. Examination of the trunk, abdomen showed detachment of large epidermal sheet [Figure 1.a and 1.b] genital area was involved extensively with scrotal and penile erosions, lower limbs examination showed maculopapular rash, no erosions.

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Figure 1: Area of detached skin involving the face (Figure 1.a) and trunk (Figure 1.b)

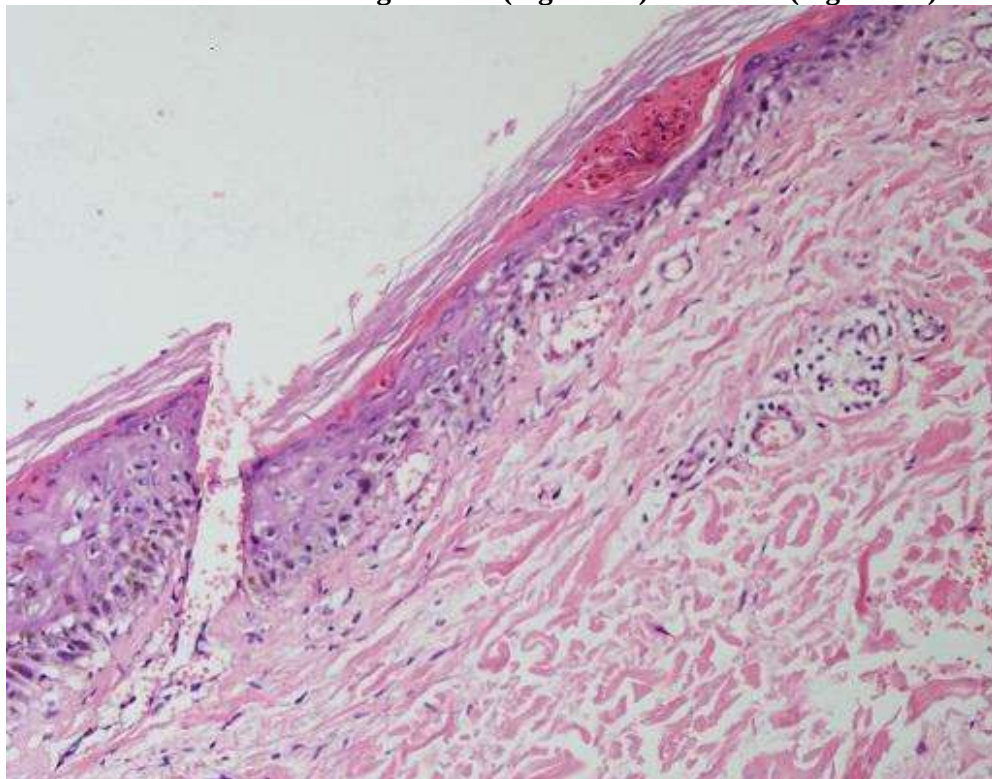


Figure 2: Interface dermatitis consistent with drug eruption.

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The initial laboratory test showed elevated liver enzyme, high renal profile, and leukocytosis. Skin biopsies were taken for hematoxylin and eosin staining, microscopic examination of the skin biopsy showed Interface dermatitis consistent with drug eruption [Figure 2]. Blood culture isolates staph aureus which is sensitive to vancomycin. The patient was diagnosed as toxic epidermal necrolysis with sepsis and the severity was monitored in order to predict the mortality with the help of the Severity of Illness Score of Toxic Epidermal Necrolysis (SCORTEN) score (Bastuji-Garin *et al.*, 2000) [Table 1].

Table 1: SCORTEN score: clinical criteria

Risk factor	0	1
Age	<40	>40
Associated malignancy	No	Yes
Heart rate (beats/min)	<120	>120
Serum Bun (mg/dL)	<28	>28
Detached body surface	<10%	>10%
Serum bicarbonate (mEq/L)	<20	>20
Serum glucose (mg/dL)	<252	>252

He was admitted to the burn unit, the culprit drug was stopped, and he received multi-disciplinary care that includes dermatologist, physicians, ophthalmologists, ENT (ear, nose and throat) and plastic surgeons. The main priorities of management were fluid therapy, analgesics, control of diabetes with insulin, antibiotics covering staph aureus, nutritional support, dressing of the skin lesions as well as adjunctive treatment in form of intravenous immunoglobulin (IVIG) and prednisolone. Remarkable clinical improvement is evident by the relief of symptoms, resolution of sepsis, and healing of the mucocutaneous lesion. The patient was discharged in good condition and given an appointment in the outpatient clinic.

DISCUSSION

Toxic epidermal necrolysis (TEN) represents the most critical form of severe cutaneous adverse drug reaction with a high mortality rate. Management of patients with TEN usually benefits from a large multidisciplinary team for both wound and medical care (Papp *et al.*, 2018). Our patient poses a challenge because of his high SCORTEN, which has several comorbidities (diabetes, hypertension, and renal impairment), and he developed sepsis because of delayed presentation to the hospital. The patient's SCORTEN (table 1) was calculated to be 4, receiving one point for each of the following: age >40 years, >10% total body surface area involvement, serum BUN >28 mg/dL, and serum glucose >252 mg/dL. His SCORTEN score of 4 correlated with a 60% chance of in-hospital mortality.

Toxic epidermal necrolysis (TEN) has unclear treatment recommendations within the literature (Roujeau JC 2005), however, multidisciplinary team of care and admission of patients to the burn unit are two basic management plan that has been recommended by several studies because of mortality benefit (Bastuji-Garin S *et al.*, 2000; Papp *et al.*, 2018; Xia P *et al.*, 2009; Boorboor *et al.*, 2008)). Early involvement of plastic surgeon, dermatologist, physician, and other specialties are essential for optimal medical care. Management of the condition varies between centers and physicians, however, general approach that prioritizes treatment according to the severity of the case is generally advocated. Airway support, optimizing fluid, correction of electrolytes imbalance, and early antibiotic therapy for suspected sepsis, all of the crucial importance (Papp *et al.*, 2018).

Concerning the efficacy of intravenous immunoglobulin (IVIG) in reducing mortality among patients with TEN, there are several studies with variable conclusions (Creamer *et al.*, 2016). However, a large cohort study concluded that the standardized mortality ratio was lowest among those receiving steroid and intravenous immunoglobulin (IVIG) (Micheletti *et al.*, 2018). Our patient received both IVIG and steroids, however, the development of sepsis urge us to reduce the duration of steroid therapy. Staph aureus is the

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most common cause of secondary infection in patients with TEN (Diao M *et al.*, 2018), therefore, it is prudent to consider empirical therapy that covers this microorganism, in our case we started vancomycin empirically which was proved effective as evident by clinical resolution of sepsis and culture result later.

CONCLUSION

Toxic epidermal necrolysis (TEN) is a life-threatening condition, therefore, early recognition of sepsis, admission to burn unit, and multidisciplinary management, are vital steps towards a favourable outcomes.

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