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Toxic Epidermal Necrolysis due to Thalidomide

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Introduction

Toxic Epidermal Necrolysis (TEN) belongs to a spectrum of life-threatening skin disorders, known collectively as Severe Cutaneous Adverse Reaction (SCAR). Although TEN is relatively rare, it carries a high risk of mortality and the immunological reaction is associated with many drugs. Therefore, survival outcome depends on the early initiation and aggressive management in the setting of an Intensive Care Unit (ICU). Our patient developed TEN as a reaction to Thalidomide and subsequently succumbed to death, despite extensive interventions.

Case:

A 62-year-old male with a past medical history of Multiple Myeloma, was referred by the Cancer Center for evaluation and presented with increased lethargy, tachycardia, and hypotension. He had recently been discharged from the hospital after he was admitted for symptomatic anemia. A month earlier, he began a new drug regimen (Daratumumab, Thalidomide, and Dexamethasone) and was scheduled for a platelet transfusion on the day of presentation. During our initial examination, a diffuse erythematous macular rash involving both upper extremities, sparing the palms, was noted. Labs were significant for

a lactic acid of 3, procalcitonin of 8.5, C-reactive protein of 8.5 and pancytopenia (with platelets of 1800). Vital signs were significant for hypotension, tachycardia and a temperature of 102.2 F. The sepsis protocol was activated; the patient was initiated on Doxycycline, Zosyn, and fluid resuscitation, while he was continued on Acyclovir and Bactrim prophylaxis. The patient also received platelet transfusions. When the patient became hemodynamically unstable, he was upgraded to the ICU. The rash progressed to his entire body, including the oral mucous membranes, sparing only the palms and soles, and it subsequently evolved into multiple bullae, with positive Nikolsky's sign. Antibiotics were discontinued, high-dose steroids and emollients were initiated, and aggressive hydration continued. Punch biopsy of a bulla revealed lysis at the dermal-epidermal junction, dissolution of the basal epidermal layer, scattered individual keratocyte necrosis and sparse dermal perivascular lymphocyte infiltration. Despite the aggressive management, the patient continued to deteriorate and transfer to a facility with a burn unit was not feasible. Subsequently, the patient was intubated, started on vasopressors, but the hemodynamic instability of the patient eventually led to his decease.

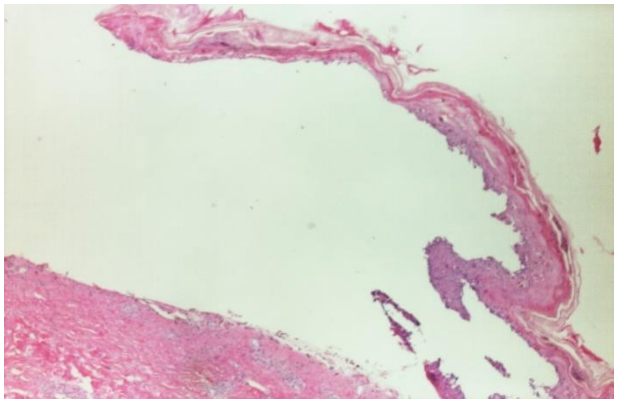

Figure 1

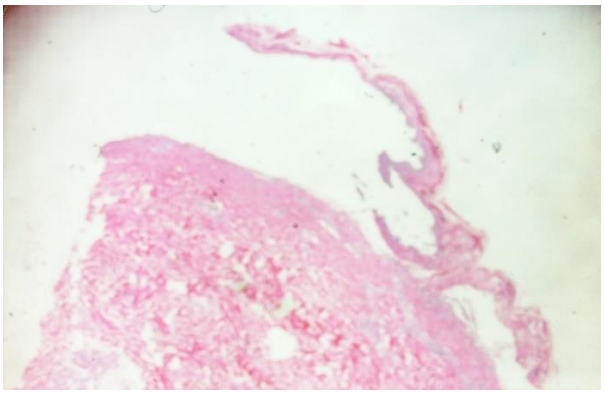

Figure 2

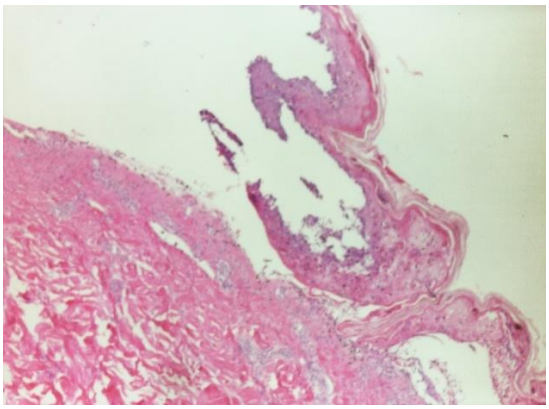

Figure 3

Discussion:

TEN, which occurs secondary to a drug in most cases, is an acute immune reaction that leads to the destruction of the epithelium of the skin and mucous membranes. TEN is similar in presentation to Steven-Johnson Syndrome (SJS) and is distinguished by the Body Surface Area (BSA) involvement greater than 30%. It is believed that immunologic hypersensitivity reactions involving T-Lymphocytes result in keratinocyte death and blister formation. Signs and symptoms of TEN typically appear anytime from a few days to 3 weeks upon exposure to the offending chemical, and as soon as the condition is diagnosed, any possible offending medications should be immediately stopped, and the patient should be managed in a burn or intensive care unit. The skin is the major organ involved, but the conjunctiva, gastrointestinal tract, genitourinary system, pulmonary system and oral cavity can be, and in most cases are, affected as well. In our patient, the skin and oral cavity were the main areas of involvement. The diagnosis of TEN can be confirmed with a punch biopsy, which would reveal necrosed keratinocytes, with full-thickness epidermal involvement; intact stratum corneum; little dermal inflammatory involvement and negative Direct Immunofluorescence of the surrounding skin.

In some cases, fever and flu-like symptoms may precede the onset of rash and blister formation **[**1-3, 6**]**. As soon as the condition is diagnosed, any possible offending medications should be immediately stopped, and the patient should be managed in a burn or intensive care unit **[**1, 2, 6, 7**]**. The first line of defense in the management of TEN is supportive care, as fluid loss and electrolyte imbalances can lead to hemodynamic instability **[**2, 3, 9**]**. Intravenous immunoglobulins and corticosteroids are two of the most commonly used interventions in patients with TEN either individually or in combination **[**1, 4, 9**]**. Although Cyclosporine, Plasmapheresis, Thalidomide and Anti-TNF drugs have shown some promise in small studies and case reports; there have been no large randomized controlled trial to date that validates their efficacy in the management of TEN **[**1, 3, 4, 9-13**]**

References

1.Eginli, A., et al., "*Stevens-Johnson syndrome and toxic epidermal necrolysis".* Annals of Allergy, Asthma & Immunology, 2017. **118**(2): p. 143-147.

2.Estrella-Alonso, A., et al., "*Toxic epidermal necrolysis: a paradigm of critical illness".* Rev Bras Ter Intensiva, 2017. **29**(4): p. 499-508.

3.Mockenhaupt, M., "*The current understanding of Stevens–Johnson syndrome and toxic epidermal necrolysis".* Expert Review of Clinical Immunology, 2011. **7**(6): p. 803-815.

4.Schneider, J.A. and P.R. Cohen, "*Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis: A Concise Review with a Comprehensive Summary of Therapeutic Interventions Emphasizing Supportive Measures".* Adv Ther, 2017. **34**(6): p. 1235-1244.

5.Sousa-Pinto, B., et al., "*Stevens–Johnson syndrome/toxic epidermal necrolysis and erythema multiforme drug-related hospitalisations in a national administrative database".* Clinical and Translational Allergy, 2018. **8**(1): p. 2.

6.Harr, T. and L.E. French, "*Toxic epidermal necrolysis and Stevens-Johnson syndrome".* Orphanet J Rare Dis, 2010. **5**: p. 39.

7.Gerull, R., M. Nelle, and T. Schaible, "*Toxic epidermal necrolysis and Stevens-Johnson syndrome: a review".* Crit Care Med, 2011. **39**(6): p. 1521-32.

8.Abe, R., "*Immunological response in Stevens-Johnson syndrome and toxic epidermal necrolysis".* J Dermatol, 2015. **42**(1): p. 42-8.

9. Zimmermann, S., et al., "*Systemic Immunomodulating Therapies for Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis: A Systematic Review and Meta-analysis".* JAMA Dermatol, 2017. **153**(6): p. 514-522.

10. Lee, H.Y., et al., "*Cyclosporine treatment for Stevens-Johnson syndrome/toxic epidermal necrolysis: Retrospective analysis of a cohort treated in a specialized referral center".* Journal of the American Academy of Dermatology, 2017. **76**(1): p. 106-113.

11. Kirchhof, M.G., et al., "Retrospective review of Stevens-Johnson syndrome/toxic epidermal necrolysis treatment comparing intravenous immunoglobulin with cyclosporine*".* Journal of the American Academy of Dermatology, 2014. **71**(5): p. 941-947.

12.Kinoshita, Y. and H. Saeki, "*A Review of the Active Treatments for Toxic Epidermal Necrolysis".* J Nippon Med Sch, 2017. **84**(3): p. 110-117.

13. González-Herrada, C., et al., "*Cyclosporine Use in Epidermal Necrolysis Is Associated with an Important Mortality Reduction: Evidence from Three Different Approaches".* Journal of Investigative Dermatology, 2017. **137**(10): p. 2092-2100.