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"TIME IS RUNNING OUT" : STEVEN- JOHNSON SYNDROME AND TOXIC EPIDERMAL NECROLYSIS OVERLAP SYNDROME, A CASE REPORT

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Introduction

Steven-Johnson Syndrome (SJS) and toxic epidermal necrolysis (TEN) are two forms of acute life-threatening dermatology emergencies. SJS and TEN are considered two ends spectrum of adverse drug reaction, both characterized by extensive necrosis and detachment of epidermis. SJS is the less severe condition, in which skin detachment is less than 10 percent of the body surface area (BSA), SJS/ TEN overlap syndrome is where skin detachment is between 10 to 30 percent, whereas TEN involves detachment of ore than 30 percent of the BSA. [2] This case report discussed on a case of SJS and TEN overlap syndrome and highlights on the rapid progression of disease observed in our emergency department.

Case Report

40-year-old lady presented with fever for past 2 days, associated with bilateral eye swelling and pruritic generalized rash. She was admitted previously for surgical procedure due to meningioma and started with steroids, antibiotics, proton pump inhibitors and anticonvulsants. These medications were last taken 2 weeks prior presentation to our centre. On arrival, patient was febrile and maculopapular rash were noted over her trunk, arms, thigh, face and back.

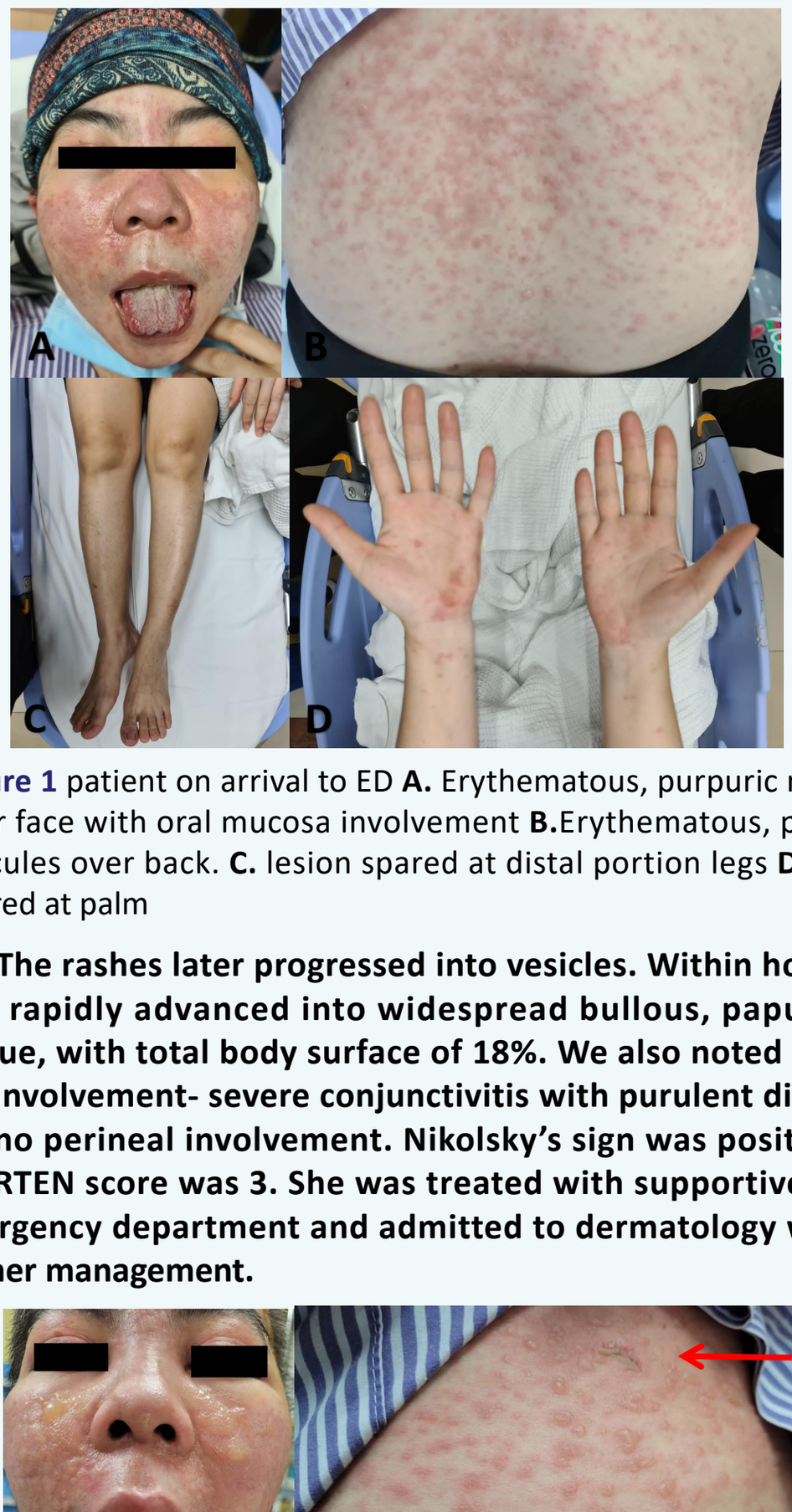

Figure 1 patient on arrival to ED **A.** Erythematous, purpuric macules over face with oral mucosa involvement **B.**Erythematous, purpuric macules over back. **C.** lesion spared at distal portion legs **D.** lesion spared at palm

The rashes later progressed into vesicles. Within hours, the rash rapidly advanced into widespread bullous, papules and plaque, with total body surface of 18%. We also noted oral and eye involvement- severe conjunctivitis with purulent discharge, but no perineal involvement. Nikolsky's sign was positive. Her SCORTEN score was 3. She was treated with supportive care in emergency department and admitted to dermatology ward for further management.

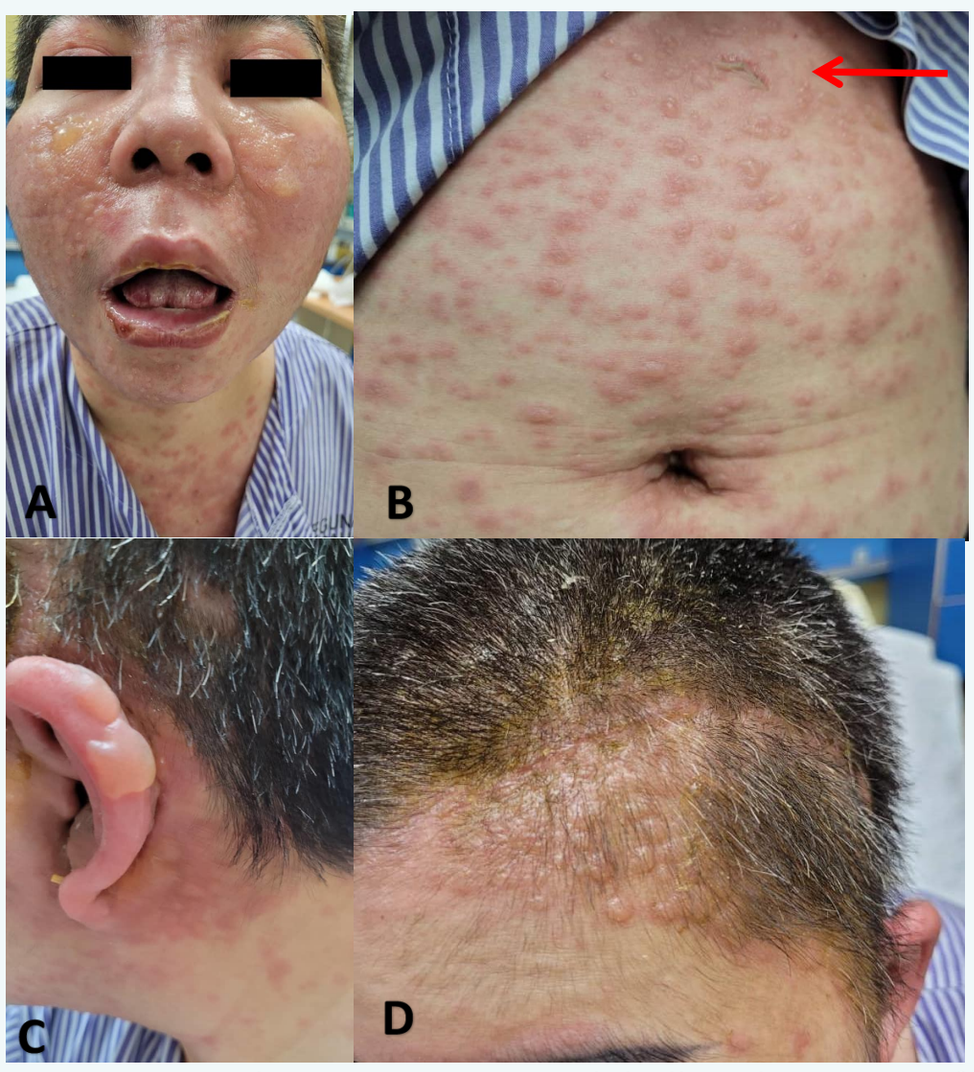

Figure 2 patient 12 hour in ED **A.** vesiculobullous lesion over face **B.** vesiculobullous lesion over abdomen. **C.** bullous over left pinna and external auditory canal **D.** vesiculobullous lesions involving scalp
Red arrow points to Nikolsky's sign

Discussion

Most literature focus on onset of the disease, but little emphasis was given to the progression of disease. Progression of disease includes the progression of the lesion morphology - from the initial skin redness to the later ruptred bullous, as well as the progression of body surface area involvement.

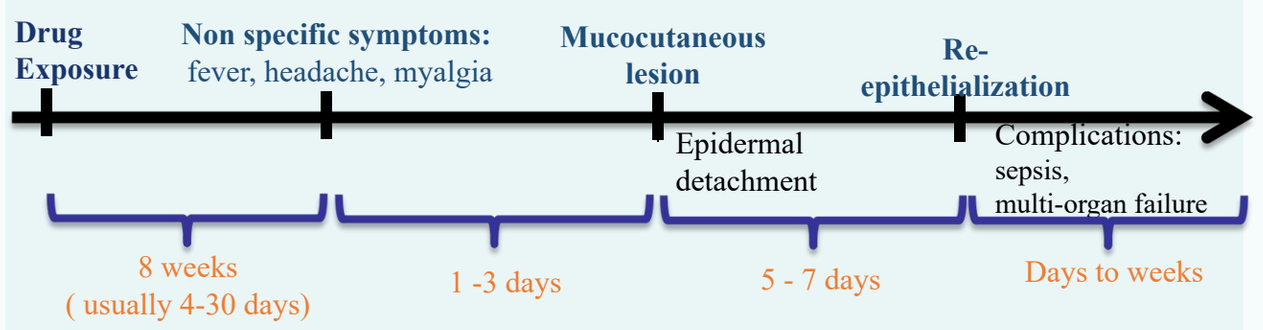

Figure 3 shows the timeline of SJS and TEN presentation

Most publications vaguely described the rash where it can rapidly extend to the rest of the body within hours to days[3], with no exact numerical range of time mentioned. This case report which demonstrated a rapid progression within 12 hour is highlighted for below learning points:

(1) Disposition

SJS and TEN patients may present during early manifestation with the main complaint of non-specific prodromal symptoms with erythematous or macules rash, which can be easily mistaken as a non-emergency condition, leading to inappropriate management and dismissal. Therefore, drug history must be explored in all patient presented with rash. Be mindful that history of drug exposure can be up to 8 weeks earlier. All patients with drug related rash is suggested to be observed for a longer period of time in the department. During observation period, rapid progression of the skin lesion should alert one as onset of severe disease. In any patient who presented with rash, if patient is to be dismissed eventually, rapid progression of disease as a red flags symptoms must be included in patient discharge advice.

(2)Prognostication

Studies have shown that mortality of SJS and TEN is determined primarily by the extend of skin sloughing.[1] A commonly used prognostication tool- SCORTEN scoring system has also concluded that epithelial detachment more than 10 percent contributes to higher mortality rate. It is well demonstrated in this case where SJS and TEN patient may initially presented with mild symptoms hence scoring a low mortality rate, but as the disease dynamically progressed into blistering and skin sloughing along with expansion of compromised body surface area, the mortality rate exponentiates significantly. It is thus important to note that SJS and TEN patient should always be managed in clinical zones where close monitoring is possible, regardless of the initial presentation that may appear trivial. Rapid progression within hours can occur. Hence, patient progression should be closely followed to ensure severe disease and complications to be picked up early and addressed instantly upon detection.

Conclusion

In conclusion, rapid progression should be used as an important indicator deciding on the disposition of any patient who presents with rash. Secondly, the disease involves wide variety of morphology appearance, thus clinician should always held high vigilance of diagnosis. Thirdly, SJS and TEN can rapidly progress, hence they must be closely monitored.

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Declaration of conflicts

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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