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Stevens-Johnson syndrome and toxic epidermal necrolysis: Vulvar and vaginal manifestations

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Keywords: Stevens-Johnson syndrome; Toxic epidermal necrolysis; Vulvovaginal epidermal necrolysis.

Abbreviations: SJS: Stevens-Johnson Syndrome; TEN: Toxic Epidermal Necrolysis; PPI: Proton Pump Inhibitor; BSA: Body Surface Area.

Description

A 75-year-old woman was admitted to our emergency department due to SJS and TEN to investigate the involvement of the vulva and the vaginal mucosa and make a consultation. The woman's medical history included hypertension, managed with a beta-blocker and a combination hydrochlorothiazide and valsartan, and gastro-esophageal reflux disease managed with a PPI which recently started.

SJS and TEN are severe mucocutaneous adverse reactions characterized by epidermal necrosis. Both are variants of the same disease process and categorized by the percentage of BSA with epidermal detachment [1]:

- SJS: <10% of BSA
- SJS/TEN: 10-30%
- TEN: >30%

There is no certain cause but many factors have been suspected such as infections or medications like non-steroidal anti-inflammatory drugs (e.g. oxicams family etc.), anti-neoplatic agents or antiepileptics. Women have higher risk to develop the disease. HLA-B15 and HLA-A31 phenotypes seem to be related with increased risk.

Treatment and proper care of the involved tissues is required to minimize the long-term effects of the disease. About 70% with SJS or/and TEN have vulvovaginal involvement and up to one third of the treated patients will have chronic complications [3]. There is no consensus on the optimal treatment. Usually a combination of topical agents such as corticosteroids and es-

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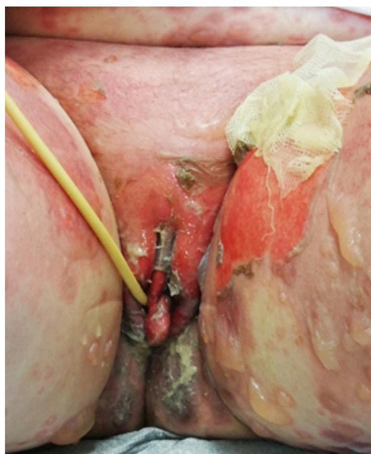

Figure 1: The characteristic lesions of SJS/TEN are evident on nearly the whole surface of the external genitalia.

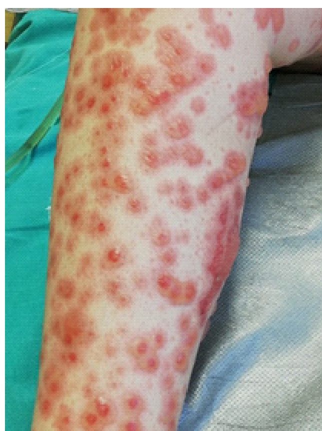

Figure 2: Typical macroscopic appearance of the SJS cutaneous lesions; target lesions, large blisters which exhibit Nikolsky's sign. Right lower extremity of the patient.

trogens, vaginal dilator therapy and Foley catheter to avoid agglutination and menstrual supression is the preferred management. The role of the gynecologist is to determine the vaginal mucosa involvement and give proper guidelines as mentioned above [2]. Most of the times speculum examination can be carried out only under anesthesia. Finally, the primary care gynecologist should not forget that any non-traumatic erosion or ulcer of the vagina or vulva might be the initial presentation of the syndrome. In our case we can only suspect that the recently started PPI was the trigger factor. The patient diascharged after long hospitalization in the Burn Unit of our hospital [5].

Declarations

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