"BLE: The Elusive Rash"

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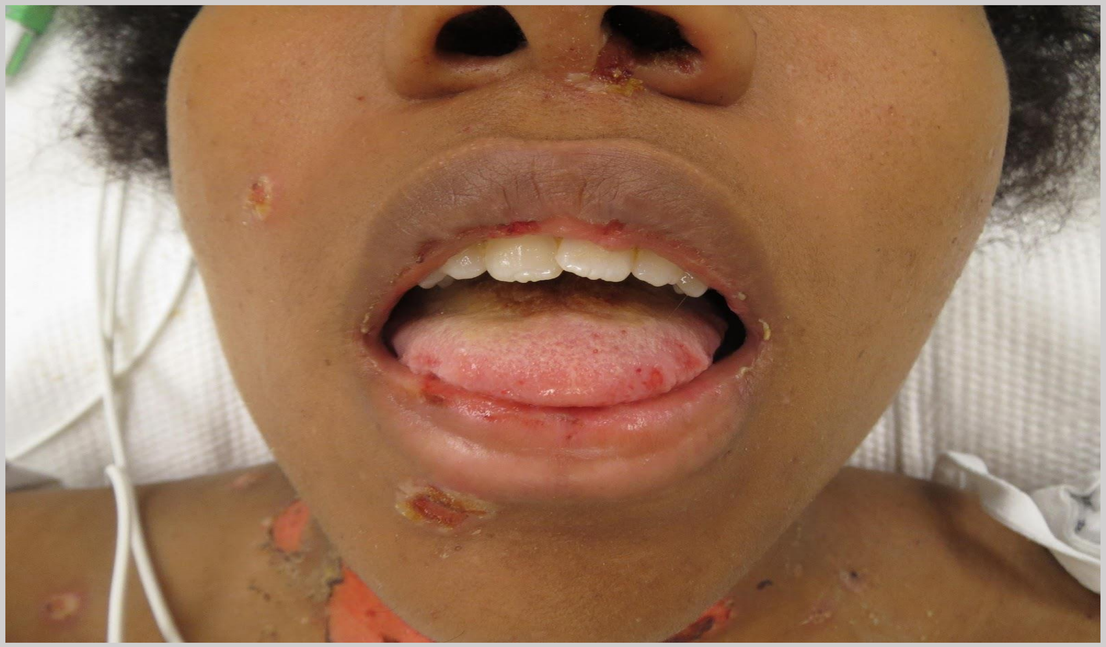
THE CASE

A 21- yr- old female presented to the Emergency Department with a chief complaint of rash, fever and generalized body pain. The patient reports that she was recently in Las Vegas for a concert about four weeks before presentation. A rash developed across her back and her abdomen. The lesions were "like hives" in nature and accompanied by itchiness. She denies any bug bites or new exposures. She visited an urgent care with the same complaints and was given clindamycin and hydrocortisone which caused worsening of the rash. Blisters began to develop as well as a fever. She had no medical problems or significant family history, with a reported sulfa allergy.



QUESTIONS

1. What is the diagnosis?
2. What work-up is needed?
3. What is the treatment?

ANSWERS

1. Bullous Lupus Erythematosus
2. Biopsy and exclusion of other possible disease processes
3. Dapsone, IVIG, and rituximab

DISCUSSION

Bullous Lupus Erythematosus (BLE) are rare lesions that mostly affect patients with a history of lupus. However, it may also be the first presenting sign of the disease. It is common in young women, and usually involves the face, upper trunk and proximal extremities. The patient in question had not been diagnosed with lupus previous to this admission, but was found to have positive complement levels, c-ANCA and p-ANCA reinforcing a Lupus diagnosis.

BLE is a result from the disruption of epidermal-dermal adhesion, caused by antibody formation against type VII collagen. These antibodies can be detected in sera in these patients. A neutrophilic infiltrate in the superficial dermis is also a common finding.

There are certain criteria needed to adequately diagnose BLE which include: acute eruption of vesicles and/or bullae; histopathologic finding of subepidermal blistering and a neutrophil-predominant infiltrate in the superficial dermis; direct immunofluorescence finding of linear or granular immunoglobulin (IgG, IgM or IgA) deposition at the basement membrane zone; elevated ANA; exclusion of other possible disease processes. The patient in question fulfilled the majority of these criteria.

Data for therapeutic options for BLE are limited, however, dapsone is the first line treatment. Past case studies and reviews have noted that dapsone has led to significant improvement in the majority of BLE patients, when not contraindicated. The improvement noted on dapsone, has now become characteristic of BLE. Corticosteroids and immunosuppressive drugs has noted to be helpful treatment adjuncts. The efficacy of these treatments varies and therefore recommendations are still highly debatable. In the patient in question, she was given dapsone, IVIG and rituximab, which showed significant improvement. Patient would later be discharged from the Burn ICU, in stable condition, with a course of dapsone to be completed outpatient. Patient continues to be followed by Rheumatology and has had no further complications since this admission.

CLINICAL PEARLS

When dealing with patients with a new rash, don't forget the must not miss diagnosis (Toxic Epidermal Necrosis, Steven Johnson Syndrome, Toxic Shock Syndrome and Staph Scalded Skin Syndrome). However, always keep in mind that the true culprit might be more elusive and require further investigation.

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