

AUTOIMMUNE BULLOUS DISEASES

LIG-AVENGERS BLISTER WARS: A CASE OF A 15-YEAR OLD MALE WITH LINEAR IGA DERMATOSIS

M. Sayo (1) - F. Jacinto-calimag (1) - M. Torres (1)

East Avenue Medical Center, Dermatology, Quezon City, Philippines (1)

Background: Linear IgA (LIGA) Dermatosis is a rare autoimmune subepidermal blistering disease that occurs bimodally either after puberty or after the 40th decade of life. It presents with lesions showing grouped bullae and vesicles in a cluster-of-jewel morphology on the trunk and extremities that may have mucosal involvement. It has an unpredictable course that may continue for years with spontaneous remissions of three to six years.

Observation: A 15-year old Filipino male presented with multiple well-defined, irregularly-shaped, clear fluid-filled tense and flaccid bullae and some vesicles on the trunk, upper and lower extremities. He also presented with irregularly-shaped erythematous plaques, some topped with hemorrhagic crust and shallow erosions over an erythematous base at the vermillion border of the lips. Nikolsky and Asboe Hansen signs were both negative and the Tzanck smear showed a prominence of neutrophils while skin punch biopsy showed findings consistent with subepidermal bullae with predominant neutrophil infiltrates. After baseline laboratory work-up and G6PD Assay, the patient was started with dapsone 50 mg/tab a day, prednisone 20 mg/day with slow tapering, omeprazole 20 mg/tab OD, and cetirizine 10 mg/tab BID. After two days of intake of dapsone, the patient was noted to have significant improvement by decrease in number and erythema of lesions.

Key Message: After a pre-requisite of a normal G6PD assay, the blisters have responded within two days after initiating dapsone with an additional low-dose prednisone to suppress blister formation as seen in our patient. This strengthens the diagnosis of LIGA but to avoid the side effects of dapsone such as hemolytic anemia, it is advisable to monitor the patient by having thorough body examination to check for recurrence of lesions and by doing serial complete blood count.





