AUTOIMMUNE BULLOUS DISEASES

UNCOVERING THE MASK: LINEAR IGA BULLOUS DERMATOSIS TREATED WITH DAPSONE IN A FILIPINO ADULT MALE

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Background: Linear IgA bullous dermatosis is a chronic, acquired autoimmune blistering disease that classically presents with tense blisters arranged in a “crown of jewels” formation.

Observation: This case presents a 41-year-old male with multiple, well-defined, clear fluid-filled, tense vesicles with crusted erosions and urticarial plaques on the face and neck that later involved the chest, back, anogenital area, and extensors in an annular pattern. Histopathology revealed a subepidermal split with neutrophil predominance supported by a direct immunofluorescence showing linear IgA deposition at the dermoepidermal junction. The patient was initially given oral corticosteroids. After adequate G6PD levels were ascertained, dapsone was subsequently started while tapering oral corticosteroid, with resolution of lesions.

Key Message: Linear IgA bullous dermatosis is an autoimmune blistering disease that presents with tense bullae and crusted erosions on the face, trunk, genitals, and extensors with a peak incidence in children and adults. The disease may mimic presentations of other autoimmune blistering diseases, and histopathology may not always yield specific results.
This case highlights the clinical heterogeinity of LABD and the failure to reach full clinical control with use of second-line agents. Quality of life can also be severely affected despite a mild disease activity. A complete history, physical exam, and diagnostic work-up that include both histopathologic findings and direct immunofluorescence are needed to accurately diagnose and unmask the disease.